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Société Belge de Pédiatrie



The changing face of pediatrics

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Dear colleagues,

The SARS-CoV2 virus continues to rock our world. Precautionary measures need to be implemented. Although an efficient vaccine can give us some hope for the future, planned activities still have to be postponed and even cancelled.

Therefore, BVK-SBP and the organizing committees have decided to organize the 49th annual meeting as a virtual congress.

Since the meeting of March 2020, hosted by ULB-Huderf, had to be canceled we decided to 'merge' the program and host this meeting by two organizing universities, ULB and UGent, which is unique in the 49 years history of our BVK/SBP-conference.

With the recent experience of joining forces in the pediatric Covid-19 taskforce, we were convinced that we could compose a scientific program, over the borders of language, academic institutions and even health professions (pediatric nurses and pediatric psychologists are welcome to participate, since we also have a theme session, hosted by them).

The topic of the meeting will be 'The changing face of pediatrics'. The cutting edge scientific program with renowned national and international speakers will address topics relevant for clinical paediatric practice, and of course Covid-related topics could not be avoided!

We also wanted to focus on the multidisciplinary approach of the chronically ill child and on patient participation and shared decision making with several patient and family testimonies.

In these special times, we decided to go for a low subscription fee, which will probably not be possible to repeat next year.

We are excited to welcome you to our first digital congress and look forward to seeing you virtually the 18th and 19th of March 2021!

On behalf of the organization

Prof. Dr. Sabine Van daele
(UGent/UZGent)
President of the congress

Prof. Dr. Pierre Smeesters
(ULB/Huderf)
President of the congress

Dr. Marc Raes
President of BVK/SBP



HET LEVEN
IS GEEN
KINDERSPEL

LA VIE
N'EST PAS
UN JEU
D'ENFANT

DE BVK BEDANKT ZIJN PARTNERS
VOOR HUN STEUN

LA SBP REMERCIE SES PARTENAIRES
POUR LEUR SOUTIEN



We care for children



BELGISCHE VERENIGING
VOOR KINDERGENEESKUNDE
SOCIETE BELGE DE PEDIATRIE

BVK/SBP Congress 2021

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Universiteit Antwerpen, UZA

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Sciensano
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UZ Gent
- PW 25. The internet use of Belgian parents relating to a physician's visit for their children: a cross sectional survey.**
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KU Leuven, UZ Leuven
- PW 26. Urinary Incontinence in The Obese Population: Characteristics and Therapy-Outcome**
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Universiteit Gent, Zeepreventorium
- PW 27. Survey on first-aid knowledge in members of Flemish youth movements.**
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KU Leuven, UZ Leuven

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Universiteit Antwerpen
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Centre for Sociological Research, KUL, Spaarne Gasthuis, UZ Leuven
- P 3. Congenital external ear malformation : key feature to the diagnosis ?**
N. Laval, N. Revencu, Y. Sznajer
Centre de génétique humaine, Cliniques Universitaires Saint Luc
- P 4. Survey about the alcohol consumption by minor in Flemish youth movements**
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KUL, UZ Leuven

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- P 6. Migrant pain, weight loss and inflammatory syndrome: did we worry too much?**
M. Rodesch, L. Goffin, F. Vermeulen
ULB - HUDERF, ULB – Erasme
- P 7. A rare cause of genital ulceration.**
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Etterbeek-Ixelles Hospital
- P 8. Abdominal Pain and Acute Cutaneous Nerve Entrapment Syndrome: Go with the gut, but use your head.**
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UZ Leuven, Imeldaziekenhuis Bonheiden
- P 9. Increase in child sexual abuse following sanitary confinement : experience of the Maltraitance Unit of the Pediatric Service of CHU Liège**
S. Pannizzotto
CHU Liège
- P 10. How to Maximize Children’s Involvement in Non-therapeutic Research—Lessons Learnt From EFFECTOR**
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Universiteit Gent, VUB, UZ Gent
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KUL, UZ Leuven
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KUL, UZ Leuven

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- LO 2. Perinatal risk factors associated with the cardiovascular risk profile of Extremely Low Birth Weight survivors**
L. Van Loo, F. Wei, Z. Zhang, J. Staessen, K. Allegaert, A. Raaijmakers
UZ Leuven, Sun Yat-Sen University, Cardiovascular Research Institute Maastricht (CARIM), Erasmus MC, ZNA Hospitals Antwerp

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UCL Saint-Luc
- SO 9. Early postnatal outcome and care after in utero exposure to lithium : a single center analysis**
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KU Leuven, UZ Leuven
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UZ Leuven, RZ Tienen
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U Liège, UZ Leuven
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UZ Leuven, KUL
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VUB, UZ Brussel
- SO 32. Respiratory outcomes in very and extremely preterm infants: what is the impact of recommended neonatal care in real-life conditions?**
A. Vuckovic, D. Mcheik, A-B Johansson
ULB - HUDERF

Poster Walk

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UZ Leuven, Sun Yar-Sen University, Research Institute Alliance for the Promotion of Preventive *Medicine*, *Erasmus MC, ZNA Hospitals Antwerp*
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UZ Gent

Posters

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ULB - HUDERF
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Clinique Saint Vincent Rocourt
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Universiteit Antwerpen, UZ Antwerpen
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KUL, UZ Leuven
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CHC Montlegia

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KUL
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- P 26. Trali in a PIMS-TS patient**
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UZ Brussel
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Centre hospitalier du Luxembourg
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V. Collet, E. Boros, V. Vlieghe, A-B Johansson
ULB - HUDERF
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CHC Rocourt
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ULB - HUDERF
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E. Goffinon, C. Vilain, J. Soblet, A. Vuckovic
ULB - HUDERF
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UZ Antwerpen, Universiteit Antwerpen, AZ Turnhout
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Universiteit Gent
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UZ Gent

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KUL
- P 38. Neonatal unexplained HUS treated with complement inhibitor Eculizumab**
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ULB - HUDERF, CHU Saint Pierre
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UCL Saint-Luc
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Universiteit Gent

INFECTIOLOGY – PNEUMOLOGY – IMMUNOLOGY

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UZ Gent
- LO 9. Beyond culture: the Molecular Assessment of Thoracic Empyema (MATE) study**
L. Fabri, J. Jacobson, J. Osowicki, S. Shanthikumar, E. Dunne, A-M Costa, A. Daley, W. Teague, J. Buttery, A. Steer, S. Ranganathan, C. Satzke
Murdoch Children's Research Institute, The University of Melbourne at the Peter Doherty Institute for Infection and Immunity, Monash Children's Hospital Victoria, Monash University, Royal Children's Hospital Melbourne, ULB

Short Oral Presentation

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L. Schiavolin, P. Smeesters, A. Botteaux
ULB
- SO 27. Multiple breath washout measurement in patients with CDH at school age compared to Chest CT score and spirometry**
M. Dirickx, F. Vermeulen, M. Boon, A. Debeer, M. Proesmans
UZ Leuven
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C. Wyndham-Thomas, N. Hammami, C. Schirvel, R. Mahieu, G. Top, M. Lesenfants, E. Wollants, S. Quoilin
Sciensano, Agentschap Zorg en Gezondheid, Agence pour une Vie de Qualité, Commission communautaire commune, Centre National de Référence des entérovirus, UZ Leuven
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UZ Gent
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ULB-CHU Ambroise Pare, CHU Saint-Pierre Hospital
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Mutualités Libres - Onafhankelijke Ziekenfondsen, ULB
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ULB - HUDERF

Poster Walk

- PW 7. Immunization budget for Belgium: Horizon scanning using an Immunisation Planning Tool**
A. Bento-Abreu, J. Vandenhaute, N. Dauby, B. Merckx, G. Bencina
MSD
- PW 9. Clinical description and outcomes of children with invasive group A Streptococcal disease**
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ULB, Royal Children's Hospital, Victoria
- PW 10. Streptococcus pyogenes related hospitalisations in a single tertiary pediatric center in Belgium: overview over 14 year**
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UZ Gent
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ULB - HUDERF
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Murdoch Children's Research Institute, ULB, University of Melbourne, Victoria, The Royal Children's Hospital Melbourne
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UZ Leuven
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L. Fabri, S. Shanthikumar, P. Robinson
Murdoch Children's Research Institute, Melbourne, Australia, ULB - HUDERF, Royal Children's Hospital, Melbourne, Australia, University of Melbourne, Parkville, Victoria, Australia
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Clinique Saint Jean, ULB-HUDERF, UZ Brussel, Sciensano, ULB-CHU Saint Pierre

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UCL, Clinique Notre-Dame de Grâce
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AZ Damiaan, AZ Sint Jan

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CHU UCL Namur Saint-Elisabeth
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HUDERF-ULB
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Centre Hospitalier epiCURA, ULB, CHU Saint-Pierre
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UZ Gent, KUL

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UZ Brussel
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University Hospital Liège, Université de Liège
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Cliniques Universitaires Saint Luc
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CHU Saint Pierre, Hôpital Civil Marie Curie
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UZ Gent
- P 70. Hepatic capillaritis in a refugee child with PICA**
S. Groignet, V. Antoons, D. Tuerlinckx, O. Chatzis, E. Sokal, E. Bottieau, R. Bradbury, D. Van der Linden
UCL
- P 71. Listeria meningoenzephalitis in an immunocompetent child**
S. Finocchiaro, M. Gillet, F-X Van Vyve, M. Creuven, C. Genin, P. Philippet
U Liège, UCL
- P 72. Refractory chylothorax in children : a case report**
E. Brandt, P. Van Laer, J. Papadopoulos, D. Francois, C. Croisier, C. Heijmans
Hôpital de Jolimont

- P 73. The prevalence of hypercapnia during an acute infection in children on chronic non-invasive ventilation: a retrospective study**
T. Beyltjens, S.R. de Leede, I. van Eekelen, F.F.W. van Ginneken, S. Installe, K. Van Hoorenbeeck, S. Verhulst
UZ Antwerpen, Lab of Experimental Medicine and Pediatrics, Universiteit Antwerpen
- P 74. HIV-infected mothers who decide to breastfeed their Infants under close supervision in Belgium: about two cases**
N. Bansaccal, D. Van der Linden, J-C Marot, L. Belkhir
UCL Saint-Luc, Cliniques Saint-Pierre
- P 75. Two cases of hemolytic uremic syndrome in the same childcare center in Brussels**
A. Revercez, R. Mahieu, S. Clément de Cléty, E. Rebuffat, T. Goetghebuer, R. Mahieu, E. Rebuffat, T. Goetghebuer
CHU Saint-Pierre, Infection Prevention and Control, Common Community Commission (COCOM), PICU, UCL Saint-Luc
- P 76. Chlorhexidine anaphylaxis after topical use in a 13-year old girl: a case report.**
L. Depoorter, J. Vanderkerckhove, M. Grosber, G. van Berlaer, Y. Vandenplas, E. De Wachter
UZ Brussel
- P 77. Benefit of genetic sequencing in the treatment of a tuberculosis disease case.**
E. Surgun, O. Stevart, V. Mathys, F. Ziereisen, F. Mouchet
ULB-CHU Saint-Pierre, Sciensano
- P 78. Cross-transmissions of Pseudomonas aeruginosa in a pediatric cohort with cystic fibrosis**
J-C Beghin
ULB - HUDERF
- P 79. Invasive Haemophilus Influenzae infections in the pediatric patient a case series**
D. Vermeulen, P. Schelstraete
UZ Gent
- P 80. Homozygous nonsense mutation in RC3H1, which encodes Roquin-1, causes a novel immune dysregulation syndrome characterized by severe hyperinflammation**
P. Verloo, S.J. Tavernier, V. Athanasopoulos, G. Behrens, D. Bogaert, M. De Bruyne, E. Parthoens, J. Ellyard, H. Van Gorp, J. Staal, G. Van Isterdael, M. Lamkanfi, P. Schelstraete, R. Van Coster, B. Menten, B. N. Lambrecht, C. Vinuesa, V. Heissmeyer, R. Beyaert, M. Dullaers, F. Haerynck
UZ Gent, VUB and many others
- P 81. Cluster of invasive meningococcal disease caused by a meningococcus serogroup B in a nursery school in Wallonia, Belgium in 2018**
S. Jacquinet, W. Mattheus, S. Quoilin, C. Wyndham-Thomas, M. Leroy, C. Martin, D. Van der Linden, A. Mulder, J. Frère, C. Schirvel
Sciensano, ULB-CHU Saint-Pierre, UCL Saint-Luc, CHC – Liège, CHU Liège, Agence pour une vie de qualité, infection prevention and control
- P 82. Phage therapy to allow liver transplantation in a toddler infected by an extensively-drug resistant Pseudomonas aeruginosa**
B. Van Nieuwenhuyse, O. Chatzis, R. Reding, S. Djebara, E. Sokal, M. Merabishvili, J-P Pirnay, P. Soentjens, D. Van der Linden
UCL Saint-Luc, Hôpital Militaire Reine Astrid
- P 83. FINCA disease presenting as Severe Childhood onset Interstitial lung disease (chILD) with neurodevelopmental disorder**
I. Van Dijck, F. Vermeulen, M. Proesmans, M. Boon
UZ Leuven

GASTROENTEROLOGY – NUTRITION

Long Oral Presentation

- LO 10. Infliximab trough levels at the end of induction will predict endoscopic remission in paediatric patients with Inflammatory Bowel Disease**
K. van Hoeve, E. Dreesen, I. Hoffman, M. Ferrante, S. Vermeire
UZ Leuven, KUL

Poster Walk

- PW 18. A systematic review on health care implications of a vegetarian diet in children**
L. Leclair, J. Toelen
KUL
- PW 19. Bowel Function in Children With Low-Type Anorectal Malformation After Surgical Repair**
F. De Baene, K. Van Renterghem, S. Van Biervliet, S. Vande Velde, M. Van Winckel, D. Van de Putte, L. Matthyssens, R. De Bruyne
UZ Gent
- PW 21. Impact of Nutrition Support Team (NST) funding on the quality of care of Home Parenteral Nutrition (HPN) in children with benign diseases**
N. Umbrain, D. Hermans, P. Schlessers, M. Hiele, M. Van Winckel, D. Ysebaert, A. Van Gossum
ULB - ERASME, UCL Saint Luc, CHC Chrétien, KUL, UZ Leuven, UZ Gent, UZ Antwerpen

Posters

- P 84. Acute cholecystitis in a patient with normal imaging findings on initial presentation**
B. Boon, K. Poschet, M. Claeys, A. Calders, A. Delmotte, F. De Meulder
GZA Sint-Vincentius
- P 85. Acholic stools and the diagnosis of a choledocholithiasis**
A. Lambiri, M. Maka
EpiCURA Hôpital
- P 86. Seronegative autoimmune hepatitis in children: An atypical entity to mention**
S. Mortaji, E. Bequet, O. Guidi, M-C Seghaye
CHU de Liège, U Liège
- P 87. Food Protein-Induced Enterocolitis Syndrome: an emergency diagnosis we must know!**
C. Micin, O. Guidi, M-C Seghaye, E. Bequet
U Liège, CHU de Liège
- P 88. When the bowel dances the twist: A rare case of sigmoid volvulus in a child**
A. Ouald Chaib, E. Levy, K. Huysentruyt, E. De Greef, G. Veereman, B. Hauser, T. Devreker, M. Aerts, C. Ernst, C. Vercauteren, Y. Vandenplas
UZ Brussel, VUB
- P 89. Rectal bleeding in infants: a clinical sign not to be neglected**
M. Lahr, A. Taxhet, M. Demarche, M-C Seghaye
CHU de Liège, U Liège, CHR de la Citadelle
- P 90. An interesting case “out of paper” with celiac disease leading to xylophagia: a case report**
E. Box, T. Claeys
UZ Leuven

- P 91. Constipation as the only symptom of ovarian teratoma: about a case.**
M. Saliba, A. Okroglic, M. Demarche, M-C Seghaye
University Hospital Liège, University of Liège, Regional Hospital La Citadelle
- P 92. Acute abdominal pain in teenager**
O. Leclercq, L. Lecomte
CH Jolimont
- P 93. Scurvy in a toddler - A case report**
Z. Ouchinsky
ULB - HUDERF
- P 94. Case report of a rare cause of jaundice in children: autoimmune pancreatitis**
J. Van Huffel, M. Renard, O. Guidi, J-L Lismonde, E. Bequet
CHR Verviers, U Liège, CHR de la Citadelle, CHU de Liège
- P 95. Mind the liver when Coombs comes positive in toddler.**
F. Henssen, A. Bobarnac, S. Colinet, N. Blétard, P. Philippet
U Liège, CHC Liège
- P 96. Acute right iliac fossa pain, about a case**
E. Hoornaert, C. Koutny, G. Van De Poel
Clinique Notre Dame de Grace Gosselies
- P 97. Hydrogen and methane breath test in the diagnosis of lactose intolerance.**
C. De Geyter, K. Van De Maele, B. Hauser, Y. Vandenplas
UZ Brussel, VUB Brussel
- P 98. First successful use of ustekinumab in an infant with very early onset inflammatory bowel disease: outcomes at 18 months follow-up**
C. De Geyter, B. Hauser, K. Huysentruyt, E. De Greef, G. Veereman, Y. Vandenplas
UZ Brussel, The Hospital for Sick Children, University of Toronto
- P 99. The search for information in media (websites and apps) by children with coeliac disease and their parents. A pilot study**
E.I. Levy, T. Devreker, E. De Greef, Y. Vandenplas
UZ Brussel
- P 100. The inter-rater reliability of the Cow's Milk-related Symptom Score (CoMiSS) between parents and a trained healthcare professional**
Y. Jaddioui, E. Carvajal, C. Ribes-Koninckx, Y. Vandenplas
UZ Brussel, LA FE Hospital
- P 101. Novel DGAT1 gene mutation in infant with protein-losing enteropathy (PLE)**
L. Bijker, S. Uyttebroeck, K. Keymolen, B. Hauser, F. Hes, Y. Vandenplas
UZ Brussel
- P 102. Diagnosis of vitamin B12 deficiency through newborn screening**
A. Empain, L. Marcélis, C. De Laet
ULB - HUDERF
- P 103. Nasobiliary drainage prior to surgical biliary diversion in patients with progressive familial intrahepatic cholestasis type II**
G. Jannone, X. Stephenne, I. Scheers, F. Smets, E. M Sokal
UCL Saint-Luc
- P 104. Is shit in real life the same as on photo?**
B. Aman, Y. Vandenplas, K. Huysentruyt
UZ Brussel, The Hospital for Sick Children, University of Toronto

ONCOLOGY – HEMATOLOGY

Long Oral Presentation

- LO 1. Interdisciplinary rehabilitation after cancer in children and adolescents**
 R. Van Der Looven, L. Hermans, K. Meylemans, H. Capiiau, K. Vandekerckhove, I. Coomans, S. Meerhaeghe, H. Morren, C. Dhooge, G. Laureys
UZ Gent

Short Oral Presentation

- SO 14. Is the preoperative preparation of Sickle Cell Disease patients optimal: Assessment of practices and post-operative complications.**
 A. Therer, A. Ferster, P. Van Der Linden, S. Wambacq
Hôpital des Enfants de la Reine Fabiola, Université Libre de Bruxelles, CHU-Brugmann
- SO 19. Physical functioning in children surviving a brain tumour**
 M. Gielis, V. Dirix, E. Vanderhenst, A. Uyttebroeck, C. Sleurs, H. Feys, S. Jacobs
UZ Leuven

Poster Walk

- PW 4. The risk of rebound hypercalcaemia after denosumab treatment in paediatric oncology**
 Y. Eelen, N. Knops, E. Levtschenko, C. Politis, M. Renard, V. Labarque, A. Uyttebroeck, H. Segers
UZ Leuven
- PW 6. Neonatal screening program for sickle cell disease : comparison of patients born 2000-2005 and 2010-2015, single center experience**
 C. Peresse, A. Ferster
CHU Charleroi, ULB - HUDERF

Posters

- P 105. A limping hiding a metastatic neuroblastoma**
 R. Kinuani, B. Florquin, M-F Dresse, K. Nyamugabo, M-C Seghaye
CHU de Liège, CHR de la Citadelle, Notre Dame de Bruyères
- P 106. A case report of Ileal Burkitt Lymphoma revealed by an intestinal invagination in a young child**
 C. Charlier, B-A David, M-C Seghaye, M-F Dresse
ULiège, University Hospital Liège
- P 107. Case report : a painful pelvic mass in a fourteen years old girl.**
 G. Conti, C. Devalck, C. Versteegh, A. Klein, F. Praet
CHIREC, HUDERF-ULB
- P 108. A rare case of concomitant Hodgkin Lymphoma and Langerhans cell hyperplasia in an 8-year-old child**
 M. Lahr, M-F. Dehou, K. Wilgenhof, L. Rozen, A. Demulder, V. Schlessler, J-H François, C. Devalck, S. Huybrechts
Centre Hospitalier de Luxembourg, Université de Liège, CHU Brugmann, Laboratoire CMP, UZ Brussels, LHUB-ULB, HUDERF-ULB

- P 109. Favism – A yellow boy and his beans**
Z. Casier, S. Van Molhem, A. Verschelde, K. Willième
AZ Damiaan, AZ Sint-Jan
- P 110. Neonatal leukemia : Report of a case with Blueberry Muffin Syndrome**
J. Baudoin, S. Gatineau, M-C Seghaye, M-F Dresse
Uliège, University Hospital Liège
- P 111. Pulmonary embolism, an unusual presentation of Fanconi anemia**
P. François, P. Philippet, S. Schiffers, L. Rouffiange, S. Eiras da Silva, M. Thimmesch , C. F. Chantrain
CHC Liège
- P 112. Infantile hemangioma : watch out for the hidden part of the iceberg...**
A. Fohn, S. Cao, M-C Seghaye, J. Longton
CHU de Liège, U Liège, CHR de la Citadelle
- P 113. A successful management of generalized kaposiform lymphangiomatosis manifested by chylothorax in an 11-month-old boy: a case report**
F. Lebrun, P. Schlessler, J. Khamis, P. Philippet
CHC Liège-Belgium, University of Liège
- P 114. Severe phototoxicity associated with concomitant use of methotrexate and voriconazole, an overlooked drug-drug interaction**
D.J. Bogaert, L. Verlinden, E. Vandecruys, G. Laureys, E. Verhaeghe, T. Bauters
UZ Gent
- P 115. Atypical presentation of a precursor B-cell Lymphoblastic Lymphoma**
T. Alliet, P. Van der Speeten, Y. Eelen, L. De Somer, S. Pans, M. Renard, H. Segers
UZ Leuven
- P 116. Recurrent cholesteatoma in a child with Fanconi anemia**
M. Mathieu, S. Gobert, T. Robillard, M. Melchior, S. Schiffers, P. Philippet, C.F. Chantrain
CHC Liège, U Liège, CHU-UCL Namur

NEUROLOGY – GENETICS – NEUROORTHOPEDICS

Long Oral Presentation

- LO 5. Clinical and biochemical phenotype of 11 patients with bi-allelic mutations in TANGO2**
P. Verloo, E. Jennions, A. Vanlander, L. De Meirleir, S. Seneca, N. Darin
UZ Gent, Queen Silvias Child Hospital, UZ Brussel
- LO 6. New FDG-PET analysis confirm previous evidence for the epileptic network of Lennox-Gastaut Syndrome**
T. Balfroid, A. Warren, L. Dalic, C. Rowe, J. Archer
University of Melbourne, Melbourne, Australia, Austin Health Hospital, Melbourne, Australia, ULB - HUDERF, Florey Institute, Melbourne, Australia,
- LO 7. First epileptic EEG activity in infants with Tuberous Sclerosis Complex is associated with neurodevelopmental outcome at the age of 2 years**
J. De Ridder, B. Verhelle, J. Vervisch, K. Lemmens, K. Kotulska, R. Moavero, P. Curatolo, B. Weschke, K. Riney, M. Feucht, P. Krsek, R. Nabbout, A.C. Jansen, D. Domanska-Pakiela, M. Kaczorowska-Frontczak, C. Hertzberg, H.M. Hulshof, S. Samuelli, B. Benova, E. Aronica, D.J. Kwiatkowski, F.E. Jansen, S. Jó wiak, L. Lagae

Short Oral Presentation

- SO 1. Executive function assessment in 2-year-olds born preterm.**
A. Van den Brande
UZ Leuven

Poster Walk

- PW 12. Pediatric neuroborreliosis**
J. Vanbekbergen
UZ Leuven
- PW 14. A first case of acute flaccid myelitis associated with enterovirus D68 in Belgium**
I. Delpire, M. Rodesch , F. Vermeulen , F. Christiaens
ULB - ERASME
- PW 36. Interest of Positron Emission Tomography in small vessel primary angiitis of the central nervous system**
M. Belcour, P. Dontaine, A. Monier, L. Lebrun, I. Salmon, A. Van Hecke, C. Fricx, O. De Witte, G. Boitsios, S. Goldman, X. De Tiège, A. Aeby
ULB - HUDERF, ULB - ERASME

Posters

- P 117. A rare presentation of congenital spinal dermal sinus**
D. Blommaert, W. van Herk, S.A. de Man, S. Hammer, J. K.H. Spoor, T.R.H. de Jong
Amphia Ziekenhuis Breda, Erasmus Rotterdam
- P 118. Obstructive hydrocephalus leading to the diagnosis of diencephalic syndrome**
M.A Tenabene, Z. Lamtiri, L. Vanden Brande, M-F Dresse, P. Leroy, M-C Seghaye
U Liège, CHU de Liège

- P 119. Sylvian stroke in a 12-year-old patient secondary to varicella arteriopathy**
Y. Lounis, C. Jacquemart, N. Cajgfinger, S. Alkan, S. Vaessen, P. Leroy, M-C Seghayé
CHU de Liège, U Liège
- P 120. A case report of dominant COL4A2 mutation leading to progressive cerebellar atrophy, porencephaly and leukoencephalopathy.**
E. Everard, L. Fabri, E. Gueulette, D. Beckers, A-S Marchand, D. Lederer, M. Mathot
CHU-UCL Namur
- P 121. Life as it is, from a different angle: Congenital laterocollis caused by trochlear nerve paresis.**
A. Wynsberghe L. Vallaëys
UZ Leuven, AZ Groeninge
- P 122. Startling cause of abdominal pain in an 11 year-old child**
V. Nguyen, N. Marcu
University of Liège, Regional Hospital Val de Sambre
- P 123. Sudden speech impairment: the case of a child with arterial ischemic stroke associated with focal cerebral arteritis**
C. Ooms, N. Demonceau, P. Philippet
CHC MontLegia, KUL
- P 124. An unexplained cause of status epilepticus associated with anti-TPO antibodies: could it be an Hashimoto's encephalopathy ?**
K. Tazi, X. Beretta-Picolli, S. Rooze, A. Aeby, D. Vens, A. Vicinanza, V. Masy, C. Voglet, D. Biarent
ULB - HUDERF
- P 125. A recurrence of Giant Bilateral Inguinal Hernias in an Infant with Pallister-Killian Syndrome (PKS).**
M.K.F. Docx, S. Heyman, M. Meuwissen
Koningin Paola Kinderziekenhuis Antwerp, UZ Antwerpen, Universiteit Antwerpen
- P 126. A case of GLUT1 deficiency syndrome – benefits of genetic testing in childhood epilepsy**
S. Neuens, A. Van Hecke, J. Soblet, C. Vilain
HUDERF-ULB
- P 127. Gut and liver dysfunction: a surprising diagnostic clue for metabolic disease**
S. Dejonckheere, P. Verloo, R. De Bruyne
UZ Gent

CARDIOLOGY

Short Oral Presentation

- SO 2. Recovery kinetics of gas exchange parameters and heart rate after maximal exercise in children with repaired Tetralogy of Fallot compared to controls**
I. Coomans, S. De Kinder, H. Van Belleghem, D. De Wolf, K. De Groote, J. Panzer, L. Muino Mosquera, H. De Wilde, T. Bové, K. François, J. Boone, K. Vandekerckhove
UZ Gent
- SO 7. The long term fate of subaortic stenosis in childhood**
R. De Wolf, K. François, T. Bové, I. Coomans, K. De Groote, H. De Wilde, J. Panzer, K. Vandekerckhove, D. De Wolf
VUB, UZ Gent
- SO 8. Oxygen uptake kinetics and local muscle oxygenation during submaximal exercise in children after the Fontan procedure compared to healthy peers**
Y. Bagdasarian, I. Coomans, D. De Wolf, K. De Groote, J. Panzer, L. Muino, H. De Wilde, T. Bové, K. François, J. Boone, K. Vandekerckhove
UZ Gent
- SO 29. Endocarditis prophylaxis in the “real life” of the general pediatrician and/or dentist**
D. De Wolf, A. Genouw, C. Standaert, A. Victor, N. Vanoverbeke, K. De Groote, L. Martens
UZ Gent, UZ Brussel

Poster Walk

- PW 16. Melody valve in mitral position in very young children with atrio-ventricular septum defect and severe mitral valve dysfunction**
J. Hubrechts, B. Cools, R. Heying, B. Eyskens, R. Rega, B. Meyns, M. Gewillig
UZ Leuven
- PW 17. Direct paratracheal lymphosclerosis for plastic bronchitis after Fontan: percutaneous versus endoscopic transtracheal technique**
J. Hubrechts, B. Eyskens, C. Dooms, G. Maleux, B. Cools, R. Heying, D. Boshoff, M. Gewillig
UZ Leuven
- PW 37. Outcome of strategy in Pulmonary Atresia, ventricular Septal Defect and Major Aortopulmonary Collateral Arteries (PA, VSD, MAPCA's)**
B. Eyskens, B. Cools, R. Heying, J. Hubrechts, D. Boshoff, F. Rega, B. Meyns, M. Gewillig
UZ Leuven

Posters

- P 129. Unilateral lung whiteout during neonatal period, about a case**
E. Hoornaert, J-L Hennecker, Z. Van Lier
Clinique Notre Dame de Grace Gosselies
- P 130. Kingella Kingae endocarditis with vegetation and mitral valve perforation in a 7 month old infant.**
S. Verbeeck, S. Van Nuijs, F. Slap, B. Suys
GZA Sint Augustinus
- P 131. How to detect incomplete forms of Kawasaki disease?**
C. Creutz, C. Jacquemart, M-C Seghaye
University of Liege, University Hospital Liege

- P 132. Long QT interval in patients with Turner syndrome**
J. Dauby, A-S Crochelet, M-C Lebrethon, J. Harvengt, S. Bulk, M-C Seghaye
U Liège
- P 133. Convulsive seizures in LQTS : think about malignant tachyarrhythmias**
J. Levieux, I. Astadicko, C. Jacquemart, S. Bulk, L. Van Casteren, M.-C. Seghaye
U Liège
- P 134. When respiratory symptoms hide a double aortic arch**
S. Finocchiaro, N. Farhat, M-C. Seghaye
University of Liège, University Hospital of Liège
- P 135. Hypertrophic cardiomyopathy : A Breathtaking disease**
J. Uwayezu, J. Van Huffel, N. Farhat, MC. Seghaye
University of Liège, University Hospital of Liège
- P 136. Establishment of a predictive model of VO₂ in patients with total cavo-pulmonary anastomosis (FONTAN)**
C. Rojas, M. Roggen, C. Vô, L. Vanhoutte, J. Hubrecht, K. Carbonez, C. Barrea, T. Sluysmans, G. de Beco, A. Poncelet, S. Moniotte
Cliniques Universitaires Saint Luc, UCL

NEPHROLOGY

Long Oral Presentation

- LO 11. Dietary fiber intake and gut-derived uraemic toxins in a Belgian paediatric CKD cohort**
A. El Amouri, E. Snauwaert, A. Desloovere, A. Foulon, C. Vande Moortel, G. Glorieux, W. Van Biesen, J. Vande Walle, S. Eloot, A. Raes
UZ Gent
- LO 12. The impact of chronic kidney disease on the quality of life and psychosocial functioning of children and their family**
E. De Bruyne, S. Leroi, S. Vankerckhove, P. Dewinter, S. Reynaert, C. Boever, J. Van de Walle, A. Raes, L. Collard, B. Adams, D. Trouet, K. Van Hoeck, N. Godefroid, N. Knops, D. Mekahli, E. Levtchenko
UZ Leuven

Short Oral Presentation

- SO 3. Kinetic modeling as guide for dialysis prescription in acute neonatal hyperammonaemia: an example using CarpeDiem and Fresenius 4008 machine**
J. De Rudder, P. Verloo, A. Prytula, L. Dossche, J. Dehoorne, J. Vande Walle, E. Dhont, A. Raes, W. Van Biesen, S. Eloot
UZ Gent
- SO 4. Perinatal determinants of renal function and blood pressure in former extremely low birthweight infants in late childhood**
M. Colleman, E. Levtchenko, Z. Zhang, J.A. Staessen, K. Allegaert, A. Raaijmakers
KU Leuven, Research Institute Alliance for the Promotion of Preventive Medicine, Erasmus MC, ZNA hospitals
- SO 13. FCGG Renal Biopsy Network: first epidemiological report on pediatric renal diseases**
S. Karamaria, J. De Meester, A. Dendooven, E. Levtchenko, N. Knops, K. Van Hoeck, D. Trouet, R. Mael, B. Sprangers, W. Laurens, M. Couttenye, J. Vande Walle
UZ Gent, NBVN, UZ Leuven, UZ Antwerpen, UZ Brussel, UZ Leuven, AZ Nikolaas
- SO 15. Genotype – phenotype correlation in a pediatric autosomal dominant polycystic kidney disease (ADPKD) cohort**
D. Van Giel, S. De Rechter, L. Breyssem, A. Hindryckx, P. Janssens, J-P Decuypere, B. Bammens, A. Corveleyn, C. Férec, R. Vennekens, M.P. Audrezet, P. Harris, D. Mekahli
KUL, UZ Leuven, Institut National de la Santé et de la Recherche Médicale, VIB Center for Brain and Disease Research, Mayo Clinic College of Medicine
- SO 17. Comparative analysis from 2005 to 2020 of outcome parameters in Belgian children with a kidney allograft**
B. Adams, L. Collard, N. Godefroid, K. Van Hoeck, E. Levtchenko
UZ Leuven
- SO 20. ADPedKD: A global online platform to explore the childhood phenotype of Autosomal Dominant Polycystic Kidney Disease**
A. Dachy, S. De Rechter, L.M. Guay-Woodford, A.J. Mallet, T. Harris, D. Bockenbauer, F. Schaefer, M.C. Liebau, D. Mekahli
KU Leuven, UZ Leuven, Children's National Health System, Washington, Royal Brisbane and Women's Hospital, The University of Queensland, Brisbane, The KidGen Collaborative et Australian Genomics Health Alliance, PKD International, PKD Charity, UCL, Great Ormond Street Hospital NHS Foundation Trust, Heidelberg University Medical Centre, University Hospital of Cologne

SO 21. Dietary fibre is associated with serum levels of uraemic toxins in children with chronic kidney disease
 A. El Amouri, E. Snauwaert, A. Foulon, C. Vande Moortel, M. Van Dyck, K. Van Hoeck, N. Godefroid, G. Glorieux, W. Van Biesen, J. Vande Walle, A. Raes, S. Eloit
UZ Gent, UZ Leuven, UZ Antwerp, Cliniques Universitaires St. Luc, Université Catholique Louvain

SO 30. Reaffirmation of empiric antimicrobial treatment of febrile urinary tract infections in infants and children in Tournai-Belgium.
 J. Malvaux, E. Hennaut, J-P Stalens
CHwapi Tournai

Poster Walk

PW 2. The choice between deceased and living donor renal transplantation in children: a multicentric cross-sectional study
 L. Dierickx, J. Vande Walle, A. Prytula, A. Raes, N. Knops, L. Willem, E. Levtchenko, K. Van Cauwenbergh
Universiteit Gent, Universiteit Antwerpen, KUL

PW 15. Effects of fecal microbiota transplantation for recurrent clostridium difficile in children with renal replacement therapy
 P. D'hondt, N. Knops, C. Caenepeel, J. Francisco Vázquez Castellanos
UZ Leuven

PW 20. Effects of faecal microbiota transplantation for recurrent clostridium difficile infection in children with chronic kidney disease
 A. Samaey, J.F. Vázquez Castellanos, C. Caenepeel, P. Evenepoel, S. Vermeire, N. Knops
UZ Leuven, KU Leuven

PW 24. The effect of a multidisciplinary weight loss program on renal circadian rhythm in obese adolescents
 S. Dejonckheere, K. Pauwaert, E. Bruneel, J. Van Der Jeugt, L. Keersmaekers, S. Roggeman, K. Everaert, J. Vande Walle, A. De Guchtanaere
UZ Gent, Zeepreventorium

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UZ Gent, Wilhelmina Children's Hospital, University MC Utrecht, Emma Children's Hospital, Amsterdam University MC, Erasmus Medical Center- Sophia Children's Hospital, UZ Leuven

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KU Leuven, UZ Leuven, CHU

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 L. Sandra, E. Degraeuwe, E. Gasthuys, A. Vermeulen, P. De Bruyne, A. Raes, J. Vande Walle
Universiteit Gent, UZ Gent

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Vivalia, Centre Hospitalier de l'Ardenne (Libramont), U Liège
- P 139. Hyperkalemia in a patient with nephrotic syndrome**
C. De Cordt, N. Segers
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- P 140. Urinary tract infection, even though urinalysis and culture are negative: a case report of a 5-year-old girl.**
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- P 141. Post-transplant focal segmental glomerulosclerosis recurrences associated with bowel obstruction: a case report.**
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- P 143. Diagnostic challenges in a boy with tubulointerstitial nephritis and uveitis (TINU) syndrome: A case report and review of literature.**
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- P 145. Gastrointestinal protein loss in a child with polycystic kidney disease and SARS-CoV2 infection**
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G. Pauwels, T. Claeys, E. Vanlaecke, A. Raes, A. Prytula, J. Vande Walle
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- P 153. Acceptance & Commitment Therapy with parents of children with chronic kidney disease**
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S. Karamaria, N. Ranguelov, P. Hansen, V. De Boe, P. Verleyen, J. Vande Walle, L. Dossche, A. Bael
UZ Gent, Universiteit Gent, Cliniques Universitaires St-Luc, KUL, CHU Tivoli, UZ Brussel, AZ Groeninge, ZNA Koningin Paola Kinderziekenhuis, Universiteit Antwerpen
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Y. Dejonckheere, N. Knops
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C. Gernay, C. Brachet, E. Boros, C. Libiouille, S. Tenoutasse, C. Heinrichs
Hôpital Universitaire des Enfants Reine Fabiola, Université Libre de Bruxelles, Centre Hospitalier
Universitaire du Sart-Tilman, Université de Liège
- LO 4. Partial CRISPR/Cas9 IL1R1 & IFNGR1 Knock-Down Improves γ -cell Survival And Function Under Cytokine-Induced Inflammation**
C. Daems, J. Vanderroost, E. Sokal, P. Lysy
UCL Saint-Luc

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K. Van De Maele, A. Bogaerts, J. De Schepper, S. Probyn, D. Ceulemans, I. Guelinckx, I. Gies, R. Devlieger
UZ Brussel - VUB, UZ Leuven, Universiteit Antwerpen, Danone Research
- SO 22. Long-term endocrine and reproductive outcome of adolescent and young adult men born appropriate for gestational age with non-syndromic hypospadias**
L. Tack, A-F Spinoit, S. Riedl, A. Springer, U. Tonnhofer, M. Hiess, J. Weninger, A. Mahmoud, K. Tilleman, E. Van Laecke, P. Hoebeke, A. Juul, J.C. Albrethsen, E. De Baere, H. Verdin, M. Cools
Universiteit Gent, UZ Gent, Medical University of Vienna, St Anna Children's Hospital, Medical University of Vienna, University of Copenhagen, Rigshospitalet
- SO 23. Eating habits of children born after maternal bariatric surgery**
K. Van De Maele, C. De Geyter, Y. Vandenplas, R. Devlieger, I. Gies
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UZ Gent, Universiteit Gent, Erasmus MC Rotterdam, Hadassah Hebrew University Medical Center

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- PW 29. Long-term endocrine and reproductive outcome of adolescent and young adult men born small for gestational age with non-syndromic hypospadias**
L. Tack, A-F Spinoit, S. Riedl, A. Springer, U. Tonnhofer, M. Hiess, J. Weninger, A. Mahmoud, K. Tilleman, E. Van Laecke, P. Hoebeke, A. Juul, J.C. Albrethsen, E. De Baere, H. Verdin, M. Cools
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- PW 30. Diagnostic dilemma of a suppressed serum TSH in a female teenager with an asymptomatic goiter.**
S. Ryckx, B. Mahieu, M. Piqueur, F. Van Acker, D. Klink, J. De Schepper
ZNA Koningin Paola Kinderziekenhuis Antwerpen, ZNA Middelheim, UZ Brussel

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A. Rochtus, J. Vinckx, F. de Zegher
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S. Touzani, D. Avino, T. Cos Sanchez, E. Boros
ULB - HUDERF, CHU Brugmann
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E. Nauwynck, K. Van De Maele, J. Vanbesien, J. De Schepper, I. Gies
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- P 161. Normosmic Congenital Hypogonadotropic Hypogonadism (nCHH) due to GNRH1 mutation: a rare etiology**
C. Gernay, E. Boross, C. Vilain, H. Steyaert, C. Heinrichs, C. Brachet
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Q. Jordens, K. Casteels, F. de Zegher
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- P 163. Impaired Hypoglycemia Awareness in children and adolescents with type 1 diabetes**
A. Messaaoui, S. Tenoutasse, L. Hajselova, L. Crenier
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S. Daelemans, A. Helsmoortel, C. Donner, P. Pelc, C. Heinrichs, I. Vandernoot, J. Desir, A-L Mansbach, P. Deltendre, C. Brachet
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O. Pollé, P. Lysy, A. Lourtie, P. Bernard
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K. Van De Maele, D. Klink, J. De Schepper
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- P 169. Renal Pseudohypoaldosteronism type I in a 6-day-old neonate**
F. Chalon, S. Balbeur, B. Brasseur
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- P 170. Rare cause of Cushing syndrome in a 12-year old girl with striae and weight gain**
A. Jacobs, K. Casteels, A. Rochtus, H. Segers, D. Hompes, F. de Zegher, M. Bex
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- P 171. Mediastinal immature teratoma leading to a supraphysiological serum testosterone concentration in an adolescent with Klinefelter syndrome**
A. Boret, V. Bordon, E. Vandecruys, J. De Schepper
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- P 172. Are low-grade inflammation or testosterone aromatization in adolescent males with overweight or obesity associated with pubertal timing?**
E. Berlangier, I. Gies, M. Van Helvoirt, A. De Guchteneere, E. Anckaert, E. Bassle, J. De Schepper
UZ Brussel, Zeepreventorium
- P 173. Algorithmic adaptation of insulin therapy and carbohydrate consumption during exercise in children with type 1 diabetes: results of the CAR2DIAB study**
E. Gasser, H. Boughaleb, T. Barrea, G. de Valensart, A. Robert, P. Lysy
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OTHER

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T. Alliet, T. Gestels, S. Daelemans, L. Hoste
UZ Leuven, UZ Antwerpen, UZ Brussels, Universiteit Gent

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PW 8. The virulence of a highly necrotizing clinical strain of Streptococcus pyogenes

G. Deneubourg, D. Lakhoulfi, C. Neerdaels, V. Imbault, V. Delforge, M. Davies, D. Communi, P. Smeesters, A. Botteaux
VUB, Doherty Institute, University of Melbourne, Australia

PW 28. The European paediatric clinical trials network conect4children: activities of the Belgium National Network and its progression

E. Degraeuwe, L. De Taeye, M. Turner, L. Nuytinck, D. Christiaens, D. Vanderschaeghe, E. Gasthuys, S. Karamaria, C. Vanderstraeten, J. Vande Walle
Universiteit Gent, UZ Gent, University of Liverpool

PW 31. Eczema herpeticum, a dermatological emergency

J-C Beghin, S. Pohlen
ULB - HUDERF

PW 32. Some trainees are more equal than others - the pediatric residency pay gap

L. Hoste, S. Daelemans, A. De Guchtanaere, A. Bael
Jong VVK, UZ Gent, UZ Brussel, VVK, Zeepreventorium, ZNA Koningin Paola Kinderziekenhuis

PW 39. Belgian CAP48 cohort of juvenile idiopathic arthritis (JIA): What is the role of the emergency department in the early management of JIA?

F. Faron, P-Q Lê, L. Goffin, A. Ferster, V. De Maertelaer, T. Sokolova, V. Badot
Centre Hospitalier Epicura, ULB - HUDERF, CHU Brugmann

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S. Sommen, E. Demandt, K. Coppens
UZ Leuven, Imeldaziekenhuis Bonheiden

P 175. Recurrent vulvar ulcers in young girls

O. Van Reeth, V. Bernier, F. Piérart, P. Philippet
CHC Montlegia, Université de Liège

P 176. A rare case of subcutaneous emphysema after video-assisted thoracoscopic surgery for pleura empyema

S.A. Kluijfhout, M. Koninckx, D. Vervloessem, S. Heyman
UZ Brussel, ZNA Koningin Paola Kinderziekenhuis

P 177. Linear IgA bullous dermatosis

M-C Rousseau, M-C Seghaye, A. Chamouine, A. Benureau
CHM Mayotte, CHU Liège

P 178. Time to diagnosis : about a case of juvenile dermatomyositis

F. Faron, L. Goffin, P-Q Lê, V. Badot, M. Maka-Abomake
Centre Hospitalier Epicura, ULB - HUDERF, CHU Brugmann

- P 179. Infantile hemangioma : Don't miss the PHACE syndrome**
S. Gobert, J. Pêtre, J. Khamis, C. Christiaens, M-C Seghaye, V. Bernier, P. Philippet
CHC, U Liège
- P 180. Lipschütz ulcer: diagnosis and management. About an unusual case of recurrence and a rapidly favorable course with topical corticosteroid therapy.**
A. Du Mortier, V. Bernier, A. Delfosse, M. Creuven, J. Pêtre, P. Philippet
CHC Liège
- P 181. Macrodystrophia Lipomatosa: a little bit "toe" much.**
Q. Jordens, D. Van Gysel, H. De Maeseneer, H. Brems
OLV Ziekenhuis Aalst-Asse-Ninove, KUL
- P 182. Infantile transient smooth muscle contraction**
J. Vanbekbergen, L. De Somer
UZ Leuven
- P 183. Ankyloblépharon filiforme adnatum : a case report.**
A. Louagé, J. Debuyst, A-L Dussart, G. Malfilatre, J. Van Poucke, C. Djeunang, E. Strebelle, G. Kazemy, C. De Jong
CHU Tivoli La Louvière, ULB-HUDERF
- P 184. A pediatric case of aerodigestive thermal burn after ingestion of hot milk**
E. Surgun, V. Masy, X. Beretta-Picolli, S. Rooze, D. Vens, A. Vicinanza, K. Kotilea, C. Laurent, B. Devroede, D. Biarent
ULB - HUDERF
- P 185. The European paediatric clinical trials network beyond the Atlantic: the collaboration between I-ACT for children (US) and European network activity**
E. Degraeuwe, L. De Taeye, M. Turner, L. Nuytinck, D. Christiaens, E. Gasthuys, C. Hovinga, C. Stanley, J. Vande Walle
Universiteit Gent, UZ Gent, I-ACT for Children, University of Liverpool
- P 186. Linear IgA Bullous Dermatitis challenging recurrent bullous impetigo**
H. Van Parys, V. Bernier, J. Pêtre, A. Servais, P. Van Eeckout, C. Genin, P. Philippet
CHC Liège, CUSL Bruxelles
- P 187. Multiple cutaneous abscesses due to Pseudomonas aeruginosa**
C. Dupriez, V. Bernier, M. Creuven, P. Philippet
CHC Montlegia, CHC Liège, UCL
- P 188. Development of a rapid molecular diagnosis test for acute otitis media**
D. Laho, D. Lakhroufi, A. Botteaux, P.R. Smeesters
ULB - HUDERF

SO 24.**Obstructive sleep apnea syndrome and non-alcoholic fatty liver disease in an obese paediatric population.**

A-S Viskens, R. Basstanie, S-A Vandembroucke

Universiteit Antwerpen, UZA

Background

Paediatric obesity is highly prevalent and can be associated with non-alcoholic fatty liver disease (NAFLD) and obstructive sleep apnea syndrome (OSAS). A relation between those has already been constituted in children, but often with invasive methods such as liver biopsy that are unfeasible for use in daily clinical practice. This study is the first to investigate the relation between OSAS and NAFLD in a Belgian obese paediatric population using standard diagnostic tools, extended with the Fibroscan®, a non-invasive technique to quantify liver stiffness and steatosis.

Methods

Obese children aged 8-18 years were included at the paediatric obesity clinic of the Antwerp University Hospital between May 2017 and August 2020. Waist circumference and waist-hip ratio were measured. All patients received a polysomnography to diagnose OSAS. NAFLD was measured by liver transaminases determined on a fasting blood sample, by an abdominal ultrasound and a Fibroscan®. The latter expresses liver stiffness as a controlled attenuation parameter (CAP) and a median value.

Results

105 children were included at baseline with a mean age of 12 ± 2 years and an average BMI Z-score of $2,44 \pm 0,36$. OSAS was diagnosed in 19% of the children and liver steatosis determined by ultrasound in 42,9%. Children with OSAS had a higher median ALT value (29 U/l (16-68) vs. 25 U/l (12-108); $p < 0,046$), a lower mean AST/ALT ratio ($0,70 \pm 0,19$ vs $0,82 \pm 0,26$; $p < 0,040$) and a higher Fibroscan median value (6,50 kPa (3,70-13,30) vs 4,45 kPa (2,70-26,30); $p < 0,01$) compared to children without OSAS. A significant correlation was found between the HI and ALT value ($r = 0,223$; $p = 0,025$) and between the Fibroscan CAP value and the average saturation ($r = -0,277$ and $p = 0,045$). A binary logistic regression showed an association between the presence of liver steatosis on ultrasound and gender (OR=0,346; 95% CI=0,142-0,841; $p = 0,019$), ODI (OR=1,798; 95% CI=1,078-2,999; $p = 0,025$) and waist circumference (OR=1,061; 95% CI=1,017-1,108; $p = 0,006$).

Conclusion

This study confirms the relation between OSAS and NAFLD in an obese paediatric population. Correlation analysis shows that the degree of liver steatosis, measured by Fibroscan®, is correlated with the severity of OSAS, expressed as mean saturation. Further research is required to validate non-invasive measurement tools to diagnose and stage NAFLD and to investigate the influence of weight loss on the relationship between OSAS and NAFLD.

PW 22.**Halting the measles epidemic in Belgium: child's play?**

L. Cornelissen, V. Hutse, S. Leenen, R. Mahieu, G. Top, P. Carrillo, S. Quoilin

Sciensano

Background

In the (WHO) European Region 93,588 measles cases were registered in the first half of 2019. This is the highest number in over a decade, despite the pledge to eliminate measles by 2020. To reach elimination, WHO recommends a 95% vaccine coverage rate for two doses. Flanders reached this threshold for the first dose (MMR1) in 2008, Wallonia in 2015.

To assess the role of the pediatric population in the current outbreak in Belgium, we examined data on measles cases for the first six months of 2019 and used available coverage data for MMR1 to look at routine coverage in <20y olds.

Methods

Data from mandatory notification, Pediatric Surveillance network (PediSurv), sentinel labs and the National Reference Centre (NRC) were analyzed with SASv9.4 and Excel. Proportions were compared with Fisher Exact test. Regional coverage data for MMR1 and regional population data per birth year (from 2000 upwards) were used to calculate the proportion of individuals that had not received MMR1 before the age of 18-24 months. Coverage was extrapolated for the years in which no coverage surveys were conducted, assuming a linear trend stagnating after the last survey.

Results

In the first half of 2019, 357 cases were registered. Sixty-six cases (18%) needed hospitalization, with a higher rate in children <5y and adults ≥20y (58/274 = 21%) compared to patients between 5-19y (8/83, 10%, p=0.02). Incidence remains highest in infants <1y (39.4/100,000 person-years), but the majority of cases (n=190, 53%) occurred in the population ≥15y. Median age of cases was 19y (IQR 4-32). Vaccination status was unknown for 51% of all cases and 32% was unvaccinated.

An estimated 8% (167,454 persons) of the Belgian population 1-20y did not receive MMR1 at the recommended age.

Conclusions

With a median age of cases of 19y, measles is no longer a typical childhood disease. Current catch-up vaccination campaigns therefore focus mainly on adults aged 20-49 years, many of which remain unvaccinated. However, even in children and young adults aged 1-20y, almost 170,000 individuals did not receive MMR1 at the recommended age, despite high vaccination coverage in recent years. Every medical contact should thus be used to check their vaccination status and administer catch-up doses if necessary. This vaccination status should also be clearly documented, as more than half of all measles cases had undocumented vaccination status.

Poster Walk**PW 23.****Parental experience and emotional well-being in families with a child with a chronic disease during the COVID 19 pandemic**

E. Van Hoecke, K. Dhondt, A. Raes, S. Van Biervliet, B. De Moerloose, S. Van Aken, F. Haerynck, L. Hoste, H. Verhelst, B. Soenens, T. Vervoort, J. Vande Walle

UZ Gent

Background/Aims

The current COVID-19 pandemic and the associated quarantine measures have a major impact on the psychosocial well-being of our population. A particularly vulnerable group are parents of children with chronic diseases as they already face multiple challenges in caring their child. The aims of the current study were twofold; 1) to examine whether parents of children with chronic diseases experience worse well-being (i.e. anxiety and depression) compared to a control sample of parents of children without chronic diseases and 2) examine a series of risk factors for worse well-being within the clinical sample including pre-existing conditions, COVID-19 specific variables and psychosocial variables.

Methods

Participants consisted of 664 parents of children with a chronic disease and 493 parents of children without a chronic disease. Both groups were requested to report on experienced anxiety and depression (1-item PROMIS). A series of potential risk factors and additional outcomes were assessed in the group of parents of children with a chronic disease. Specifically, we assessed additional demographic data/pre-existing conditions (e.g., number of children, diagnosis,...); COVID-specific variables (living space, current health care experience and financial worries) and parenting experience during during COVID-19 (Parental Burnout Assessment (PBA)). The clinical group also filled out a prolonged measure of anxiety and depression (PROMIS -SF) and a measure of sleep quality (Insomina Severity Index).

Results

Findings demonstrated that parents in the clinical group reported higher levels of anxiety and lower positive parenting experiences compared to parents in the control group. Analyses within the clinical sample indicated that COVID-19- specific variables and parental burn-out contributed to higher levels of anxiety and depression and worse sleep quality. Mediation analyses furthermore indicated that the impact of COVID-specific variables upon all outcomes measures was mediated by higher parental burn-out and lower levels of positive parenting experiences.

Conclusion

Parents of children with a chronic disease are, in comparison to a control sample, a vulnerable sample for worse well-being during this COVID-19 pandemic. Findings suggest interventions directly targeting parental experience during the COVID-19 pandemic are warranted for this vulnerable sample.

PW 25.**The internet use of Belgian parents relating to a physician's visit for their children: a cross sectional survey.**

K. Berteloot, J. Toelen

KU Leuven, UZ Leuven

Objectives

The internet is an important source of information, also for medical purposes. As a result, parents are increasingly consulting the internet in relation to their child's health. This study examines the internet use of Belgian parents in relation to their visit to a physician. It investigates why and how parents search for information and how they value it.

Methods

An online questionnaire consisting of 16 questions was distributed via online media between May 11 to August 27, 2020. Only parents with one or more children (aged 16 years or younger) were asked to participate in the study.

Results

A total of 563 questionnaires was completed. Belgian parents frequently consult the internet: 67% of parents seek information before the medical visit, 42% after and 32,5% both before and afterwards. Young parents (<40 years) consult the internet significantly more before visiting the physician ($p = 0,01$). Parents with a higher education consult the internet significantly more after their visit to the paediatrician ($p = 0,04$). The main reason parents consult the internet before the medical visit is to determine whether they should consult a physician (71%). After their visit to the physician, they usually search for additional information about the diagnosis (76%). The online information corresponds to the physician's information in 81%, but only 50% of parents discuss the online information with their doctor. Parents have more confidence in their physician than in the internet ($p < 0,001$). In only 3% of cases the physician spontaneously recommended a reliable website to a parent.

Conclusion

Belgian parents frequently use the internet for medical purposes, generally consulting reliable sources. This study shows that only half of parents discuss the online information with their doctor and parents receive little guidance in their search for reliable and understandable medical information. Guiding parents in their search for reliable information and making it discussable can be an added value in modern health care.

Poster Walk**PW 26.****Urinary Incontinence in The Obese Population: Characteristics and Therapy-Outcome**

L. Dierickx, A. De Guchteneare, A. Raes, M. Smedts

*Universiteit Gent, Zeepreventorium*Introduction

Obese children with urinary incontinence are more likely to be resistant to therapy. The objective of the study was to evaluate the characteristics of lower urinary tract symptoms (LUTS) in obese children in a tertiary obesity clinic in Belgium and to investigate the response to urotherapy.

Methods

All patients admitted at a residential tertiary revalidation centre for morbidly obese children in Belgium between September 2015 and August 2018 were evaluated. Only obese children with symptoms of urinary incontinence answered a standard inquiry to evaluate their eating and drinking habits, voiding frequency, LUTS, severity of symptoms and stool pattern. All children started standard urotherapy and a strict drinking and micturition schedule. The success-rate was evaluated three months after achieving continence or at the end of the obesity program. Data were registered using computer software Statistical Package for the Social Sciences.

Results

Fifty-four (12.3%) children (mean 13 yr, range 7-19 yr) were diagnosed with urinary incontinence, 42 females (77.8%) and 12 males (22.2%). At admission all patients were morbidly obese. The majority of the patients had a decreased fluid intake (66.7%), a decreased (40,7%) or normal voiding frequency (42.4%). Almost half (48.1%) of the patients had urgency symptoms. Uroflowmetry showed a normal maximum voided volume in 68.5%. Nine patients (16.7%) had a large and only 1 patient (1.9%) had a decreased maximum voided volume. Remarkable thirty-two (59.3%) patients had an overdistended bladder (>115% EBC). Seventeen patients (28%) were excluded because of a lack of motivation for therapy and /or early departure. All other patients started urotherapy, followed by a strict drinking and micturition schedule. Eleven (29.7%) patients needed additional biofeedback training. All these patients achieved complete continence.

Conclusion

Morbidly obese children are more prone to urinary incontinence with a prevalence of 12.4%. The main characteristics of urinary incontinence in this population are an overdistended bladder, a decreased or normal voiding frequency which in half of the patients is accompanied with urgency symptoms. These findings suggest that obese children suffer more from an underactive bladder. Obese children with urinary incontinence are a challenge to treat not because of the complexity of their problem, but rather because of a lack of compliance influencing the success rate of therapy

PW 27.**Survey on first-aid knowledge in members of Flemish youth movements.**

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KU Leuven, UZ Leuven

Objectives

First-aid providers can play a vital role in life-threatening situations such as sudden illness or severe injuries. Every week almost 200.000 children attend a meeting of their youth movement and every year they participate in activities during the annual camp, which exposes them to possible situations that may require first aid. In the current survey we assessed the first aid knowledge of the leaders of the different youth movements (Scouts, Chiro, KSA...).

Methods

This study is an explorative cross-sectional study based on an online survey. Leaders of Flemish youth movements were questioned voluntarily and anonymously on their knowledge of first aid using 15 fictional scenarios (including: wound care, basic CPR, heat stroke, bee stings, splinter removals, venous and arterial bleeding,...).

Results

Of the 4405 respondents, 2784 completed the survey in full. The median score was 8/15 [IQR 7-10], 43% of the respondents answered more than half of the cases correctly. A higher age, studying or working in a healthcare field and having a first aid certificate resulted in a significantly higher score. The questions about treating a bee sting, removing a splinter and stopping an arterial bleed had the lowest correct scores.

Conclusion

The first aid knowledge of leaders in our youth movements could still be improved, especially regarding fairly common problems such as stings and splinter removals. Overall basic CPR is well understood, especially by those with first aid experience or training.

Posters**P 1.****Case report: Therapeutic challenges in an infant with trisomy 21, tracheomalacia and cystic fibrosis**

S. Liefoghe, M. de Lausnay, S. Verhulst

Universiteit Antwerpen

Trisomy 21 (T21) and cystic fibrosis (CF) are two relatively common genetic disorders. Nevertheless, the coexistence of genetically confirmed diagnoses of trisomy 21 (T21) and cystic fibrosis (CF) is rare in literature. We found seven cases documented. We would like to present an infant who suffered from complications of both diseases with important clinical consequences from a very young age. The prognosis of patients diagnosed with both these diseases is poor. A multidisciplinary approach is of significant importance for the treatment of these children

P 2.

Late Adolescents' Own and Assumed Parental Preferences Towards Health-Care Related Confidentiality and Consent

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Centre for Sociological Research, KUL, Spaarne Gasthuis, UZ Leuven

Objective

To investigate late adolescents' own and assumed parental preferences towards health-care related confidentiality and consent.

Methods

We analysed online survey data of four vignettes from 463 first-year university students at KU Leuven (Flanders, Belgium). We use paired samples t-tests to assess the (in)consistency between attitudes of late adolescents and their assumed parental attitudes, independent samples t-tests to estimate gender differences, and multinomial logistic regressions to analyse the association of assumed parental preferences with late adolescents' own preferences.

Results

Attitudinal inconsistencies are present in all vignettes. Late adolescents are significantly more in favour of confidentiality and non-parental consent than what they believe their parents are. Gender differences are small. Multinomial logistic regressions indicate that assumed parental preferences are strongly associated with late adolescents' own preferences.

Conclusions

Findings suggests a clear disjunction between late adolescents' preferences and assumed parental preferences: adolescents believe that their parents are less inclined to favour confidentiality and treatment without parental consent. We also find that this disjunction differs by case, indicating that there is no such thing as general 'confidentiality preferences'. Rather, a decision- and/or context-specific perspective should be adopted.

Posters**P 3.****Congenital external ear malformation : key feature to the diagnosis ?**

N. Laval, N. Revencu, Y. Sznajer

*Centre de génétique humaine, Cliniques Universitaires Saint Luc*Introduction

External ear malformation is a rare condition. EUROCAT 2018 update, prevalence is reported in 1,8 per 10 000 living birth and remains stable. Outer ear malformations can be described using Weerda gradation or using standard terminology of American Journal of Medical Genetics. It is reported to be part of a syndrome in 30 % (i.e. renal anomalies, heart defects ...). Ear abnormalities can sometimes be seen in utero due to improvement of 3D ultrasonography. It can be an orientation to the work-up because genetic cause is identified in 50%. Pediatricians in the delivery rooms or in maternity are in first line to evaluate for the diagnosis.

Discussion

We report the terminology as the recommendation for external ears presentation as distinctive anomalies and their related syndromes: those related to chromosomal anomalies (trisomies 13, 18 and microdeletion 22q11.2, namely) and those related to monogenic causes: CHARGE syndrome, mandibulo facial dysostosis (i. e. Treacher Collins or Guion Almeida type); auriculo condylar syndrome with 'question ear' mark; BOR syndrome and RASopathies. Key features and images are pinpoint to help the physician for the management. Presence of an external ear anomaly requires a complete clinical examination, having in mind these entities, and to also look for specific additional features. Several studies have shown excessive associated urinary tract malformations. Present guidelines recommend renal ultrasonography in patients with isolated preauricular pits, cup shape ear or any external ear malformation associated with dysmorphic features, history of deafness or maternal diabetes. It should be kept in mind that some presentations are part of a spectrum (no known chromosomal neither as gene mutation so far): external ear malformation with vertebral defect is present in oculo auriculovertebral association (Goldenhar). Finally, it is not recommended to perform full spine x-ray among children with isolated microtia. External ear malformations are also correlated to middle or inner ear defects. Auditory evoked potentials should be performed to evaluate presence of hearing loss.

Conclusion

Presence of external ear malformation is always worrisome for the parents and a challenge for the pediatrician. Complete clinical examination should be performed to every child with this congenital malformation as well as auditory evoked potentials. Renal ultrasonography should be discussed as well as genetic investigations.

P 4.

Survey about the alcohol consumption by minor in Flemish youth movements

M. Papy, J. Toelen

KUL, UZ Leuven

Objectives

Alcohol consumption by Belgian minors remains a major societal issue. It can have harmful consequences in various areas of somatic and mental health. Young people are a high-risk group, because they are in an age-period of intense somatic and psychological changes and developments. We surveyed the drinking behaviour of the 14 to 16-year-old Flemish youth. Especially when they are not under parental supervision, as is the case when they attend the regular meetings of the youth movement.

Methods

This study is an explorative cross-sectional study based on an online survey. Leaders of 14 to 16-year-olds within the Chiro and Scouts were questioned voluntarily and anonymously. The results were compared to data from the current literature.

Results

Of the 476 respondents, 84% (n=400) indicated that 14 to 16-year-olds have been consuming alcohol at least at one occasion at the youth movement. In the majority of the cases, this is done occasionally. They mainly drink beer (84.3% (n=401)), on average 2-3 glasses. However, 'binge-drinking' is also observed. The adolescents agree that there should be clear rules about alcohol consumption. Nevertheless, half of the respondents think that their youth movement should tolerate occasional consumption within this age group, since it is a good environment to learn to deal with alcohol.

Conclusion

The majority of the Flemish youth who attend a youth movement drinks alcohol before the age of 16 without parental supervision. They drink mostly 'occasionally', but sometimes, 'binge drinking' occurs as well. This research shows that the preventive measures of the youth movements for alcohol consumption are not yet fully utilized

Posters**P 5.****Vitamin D: Extremes are dangerous**

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*ULB - HUDERF, CHU Charleroi*Introduction

Vitamin D is a key hormone for bone mineral metabolism and calcium homeostasis. Excess or deficiency can lead to severe complications.

We report two cases of infants with severe vitamin D related diseases.

Cases reports

Case 1: A previously healthy 7-month-old boy was admitted for polyuria, poor appetite and failure to thrive. Vital signs and physical examination were unremarkable. Biochemistry analyses highlighted huge hypercalcemia (4,72 mmol/L; N: 2,1-2,55 mmol/L), high levels of 25-OH-vitamin D (584 mcg/L; N: 25-80 mcg/L) and 1-25-OH²-vitamin D (118 ng/L; N: 29-83,6 ng/L), hypoparathyroidism (< 10 ng/L; N: 14-72 ng/L) and hypercalciuria (urine calcium/creatinine ratio 10,8 mmol/mmol; N < 1,7 mmol/mmol). Renal ultrasound found nephrocalcinosis. Medical history revealed that the child received six drops a day of a vitamin D purchased on internet and containing 1000 UI per drop.

Case 2: A previously healthy 14-month-old boy was admitted in intensive care unit for seizure and QT interval prolongation on EKG. Biochemistry revealed deep hypocalcemia (1,10 mmol/L; N: 2,1-2,55 mmol/L), low levels of 25-OH-vitamin D (< 4,0 mcg/L; N: 25-80 mcg/L) and hyperparathyroidism (118 ng/L; N: 14-72 ng/L). Calciuria and renal ultrasound were normal. X-ray wrist of the left hand showed features of rickets. Patient history highlighted that he was exclusively breast-fed without any vitamin D supplementation.

Comments

Vitamin D has a key role in the regulation of phosphocalcic homeostasis and is essential for the maintenance of bone mineralization in children.

Severe deficiency leads to rickets but also to threatening clinical situations including seizures and/or cardiac arrhythmias. Conversely, intoxication leads to hypercalcemia and hypercalciuria with nephrocalcinosis, severe gastrointestinal (nausea, anorexia) and neurologic (muscle weakness, evolution from lethargy to coma) symptoms.

There is no clear consensus for vitamin D supplementation but according the American Academy of Pediatrics, the IOM and the ESPGHAN Committee on Nutrition, the recommended daily doses are at least 400 UI during the first year of life for all exclusively or partially breast-fed infants, and 600 UI from 1 to 18 years of age for a plasmatic target above 20 mcg/L.

Pediatricians should be cautious on the prescribed doses as threatening complications may occur in low and high vitamin D intake.

P 6.

Migrant pain, weight loss and inflammatory syndrome: did we worry too much?

M. Rodesch, L. Goffin, F. Vermeulen

ULB - Huderf, ULB - Erasme

We present here the case of an 8-year-old girl, complaining of a sharp and strong pain in a precise area of her inner left thigh, followed by pain of both thighs and belly with loss of 10% of body weight. The complementary examinations showed a bilateral femoral periostitis, ascites with hypermetabolic abdominal lymphadenopathies and a high inflammatory syndrome. Lymphoma with paraneoplastic periostitis, Crohn's disease or tuberculosis (TB) were the most likely diagnosis. We performed some invasive or less invasive tests to rule out those diagnosis: faecal calprotectin was negative, gastro- and colonoscopy and bone marrow puncture were both normal, tuberculin skin test and QuantiFERON were negative.

A non-granulomatous uveitis at the slit-lamp, and an HyperIgE (max 2536kU/l) syndrome were recognized. An exploratory laparoscopy with biopsies of the hypermetabolic lymphadenopathies revealed multiple adherences compatible with Fitz-Hugh-Curtis syndrome. No anatomopathological signs were observed except an unspecific inflammation, cultures and PCR remained negative. At that point, the patient had already gone through multiple invasive exams and the diagnosis was still unknown. Three approaches were proposed: Test treatment with anti-TB drugs for a hypothetic TB infection, test treatment with steroids for a hypothetic autoinflammatory disease or a watchful waiting attitude which was our final choice. After a few months of wait-and-see attitude and a treatment with diclofenac, she did not have any complain anymore, was gaining weight and the inflammatory syndrome dropped completely. The final diagnosis is unknown and the gene panel for autoinflammatory disease returned negative. The hypothesis of a chronic recurrent multifocal osteomyelitis (CRMO) in which the peritonitis and the uveitis may be explained as well as the migrant periostitis is still in our minds but does not really fit the clinical picture. The broad spectrum of autoinflammatory diseases that have not yet been discovered and therefore are not included yet in the gene panel might give us the final diagnostic in the future.

The patient will be closely followed-up but although she presented a stressful clinical picture, she recovered totally and remained healthy in the past 14 months with no specific treatment.

This case taught us that making a choice between numbers of complementary examinations might be difficult in those challenging cases and raise the question: did we worry too much?

Posters**P 7.****A rare cause of genital ulceration.**

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Etterbeek-Ixelles Hospital

Lipschütz ulcer (LU) or acute vulvar ulcer is a rare and unknown cause of genital ulcers specifically affecting adolescents and young women sexually inactive. Their onset is acute and preceded by a prodromal influenza-like phase, including fever, skin rash and odynophagia. The exact pathophysiology remains unknown but non-venereal infectious and idiopathic causes are highly suspected. The diagnosis of LU remains difficult and challenging after consideration and exclusion of other causes of genital ulceration such as venereal and non-venereal infections, autoimmune diseases, traumatic causes and vulvar tumors.

A twelve years old Moroccan girl was admitted to our pediatric emergency room for acute vulvodynia associated with fever, preceded by sore throat and macular skin rash few days prior the labial swelling and pain. The patient was sexually inactive, without any relevant medical history. Gynecologic examination showed an erythema and edema of the left labia majora with a painful ulcerative lesion of about 1cm of diameter. No extra-genital signs have been objectified. Complementary investigations including blood count, viral (EBV, CMV, HSV, HIV) and treponemal serologies, were negative except a moderate inflammatory syndrome without hyperleukocytosis (CRP: 43.45 mg/L) and a slightly high EBV IgM level at 25 U/mL. Furthermore additional tests for immunodeficiency or autoimmunity were also negative. During the hospitalization, a second small ulcerous lesion on the clitoris was appeared without any other complication. The patient was managed conservatively with analgesia (step I-II), and local barrier ointment. The lesions were fully healed 2 weeks later. LU was diagnosed at this time.

Although the etiology of LU is still unclear, infectious and idiopathic causes seem to be associated with their onset. Indeed, some studies considered its association with some pathogens such as EBV, CMV, influenza, paratyphoid fever, toxoplasmosis, M.pneumoniae, and mumps. Moreover, the availability of serological evaluation is limited and, therefore, it is common that some patients remained idiopathic. Nowadays there is no consensus yet about the diagnostic criteria for this disease. Their diagnosis is predominantly clinical, after excluding other causes of genital ulcer.

LU is a poorly understood illness and could be underdiagnosed. It should be evoked since this condition generates distress and concern, for virgin and young patients, for whom sexual abuse may be suspect

P 8.

Abdominal Pain and Acute Cutaneous Nerve Entrapment Syndrome: Go with the gut, but use your head.

S. Sommen, K. Coppens

UZ Leuven, Imeldaziekenhuis Bonheiden

Background

Abdominal pain in children is a frequent complaint at presentation to the Emergency Department. To rule out serious disease laboratory and radiological investigations are often carried out, but they do not always lead to a diagnosis.

When no cause is found, patients are often diagnosed with functional abdominal pain (FAP). Recent evidence suggests that up to 10% of FAP patients suffer from Acute Cutaneous Nerve Entrapment Syndrome (ACNES), an abdominal wall condition that can be easily diagnosed and treated.

Methods

We present the case of a 15-year-old patient who repeatedly presented at the emergency department with severe abdominal pain. Laboratory and radiological work-up did not yield any results. Careful history taking and physical examination revealed the clinical characteristics of ACNES. Subsequent treatment was initiated. Pain relief with local administration of lidocaine confirmed the diagnosis.

Result

A simple treatment regimen was highly successful in a child with severe abdominal pain and clinical characteristics of anterior cutaneous nerve entrapment syndrome.

Conclusion

ACNES, an abdominal wall pain syndrome, is not routinely considered as a cause of abdominal pain in children. This case report should increase awareness of paediatricians regarding the diagnosis of ACNES in children with abdominal pain.

Posters**P 9.****Increase in child sexual abuse following sanitary confinement : experience of the Maltraitance Unit of the Pediatric Service of CHU Liège**

S. Pannizzotto

*CHU Liège*Goals

Among the somatic and medical consequences of the pandemic linked to Covid19, the increase in the rate child abuse is one of the most serious and significant encountered in pediatrics. Sexual abuse was among those most encountered by the team Cellule Maltraitance after sanitary confinement in may 2020.

Methods

We want to describe and analyze here the situations handled by our team between May and November 2020 and compare them to the situations handled at the same time in 2019. When we resumed our activities in mid-May 2020 until this article was written in mid-November 2020, we admitted to the Maltraitance Unit 28 children including 12 boys and 16 girls (sex ratio 0.75) for medico-psycho assessment. -social is 8 more than the previous year over the same period, which corresponds to an increase of 40%..

Results

Sexual violence is clearly emerging as the most frequent form of abuse, particularly among girls (62.5%) and in the 3 to 6 year age group. We are also seeing a much higher percentage of this form of abuse compared to previous years. Of the perpetrators, 100% were close relatives of the child, having lived or living under the same roof at the time of the facts. The disclosure modes were, in 50% of the situations, the collection of the child's speech or the observation of hyper sexualized behaviors observed in school after sanitary confinement. The other disclosures of sexual abuse turned out to be a complaint lodged or a request for management by one of the parents following the disclosure of the abuse, and finally in one case, the diagnosis of gonorrhoea in an 8.5-year-old patient was allowed to guide the rest of the care. The isolation of families, physical and psychological promiscuity, the absence of a physical protective framework and psychic container (individual therapeutic work, school, psycho-social teams, etc.) have certainly contributed to these transitions or their intensification.

Conclusion

Child abuse is a severe, multimodal and multifactorial pathology. Especially we observed an increase in incidence of sexual abuse in the weeks following the spring confinement of 2020 and the isolation of families that the latter engendered.

P 10.

How to Maximize Children's Involvement in Non-therapeutic Research—Lessons Learnt From EFFECTOR

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UZ Brussel, KUL, VUB, UZ Leuven

Background

Children are vulnerable study subjects, especially in non-therapeutic research. Nowadays more attention is paid to the children's voice in both decision-making on participation and their experience of clinical research procedures.

Methods

We share our experiences from the EFFECTOR study. This is a long-term, cross-sectional, non-therapeutic follow-up study in the offspring (aged 4 to 11 years) of mothers who participated in scientific research during their pregnancy. The data was collected during a single study visit at home.

Results

During the data collection process, different strategies were developed to achieve a satisfactory participation rate with a focus on the involvement of the children. All study documents and measurements were assembled into a superhero framework. This theme is flexible and attracts children of a wide age-span. In order to inform the children before the study visit, a visually attractive assent was created as well as a superhero video (available through YouTube). During the study visit, a sticker diploma was used with similar visuals from the assent and all by the study doctor is framed in the superhero theme and tailored to the age of the children. The toddlers received a superhero-cape. The children were involved in the decision-making process during the whole process.

Conclusion

From our experience during the EFFECTOR data collection process, parents and their children can be motivated to participate in a long-term, non-therapeutic, follow-up study when child-friendly and adequate communication is used. Framing in a superhero theme is simple and suitable for children of a wide age-span.

Posters**P 11.****Physical activity in children with a chronic illness during the COVID-19 outbreak – the role of motor competence and motivation towards sport**

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Universiteit Gent, VUB, UZ Gent

In children, adequate motor competence is essential for the acquisition and maintenance of a physically active and healthy lifestyle. Clearly, children with a chronic illness are severely challenged in this respect, due to physical difficulties and psychosocial obstacles. Furthermore, it is known that these challenges are even greater in families with a low socio-economic status. The quarantine measures associated with the COVID-19 pandemic have led to an increase in online activities that potentially foster physical activity, however, we hypothesise that those may not have reached the vulnerable group of children with chronic diseases. Therefore, the aim of the current study was to investigate if the level of physical activity in the quarantine had changed in comparison with the previous year in a group of children with a chronic disease and in a healthy control group. Furthermore, we aimed to explore the role of motor competence and motivation towards sport in the potential change of physical activity.

During the Summer of 2020, seventy children with a chronic disease (31 girls, age range: 6-18), including 43 with renal failure, 15 with cystic fibrosis, 9 with diabetes and 3 with inflammatory bowel disease, completed the Flemish Physical Activity Questionnaire to measure the level of physical activity during the pandemic and at a similar time the year before. Motor competence was evaluated using a qualitative video assessment of core movement activities and motivation towards sports was assessed with a validated questionnaire. The same data were collected on a control group of 136 healthy control children (75 girls, age range: 6-18).

The preliminary results of this study (based on 31 patients, of which 18 with renal failure, 11 with cystic fibrosis and 2 with diabetes and 30 control children) show a considerable decrease in the total amount of physical activity during the pandemic relative to the previous year in both groups. In the patient group, the level of sedentary activity remained unaltered and high, but the amount of walking and cycling during transport increased. When controlled for age, children with a chronic illness demonstrate a lower level of motor competence and less autonomous motivation towards sport than the control group. These preliminary findings highlight the importance of stimulating and monitoring motor development and physical activity in children with chronic diseases, during as well as after the pandemic.

P 12.

The parental perception of pediatric disease symptoms: a cross sectional survey.

R. Knapen , J. Toelen

KUL, UZ Leuven

Objectives

The parental perception of pediatric disease symptoms is of great value to the decision process. Mainly parental concern appears to be important for this symptom perception. We investigated which symptom combinations particularly worry parents and if specific parent-related factors affect this concern.

Methods

We performed a cross-sectional study design using an online survey. In this we assessed the parent-related factors gender, age group, number of children and level of education. 20 symptom combinations were presented (4 cardinal symptoms 'cough, abdominal pain, fever and headache' each attached to 5 additional symptoms) and scored regarding degree of concern.

Results

We analysed 563 responses on the survey. The symptom combinations for every cardinal symptom perceived with the most concern were 'cough + wheezing', 'abdominal pain + blood in the stool', 'fever + skin rash' and 'headache + neck pain/neck stiffness'. The parent-related factors which affected parental concern were level of education (diploma of secondary education more concerned OR = 1,233; $p < 0,001$) and number of children (1 child more concerned, OR = 1,213; $p < 0,001$). There was no difference in parental concern for the dual division of age group (cut-off 40 years).

Conclusion

Parental concern about disease severity is affected by level of education and number of children. This insight is a first step towards better physician-parent communication and relevant parental recognition of severe pediatric pathology.

Posters**P 13.****Acute paediatric nicotine intoxication due to e-liquids: a novel and growing health risk in Belgium.**

J. Truyers, J. Toelen

*KUL, UZ Leuven*Objectives

Nicotine intoxication is a novel a rising phenomenon facing clinicians and poison control centres. The aim of this study is to provide the clinician with a practical tool to approach paediatric intoxications. We provide discuss this topic within the context of the Belgian legislation and situation.

Methods

For this study we collaborated with the Belgian poison control centre that provided us with all documented data. We supplemented this with data from a literature study. The focus of this review is the paediatric population.

Results

We found a parallel trend between the popularity of vaping and the number of suspected nicotine intoxications in Belgium. We analysed 110 contact with the Belgian poison control centre related to children with suspected nicotine intoxication involving e-liquids. Our main finding is that the majority of patients are asymptomatic (69,1%) and the most reported symptom is vomiting (8,2%). Almost all paediatric intoxications were accidental (99,1%). This is in line with published studies on paediatric intoxications.

Conclusion

Despite the high toxicity of nicotine, most patients are asymptomatic. Some preventive measures concerning e-liquids are already in place in Belgium. However, adjusting the packaging and informing the general public about possible dangers of e-liquids may offer additional protection.

LO 2.

Perinatal risk factors associated with the cardiovascular risk profile of Extremely Low Birth Weight survivors

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UZ Leuven, Sun Yat-Sen University, Cardiovascular Research Institute Maastricht (CARIM), Erasmus MC, ZNA Hospitals Antwerp

Background

Extremely Low Birth Weight survivors (ELBW, birth weight < 1000 grams) have a higher cardiovascular risk in later life. The objective of this study was to compare ELBW survivors to healthy controls and search for differences in microvascular structure, macrovascular environment and lipid concentrations at prepubescent age as biomarkers of such cardiovascular risk profile. We hereby aimed to evaluate whether three perinatal risk factors, retinopathy of prematurity (ROP), bronchopulmonary dysplasia (BPD) and intraventricular hemorrhage (IVH) are associated with these cardiovascular risk biomarkers.

Methods

This cohort analysis used the PREMATCH cohort, a group of 140 ELBW survivors treated in UZ Leuven, Belgium between 2000-2005. The 93 eligible index patients were age- and gender-matched to 87 healthy controls. These children underwent an extensive series of tests at 9-14 years of age to examine micro- and macrovascular function. Outcome parameters are blood pressure (SBP, DBP, MAP), pulse wave velocity (carotid-femoral PWV, carotid-radial PWV), perfused boundary region (PBR) and blood lipid concentration (total cholesterol, triglycerides, high-density lipoprotein, low-density lipoprotein) as markers for cardiovascular risk.

Results

ELBW children had significantly higher SBP, DBP and MAP compared to the control group (mean difference 7.04, 3.96 and 4.92 mmHg respectively, all $P < 0.001$). Unexpectedly, cf-PWV was on average 0.39 m/s lower in cases ($P = 0.005$). There was no significant difference in PBR between both groups ($P = 0.285$). Mean triglyceride concentration in ELBW children was 63.56 mg/dL compared to 53.31 mg/dL in healthy controls ($P = 0.003$). In ELBW cases, BPD was positively correlated with PBR ($r = 0.246$, $P = 0.048$). ELBW children with one or more perinatal comorbidities had significantly higher PBR (mean difference 0.21 μm , $P = 0.020$). Children with all three comorbidities had significantly higher total cholesterol (mean difference 20.82 mg/dL, $P = 0.043$).

Conclusion

At eleven years of age, ELBW survivors have significantly higher blood pressure and serum triglycerides compared to healthy controls. When faced with one of three perinatal comorbidities (ROP, BPD or IVH), they had a larger PBR, reflecting microcirculatory changes that correspond with endothelial dysfunction.

Short Oral Presentation

SO 5.

A single dose of budesonide/surfactant prevents hyperoxia induced lung injury in preterm rabbits

A. Gie, Y. Regin, T. Salaets, J. Vanoirbeek, J. Deprest, J. Toelen

*KUL, UZ Leuven*Objective

The role of postnatal steroids in the prevention of BPD is controversial because of non-pulmonary side effects, such as neurodevelopmental delay. Co-administration of budesonide with surfactant is an attractive strategy to maximize local delivery and minimize distant side effects. A repetitive dosing schedule of 0.25mg/kg budesonide in surfactant has been shown to prevent BPD. In this work we explore whether a single administration improves lung function, cytokine expression and lung structure in preterm rabbits exposed to hyperoxia.

Methods

Rabbit pups were delivered prematurely (day 28 of gestation) and exposed to hyperoxia (95% O₂). Pups were randomized to either a hyperoxia control group or to treatment with a single (on day 0) dose of 0.25mg/kg budesonide plus 100mg/kg surfactant with assessment of lung function, cytokine expression and lung structure on day 7.

Results

A single co-administration of surfactant with budesonide on day 0 improved lung function (normalization of lung damping, elastance, compliance and P/V loops), lung structure (Lmw) and CCL2 and IL8 expression at day 7, in comparison to untreated pups.

Conclusion

A single dose of budesonide plus surfactant had persistent effects on lungs of preterm rabbit pups exposed to hyperoxia. This indicates that a further dose reduction could be envisaged in clinical trials.

SO 6.

Mid-trimester premature prelabor rupture of membranes : neonatal evolution and long-term outcome

B. Van Grambezen, C. Hocq, F. Piersigilli, O. Danhaive

*UCL Saint-Luc*Background/Aims

Mid-trimester premature prelabor rupture of membranes (pPPROM) is a rare pregnancy complication. Thanks to the neonatal progress, the survival rate has increased but little is known about long-term outcome of these patients.

Methods

A retrospective study conducted between 2006 and 2019 at Cliniques Universitaires Saint-Luc in Bruxelles including all preterms born after PPRM before 25 weeks of pregnancy, with a latency between PPRM and delivery of at least 2 weeks and with severe oligohydramnios (amniotic fluid index (AFI) < 5). Each infant with PPRM was matched with a control of the same gestational age (+/- 1 week) and gender. The primary outcome was psychomotor development determined by the Bailey Score at 2 years follow-up. The secondary outcomes were perinatal hypoxic respiratory failure (severe pulmonary hypertension by echo, need for inhaled nitric oxide (iNO)), bronchopulmonary dysplasia (BPD) (respiratory support at 36 weeks), IVH grade>2 and mortality.

Results

90 infants were included : 45 in group 1 (PPROM group) and 45 in group 2 (control group). All infants had completed an antenatal steroid course. The mean gestational age was 28 weeks 3/7 in group 1 and 28 weeks 4/7 in group 2 (NS). Mean birthweight was 1210 gr and 1090 gr respectively (NS). 28/45 patients from group 1 (62%) required iNO for severe pulmonary hypertension versus none from group 2 (p<0,001). 37 PPRM infants have survived the neonatal period : 14 of them (38%) had BPD at 36 weeks versus 6 in control group (16,2%) (p = 0,05). One PPRM baby had a HIV grade 3; there was no HIV > grade 2 in group 2. The mortality rate was 18% in PPRM group (8 patients/45). There was no significant difference in Bayley scores between group 1 and group 2 (PDI Bayley 94,8 versus 95,2, MDI Bayley 97,6 versus 101,1 and language Bayley 92,8 versus 93,3). When comparing the 2-year Bailey scores of children with PPRM occurring before vs. after 20 weeks, there was no significant difference.

Conclusion

Preterm infants with mid-trimester PPRM had a higher rate of hypoxic respiratory failure in the perinatal period. After a few days, their evolution was similar to those of their counterparts. The 2-year psychomotor outcome was comparable with and without extreme PPRM. These results are in support of offering full resuscitation and perinatal care to these critically ill infants, but larger multicentric studies with longer follow-up have to be performed to confirm our data.

Short Oral Presentation

SO 9.

Early postnatal outcome and care after in utero exposure to lithium : a single center analysis

M. Torfs, T. Hompes, K. Van Calsteren, A. Smits

*KU Leuven, UZ Leuven*Background/aims

Knowledge on the impact of in utero exposure to lithium during the postnatal period is limited. Besides a possible teratogenic effect during the first trimester, exposure during the second and third trimester might lead to neonatal lithium effects. Today, uniform guidelines for postnatal management of neonates with this condition are lacking. The aim of this study was first to retrospectively describe all neonates admitted to the University Hospitals Leuven after in utero exposure to lithium over the past 10 years. Second, we aimed to propose a medical postnatal care protocol.

Methods

Based on a retrospective search, neonates with in utero exposure to lithium, admitted to the neonatal medium and intensive care unit of the University Hospitals Leuven (January 2010-April 2020) were included. Descriptive statistics were performed and data were presented as median (interquartile range, IQR) or incidence. For continuous parameters with serial measures, median population values were calculated.

Results

Ten neonate-mother pairs were included. Maternal diagnosis for each case was bipolar disorder. Median maternal lithium TDM at delivery was 0.62 (IQR 0.54-0.72) mmol/L, but lithium doses varied between and within pregnancies. The neonates had a median gestational age of 37 (IQR 36-39) weeks, with 3 neonates born prematurely. Median birth weight was 3000 (IQR 2620-3440) grams. At birth, neonatal lithium TDM was 0.65 (IQR 0.56-0.83) mmol/L with a median neonate/mother TDM ratio of 1.02 (IQR 0.87-1.08). Three of 10 neonates needed respiratory support, and 7/10 started with full enteral (formula) feeding at the day of birth. Biochemically, 1 neonate displayed hypoglycemia, and no major abnormalities in early electrolyte levels, liver- and thyroid function were seen in this cohort. One neonate developed nephrogenic diabetes insipidus with a maximum diuresis of 16 ml/kg/h, and had a congenital diaphragmatic hernia. Overall, median length of neonatal stay was 8.5 (IQR 8-12) days.

Conclusion

We reported in detail on the postnatal characteristics and short-term neonatal outcome after in utero exposure to lithium. A postnatal care protocol was proposed, including suggestions for cardiorespiratory monitoring, lithium TDM, electrolyte follow-up, and observation of a least 5 days (extension on individual base). Our results might enhance the quality of care for future neonates with this condition, and can be useful for prenatal counseling.

SO 10.

Umbilical venous catheter-related complications

E. Exelmans, V. Cossey, S. Van Lierde

UZ Leuven, RZ Tienen

Aims

Assess the complication rate of umbilical venous catheters (UVC) in newborns and identify risk factors.

Methods

A retrospective observational study at the Neonatal Intensive Care Unit (NICU) of the University Hospitals Leuven, in newborns admitted between January and December 2016. Central line-associated bloodstream infection (CLABSI) was defined as one positive blood culture in a symptomatic neonate 48 hours after insertion and up to 48 hours after removal of the UVC. Portal vein thrombosis was characterized as the presence of an occlusive clot in the vena portae, diagnosed by doppler ultrasound. Descriptive statistics were used to calculate incidences. The significance of results was valuated with Pearson's chi-squared test and Fisher's exact test.

Results

In 291 neonates, i.e. 58.2% of all neonates, an UVC was inserted on admission. Portal vein thrombosis occurred in 3.4% of patients with UVC. In the ten patients with a thrombosis twelve UVCs were placed, of which nine in the low position (75.0%). In newborns, examined by abdominal ultrasound (33.3%), the incidence of portal vein thrombosis increased to 10.3%. Extremely low birth weight (ELBW, ≤ 1000 g) is a significant risk factor (RR = 6.8; 95% CI: 1.9 to 23.9) compared to neonates with a higher birth weight (> 1000 g).

The second most frequent complication is CLABSI with an incidence of 2.4% or 3.8 per 1000 catheter days. ELBW is again a significant risk factor compared to neonates with a higher birth weight (RR = 6.5; 95% CI: 1.5 to 27.8) .

Conclusion

Neonates weighing 1000 grams or less had the highest risk of developing CLABSI or thrombosis during UVC use. Lack of screening for portal vein thrombosis possibly causes an underestimation of the incidence of thrombosis. Improvements can be made regarding prevention and follow-up of UVC-related complications.

Short Oral Presentation**SO 11.****Stenting of aortic coarctation in VLBW neonates**

J. Dauby, V. Rigo, C. Jacquemart, I. Broux, M-C Seghaye, M. Gewillig

U Liège, UZ Leuven

Introduction

Coarctation of the aorta (CoA) is a frequent congenital heart disease (CHD) leading to significant morbidity and mortality if left untreated. In extremely preterm infants, therapy remains especially challenging. Until recently, surgical correction was the only way to manage critical CoA in neonates but was associated with a high complication rate or was not available for the smaller infants. In order to bridge patients until a lower risk surgery, stent implantation, previously reserved for older children, has been suggested. Three cases of stenting through sternotomy are presented.

Cases presentation

All 3 preterm neonates considered (N1-N3) were born from a monochorionic diamniotic pregnancy, a known risk factor for CoA. N1 was born at 29w and 900g. Patent ductus arteriosus (PDA) screening by echocardiography on day 2 showed both a PDA and a narrowing of the aortic isthmus. Prostaglandin infusion allowed duct maintenance and transfer until aortic stenting 5d later. A post-operative pericardial effusion was drained and NSAID was introduced for 3d. Secondary restenosis appeared gradually and was confirmed by echography. Stent expansion by introduction of a second stent was performed at 30d and 1210g. Hypertension and restenosis appeared again and required an additional stent expansion at 13w and 2845g. No further re-CoA is observed until the age of 4m. Surgical reparation is scheduled at ~6kg. N2 was born at 34w, 1380g, after an antenatal diagnosis of CoA and ventricular septal defect. Prostaglandin therapy is started at birth until stenting on d9. Stent expansion through a second stent is required at 10w due to medication resistant hypertension and was complicated by a pulmonary edema. A second dilatation was performed at 19w. Surgery is planned at the age of 1y. For N3, born at 26.5w, 640g, PDA screening on day 1 showed a large PDA and a suspicion of CoA, which was confirmed on day 21 when PDA became restrictive. The infant was transferred for aortic stenting with pulmonary artery banding. The patient died from respiratory insufficiency due to bronchopulmonary dysplasia.

Conclusion

Three cases of CoA in VLBW neonates are presented. Early stenting by sternotomy was performed and allowed bridging very preterm infants, for whom correction was not an option, to a delayed and safer surgery. Because of rapid growth, stent diameter may not remain sufficient and secondary stenting was required in both surviving patients.

SO 12.

Point-of-Care Ultrasound in the Neonatal Intensive Care Unit

R. Demerre, L. Thewissen, A. Debeer

*UZ Leuven, KUL*Background

Point-of-care ultrasound (POCUS) refers to an ultrasound performed and interpreted by a bedside clinician, which aims to answer a specified question or to guide a clinical procedure. The use of point-of-care ultrasound (POCUS) by neonatologists is rapidly increasing, but there is a lack of evidence and standardization about its implementation in the neonatal intensive care unit (NICU). The purpose of this literature study was to summarize and compare the studies about specific applications of POCUS in the NICU, performed by neonatologists.

Methods

We conducted a literature search in the PUBMED and EMBASE database to identify clinical studies in neonates where POCUS was applied. The outcomes were defined as 1) the accuracy of POCUS when performed by a neonatologist, compared with the golden standard or 2) change in management due to the POCUS results. Risk of bias and level of evidence was assessed for each study.

Results

A total of 30 studies were selected. Indications could be divided into four main categories: echocardiography, line placement, respiratory distress and endotracheal tube placement. For echocardiography, there was a high agreement between the cardiologist and neonatologist. A change of management was observed in multiple cases, of which most changes were related to management patent ductus arteriosus (PDA). During line placement, POCUS showed a similar accuracy compared to X-ray, which is considered as the golden standard. It resulted in a reduction of time, radiation exposure, manipulation and malposition. Concerning respiratory distress and endotracheal tube placement, less is known about the use in the NICU, but several studies indicated an adequate accuracy and a reduction in time and radiation exposure. A level of recommendation was made for all four indications of POCUS. Line placement had the highest level of recommendation, the other indications had a moderate level of recommendation.

Conclusions

There is evidence that POCUS can ensure faster clinical decision-making and less radiation exposure to the neonate, resulting in less complications. It is important to develop a proper training protocol and evidence-based guidelines so safe implementation in the NICU can occur.

Short Oral Presentation**SO 16.****Safety and feasibility of S-Caine patch use in children under the age of three: a pilot study**

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VUB, UZ Brussel

Background

Local anesthesia of intact skin before performing painful punctures in children is considered essential. Several drugs and devices have proven to be safe and efficient, including the topical S-Caine patch (containing a 1:1 eutectic mixture of 70mg lidocaine and 70mg tetracaine, and an air-activated heating element), yet until today only approved for use in children over the age of three years. This study aims to evaluate the safety and feasibility of the use of S-Caine patches in children under three.

Methods

In this open-label, single-dose pharmacokinetic pilot study, all children younger than three admitted to the Paediatric Intensive Care Unit with central catheters, were eligible for inclusion. During four hours following a 30 minutes skin application of one S-Caine Patch, plasma levels of lidocaine were determined with a validated liquid chromatography Phospho Diode Array method. Safe plasma concentration threshold was defined as 0.1mg/L, or one-tenth of the lowest concentration reported to be clinically relevant. The maximum concentration (C_{max}; mg/L) and the associated time (T_{max}; minutes) were measured. Local and systemic adverse events (AE) were monitored, and compared among three age groups using ANOVA. Ease of application and overall satisfaction were scored by the nurse, using a Likert scale.

Results

Nineteen patients were included and stratified into three age-groups: 0-5 months (n=6), 6-11 months (n=4) and 12-36 months (n=9) old. Mean concentrations were 0.010; 0.013; 0.029; 0.024; 0.023; 0.019 mg/L at 0; 15; 30; 60; 120; 240 minutes after application respectively. Regardless of age, lidocaine plasma concentrations did not exceed the safe threshold, except in one patient (C_{max} 0.11 mg/L at 30 minutes) without any clinical repercussions however. Eleven subjects (58%) demonstrated mild to moderate local effects (erythema, blanching and edema), which all spontaneously disappeared within 30 minutes after patch removal; except for one patient demonstrating longer erythema at the application site. No serious treatment-related AE were noted. In 17 patients (89%), the patch was easy to apply, in two patients (11%) patches released too early. Overall satisfaction was excellent or good.

Conclusions

This pilot study suggests that the use of S-Caine Patch in children younger than three years old could be safe and feasible. This study continues on a larger scale to confirm these results.

SO 32.

Respiratory outcomes in very and extremely preterm infants: what is the impact of recommended neonatal care in real-life conditions?

A. Vuckovic, D. Mcheik, A-B Johansson

*ULB - HUDERF*Background and aims

The European guidelines for the management of respiratory distress syndrome in preterm infants have evolved in the last decade in parallel to evidence regarding stabilization in delivery room, oxygen saturation targets, timing and indication of surfactant therapy, as well as the use of caffeine and corticosteroids. The aim of this study was to assess the impact of updated recommendations on the incidence of bronchopulmonary dysplasia (BPD), mortality and respiratory outcomes in a cohort of preterm infants during two recent epochs.

Methods

This was a retrospective cohort analysis of neonates born before 32 weeks of gestation and admitted in a single neonatal intensive care unit (NICU) between 2007-2010 (era 1) or 2013-2016 (era 2). Patients were not eligible in case of initial management in another NICU, congenital anomaly, postnatal transfer or lost to follow-up. Demographics, neonatal management, short-term outcomes including BPD and mortality, and respiratory outcomes until 24 months of corrected age were collected from medical records. BPD was defined as the requirement of supplemental oxygen and/or any respiratory support at 36 weeks of postmenstrual age. Univariate analysis was performed.

Results

Demographic characteristics were similar in both eras. As compared to the 150 infants born in era 1, the 131 infants born in era 2 were less often intubated in the delivery room. Surfactant was also administered later, reflecting rescue administration, and was more often given via INSURE technique. Days on noninvasive support increased, but duration of mechanical ventilation and supplemental oxygen did not vary. There was a rise in early use of caffeine. The use of ibuprofen for patent ductus arteriosus was twice lower in era 2. BPD incidence increased (OR = 4.11, 95%IC: 2.1,8.0). Mortality rates and other comorbidities did not change except a lower rate of late onset sepsis in era 2. At 24 months of corrected age, 114 and 99 surviving patients had a complete follow-up in era 1 and 2 respectively. In a similar fashion in both eras, BPD patients had more chronic respiratory symptoms requiring inhaled therapy and more hospitalizations for acute respiratory illness.

Conclusions

In this study, a rise in BPD incidence occurred despite implementation of the European guidelines for the management of respiratory distress syndrome. However, midterm outcomes after NICU discharge did not worsen over time in BPD patients.

Poster Walk**PW 1.****Associations of perinatal characteristics in extremely low birthweight children with body composition and cognition in early adolescence**

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UZ Leuven, Sun Yar-Sen University, Research Institute Alliance for the Promotion of Preventive Medicine, Erasmus MC, ZNA Hospitals Antwerp

Background/Aims

Children born with extremely low birth weight (ELBW) have a different body composition at 40 weeks post menstrual age and remain smaller compared to children born at term. They are also at risk for impaired neurocognitive development. The aim of this study was to associate anthropometrics and cognitive performance outcome in 11 year old children with perinatal factors within ELBW children.

Methods

We used data from a retrospective study, which collected perinatal data on live-born ELBW infants between 2000 and 2005. At 11 years of age, in 93 ELBW children, cognitive function was assessed by the Wechsler-Non-Verbal test and height, weight, skinfolds, and fat-, lean- and water percentage were measured, using the BodyStat device.

Results

Ninety-three cases were included. Children treated with antenatal steroids were smaller, 145.0 vs 148.6cm ($p=0.047$), had a higher fat percentage, 25.6 vs 19.5% ($p=0.045$), and matrix score, 48 vs 38 ($p<0.001$). Earlier achievement of full enteral nutrition was associated with higher fat percentage ($p=0.045$, fat % $r=-0.252$). Body mass index, water percentage and iliac skinfold were correlated with bronchopulmonary dysplasia (respectively, $p=0.035$ with $r=-0.235$, $p=0.025$ with $r=0.255$, $p=0.023$ with $r=-0.246$). Children who suffered from an intraventricular hemorrhage had a higher water percentage, 76.1 vs 71.6% ($p=0.045$), smaller subscapular skinfold, 0.73 vs 0.87 cm ($p=0.040$), and a lower matrix score, 44 vs 48 ($p=0.029$). Children with retinopathy of prematurity had a lower fat percentage, 17.7 vs 26.2% ($p<0.001$), and a lower spatial score, 42 vs 47 ($p=0.038$).

Conclusion

Fat percentage was higher in children who received antenatal steroids and earlier nutrition. It was lower in children with retinopathy of prematurity. Antenatal steroids were also correlated with smaller children. A higher total body water percentage was found in children who experienced an intraventricular hemorrhage and bronchopulmonary dysplasia. No significant differences were found on the total IQ score. However, children born with retinopathy or who had experienced an intraventricular hemorrhage had lower scores on matrix or spatial reasoning separately. Children born after administration of antenatal steroids had higher matrix reasoning scores.

PW 3.**Delayed cord clamping till 3 minutes in term caesarean sections: an audit of maternal safety**

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UZ Gent, Erasmus Rotterdam

Background

The World Health Organization (WHO) currently recommends to postpone cord clamping for healthy newborns to at least 1-3 minutes after birth to reduce the incidence of iron deficient neonatal anaemia. During a caesarean delivery most clinicians are hesitant to delay cord clamping longer than 1 minute to avoid severe bleeding complications. On the other hand, placental transfer may not be completed until 3 minutes after birth. The aim of the current study was to assess maternal and neonatal safety outcomes after a local protocol adjustment to prolong the interval to at least 3 minutes after caesarean delivery.

Methods

A retrospective cohort study was performed in pregnant women who delivered via a caesarean section (CS) after ≥ 37 weeks at Erasmus MC, Rotterdam, The Netherlands. A cohort (Nov 2016 – Oct 2017) prior to the protocol adjustment was compared to a cohort after its implementation (Dec 2017 – Dec 2018). Maternal outcomes include: total estimated blood loss, severe postpartum haemorrhage, and need for blood transfusions. Neonatal outcomes include: body temperature, admission to neonatal intensive care unit (NICU), hyperbilirubinemia, and use of phototherapy.

Results

The total study population consisted of 789 women (n=376 pre-cohort; n=413 post-cohort). In the pre-cohort group 100% underwent immediate cord clamping; in the post-cohort group 70% underwent delayed cord clamping (DCC) up till 3 minutes. The estimated maternal blood loss was not significantly different between the two groups (400 ml vs 400 ml; $p = 0.52$). Neonatal haematocrit values were higher after DCC (0.51 l/l vs 0.55 l/l; $p =$) but more infants required phototherapy (3.5% vs 6.5% ; $p=0.05$). There was no difference in NICU admissions.

Conclusions

DCC after 3 minutes in term caesarean section was not associated with maternal bleeding complications and other safety outcomes and could be beneficiary for neonates

Poster Walk**PW 5.****Accuracy of renal function estimating equations compared to iohexol clearance in critically ill children**

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UZ Gent

Introduction

Accurate assessment of renal function is crucial in intensive care. Both acute kidney injury and augmented renal clearance may compromise outcome. Common formulas to estimate glomerular filtration rate (GFR) are proven unreliable in critically ill adults. Comparison of a gold standard technique to assess GFR with formula-based GFR estimations (eGFR) has never been reported in pediatric intensive care (PICU) patients. Our aim was to evaluate the performance of 25 currently used GFR estimating formulas based on creatinine (Screat), cystatin C (CysC) and betatrace protein (BTP) in comparison with measured plasma iohexol clearance (CLiohex) in critically ill children.

Methods

A prospective, interventional study was conducted at the PICU of the Ghent University Hospital. 40 Critically ill children, median age 16 months (range: 15 days -13,6 years), 29 males, without chronic renal disease were included. After injection of a weight-dependent bolus of iohexol, 6 serial blood samples were taken over a 360-minutes interval. Measured CLiohex was compared with 10 Screat-based, 10 CysC-based and 2 BTP-based eGFR formulas and 3 eGFR formulas combining the above biomarkers. Correlation between methods was described using Passing-Bablok regression analysis with determination of bias and precision. Agreement between the methods was assessed visually using Bland-Altman plots. Accuracy of GFR estimating equations was determined as the percentage of GFR estimates within $\pm 10\%$ and $\pm 30\%$ of measured GFR by CLiohex (P10 and P30). P30 accuracy $>75\%$ is considered sufficient, however, ideally, P30 should be $>90\%$. Performance of eGFR formulas was also assessed separately in subgroups with CLiohex <100 ml/min/1.73m² and with CLiohex >100 ml/min/1.73m².

Results

No adverse effects related to iohexol were observed. Median CLiohex was 121ml/min/1.73m² (range: 43-221ml/min/1.73 m²). Only 5 eGFR formulas showed an overall P30 $>75\%$. None of the eGFR formulas reached a P30 $>90\%$ for the entire study population. Almost all eGFR formulas tended to overestimate true GFR. Surprisingly, all formulas showed higher accuracy in the subpopulation with higher GFR compared to the lower GFR range. Of the Screat-based formulas, the 2012 Schwartz formula and De Souza formula showed the highest accuracy, with P30=75% and P10=35% and P30=75% and P10=20% respectively. Of the CysC-based formulas, the Schwartz(cys) formula performed best with P30=80% in the overall population and P30=92.3%

P 14.

Effect of CPAP therapy on respiratory control of an infant born prematurely: a case study

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ULB - HUDERF

Background

Sleep analysed by polysomnography in the prematurely born infant is often characterised by the presence of apnea. These can be central, mixed or obstructive apnea, all resulting from unstable ventilator control. The central apneas can be part of a breathing pattern called periodic breathing, which is often accompanied by intermittent hypoxia. This is a report of an infant born after 26 weeks of gestation, ready for discharge from the neonatal unit (HUDERF).

Method

A sleep study using polysomnography was requested as she still exhibited desaturations during sleep on the ward monitor. A control polysomnography was also performed, once treatment had been started.

Results

She exhibited all types of apnea during sleep as well as a high amount of periodic breathing, which was accompanied by fluctuating oxygen saturation. The obstructive apnea (OA) index was also high (21.3 obstructive events per hour of sleep), for which a CPAP treatment was initiated during sleep. The control PSG with CPAP showed a disappearance of the obstructive events, as well as the disappearance of the periodic breathing.

Conclusion

CPAP treatment insures upper airway patency, explaining the disappearance of the OA. An unexpected effect was the disappearance of the periodic breathing, as this pattern of breathing is thought to be centrally controlled. It is proposed that CPAP treatment, by maintaining the upper airways open during sleep, favours better oxygenation, thereby reducing overall hypoxia, resulting in a beneficial effect on the respiratory control centers.

Posters**P 15.****A rare cause of persistent cholestasis in a neonate after surgery for an intrathoracic liver in a case of a congenital diaphragmatic hernia**

D. Dinneweth, K. Van Renterghem, L. Defreyne, T. Van Genechten, L. Goossens

*UZ Gent*Background

A term neonate of healthy non consanguineous parents was admitted at the Neonatal Intensive Care unit with a prenatal diagnosed right-sided diaphragmatic hernia. Uncomplicated start with gentle ventilation and surgery at day 2. Procedure was complicated with an unexpected severe haemorrhage. Because of increasing conjugated hyperbilirubinemia, thrombocytopenia and bleeding diathesis, further investigations revealed a large hypervascular tumour with multiple arteriovenous fistulas.

Methods

Blood and genetic testing and radiographic imaging were performed for quick diagnosis. No biopsy was done.

Results

Severe cholestasis (total bilirubin 51,4mg/dL (0,2-1,3mg/dL), mostly conjugated), persistent thrombocytopenia and coagulopathy developed after surgery. Echocardiography showed an ASD and a dilated vena cava inferior with detectable flow from the liver veins, due to the vascular malformation. CT imaging confirmed the diagnosis. Prenatal ultrasound was negative, but in retrospect the malformation was seen on the prenatal MRI performed for assessment of the diaphragmatic hernia. Surgery was not an option mainly because of the size of the tumour. Partial embolization via the umbilical artery was successful. Propranolol 3mg/kg/d and short-course corticosteroids were associated for further eradication. Genetic testing was inconclusive. Follow-up shows involution of the haemangioma, disappearance of the cholestasis and a good clinical condition.

The exact mechanism of the cholestasis is unknown. Hepatocyte dysfunction due to liver damage because of surgery and large tumour size could be responsible for the inability to excrete conjugated bilirubin. Also intravascular haemolysis with massive destruction of erythrocytes can be responsible for increased production of direct bilirubin.

Hepatic haemangioma is a common benign vascular neoplasm in childhood that can cause early congestive heart failure by arteriovenous shunting and consumptive hypothyroidism. It can be associated with Kasabach-Merritt-like syndrome with profound thrombocytopenia and consumptive coagulopathy. Treatment strategies include medical, surgical and/or interventional approach.

Conclusion

This case shows a very rare combination of a diaphragmatic hernia and a congenital hepatic haemangioma associated with a Kasabach-Merritt-like syndrome. Few cases have been described in literature. Combination of interventional radiology and drug therapy were needed for treatment of the haemangioma.

P 16.

Neonatal adrenal insufficiency : always listen to parents

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ULB – HUDERF, ULB - ERASME

Background/Aims

Neonatal adrenal insufficiency is a rare disease. Diagnostic delay can lead to both dreadful early complications such as a hemodynamic shock and late complications such as intellectual disability due to hypoglycemic episodes.

The most frequent cause of adrenal insufficiency is congenital adrenal hyperplasia, usually due to 21Hydroxylase deficiency. This is easily diagnosed in a female newborn because of atypical genitalia and elevated 17OHP. Adrenal insufficiency can also arise from rare enzymatic blocks or adrenal hypoplasia.

Methods: case report

Results:

We report on a female newborn born at term after an uneventful pregnancy with normal birth parameters and examination. Her parents are from Conakry, non-consanguineous and both healthy. Since birth, the parents had noted an unusual dark skin pigmentation. At 20 hours of life, she presented with profound hypoglycemia and was admitted to the NICU. She received naso-gastric feeding (maternal and artificial milk). She was sleepy and hypotonic. After 7 days of naso-gastric feeding, while she started to drink by herself, other episodes of profound hypoglycemia occurred. Adrenal insufficiency was suspected on the basis of hypoglycemia, hypotonia, hyperpigmentation and a tendency for salt loss. Her laboratory results confirmed the primary adrenal failure: cortisol <3 nmol/L, ACTH > 2000 ng/L, SDHEA <0,08 umol/l, 17-OHP 0,30 ng/ml. A central hypogonadism was also noted with a pathological LHRH test (no LH response). Plasma renin was only slightly elevated 219.6 mUI/L. Hydrocortisone and 9alpha fludrocortisone substitution was started. The genetic diagnosis is pending.

Conclusion

This case report illustrates the importance of an early diagnostic work-up for neonatal hypoglycemia, especially when in the absence of any risk factor such as intra-uterine growth retardation, prematurity or gestational diabetes. The absence of atypical genitalia and normal-low 17 OHP dosage are falsely reassuring: this allows to exclude congenital adrenal hyperplasia but not other rarer forms of adrenal insufficiency. Last but not least, the parents remark about skin pigmentation were judicious and a very early sign of disease. Parents opinions should always be taken very seriously.

Posters**P 17.****« Ping-pong » skull fracture**

S. Cahen, M. Rodesch, C. Fricx, P. David, O. De Witte, F. Vermeulen

*ULB - ERASME*Introduction

Depressed skull fracture in newborns are rare (estimated incidence 4 -10/100 000). They are often associated with instrumental deliveries but they can be spontaneous. Spontaneous depressed skull fractures are caused by the bones of the skull sinking inward without cortical break. We will hereafter report the case of a newborn who presented a “ping pong” fracture.

Case

A 9-days-old boy was brought to the outpatient clinic because his parents noticed from his third day a large right-sided parietal depression of his skull (5cm x 3cm). The birth report did not mention it. It was an uncomplicated second pregnancy. Five prenatal ultrasounds were normal. The child was born at 38 weeks of gestation spontaneously vaginally delivered. Since birth, no trauma was reported. The physical examination showed a large depressed parietal fracture. The rest of the skull was normally shaped. X-ray was performed and confirmed a depressed skull fracture with no cortical impairment. The magnetic resonance imaging did not show any associated encephalic lesion. Biological check-up was normal. Fundoscopic examination revealed no indirect sign of trauma nor intracranial hypertension. The diagnosis was a spontaneous depressed skull fracture. A surgery of reshaping of the skull through a drill hole was performed at 12 days of life. The clinical and sonographic courses were good.

Discussion

A ping pong fracture is a type of spontaneous depressed skull fracture that can occur in neonates due to the soft nature of the newborn skull. During vaginal delivery, the fetal head is compressed by the mother’s pelvis and it may lead to depression of the parietal bones. There are multiple risk factors as a prolonged delivery, trauma during pregnancy, multiple gestations and disorders of osteogenesis. Those fractures can be identified clinically at delivery. Radiographs of the skull can demonstrate the degree of deformation. Cranial ultrasound or MRI can be used to see if there is intracerebral bleedings associated.

The prognosis of those fractures is good.

There are no guidelines for treatment, which usually depends on the severity of the fracture, the underlying brain injury and the use of instrument during delivery. Some cases of spontaneous resolutions have been described recently.

Conclusion

Neonatal depressed skull fractures are rare and more often associated with obstetrical trauma but some spontaneous cases are described. Both surgical and conservative treatments are possible.

P 18.

Recurrent E. Coli Meningitis in a preterm infant

E. Hoornaert, S. Smeets, C. Genin , P. Maton

Clinique Saint Vincent Rocourt

Introduction

Neonatal bacterial meningitis is a life-threatening infection that is associated with high rates of mortality (20-25%) and morbidity. A prompt and adequate treatment helps reducing sequelae but does not prevent them. Escherichia Coli is the second most common pathogen.

Case report

A female infant was born by vaginal delivery at 27.5 weeks. Birth was induced due to a premature rupture of membranes and an increase of the patient's mother CRP. The mother was treated with ampicillin before delivery and her vaginal swab returned positive for an E. Coli resistant to Ampicillin. Infant's adaptation was good but she presented a neonatal respiratory distress that required a CPAP support. Empiric antibiotics were initiated at birth (Penicillin and Amikacin). However, negative blood culture (collected prior to antibiotics) and normal lab results justified the interruption of the antibiotics after 3 days. After 7 days of life, the infant presented low grade fever, tachycardia and irritability. CSF analysis revealed elevated pleocytosis and protein levels (10g/L). E. Coli resistant to ampicillin and susceptible to cefotaxime was isolated from blood and CSF samples. Cefotaxime treatment was initiated for 21 days with good clinical improvement and normal cerebral ultrasounds. Blood cultures on day 2 and CSF culture on day 5 were all negative. 10 days after interruption of the antibiotics, the patient turned back irritable with marked tachycardia and presented diarrhoea. There was again an elevated pleocytosis and protein levels in CSF analysis and an identical drug-resistant E. Coli phenotype was isolated. No anatomic defects or abscesses were detected by cranial and spinal MRI. Primary immunodeficiency testing is underway to explain the recurrent meningitis.

Conclusion

Recurrent meningitis is a rare condition (1.3-5% of meningitis cases). Early diagnosis of an underlying pathology responsible of recurrent meningitis (congenital or acquired anatomic defects or immunodeficiencies, or chronic parameningeal infections) is crucial to prevent further episodes and improve the outcome of the patient. Therefore, an MRI and an immunodeficiency screening must be done to each patient presenting a recurrent bacterial meningitis. This case report also highlights the question if a lumbar puncture is needed in each patient after 3 weeks of correct treatment to be sure the bacteria has disappeared. No definite answer can be found at present in the medical literature.

Posters**P 19.****A premature infant with congenital anomalies and respiratory failure**

S. Verhulst, A. Mulder, K. Van Hoorenbeeck

*Universiteit Antwerpen, UZ Antwerpen*Case history

This case depicts the clinical course of a preterm boy admitted to our NICU. Prenatal ultrasounds showed a megacystis with wall thickening, bilateral hydronephrosis and hydroureter, ascites and the absence of abdominal musculature. The infant was delivered at a gestational age of 28 weeks and 3 days; after preterm contractions not responding to tocolysis and incomplete prenatal corticosteroid therapy. At birth, the diagnosis of Prune-Belly syndrome was confirmed. Additionally, clinical examination showed chest wall deformities and severe thoracolumbar scoliosis. The infant was intubated in the delivery room and surfactant was administered because of RDS grade 4. Over the next weeks, there was a persisting need for high FiO₂ and the boy eventually developed severe BPD. Despite optimizing supportive treatment and a course of hydrocortisone, extubation failed and the boy remained ventilator-dependent. Several episodes of ventilator-associated pneumonia occurred in the following weeks and there was recurrent atelectasis of the right upper lobe. At the postnatal age of 3 months, there was no extensive nephrological or neurological damage. After multidisciplinary consultation and conversations with the parents, a tracheostomy was made. Ventilation was switched to settings compatible with transfer to a revalidation center. Although this was initially not tolerated well, stable ventilation was established after 10 weeks and the infant was transferred with continuous invasive ventilation.

Conclusion

This case highlights the respiratory management of a preterm infant with restrictive lung disease, severe BPD and congenital anomalies. Eventually, continuous invasive ventilation was established. Ethically important; the overall and neurological prognosis remain unclear.

P 20.

Influence of age on clinical presentation, diagnosis delay and outcome in preschool children with acute appendicitis

Y. Lounis, J. Hugo, M. Demarche, M-C Seghaye

*CHU de Liège, Université de Liège, CHR Citadelle Liège*Background

Unusual clinical presentation of acute appendicitis in preschool children leads to misdiagnosis and complications.

We aimed to analyze the influence of age on clinical presentation, laboratory findings and complications in preschool children with acute appendicitis.

Patients and methods: From January 2012 until December 2017, 29 children younger than 6 years of age (median 50 months) with acute appendicitis were enrolled in this retrospective study. Patients were grouped according to their age: group 1: <48 months (n=13); group 2: > 48 months (n=16), their clinical data, laboratory results and complications were compared.

Results

In group 1, duration of nausea and vomiting was longer, alteration of general state was more frequent and pain in the right fossa iliaca less frequent than in group 2 ($p = 0.026$, $p=0.0001$ and $p=0.029$, respectively). Heart rate was higher in group 1 than in group 2 ($p=0.012$). Leucocyte and polynuclear neutrophil counts were lower in group 1 than in group 2 ($p=0.03$ and 0.004 , respectively) but C-reactive protein levels were not different between groups. In the whole cohort however, C-reactive protein values at admission correlated negatively with age ($p=0.025$).

Abdominal ultrasound allowed diagnosis in 19/29 patients (65.5%), without any difference between groups. Appendicular perforation was more frequent in group 1 than in group 2 ($p=0.003$). Perforation was also related to longer hospital stay ($p=0.02$). Peritonitis occurred in 21/29 (72%), post-operative ileus in 5/29 (17%) and sepsis in 4/29 (14%) patients without any difference between groups. In the whole cohort, hospital stay correlated negatively with age ($p<0.0001$). There was no mortality.

Conclusions

Among preschool children, those younger than 48 months present with longer duration of pre-admission symptoms indicating longer infection course than in older children. Altered general state and higher degree of tachycardia in the younger reflect higher systemic repercussions of the illness. Less specific abdominal pain and dissociation of the inflammatory markers with lower leucocyte- and neutrophil counts and higher C-reactive protein levels in the younger may contribute to further diagnosis delay and higher rate of perforation in these patients.

Posters**P 21.****Assessment of pediatric CPR training videos on YouTube**

A. Goossens

*KUL, UZ Leuven*Objective

The interval to initiate cardiopulmonary resuscitation (CPR) in patients with cardiac arrest is crucial for survival and a proper education is a prerequisite for CPR conducted by lay bystanders. The contemporary habit of using the internet and social media explains the growth of the online medical education. CPR is a good example for the use of educational videos. The goal of this assessment is to determine the medically justified information and the educational quality YouTube videos regarding CPR on children in accord with the 2015 CPR guidelines. The objective of this study is also to investigate if this information is acceptable for laymen.

Methods

YouTube was accessed during February and March 2019. Twelve search strings were entered in the search field. The final videos were selected based on specific inclusion and exclusion criteria. The criteria used to evaluate the videos were included into four checklists. Two experts and two laymen scored the videos by watching each video independently. The checklist reliability was chosen as reference.

Results

99 videos were included out of the 451 videos found following a YouTube search. The popularity of videos about CPR on children is low compared to other instruction videos. The source 'healthcare professional' contained the highest scored reliability. There was a statistically significant positive impact found of two specific search strings ('Basic life support pediatric' and 'CPR pediatric'), comment count/day and duration on the reliability.

Conclusion

The use of medical scientific words in the search string such as 'pediatric', a reliable source, high number of comment count/day and a longer duration are important predictors for the overall quality of a video on YouTube concerning CPR on children. This study can help laymen choosing the right video to supplement their knowledge concerning CPR on children

P 22.

Parechovirus encephalitis: an overlooked entity

S. Magotteaux, N. Demonceau, F. Lebrun

*CHC Montlegia*Case

A 9-days old infant with no medical history comes to the ER for a sudden irritability and difficulty to maintain normal temperature. The clinical examination is normal except for the irritability. The laboratory blood work shows no significant inflammatory syndrome, normal urine analysis and negative NPA. The CSF is positive for parechovirus (HPeV) with a normal leukocyte count, later the serotype is identified as HPeV 3. Antibiotics (Amoxicillin and Cefotaxime) are started pending cultures. Later that day, she has status epilepticus which requires a total of four anti-epileptic drugs to stop: Levetiracetam, Midazolam, Phenytoin, Phenobarbital. The cerebral MRI shows deep white matter lesions as well as lesions in the corpus callosum and caudate nucleus. Levetiracetam and Midazolam are stopped before discharge. The 2 months follow up EEG is normal. Phenytoin is slowly reduced and stopped.

The final diagnosis is of a parechovirus 3 encephalitis.

Discussion

Parechovirus is a nonenveloped RNA virus of the Picornaviridae family which includes enterovirus, hepatovirus and rhinovirus. First description of HPeV dates from 1956. The common triad in infants is high fever, skin rash and irritability (red hot angry babies). In neonates (< 3 months), HPeV infection tends to be more severe with high similarities to bacterial (sepsis-like) or extended HSV infections. Mortality is highest in neonates (up to 83%). Laboratory findings are nonspecific (low inflammatory response, normal CSF counts). Rapid diagnosis relies on PCR detection. HPeV 3 has a neurotropism and can cause encephalitis, transient paralysis and white matter lesions. Other complications are hepatitis and myocarditis. Long term follow up is necessary particularly in CNS infection with risk of language and speech dysfunctions, lower intelligence and seizure.

Undergoing investigations for treatment suggests immunotherapy with immunoglobulins injection (neutralizing antibodies) and/or antiviral drugs. Capsid inhibitors (pleconaril, pocapavir) have showed promise in neonatal HPeV infections.

Conclusion

Neonatal HPeV infections have a high rate of morbi-mortality. Diagnosis should be evoked in infants with overwhelming sepsis or serious symptoms, especially with nonspecific laboratory findings. Diagnosis is based on PCR detection. Suggested therapies are immunoglobulins and capsid inhibitors. Long term follow up is necessary due to potential severe sequelae, especially with neurological impairment.

Posters**P 23.****High Speed Chest CT for the diagnosis of bronchopulmonary dysplasia**

A. Jeunen, M-R. Van Hoestenbergh, W. Gysemans

*KUL*Background

Bronchopulmonary dysplasia (BPD) is a well-known long-term complication of prematurity. Despite advances in medical care and improvement in survival of preterm babies, the incidence of BPD has remained stable or has even increased over the past decades, which likely reflects the impact of increased survival of extremely preterm infants. A shift though, has been made from what is now called 'old' BPD to the 'new' BPD. The 'old' BPD is attributed to the use of high oxygen levels and mechanical ventilation and is characterized by prominent airway injury, epithelial metaplasia, smooth muscle hypertrophy and parenchymal fibrosis. The 'new' BPD is rather an aberration of normal lung development. This interruption of normal lung development compromises both alveolar and vascular development in the small airways and is influenced by prenatal as well as postnatal factors, such as antenatal glucocorticoids, stress, cytokines, infections, ventilation, oxygen exposure.

Methods

In this report we describe the case of a small for gestational age (SGA) preterm, born at 31 weeks five days post menstrual age (PMA). Initially the X-Ray of the chest shows only mild hyaline membrane disease. Later on, we see a gradual onset of intermittent hypoxic episodes with cyanosis requiring high flow nasal cannula therapy without supplemental oxygen requirement. The need for ventilatory support is still present at 38 weeks PMA, which by definition sets the diagnosis of BPD.

Results

Chest X-ray at post menstrual age of 38 weeks PMA showed relatively small lung volumes without abnormalities of the lung parenchyma. High speed Chest CT (HSCT) at near term age however revealed marked changes both of the vascular and pulmonary compartment.

Conclusion

Chest CT is a helpful tool in the diagnosis and management of BPD. There are multiple quantitative scoring systems but a standardized protocol is missing so further research is necessary. HSCT has even more advantages since it has an excellent spatial resolution which helps to understand the complexity of BPD and helps to refine the therapeutic management.

P 24.

A spontaneous arterial thrombosis in a preterm infant

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CHU de Liège, U Liège

Case report

Our infant is a preterm baby born at 30 weeks of gestational age by a cesarean section for breech presentation and spontaneous labour caused by a chorioamnionitis. Pregnancy is marked by a gestational diabetes and a premature rupture of membrane at 29 weeks with subsequent oligoamnios. The birthweight is 1440 grams. The newborn adapts well, and needs a non-invasive ventilation. At delivery, the left lower limb appears very pale with a prolonged capillary refill time but presents normal spontaneous motricity. The rest of the physical examination is normal. The doppler ultrasound shows an organized thrombosis of the left iliac artery with arterial vicariences, which confirms the old character of the thrombus.

A treatment by low molecular weight heparin (LMWH) is initiated. Before the beginning of the treatment, laboratory testing for thrombophilia is performed and shows a deficit of C protein, S protein and antithrombin. These abnormalities still need to be confirmed at the age of 3 months as they aren't found in both parents. The evolution under treatment is favorable with a progressive regression of the clot.

Discussion

Neonatal thromboembolism is a rare condition with an incidence of 5.1 events per 100000 births. The majority of the thrombosis are due to the presence of an arterial or venous access device. Other risk factors include maternal lupus, gestational diabetes, birth asphyxia, neonatal polycythemia, sepsis, dehydration, cardiac disease, prothrombic disorders and surgery. The symptoms of an arterial thrombosis depend on the thrombus size and location. The doppler ultrasound is used to confirm the diagnosis and to evaluate the efficacy of the treatment.

Before any treatment is started, laboratory testing for thrombophilia must be realized. Well-designed randomized controlled trials evaluating the efficacy, safety and dosage regimen of anticoagulant therapies are lacking. Therefore, the management of neonatal thrombosis is extrapolated from the adult. Every case should be assessed individually. When an anticoagulant therapy is initiated, LMWH appears as a first line strategy for most of the situations. The use of thrombolytic agents can be considered in case of life-threatening conditions.

Conclusion

Neonatal thrombosis is rare but early diagnosis and assessment is required to initiate early management. Physical examination and doppler ultrasound lead to diagnosis. The treatment has to be discussed on individual risk/benefit ratio basis.

Posters**P 25.****Complication of umbilical venous catheter: TPN leakage in the abdominal cavity.**

A. Ouald Chaib, J. Lefevre, M. Sonnaert, F. Cools

*UZ Brussel*Introduction

Umbilical venous catheter(UVC) is a frequently used technical act in intensive neonatal care. These catheters allow administration of parenteral feeding and medication. Although rare, low position of the UVC and its prolonged use can predispose for complications such as portal vein thrombosis, infection, vascular or hepatic injury, arrhythmia, extravasation, and sepsis.

Case

A male infant was born at a gestational age of 30+4/7 weeks and a birthweight of 970 grams. An UVC was inserted 2 hours after birth for parenteral feeding. The insertion distance was calculated at 8 cm, but as there was no return of blood on aspiration, the catheter has been repositioned to a less deep position (6.5 cm), where only a small quantity of blood could be withdrawn. A chest X-ray confirmed the non deep position of the UVC, 12 mm below the diaphragm.

On day 1 a slight abdominal distension and scrotal edema was observed, but a conservative attitude was adopted. On day 2, the volume of the abdomen and scrotum was exponentially augmented with clinical signs of peritonitis, which led to the suspicion of TPN-leakage in the abdominal cavity. The UVC was immediately removed and a peripherally inserted central venous catheter was placed to continue parenteral nutrition. Abdominal X-ray and ultrasound confirmed a large amount of intra-abdominal fluid. Diagnostic and therapeutic paracentesis was performed, and 100 ml of a milky fluid could be drained. Biochemical analysis of the fluid confirmed that the composition was equal tot the administered TPN. The hypothesis is that with placement of the UVC, the umbilical vein was perforated which caused the TPN to leak into the peritoneal cavity. Besides a hyponatremia, which could be corrected with an NaCl-solution, there were no other sequels. There were no signs of infection following the incident.

Remarkably, despite inadequate feeding for a period of 36 hours, glycemic controls stayed within normal range.

Conclusion

UVC is a common way to gain IV-access in neonates. Although rare, complications are possible. In this case, the insertion of the UVC caused a perforation of the vein and extravasation of TPN in the peritoneal cavity. When in doubt about correct position, the UVC should be removed and an alternative access way should be gained if necessary.

P 26.

Trali in a PIMS-TS patient

K. Vergaelen, G. Van Berlaer, D. Bulckaert

UZ Brussel

Introduction

Transfusion-related acute lung injury (TRALI) forms the leading cause of transfusion-related deaths. This clinical diagnosis consists of acute non-cardiogenic pulmonary edema with hypoxemia and bilateral pulmonary infiltrates, developing within six hours after transfusion. We present a pediatric case of TRALI after administration of intravenous immunoglobulins (IVIG).

Case

A four year old boy presented with fever, high inflammatory markers and Kawasaki-like symptoms. PCR and circulating antibodies were positive for COVID-19. Cardiac ultrasound showed myocarditis with decreased ejection fraction. He was admitted to the Pediatric Intensive Care with the diagnosis of pediatric inflammatory multisystem syndrome (PIMS) temporally associated with COVID-19. After stabilization with non-invasive ventilation and inotropes, treatment with IVIG and corticosteroids was initiated. Good clinical response was seen and cardiorespiratory support could be weaned. However, because of persistent fever after 48 hours he received a second dose of IVIG. Four hours after administration, he developed acute respiratory deterioration, necessitating intubation and mechanical ventilation. Fluid overload was excluded and since cardiac ultrasound showed no cardiac decompensation as the main contributor to the hypoxemia, the diagnosis of TRALI was established.

Discussion

PIMS is an emerging entity, first described in April 2020. Mainstay of treatment is IVIG, resulting in an important increase of its application. Although all blood products can provoke TRALI, its occurrence after IVIG is rare with only scarce pediatric case reports. Our case represents the first documented case of TRALI in a patient with PIMS.

Conclusion

Although rare, TRALI should always be suspected in acute respiratory failure after transfusion in the pediatric population.

Posters**P 27.****Don't forget to breathe !**

A. Marcuzzi, J. Bottu, C. Pierron

*Centre hospitalier du Luxembourg*Background

What should we think about when faced of a child with persistent hypoventilation, normal clinical exam and normal initial assessment?

Case report

A full-term child is born after decreased foetal movements of several hours. At birth she was hypotonic and didn't breathe except during vigorous stimulation. She was intubated, after an attempt of CPAP, under the hypothesis of perinatal asphyxia. The extubation was performed after a few hours in sight of the normal clinical exam once she was awake. Unfortunately, as soon as she fell asleep, she presented apneas and was reintubated. There was no infectious risk factor or specific family history. The pregnancy was physiological. The initial work-up including EEG, chest x-ray, brain MRI, toxic and metabolic screening was normal. Therefore, a congenital central hypoventilation (CCHS) was suspected. The genetic studies confirmed the diagnosis with a heterozygous expansion of 25 alanine repeats in the PHOX2B gene.

Discussion

Children with a normal clinical exam except the central hypoventilation, severe during sleep, sometime present during wakefulness, are suspect of disorder of ventilatory control. The CCHS also know as Ondine's syndrome is one of the causes of CSA in neonatology. CCHS is a rare condition characterized by an alveolar hypoventilation due to impaired autonomic control of ventilation with an intact voluntary control. An abnormally low or absent ventilatory responses to hypercapnia and hypoxia, observed on the polysomnography, is present at both states of sleep and wakefulness. Dysautonomia can also affects other functions which can lead to digestive, especially Hirschsprung disease, cardiovascular, eye, endocrine disorders etc. A polyalanine expansion in gene PHOX2B is found in most patients, its length is associated with the severity of the autonomic dysfunction.

Conclusion

The diagnosis of CCHS is based on the combination of two major criteria: central hypoventilation or CSA and the presence of a phox2B gene mutation. CCHS is therefore a severe, disabling and potentially fatal disease. A multidisciplinary follow-up is mandatory during all life. The quality of life of these patients has improved past years, thanks to earlier diagnosis and evolution of non-invasive techniques, which makes sometimes possible to avoid tracheotomy. However, it affects the whole daily life of the family. Parents must be able to control the respirator and give first aid if necessary.

P 28.

Metabolic bone disease in premature neonates : implementation of new diagnosis and treatment protocol

V. Collet, E. Boros, V. Vlieghe, A-B Johansson

ULB - HUDERF

Metabolic bone disease (MBD) is defined as bone demineralisation and accounts for significant comorbidity in preterm neonates. MBD occurs in 15 to 40% of infants with very low birth weight (VLBW < 1500 grams). It is a multifactorial disorder caused by insufficient mineral accretion due to the premature birth combined with post-natal factors such as medication, prolonged parenteral nutrition and chronic immobilisation. Clinical manifestations depend on the degree of demineralisation and can result in bone deformation and fractures. Biochemical markers help to detect early mineral deficiency as of the second week of life. The predominant biochemical change includes hypophosphatemia and elevated alkaline phosphatases (ALP). Optimisation of parenteral and early enteral nutrition with adequate provision of calcium, phosphate and vitamin D prevents MBD of prematurity. But there is no evidence-based consensus regarding its diagnosis and treatment. Current practice consists in phosphate supplements alone to lower ALP levels. However, this approach can reduce serum calcium levels, further stimulate secondary hyperparathyroidism, increase bone metabolism and deteriorate osteopenia of prematurity. Our new screening concerns VLBW neonates and/or neonates born before 32 weeks of gestation. The management approach will be based on the plasma parathormone level to distinguish the underlying calcipoenia or phosphopaenia causing the MBD and treat it with adequate mineral supplements.

The aim is implementing a new protocol of MBD in the neonatal unit of the children's hospital in Brussels based on recent review of the literature and assess its impact by conducting a prospective study.

Posters**P 29.****Treatment of Severe OSAS due to Pierre Robin Sequence by placement of a nasopharyngeal airway**

Y. Jaddioui, K. Vergaelen, V. Vanderstraeten, L. Peeters

*UZ Brussel*Introduction

We report a case of a girl with severe obstructive sleep apnea syndrome (OSAS) due to her craniofacial anomalies associated with Pierre Robin Sequence. Pierre Robin Sequence is a condition which presents with the clinical features of micrognathia, glossoptosis and cleft palate. The tendency of the tongue to prolapse backwards can lead to airway obstruction, which can manifest itself as hypoxia, feeding difficulties and failure to thrive.

Case description

Our patient was born at an age of 38 weeks after a normal pregnancy. The diagnosis of Pierre Robin Sequence with a cleft palate was made postnatally. Cardiorespiratory home monitoring was required due to the risk of apnea by an obstructive breathing pattern. At the age of 2 weeks she presented herself with poor weight evolution and breathing difficulties during feeding. Blood gas tests showed important hypercapnia and a performed polysomnography confirmed a severe OSAS.

To relieve airway obstruction a nasopharyngeal airway (NPA) was placed at the age of 2 weeks. The hypercapnia disappeared rapidly (within 24 hours). A favorable evolution in weight and improvement of feeding was seen in the weeks after the placement of the NPA. The beneficial effect of the treatment is confirmed with improved results of the blood gas tests and the polysomnography. At the age of nearly 3 months the NPA was removed because of clinical intolerance. A polysomnography without the NPA showed a continuous improvement of the OSAS. Two months after the removal of the nasopharyngeal tube the polysomnography was completely normalized.

Discussion

An upper airway obstruction is one of the primary issues in patients with Pierre Robin sequence. A non-invasive option for the treatment of upper airway obstruction is the placement of a nasopharyngeal tube to relieve airway obstruction by keeping the tongue from falling back on the posterior pharyngeal wall and occluding the airway. In this case there was noticeable weight gain, less feeding difficulties and less obstructive apneas and hypopneas, after the placement of the NPA. This case shows that NPA is an effective treatment in patients with Pierre Robin Sequence, until the expected facial growth occurs, that leads to the resolution of the airway problems.

P 30.

Supplemental oxygen improves respiratory stability in ex-early preterm infants, but not as well as non invasive ventilation: a case study

S. Scaillet, D. Qachri, C. Lelong, C. Haggemacher, D. Vens

*ULB - HUDERF*Background

It is common practice to discharge ex-early preterm infants from NICU with supplemental oxygen (SO) in case of persisting cardiorespiratory immaturity characterized for example by periodic breathing. This case report describes the effect of SO versus NIV on respiratory stability, using polysomnography.

Case report

A 25 weeks 6/7 new born was discharged from the NICU at 40 weeks postmenstrual age (PM), after having had a polysomnography. It did not show periodic breathing. Most of the apnea were well tolerated. Mean respiratory rate was 72 cycle per minute. She received a cardiorespiratory monitor as additional surveillance during sleep. She was re-admitted a week later for multiple alarms on the monitor. Clinically, she presented intermittent hypoxia, for which SO was administered. No germs were identified. As she remained oxygen dependent, a second polysomnography was performed at 43 weeks PM. It showed pronounced respiratory instability in the absence of SO with a high amount of obstructive apnea (OA), as well as periodic breathing. Sub-optimal oxygen saturation was observed. This respiratory instability was partly reduced when SO (0,5 liter) was introduced during the sleep. It was decided to try NIV by bi-level positive airway pressure treatment during sleep, to wean the child from oxygen. A third polysomnography was performed at 44 weeks PM with NIV, which showed complete resolution of all signs of respiratory instability during sleep.

Discussion

One study suggests that administration of SO improves cardiorespiratory stability in asymptomatic preterm infants when assessed in the sleep laboratory. To date, no similar study with NIV has been reported. This case report shows that NIV therapy improves cardiorespiratory immaturity during sleep better than SO. The hypothesis proposed is that NIV improves residual functional capacity and recruits the various peripheral mechanoreceptors, thus improving ventilatory oscillations. This in turn results in better overall oxygenation, optimizing central control.

Conclusion

This particular case suggests that NIV by bi-level positive airway pressure improves respiratory stability during sleep better than supplemental oxygenation in an ex-early preterm infant. Prospective studies with more subjects need to be done to confirm this observation

Posters**P 31.****Turner's syndrom monosomy X in a 32 weeks preterm with multiple organ failure: case report**

P. Maton

*CHC Rocourt*Case presentation

A premature girl was born at 32 weeks of gestational age by urgent C-section for decreased foetal movements and abnormal Doppler discovered incidentally on a routine outpatient visit.

Initial foetal adaptation was good (Apgar 8/10/10). Physical examination was normal except for a disproportionate stature and lymphoedema on the inferior limbs, associated with hematomas.

Shortly after birth, she developed a neonatal respiratory distress requiring CPAP support as well as severe recurrent hypoglycaemias (<20mg/dL) associated with hyperinsulinism. As hypoglycaemias were not responding to glucose boluses and to increased parenteral nutrition a treatment with diazoxide was started with good response as her glucose blood level stabilized after 48 hours of life.

Thereafter, an important weight gain due to increased fluid retention was observed. On day 6, her respiratory distress increased, and we observed a deterioration of her general condition. A control cardiac ultrasound brought out pulmonary hypertension and a patent ductus arteriosus, mitral insufficiency, a small aortic bicuspid valve and quickly progressing left heart failure on the same day. Diazoxide was stopped.

Lab tests showed hyperkalaemia that led to cardiac dysrhythmia and a renal insufficiency with progressive anuria. Peritoneal dialysis was started to palliate fluid overload and ionic disturbance. She was also treated with broad-spectrum antibiotics.

The infant progressed quickly towards multiple organ failure and presented disseminated intravascular coagulation, metabolic acidosis and cardiac failure causing her death 9 days after birth.

Autopsy results showed : mild dysmorphic signs (hypertelorism, flat root of nose, large philtrum, abnormal auricle and large ears), disseminated intravascular coagulation with multiple thrombi in the lungs, liver and kidneys and secondary signs of organ failure. Turner syndrome with monosomy X was diagnosed through CGH array analysis.

Discussion

Turner syndrome is caused by a partial or complete loss of the second sexual chromosome. XO Monosomy is the most frequent genetic abnormality as it is present in 2% of all conceptions, although 99% are spontaneously miscarried during first trimester and approximately 80% between the 10th week and term. Here, we report an unusual association of hyperinsulinism, acute renal failure and heart dysfunction with minimal anatomical anomalies. Among the questions raised by the case, could pure cell li

P 32.

PENTALOGY OF CANTRELL : Patience is a virtue.

M. Karimi, D. Avino, A. Poupalou, H. Dessy, H. Demanet, A-B Johansson

ULB - HUDERF

Background

Pentalogy of Cantrell (PC) is a rare congenital syndrome including defects of the supraumbilical abdominal wall, the anterior diaphragm, the lower sternum, the diaphragmatic pericardium as well as various intracardiac congenital abnormalities. Its management requires a multidisciplinary team and is particularly challenging for surgeons.

Case Report

We report a case of a 3060g female infant born from a G2 P1 mother at 40 weeks by caesarean section. At 23 gestational age, prenatal ultrasound showed a cardiopathy associated with an anterior diaphragmatic defect and an absence of lower sternum – suggesting a PC. The results of the amniocentesis were normal. At delivery an omphalocele totally covered by skin was noted and a pulsatile mass in the lower chest was palpable. The echocardiography showed a double-outlet right ventricle without pulmonary protection. A CTscan was carried out in order to define the parietal defect and revealed an ectopia cordis with a small ventricular diverticulum. Cardiorespiratory evolution and food tolerance were favourable. Following a multidisciplinary discussion, cardiac and digestive surgeons as well as neonatologists decided to postpone the surgical repair and the patient was discharged home. Currently the clinical course is adequate. A one-step surgery (cardiac and digestive) is scheduled at 3 months old.

Discussion and conclusion

PC appears to be the consequence of an early embryologic developmental failure of the mesoderm segments resulting in complex malformation of the anterior diaphragm and the upper abdomen associated with intracardiac defect. The prognosis is mainly poor given the severity of the heart malformation. The management of this severe pathology requires the collaboration of different specialties and a patient-centered approach. The attitude will depend upon the severity of the malformations (especially the presence of an ectopia cordis), the patient's hemodynamic stability and his or her ability to tolerate the intervention. The morbidity and mortality may be increased by an early surgery. As a matter of consequence, the surgical timing and the therapeutic strategy—in a single-stage or a multi-stage repair—have to be determined on the basis of a comprehensive evaluation. In the reported case, it was decided to postpone the surgery based on the assessment that the omphalocele and the cardiac defects did not require an urgent correction. This allowed for better planning of the intervention.

Posters**P 33.****Overgrowth syndrome: overlap between Sotos and Beckwith-Wiedemann syndromes**

E. Goffinon, C. Vilain, J. Soblet, A. Vuckovic

*ULB - HUDERF*Background

The Sotos syndrome (SS) and the Beckwith-Wiedemann syndrome (BWS) are the two most frequent neonatal overgrowth syndromes. Clinical overlaps between both conditions have been reported, making the diagnosis difficult.

Clinical case

A 36-week female infant presented at birth with weight, length and head size above the 97th percentile in the context of an unbalanced gestational diabetes mellitus. Early hypoglycemia was solved under enteral feeding competed with intravenous glucose. Facial dysmorphism included retrognathia and low implanted, hairy and fleshy ears, with protruding earlobes. The infant remained severely hypotonic with feeding difficulties. At day 11 of life cardiac ultrasound was realized for persisting tachypnea and heart murmur, revealing atrial and ventricular septal defects. Five days later the infant was referred to our center for cardiac management and further neurologic assessment. Brain MRI indicated paucity of gyration. Evoked potentials and electroencephalogram were normal for gestational age. Recurrent hypoglycemia was consistent with hyperinsulinism. As clinical presentation was not pathognomic of any overgrowth syndromes, an in trio mendeliome sequencing together with analysis of the methylation profile at 11p15 were conducted. The former revealed a de novo likely pathogenic p.Glu1966* variant in NSD1, which was consistent with SS. The infant was discharged at 2 months of life with enteral feeding and neurological and cardiac follow-up.

Discussion

SS and BWS are two distinct overgrowth conditions. Most cases of BWS are associated with deregulation of imprinted growth-regulatory genes in the 11p15 region, while abnormalities of NSD1 gene are commonly described in the SS. Beside overgrowth, features of SS include macrocephaly with facial dysmorphism, brain formation anomalies, neonatal hypotonia, intellectual disability and congenital heart defects. Major findings in BWS are neonatal hyperinsulinemic hypoglycemia, hemihyperplasia, macroglossia, abdominal wall defects, unusual ear pits, and increased risk for embryonal tumors. However, recent literature suggests clinical overlaps between SS and BWS, as illustrated in the present case.

Conclusion

Phenotypic overlaps between SS and BWS might render the genetic diagnosis of neonatal overgrowth difficult. Enlarged genetic analyses are indicated in overgrowth syndromes with atypical presentation.

P 34.

Non-primary CMV Infection Not Always Innocent. A Case-report And Literature Review.

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UZ Antwerpen, Universiteit Antwerpen, AZ Turnhout

Background

Cytomegalovirus (CMV) is the most common infectious cause of congenital malformations. Anytime during pregnancy, primary or non-primary infection (reactivation of latent infection or re-infection by a different strain) can occur, with the risk of transplacental transmission. Ten to fifteen percent of infected fetuses have a symptomatic congenital infection at birth. CMV is the main cause of brain damage and sensorineural hearing loss (SNHL) due to prenatal infection. Primary maternal infection has a higher hazard of transplacental transmission than non-primary infection, 32% versus 1.4% respectively. In addition, primary infection is more likely to have clinical significance in comparison with non-primary infection.

Methods

We present a neonatal case of non-primary congenital CMV infection with a severe clinical presentation.

Results

A full-term Caucasian girl was born with diffuse petechia, facial dysmorphisms, respiratory distress, hepatomegaly and hypotonia. Her biometry was within normal ranges. Blood results pointed out thrombocytopenia and elevated transaminases. Chest X-ray showed pneumonitis. Cranial ultrasound and MRI of the brain revealed abnormalities (striatal vasculopathy, subependymal germinolytic cysts, periventricular pseudo-cysts) compatible with CMV. Auditory brain stem response testing was abnormal. CMV culture of saliva was positive. Serology of the mother before and during pregnancy showed immunity for CMV. Serology for other congenital infections was negative. The diagnosis of a severe congenital CMV infection due to a non-primary maternal infection was made. Antiviral treatment with valganciclovir was initiated immediately and continued for 6 months.

Generally, maternal immunity protects the newborn against symptomatic congenital CMV infection. But symptomatic CMV infection due to a non-primary infection is not uncommon. Sensorineural hearing loss in non-primary CMV infection varies between 0 and 11.8%; an abnormal cranial ultrasound is found in 8.3%; mental impairment in 0-57% and small for gestational age in 16.6-37.5%.

Conclusion

Our case illustrates the need for clinicians to take congenital CMV infection in the differential diagnosis, even in CMV immune mothers.

Posters**P 35.****Additional value of microdialysis in studying PK/PD at tissue level in critically ill children.**

E. Hermans, P. De Cock, P. De Paepe, M. Devreese, J. Vande Walle

*Universiteit Gent*Background

Only a small number of drugs used in critically ill children are licensed for use in this specific population. In daily practice these children are generally started on the same dose of therapy as their non-critically ill counterparts, which is mostly extrapolated from adult dosing schemes based on allometric data. However, the combination of dynamic maturational processes and pathophysiological and treatment induced changes can significantly impact drug disposition in diseased children. Worryingly, multiple reports have demonstrated that conventional antibiotic dosing strategies consistently fail to achieve therapeutic plasma concentrations of these drugs in critically ill children. In addition to the challenge of achieving therapeutic antibiotic blood concentrations in this population, adult data show that blood concentrations may not always adequately predict target tissue concentrations.

We investigated the possible solutions to optimize antibiotic dosing in the critically ill child.

Methods

A literature search was conducted on the current knowledge and future perspectives on the optimization of antibiotic dosing in critically ill children.

Results

Individualized dosing, based on population pharmacokinetic models and patient factors known to influence antibiotic pharmacokinetics (PK), increases the probability of achieving therapeutic drug exposures while at the same time avoiding toxic concentrations. However up until now, these models have been based on plasma drug concentration profiles. The effect of developmental and disease-related changes (e.g. sepsis) on tissue penetration of antibiotics is still poorly understood, even though tissue is the target site for most antibiotics. Microdialysis (MD) is currently considered as the golden standard for studying drug disposition in tissues. It is a well-established technique through which unbound drug concentrations in the target tissue can be measured. The application of MD in PK research in children, however, is scarce. Regarding antibiotics, only two studies with a limited number of patients have been published, both showing the limitation of plasma levels as a surrogate for target tissue concentrations.

Conclusion

A shift towards individualized dosing in critically ill children is required to optimize the efficacy of antibiotic therapy. Future PK modelling should be based on drug tissue concentrations. MD appears to be a promising tool to conduct studies on tissue PK in children.

P 36.

Severe ethylene glycol intoxication successfully treated with fomepizol and early hemodialysis - a case presentation

G. Mets, R. Van De Wielle, A. Verrijckt, A. Raes

UZ Gent

Ethylene glycol intoxication is associated with severe neurological outcomes, renal impairment and mortality. Prompt recognition and management increase the chance of good outcome.

Case

Brother (2y) and sister (4y) were admitted to the emergency unit with altered consciousness and vomiting. Symptoms had started less than 3 hours before admission. At that time both are tachypneic but hemodynamically stable. Work-up showed severe metabolic acidosis (pH 7.09 and 7.08) with high-anion gap (27 and 26 mmol/l) and very high lactate levels (29 and 28 mmol/l). While waiting for the toxicology report, acidemia correction was attempted through bicarbonate infusion. After identification of ethylene glycol in urine, both children were transferred to our tertiary centre for administration of fomepizol and initiation of hemodialysis. Blood levels for ethylene glycol in both children (0.87 g/l and 0.56 g/l) met criteria for severe intoxication (>0.5 g/l).

Fomepizol (inhibiting metabolism of ethylene glycol to glycoaldehyde) was started 4 hours after initial presentation. In addition a single dose of thiamin and pyridoxine (cofactors stimulating conversion of glycolate and glyoxylate into non-toxic metabolites) were administered. As thresholds for dialysis (increasing elimination of ethylene glycol, glycolate and oxalate) were met, hemodialysis was initiated within 2 hours after admission at our centre (6 hours after first presentation). High efficiency dialysis was performed with Genius 90 (Fresenius). After 3 hours of dialysis, ethylene glycol levels had decreased significantly to 0.14 g/l and 0.13 g/l, with normalization of pH. Hemodialysis was continued until ethylene glycol levels were no longer toxic (0.03 and 0.05 g/l) and stopped after 12 hours. There were no complications. After extubation, both showed rapid recovery towards normal behaviour and clinical parameters. In follow-up renal function remained normal as well.

Conclusion

We report on two siblings successfully treated for a severe ethylene glycol intoxication, presenting with altered consciousness, severe high-anion gap metabolic acidosis and very high lactate levels. Rapid recognition of this toxidrome allowed for early initiation of antidote (fomepizol) to block further enzymatic conversion to toxic metabolites and early hemodialysis ensuring rapid elimination of the toxic alcohol and its metabolites, resulting in full clinical and biochemical recovery of a potentially life-threatening intoxication

Posters**P 37.****Management of pulmonary interstitial emphysema in a neonate requiring prolonged respiratory support**

E. Vandendriessche, C. Theyskens

*KUL*Background

Pulmonary interstitial emphysema (PIE) is a serious condition mainly seen in mechanically ventilated preterm infants with respiratory distress syndrome. We present a case of localized PIE with cardiomeastinal shift requiring prolonged supplemental oxygen and noninvasive ventilation (NIV). Conservative therapy is generally accepted as initial management in PIE, especially if the patient has no or few symptoms. In our case, cystic lesions were however growing over a prolonged period with persistent moderate respiratory distress and no opportunity in reducing respiratory support. Therefore, a review of literature on the therapeutic options in PIE is performed and discussed.

Methods

A literature search in Pubmed, Cochrane and Embase was performed.

Results

26 studies were included, predominantly single-case studies. The majority of case reports discuss very low birth weight infants born before the gestational age of 30 weeks, suffering from unilateral PIE which could be managed conservatively. Non-surgical treatment includes decubitus positioning with the affected side down, and in ventilated patients selective intubation of the main bronchus on the uninvolved side or bronchial occlusion on the affected side. High-frequency ventilation has been shown to be superior to conventional ventilation. Percutaneous catheter drainage of an enlarging pneumatocele is also described. When conservative management fails, surgery (i.e. lobectomy) should be considered. The increasing size of lung lesions over a prolonged period in symptomatic patients is mentioned as an indication for surgery in literature, yet no clear statement about the recommended duration of an expectant management is made. Our case shows that localized PIE, even with cystic lesions growing for more than 5 weeks, causing cardiomeastinal shift and requiring prolonged supplemental oxygen and NIV for up to 9 weeks, can be managed conservatively.

Conclusions

Current knowledge about the treatment of PIE is mainly based on individual case reports, making it difficult to formulate general recommendations. Risks inherent to invasive procedures have to be weighed against the chance on spontaneous resolution. Our case shows that localized PIE, even with cystic lesions growing for several weeks, causing cardiomeastinal shift and requiring prolonged supplemental oxygen and NIV, can be managed conservatively.

P 38.

Neonatal unexplained HUS treated with complement inhibitor Eculizumab

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*ULB – ERASME, ULB - HUDERF*Background

Hemolytic uremic syndrome (HUS) is rarely diagnosed in the perinatal period. It may present similarly to perinatal asphyxia (PA), with multi-organ failure (MOF), disseminated intravascular coagulopathy (DIC) and platelet consumption.

Case description

A boy was born at 35 weeks by caesarean section due to placenta praevia with normal birth weight, and good Apgar scores. At birth, he presented normal blood gas analysis and moderate anemia. Soon anemia worsened complicated by hypotension, deteriorating oxygenation, and severe metabolic acidosis. A blood transfusion was administered and acidosis rapidly corrected. Neurological examination was reassuring. Ten hours later he developed MOF with severe renal failure, haematuria, and thrombocytopenia refractory to platelet transfusion. He also developed coagulopathy with hypofibrinogenemia, and high D-dimer level (>35.000 ng/ml FEU). He was treated with daily fresh frozen plasma (FFP) with unsuccessful results. In addition to hematuria, hemobilia was observed. No infection causes were found. Serum lactate was moderately elevated, haptoglobin non-measurable, and schizocytes present. Cobalamin, ADAMTS 13, CD46, ANCA, FAN and anti-FH antibodies levels were within the normal levels for the age. Factors C3, C4 and B, measured 48 hours after FFP administration, were extremely low, while factors I and H concentrations on the lower range of normal values. HUS was suspected and eculizumab administered on day 8. Kidney function and coagulation parameters improved in 3 and 5 days respectively. No genetic causes of HUS could be found. He was discharged home at day 29 of life with still decreased renal function and developing cholestasis.

Conclusion

Perinatal HUS and PA may present with a similar clinical picture. In front of an atypical PA presentation, complicated by anemia, renal failure, hematuria and transfusion refractory thrombocytopenia, HUS should be suspected. This is the first report of a newborn with neonatal HUS of unknown origin successfully treated by Eculizumab as early as 8 days of life.

Posters**P 39.****A rare case of antenatal hepatomegaly: congenital leukemia in a newborn with Down syndrome.**

S. Faggetter, G. de Crombrughe, I. Van Herreweghe, M. Tackoen, A. Amoruso, D. Avino, A. Ferster, A-B Johansson

*ULB - HUDERF, CHU Saint Pierre*Introduction

Fetal hepatomegaly is a specific finding of severe anemia, infection, metabolic disease or myeloproliferative disorder. When associated with Down syndrome (DS), fetal hepatomegaly is suggestive of hematological disorder. Patients with DS have an increased risk of developing neonatal myeloproliferative disorders such as transient abnormal myelopoiesis (TAM) or acute leukemia. Congenital leukemia is an extremely rare condition, often of myeloid origin. Acute megakaryocytic leukemia (AMKL) is a type of acute myeloid leukemia (AML) accounting for 50% of the AML in children with DS. We report a case of acute liver failure in a newborn with AMKL and trisomy 21.

Case report

A male infant born at 37 weeks gestation from a poorly followed pregnancy was admitted to the neonatal intensive care unit for respiratory distress at birth. No anomaly was described on the 2nd and 3rd trimester ultrasound but an isolated hepatomegaly associated with heterogeneous liver architecture was described a few days before birth, with no further investigation. No antenatal screening for chromosomal anomalies was done. At admission the newborn presented phenotype of DS, later confirmed by karyotyping, abdominal distention and hepatomegaly. The blood test revealed anemia (hemoglobin 13.4g/dL), thrombocytopenia (89000/ μ L) and leucocytosis (76960/ μ L) with 54% blasts, suggestive of AMKL M7 on immunophenotyping. Low dose cytarabine was started on day 1 and white blood count decreased. Despite chemotherapy and conservative treatment, the newborn developed progressive hepatic failure with conjugated hyperbilirubinemia, hyperammonemia, severe coagulation disorder and ascites, and he died on day 4.

Discussion and conclusion

Congenital leukemia is a rarity that can be associated with chromosomal defects. Neonates with DS have a propensity to develop TAM, which is usually self-limiting and resolves spontaneously in ≤ 3 months. Approximately 10% will however present severe disease and die from hepatic or multi-organ failure, probably due to blast cell infiltration. Despite the better prognosis of AMKL in DS patients, due to higher sensitivity to cytarabine caused by increased expression of a gene localized on chromosome 21, congenital leukemia remains a condition with very poor prognosis, particularly when associated with liver failure. Isolated hepatomegaly can be the only ultrasonographic antenatal finding in these patients and should therefore be investigated.

P 40.

Pitfalls of pleural drainage in extremely preterm neonates

F. Piersigilli, C. Hocq, B. Van Grambezen, K. Carkeek, O. Danhaive

*UCL Saint-Luc*Introduction

Pneumothorax is a major complication of mechanical ventilation in extremely preterm infants with respiratory failure. Chest tube drainage is an effective treatment for symptomatic pneumothorax. Chest tube placement may also be required for pleural effusion. Complications can arise from the placement of a chest tube, as infection site and pulmonary infections, phrenic nerve paralysis or hemorrhagic effusion. The pigtail catheter is a less invasive alternative to traditional chest tube insertion, especially in preterm neonates. We describe two cases of unusual complications associated with drainage with pigtail catheters in two preterm infants.

Case 1

A male infant with Trisomy 21 was born at 30 weeks gestational age. The baby was born with hydrops and the first X-ray highlighted a right pleural effusion and a duodenal atresia. A chest tube was placed to drain the effusion. The day after the placement, there was the appearance of a green liquid from the drain, resembling bile. A gastric opacification followed by and a CT scan were performed and a gastropleural fistula was supposed. To confirm the diagnosis we injected 5 ml of methylene blue in the stomach, which was short after found in the pleural drain. The gastropleural fistula was eventually generated by the drainage, due to a perforation of the stomach by the drain. After 10 days the fistula resolved spontaneously.

Case 2

A female infant was born at 24 weeks gestational age. She was intubated and ventilated due to respiratory failure. At 48 hours of life a symptomatic pneumothorax developed. A chest tube was placed on 10 cm H₂O suction for continuous pleural drainage. Nevertheless after the drainage, although baby adequate positioning and high frequency oscillation, the pneumothorax persisted. Chest tube was replaced twice, but the resolution of the pneumothorax was very transient and symptomatic pneumothorax persisted for more than 10 days. In the suspicion of a lung injury, the baby underwent surgery. A lung perforation was found and sutured, with resolution of the pneumothorax. The perforation was probably provoked by the drain at the moment of the first pleural drainage.

Conclusions

Although pleural drainage is a life-saving procedure, it is important to keep in mind that it can have severe complications. The use of a pigtail catheter is considered a less invasive technique, but it has to be used carefully in preterm neonates, as it can give rise to life threatening complications.

Posters**P 41.****Determination of relative recovery and optimal calibration technique for piperacillin-tazobactam: an in vitro microdialysis study.**

E. Hermans, J. Vande Walle, P. De Paepe, M. Devreese, P. De Cock

*Universiteit Gent*Background/aims

The mortality rate of children with severe sepsis and septic shock remains high. It is hypothesized that the treatment failure in these patients is partly due to suboptimal penetration of antibiotics from the blood into the tissue. Research on the antibiotic tissue disposition of septic children is very scarce. Microdialysis (MD) is currently the gold standard for measuring unbound drug concentrations in the interstitial fluid (ISF) of tissue. In MD, a small probe consisting of a semipermeable hollow fiber membrane is implanted into the target tissue. The fraction of the drug in the ISF that actually crosses the membrane is called the relative recovery (RR). Different calibration methods can determine the magnitude of this RR. Preceding in vivo studies, an in vitro phase is needed to finetune several experimental factors to ensure optimal RR. In this in vitro study the RR of piperacillin-tazobactam (PTZ) was determined and the suitability of two calibration techniques (retrodialysis by drug and retrodialysis by internal calibrator) were evaluated for this drug.

Methods

The experimental set-up consisted of one type of MD probe, fixed temperature and fixed perfusion flow rate. The RR was determined in the context of different PTZ concentrations and at different time points. A RR of at least 20% is recommended to be able to obtain reliable results in in vivo studies. Penicillin was chosen as the internal calibrator.

Results

The RR of PTZ was sufficiently high and invariable in time and at different drug concentrations (range: PIP 34.8–55.7 %; TAZ 45.8–70.3%). During retrodialysis the RR of PTZ (range: PIP 39.3-53.7%; TAZ 50.4-64.6%) and penicillin (range 45.9-64.5%) were in the same order of magnitude as during the forward dialysis of PTZ.

Conclusions

For PTZ, it can be concluded that MD is a reliable measuring technique. Both retrodialysis by drug and retrodialysis by internal calibrator (penicillin) can be used as calibration methods. These results will be applied in soon to start in vivo pharmacokinetic studies that will investigate the tissue pharmacokinetics of PTZ in critically ill children and in a juvenile pig model of pediatric sepsis

LO 8.

SeroCovid<19: prospective seroprevalence monitoring reveals substantially reduced SARS-CoV-2 infection rate among tertiary pediatric patients

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UZ Gent

Background

Coronavirus disease 2019 (COVID-19) is a clinically heterogeneous entity with several host, environmental, and viral risk factors identified to date. COVID-19 in children generally manifests asymptomatic or mild. Pediatric patients with chronic medical conditions are potentially at increased risk for severe COVID-19, although convincing data is lacking. Among these children, we aimed to study COVID-19 incidence and disease severity.

Methods

We initiated a prospective cohort study with longitudinal (every 3-6 months) quantification of serum SARS-CoV-2 IgG (Abbott) and questionnaires in pediatric patients (0-18y) with chronic medical conditions in follow-up at Ghent University Hospital, a tertiary referral center. This interim analysis incorporates inclusions from Nov 1, 2020 until Jan 14, 2021.

Results

Here, we present data of the first 250 included patients (mean age 10.4y). Chronic conditions required follow-up at departments of immunology (26.4%), nephrology (16.7%), rheumatology (15.4%), gastroenterology (14.5%), endocrinology (8.8%), pulmonology (8.8%) or others (9.3%). Chronic immunosuppressive drugs were used in 35.6%. Inborn errors of immunity, possibly affecting antibody responses, were present in 18.0%. Restriction from school or daycare centers (additional to general lockdown and school closures) was documented in 35.1% and 10.7%, before and after the summer break, respectively. Since the start of the pandemic, close contacts with proven COVID-19 were known in 21.0%.

An overall SARS-CoV-2 seroprevalence of 6.0% (15/250) was found, substantially lower than concurrently reported in Belgian blood donors and health care workers (14.4% in Nov 2020), and on the lower end compared to healthy children measured prior to the second epidemic wave in Flanders (4.4-14.4% in Sept-Oct).

Asymptomatic COVID-19 was documented in 7/15. One patient with cystic fibrosis (9y) required hospitalization. All other cases experienced mild COVID-19, at most. Of seropositive cases, 5/15 received immunosuppression, all experiencing mild infection.

Conclusion

A strikingly low SARS-CoV-2 seroprevalence was found in our cohort. Despite no supplementary preventive measures were imposed for the majority, this cohort seemed additionally shielded from SARS-CoV-2. Asymptomatic COVID-19 was present in half of cases. These findings may be of importance for ongoing and future protective measures, clinical management, and vaccination strategies in these populations

Long Oral Presentation**LO 9.****Beyond culture: the Molecular Assessment of Thoracic Empyema (MATE) study**

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Introduction/Aim

Pleural empyema is a serious complication of bacterial pneumonia. In children, most cases are caused by *S. pneumoniae* (SPN), *S. pyogenes* (GAS), *S. aureus* (SA) and *H. influenzae* (HIN). Pleural fluid (PF) culture has low sensitivity, usually failing to confirm a bacterial cause, in part due to antibiotic pre-treatment. Lack of identification of the causative organism leads to prolonged empiric broad-spectrum antibiotic, increasing the risk of adverse drug effects including antimicrobial resistance. The aim of the MATE study is to validate a multiplex qPCR targeting the four main bacteria responsible for pleural empyema and assess its potential clinical impact.

Method

We are conducting a prospective observational study across two tertiary paediatric hospitals, with the goal of recruiting 125 paediatric patients with pneumonia complicated by pleural empyema requiring PF drainage. PF samples are tested using a novel multiplex qPCR targeting SPN, GAS, SA and HIN. Conventional diagnostic microbiological results will be recorded, to compare the performance of our multiplex qPCR against culture. Clinical information will be recorded to assess the potential impact of this diagnostic test on clinical management.

Results

Following extensive pre-clinical work to establish the assay, we piloted the multiplex qPCR on PF samples collected from the first 43 participants enrolled (median age 3.2 years). All participants received intravenous antibiotic treatment prior to PF collection. The qPCR detected a bacterial cause in 41/43 (95%) PF samples: SPN was detected in 33/41 (80%), GAS in 5/41 (12%) and SA in 1/41 (2%). In comparison, 8/43 (19%) samples had a bacterial species detected by culture, among which the qPCR produced matching results in 8/8 (100%).

Conclusion

We have developed a multiplex qPCR targeting the four main bacteria responsible for pleural empyema in children. Preliminary data indicate that our multiplex qPCR assay improved identification of the bacterial cause of empyema when compared with traditional culture-based methods. We will further assess its performance and its potential clinical impact as a novel molecular diagnostic allowing for efficient antibiotic stewardship and improved clinical outcomes for patients

SO 26.

Interactions between Group A Streptococcus CRISPR-Cas systems and prophages in virulence

L. Schiavolin, P. Smeesters, A. Botteaux

*ULB*Background/Aims

Group A Streptococcus (GAS) are Gram-positive bacteria responsible for superficial (like pharyngitis and impetigo) to severe (like necrotizing fasciitis) infections. Emergence of epidemic GAS lineages is linked to the acquisition of new virulence traits encoded by virulence genes. These genes are horizontally acquired by different mechanisms (HGT) which mainly involve mobile genetic elements like temperate phages. These are viruses of bacteria that can stably integrate into the bacterial genome, as prophages using integrases, and increase the fitness of their host. In GAS, these prophages carry toxins like DNases and superantigens which promote infection. HGT is also one of the primary mechanisms of antibiotic resistance spread. Bacteria have also to face predatory phages (lytic) which are able to kill them. Therefore, bacteria have notably evolved 'adaptive' immune systems to prevent phages infection. The CRISPR-Cas systems track record previous infections by integrating fragments of phage genomes and degrade specifically infecting phages carrying these fragments. GAS can carry simultaneously up to two CRISPR-Cas systems (type I-C and II-A). Although these systems are present, prophages are found in GAS genome suggesting that either they are not fully functional or phages have evolved counter-defence to bacterial immunity. Here, we want to address the link between the presence of the two CRISPR-Cas systems and the numbers of prophages in GAS genome to determine their impact on virulence.

Methods

We identified and annotated correctly the prophages of <200 reference GAS genomes. We generated a tree of 462 integrases and identified 17 distinct families, i.e. that cannot be found simultaneously in a same strain. We used the consensus sequences of these 17 integrases as a proxy to count the number of prophages using DNA-seq reads from >2000 draft genomes. Using the same approach, we also determined the presence of CRISPR-Cas systems (type I-C and/or II-A) and potential phage-encoded resistance genes.

Results

The CRISPR I-C system seems to be functional in GAS. However, the presence of specific genes in prophage genomes seems to inactivate this immune system and allows the integration of more prophages.

Conclusion

We have identified potential phage-encoded anti-CRISPR genes in silico. Now, we will move forward to characterize their role in prophage-GAS interactions to further understand new virulence traits acquisition in GAS.

Short Oral Presentation**SO 27.****Multiple breath washout measurement in patients with CDH at school age compared to Chest CT score and spirometry**

M. Dirickx, F. Vermeulen, M. Boon, A. Debeer, M. Proesmans

*UZ Leuven*Introduction

Data on long term structural and functional lung abnormalities in survivors of congenital diaphragmatic hernia (CDH) are scarce. The purpose of this follow-up program of CDH survivors is to assess the structural lung sequelae with chest CT scoring at 1 year of age and again at school age with additional lung function testing and exercise testing. Lung function testing includes multiple breath washout testing (MBW) with nitrogen with measurement of the lung clearance index (LCI).

Aim

The aim is to present some preliminary data on MBW-LCI results in our cohort.

Methods

Prospective, clinical follow-up program of CDH survivors at the University Hospital of Leuven of babies admitted to our NICU, with a pre- or postnatal diagnosis of CDH between January 2007 and August 2015. Clinical follow-up includes a low dose chest CT at the age of one year and at school age (age 7-10 years). Scanning was carried out during quiet breathing and with sedation at age 1 year. The CT images were anonymized to allow blinded scoring with an adapted CT scoring system based on the revised Auckland score for chronic lung disease of prematurity (CLD).

At the second follow-up, spirometry, nitrogen MBW, as well as exercise testing (six minute walk test and a shuttle run) are performed.

Results

Thirty-five patients have been included so far of which 22 reached school age. LCI was abnormal in 13/18 children. LCI correlates significantly with FEV1% ($r = -0.688$, $p = 0.002$) and to a lesser extent with FVC% ($r = -0.469$, $p = 0.05$).

All patients with abnormal FEV1% ($n = 10$) also had an abnormal LCI. Of the 8 patients with a normal FEV1%, 5 had an abnormal LCI.

A significant correlation was found between the CT score at age 1 and the LCI (Pearson $r = 0.477$, $p = 0.042$), the FEV1% ($r = 0.506$, $p = 0.038$) but not with the FVC% nor the exercise tests. The 6 min walk test % did not correlate with lung function parameters. A significant correlation was found between the shuttle run level and the FEV1% ($r = 0.651$, $p = 0.001$) but not with the LCI.

Short Oral Presentation**Conclusion**

In this small cohort of children with CHD, MBW-LCI is abnormal in the majority of children and may be more sensitive to detect lung disease compared to FEV1%.

CT score on chest CT's performed at age 1 correlate with LCI and FEV1% at school age. Data needs to be confirmed in a larger patient group.

Short Oral Presentation**SO 28.****Poliomyelitis Surveillance in Belgium 2003 to 2018: Under reporting of Acute Flaccid Paralysis**

C. Wyndham-Thomas, N. Hammami, C. Schirvel, R. Mahieu, G. Top, M. Lesenfants, E. Wollants, S. Quoilin

Sciensano, Agentschap Zorg en Gezondheid, Agence pour une Vie de Qualité, Commission communautaire commune, Centre National de Référence des entérovirus, UZ Leuven

Background

2019 is marked by a worldwide increase in the circulation of wild and vaccine-derived poliovirus, with 19 countries affected compared to only six in 2018. Countries where polio is eradicated, including Belgium, need a strong surveillance system able to confirm the absence of the virus and promptly detect polio reintroduction.

The gold standard for polio surveillance is the notification and microbiological exclusion of the virus in all cases of acute flaccid paralysis (AFP) occurring in <15 year olds, regardless of etiology. AFP is a clinical syndrome with rapid onset of weakness without spasticity or other signs of CNS motor tract disorder. Causes of AFP are multiple, including neurotropic infections (e.g. polio/non-polio enterovirus), Guillain-Barré Syndrom (GBS) and acute myelopathies. According to WHO indicators, polio surveillance systems must annually detect 1 AFP case/100 000 children aged <15 years in order to achieve sufficient sensitivity.

We summarize the results of AFP surveillance in Belgium since its implementation.

Methods

AFP cases in <15 year olds registered from 2003 to 2018 by mandatory notification and/or by voluntary notification via PediSurv (Pediatric Surveillance network) were analyzed. Minimal Hospitalization Records (MHR) were consulted to obtain number of hospital stays for GBS from 2003 to 2017, using codes ICD-9 357 and ICD-10 G610.

Results

A total of 77 AFPs were notified, with a median of 4 cases per year (range 1 to 9) or 0.2 cases per year/100 000 children aged <15 years. 42 were reported in Flanders, 20 in Wallonia, 13 in Brussels (2 region unknown). Only 2 cases had the required polio-surveillance microbiological workup i.e. 2 stool samples collected 24-48h apart & within 14 days of disease onset.

Causal disease was known for 76 AFPs notified, with GBS as the most frequent diagnosis (n=63; 83%). That is, in average, 3.9 cases of GBS notified annually. By contrast, MHR indicate that in <15 year olds, a median of 24 hospital stays per year had GBS as primary cause of hospitalization (range 12-40).

Conclusions

Belgium remains 5 fold below the WHO indicator for sensitive surveillance, and the majority of GBS remain un-notified. Due to weaknesses in AFP surveillance, Belgium is considered at intermediate-risk for polio. Pediatricians should be better informed, during medical training and later, on the reason and modalities for AFP surveillance, as well as their central role in reporting notifiable diseases.

SO 33.

Immunoglobulin g subclass and specific polysaccharide antibody deficiency: a clinical and immunological profile in a tertiary cohort

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UZ Gent

Background and Aims

Immunoglobulin(Ig)G-subclass deficiency (IgGSD) and specific polysaccharide antibody deficiency (SPAD) are two common antibody deficiencies, mainly associated with recurrent respiratory tract infections (RTI). The clinical variability and effect of therapy in a large cohort is not well studied.

Methods

A retrospective observational study of children and adults with IgG2SD and/or IgG3SD and/or SPAD and normal total IgG.

Results

In total, 105 patients (age 2y-79y (mean 24y), 53% pediatric, 44% male) were included. To our knowledge, this is the largest cohort of IgGSD and/or SPAD patients studied. Isolated IgG3SD was both in children (16/56;29%) and adults (30/49;61%) most prevalent. Of all IgG2SD(+/-SPAD) cases, 85% were found in children (22/26). In total, isolated SPAD was less frequent (16/105;15%) than IgGSD+SPAD (21/105;20%).

Recurrent upper and lower RTI (76-82% and 38-57% respectively) were most frequently observed, similar in all groups. Gastrointestinal infections (42%;P=0.017) and fatigue (45%;P=0.005) were associated with IgG3SD. Bacterial skin infections (38%;P=0.024) were frequently observed in SPAD. Autoimmunity (25%;P=0.001), lymphadenopathy (71%;P=0.002) and fatigue (59%;P<0.001) were significantly common in adults compared to children. Autoimmunity was not associated with aberrant B-cell maturation, only present in 2.8%. IgG2SD patients (69%;P=0.046) and children (73%;P<0.001) were hospitalized more.

Monocytopenia was repeatedly observed (32/105;30%), predominantly in IgG2SD (15/31;48%) and persistent, without effect of Ig replacement therapy. In patients with Ig replacement, bacterial infection rate reduced significantly in 82%, although low IgG3 persisted.

Conclusions

RTI remain the hallmark presentation in IgGSD and/or SPAD. Our cohort revealed remarkable findings, such as frequent autoimmunity and monocytopenia. Long-term multi-centre studies are needed to better characterize these patients

Short Oral Presentation**SO 34.****Epidemiological and clinical features of malaras imported to CHU Saint-Pierre between 1998 and 2017**

M. Leys, C. Martin, S. De Wit

*ULB-CHU Ambroise Pare, CHU Saint-Pierre Hospital*Background

Malaria is a parasitic infection caused by a protozoan of the genus Plasmodium, responsible for a significant number of infections and deaths annually in endemic areas. However, non-endemic regions such as Europe are diagnosing a significant number of imported malaras, indicating a new challenge in terms of malaria prevention and management.

Methods

We conducted a descriptive retrospective study of a cohort of malaras diagnosed at CHU Saint-Pierre from 1998 to 2017. Epidemiological and clinical data were collected by studying each medical record.

Results

A total of 1000 malaria episodes were studied, for a total of 985 patients. A net increase in the number of cases has been observed over the years. Plasmodium falciparum is most often diagnosed, mostly from Central and West Africa. The proportion of severe malaras is 15% and the mortality rate is low. A large number of cases have been diagnosed in "visiting friends and relatives (VFR)" patients (50%).

Conclusion

As in other European countries, we have seen an increase in the number of imported malaria cases in recent years. This underlines the importance of a national surveillance of imported malaras in Belgium, and the reinforcement of prevention messages towards groups at higher risk of malaria (VFR) and at higher risk of severe malaria (immunocompromised patients).

SO 35.

Analysis of the consumption of drugs prescribed for the treatment of asthma in Belgian children

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Mutualités Libres - Onafhankelijke Ziekenfondsen, ULB

Background

Asthma is one of the most common chronic diseases in the world among children. It's diagnosis in (preschool-)children is difficult. The purpose of this study was to analyze the consumption of asthma medications in order to investigate asthma in children (2-18 years).

Methods

A retrospective study using anonymized administrative data for 2013-2018 from the third Belgian health insurer, Mutualités Libres - Onafhankelijke Ziekenfondsen (MLOZ), was conducted. To identify asthmatic children, two approaches based on drugs dispensations were used: (1) at least one asthmatic drug dispensation and (2) at least 2 asthmatic drugs dispensations with a minimum of 30 days between 2 purchases. This second approach allows to identify children with a more severe clinical picture. The main drugs selected were: SABA (Short-Acting Beta Agonists), SAMA (Short-Acting Muscarinic Antagonist), ICS (Inhaled Corticosteroids), LABA (Long-Acting Beta Agonists) and drug combinations of ICS-LABA and SABA-SAMA.

Results

In 2018, 12.9% of children received at least one asthma medication and 4.4% received at least two packages with a minimum of 30 days between purchases. Preschool children (2-6 years) were 3 times more likely to take asthma medication than older children (7-18 years). Only a small part of children taking asthma medication in 2013 will continue to use them for the next 5 years (5.9% to 16.7% according to age). ICS, in combination or not with LABA, were the most dispensed drugs among children. Indeed, 60 to 70% of children who take at least 1 asthma medication received ICS. More than half (50.4%) received SABA and one third (33.6%) SAMA. SABA as monotherapy was used by 22.7% of preschool children. These 2 bronchodilators were mostly administered by nebulizer in young children (72.6%) and by inhalator in older children (65.3%).

Conclusion

Caution is to be used in interpreting results as anti-asthmatic drugs can be used, for example, to treat other diseases or to test the hypothesis of asthma. Most children are taking ICS according to the GINA (Global Initiative for Asthma) 2019 guidelines. In contradiction to these guidelines, SABA was sometimes used in monotherapy. The high use of SAMA, only rarely mentioned in guidelines, can be questioned as well. Finally, high rates of nebulization in young children were observed, despite the recommendations to use an inhaler with a spacing chamber as much as possible.

Short Oral Presentation

SO 36.

Tonsillar obstructive hypertrophy and recurrent tonsillitis in children: what is the role played by the tonsillar carriage of *Streptococcus pyogenes*?

A. Vandevoorde, D. Lakhroufi, G. Botquin, V. Delforge, A-L Mansbach, A. Botteaux, P. Smeesters

*ULB - HUDERF*Background/Aims

Streptococcus pyogenes (Group A *Streptococcus*, GAS) is a human pathogen responsible for bacterial tonsillitis in children. It is also part of the commensal flora of the pharynx with ~ 20% of children carrying it asymptotically. We aim to investigate the potential role of GAS tonsillar carriage in chronic obstructive tonsillar hypertrophy and recurrent tonsillitis in children.

Methods

Monocentric prospective study including children undergoing tonsillectomy for either Obstructive Sleep Apnea Syndrome (OSAS) or recurrent tonsillitis. Microbiological throat swabs, superficial and deep tonsil fragments were collected during tonsillectomy and cultured for GAS. Multiple colonies were stored for each GAS-positive clinical sample. GAS strains were characterised by emm-typing and antimicrobial susceptibility to 5 antibiotics. The presence of GAS, the emm-type and the antibiotic susceptibility pattern were then compared inter-individually (OSAS vs recurrent tonsillitis) and intra-individually (tonsil surface vs superficial/deep tonsil crypts).

Results

Fifty-eight children were included (30 OSAS, 28 recurrent tonsillitis) and GAS was found in 32.8% of them. GAS was more frequently recovered in OSAS (40%) than in recurrent tonsillitis (25%). The culture of GAS carriers was always positive in every microbiological sample (tonsil surface, superficial and deep tonsil crypts). Ten different emm-types were identified amongst the cohort, with a preponderance of emm-type 1. No intra-individual diversity of emm-type was observed. On the contrary, 36.8% of the cases displayed intra-individual heterogeneity in their antibiotic profile, with some strains carrying antibiotic resistance genes amongst mostly sensitive strains.

Conclusion

The preliminary results of this ongoing study show a high GAS tonsillar carriage rate with a particularly high rate in OSAS, suggesting a potential role of GAS in this syndrome. About one third of GAS-positive cases shows a heterogeneity of antibiotic resistance profiles, making resistant strains difficult to detect but likely to be associated with clinical treatment failure. The bacterial virulence profile of these strains will be analysed to develop better prevention and/or treatment of chronic obstructive tonsillar hypertrophy and recurrent tonsillitis by specifically targeting their causal pathways.

PW 7.**Immunization budget for Belgium: Horizon scanning using an Immunisation Planning Tool**

A. Bento-Abreu, J. Vandenhoute, N. Dauby, B. Merckx, G. Bencina

MSD

Background/Aims

Perspective is missing on available or required budget for new or expanded vaccination schemes. Using an Immunisation Planning Tool (IPT) we aimed at quantifying the cost-per-capita of vaccines and of life-long immunization of currently implemented vaccine schedule but also upcoming vaccine.

Methods

The IPT models budgetary impact based on published demographic data, official vaccination schemes, vaccination coverage rate and cost input assumptions including vaccine list prices as conservative overestimation.

The base case preliminary results include 7 or 8 vaccines protecting against 13 or 14 infectious diseases as covered by the vaccination schemes of the Wallonia-Brussels Federation and Flanders, respectively (polio, diphtheria, pertussis, tetanus, hepatitis B, Haemophilus influenza type B, meningococcus C, pneumococcal disease in children, human papillomavirus, measles, mumps, rubella, and influenza in nursing-home residents (Flanders only)). Scenario analyses include new available and registered vaccines.

Results

The life-long immunization cost-per-capita in full compliance with current NIP in Flanders and Wallonia-Brussels is 930€ for men and women, and 1956€ for men and 2063€ for women if the vaccines for rotavirus, Men B, Men ACWY, varicella, influenza, pneumococcal and zoster for all adults 65+ are also included.

The per-capita spending on vaccines accounts for 9.5€/capita/year in Flanders and 8.5€/capita/year Wallonia-Brussels, according to their current immunization schemes. In Flanders, this cost increases to and to 17.9€/capita/year if all new available vaccines are included.

Total regional vaccines budgets for Flanders and Wallonia combined equal less than 0.4% of the federal health care budget in 2020. The federal budget for reimbursed vaccines was separately estimated around 32 Mio€.

Conclusion

The life-long immunization cost-per-capita is in the range previously reported data from other European Countries. The IPT results and scenarios allow the assessment of the current level of investment in vaccines and provide baseline quantitative perspective for immunisation horizon scanning discussions with policy makers.

Poster Walk**PW 9.****Clinical description and outcomes of children with invasive group A Streptococcal disease**

E. Thielemans, J. Oliver, A. McMinn, C. Baker, P.N. Britton, J. Clark, H. Marshall, C. C. Blyth, J. Francis, J. Buttery, P.R. Smeesters, N. Crawford, A.C. Steer

ULB, Royal Children's Hospital, Victoria

Background

Invasive group A Streptococcus disease is a severe infection with a high case fatality rate, estimated to cause more than 150, 000 deaths per year worldwide. There are few data on its short and longer term outcomes, especially in children. The clinical presentation of this infection is variable and early diagnosis can be challenging. The aim of this study was to assess the clinical presentation, management and the short and longer term outcomes of invasive group A streptococcal disease in children in Australia.

Method

We undertook surveillance of children with laboratory confirmed invasive GAS disease admitted to tertiary children's hospitals between July 2016 and June 2018. We prospectively collected demographical and clinical data. We contacted these patients 6 months after discharge to assess longer term outcomes.

Results

We enrolled 181 children, aged from 7 days to 16 years. The principal site of invasion was the blood (126 children, 69.7%), but the most frequent clinical presentation was pneumonia (46 children, 25.4%). Twenty-six children developed streptococcal toxic shock syndrome (14.4%), and 74 had severe disease (40.9%) including 71 admitted to the intensive care unit. Five children died (2.8%). At discharge and six months, 29.3% and 15.2% of the children had persisting health problems, respectively.

Conclusion

Invasive group A streptococcal infection in Australian children is frequently severe and has a high long term morbidity burden, highlighting the need for strengthened clinical care pathways, epidemiological surveillance and improved prevention strategies.

PW 10.**Streptococcus pyogenes related hospitalisations in a single tertiary pediatric center in Belgium: overview over 14 year**

J. Van Vlaenderen, P. Schelstraete, J. Willems

UZ Gent

Background

Invasive Group A Streptococcus (IGAS) infections cause high morbidity and mortality. The main objective of this study is to describe the prevalence, clinical characteristics, morbidity and mortality about invasive streptococcal pyogenes infections in a tertiary pediatric center in Belgium.

Methods

Selection of the patients was done retrospectively by using the ICD coding system. Analysis of data of all pediatric patients (0 to 15 years old) admitted to the University Hospital of Ghent with IGAS infections from January 1st, 2005 till December 31st, 2018. For inclusion in the study, we used the definition of invasive group A streptococcal infections of CDC (Centers for Disease Control and Prevention). Demographic, clinical, biochemical, microbiological data were analyzed, together with complications, treatment and sequels.

Results

In total, 95 patients met the inclusion criteria of IGAS infections. Almost 50% of the patients was younger than 2 years. For five patients the IGAS infection was fatal (5.2%), all of them were 2 years old or younger. The main clinical diagnosis of all patients were severe soft tissue infections (30.2%), empyema (18.7%), complicated ENT (ear-nose-throat) infections (16.7%), toxic shock syndrome (19.8%), meningitis (2%) and osteomyelitis (2%). Thirty percent of children identified had preceding varicella infection, data about varicella vaccination lack. Sixty percent of the patients underwent surgery. Seventeen percent (n=16) were treated with vasoactive medication. Twenty one patients (n=21) were intubated, of which 14 patients five days or longer. Intravenous immunoglobulins were given to 13 patients (13.5%). More than half of the patients received clindamycine (n=51). Serotypes were available from 37 isolates, of which 17 isolates were emm type 1.0.

Conclusion

Health workers must have a high index of suspicion of an IGAS infection in patients with varicella. An IGAS infection knows a very rapidly course and can be fatal, especially in young children. Prompt initiation of therapy is most important. Varicella immunization could potentially prevent significant number of IGAS infections. Serotype Emm 1.0 was the micro-organism most often isolated.

Poster Walk**PW 11.****Para-pneumonic effusions in children: comparison between surgical drainage by video-assisted thoracoscopy and percutaneous drainage with urokinase**

V. Weber, H. Steyaert, D. Biarent, N. Lefèvre

ULB - HUDERF

Introduction

The treatment of complicated para-pneumonic effusions in children remains controversial. We decided to compare the effectiveness, safety and comfort of percutaneous drainage associated with instillation of urokinase to surgical drainage by video-assisted thoracoscopy (VATS) at our hospital in order to establish the most appropriate approach for our patients.

Methodology

Patients aged 1 to 17 years with a complicated para-pneumonic effusion were randomized in the percutaneous drainage with urokinase group or the VATS group.

The primary objective was the duration of the drainage in days. Secondary objectives included hospitalization, fever, intravenous antibiotic therapy, and oxygen therapy durations; as well as doses and duration of morphine sulfate; complications; three months post-intervention follow-up; and mortality.

Results

Our results are based on the intermediate analysis of the first 22 patients enrolled, 11 in each group. All the patients shared the same characteristics when enrolled. Drainage duration was similar in both groups (8 days [6-9] in the urokinase group vs. 5 days [4-10] in the VATS group, $p=0,27$). Secondary objectives did not differ between groups, though we found a tendency toward higher doses of morphine in the VATS group (13 mcg/kg/h [10-16,5] vs. 10 mcg/kg/h [10-10], $p=0,06$).

Conclusion

In our center, the effectiveness and safety of both treatments appear to be similar in children with para-pneumonic effusions. However, percutaneous drainage coupled with urokinase installation showed a tendency to be less painful than VATS.

PW 13.**Quantifying *Streptococcus pyogenes* during experimental human pharyngitis**

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Background

Streptococcus pyogenes (group A *Streptococcus*, GAS) causes a spectrum of clinical manifestations responsible for more than 500,000 deaths per year worldwide. Development of a vaccine against GAS has been impeded, in part due to the limitations of current understanding of host-pathogen interactions.

A multinational collaborative group has established the Controlled Human Infection for Vaccination Against *Streptococcus pyogenes* (CHIVAS) model as a safe and reliable experimental human pharyngitis platform to accelerate vaccine development. Carefully screened healthy adult volunteers had well characterised and meticulously manufactured doses of emm75 GAS applied by swab to their pharynx. The volunteers were closely monitored and throat swabs taken regularly from before the challenge, during an inpatient period and at periodic outpatient visits for six months afterwards. We describe the molecular methods used to quantify the bacterial load in pharyngeal swabs during the course of infection and compare these results to standard culture-based methods.

Methods

Two throat swabs were taken at each time point: one for standard microbiological culture (eSWAB™, Copan), and another was immediately placed into a commercially available nucleic-acid preservation medium (eNAT™, Copan) for downstream molecular analysis. DNA and RNA from eNAT™ swabs was extracted simultaneously using a single spin-column (RNeasy®, Qiagen) and the challenge strain was quantified using an emm75-specific quantitative polymerase chain reaction (qPCR). We assessed the quality of our extraction process and performance of the emm75 qPCR.

Results

The quality and yield of simultaneous DNA and RNA extractions was comparable to separate extractions, as tested by qPCR and reverse transcriptase-qPCR, respectively. Bacterial load determined by the emm75 qPCR correlated with semi-quantitative culture results. The probe-based qPCR for emm75 provided the same dynamic range and a similar limit of detection to the widely used speB qPCR, with sensitivity down to 33 copies of emm75 GAS genome. The emm75 qPCR assay was 100% specific against a panel of 15 other GAS emm-types.

Conclusion

We have used a fast and reliable dual extraction method to isolate high quality DNA to detect the emm75 allele expressed by the GAS challenge strain. This approach can accurately measure the pharyngeal load of GAS during experimental human pharyngitis and could replace culture in future challenge studies.

Poster Walk**PW 33.****Respiratory morbidity in children with Spinal Muscular Atrophy: a single-center retrospective cohort study**

T. Smeets, M. Boon

*UZ Leuven*Background/Aims

Spinal muscular atrophy (SMA), caused by degeneration of anterior horn cells in the spinal cord, leads to progressive muscle atrophy, weakness and paralysis. Based on the age of onset and maximum motor function achieved, SMA type 1 till 4 are distinguished. In SMA 1 and 2, restrictive lung disease is the most serious complication, because of impaired cough, hypoventilation, recurrent respiratory infections and over time respiratory failure and subsequent death. New treatment options (f.e. nusinersen) drastically improve motor milestones, but it remains unclear how well they would reduce the respiratory morbidity. In this study, we investigate the inherent respiratory morbidity in patients with SMA without major influence of new treatments, in particular in SMA 2.

Methods

A single center, retrospective cohort study was conducted at UZ Leuven. General and respiratory characteristics were collected from children with SMA in a longitudinal way and the retrieved data was analyzed using descriptive statistics.

Results

A total of 51 children with SMA was included; 9 SMA 1, 25 SMA 2 and 17 SMA 3. Of those with SMA 2, 20 had at least one episode of pneumonia, 24 suffered from respiratory tract infections, with 13/112 of the hospitalizations for respiratory infections needing intensive care unit (ICU) admission and 5/112 resulting in invasive ventilation. Pseudomonas Aeruginosa was cultured from respiratory samples in 5/25 patients with SMA 2, of whom two were chronically infected. Respiratory support with assisted cough, intrapulmonary percussion ventilation and non-invasive ventilation was needed in 18/25, 22/25 and 15/25 patients with SMA 2 respectively. Maintenance antibiotics (azithromycin or other) was used in 12/25 SMA 2. Most patients with SMA 1 died at young age because of abstinence of invasive treatment. Patients with SMA 3 infrequently had rare respiratory complications.

Conclusion

We showed that the respiratory morbidity of SMA is considerable, especially in SMA 2. With the advent of new therapies, a new phenotype of “treated SMA 1” arises, which might be more similar to SMA 2 if treated at young age, with fewer deaths. However, the respiratory burden in children with SMA 2 should not be underestimated as recurrent infection, hospitalizations on ICU and need for invasive and non-invasive ventilation is not infrequent. This phenotype will require intensive monitoring for respiratory complications and a pro-active treatment.

PW 34.

Identification of a subsegmental lobar collapse as an early radiologic sign of Primary Ciliary Dyskinesia

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Introduction/Aim

Establishing the underlying cause in a child with chronic suppurative lung disease (CSLD) is beneficial as it allows for targeted treatment and screening for associated complications. One cause of CSLD is Primary Ciliary Dyskinesia (PCD), however testing for PCD requires specialist expertise which is not widely available. Computed Tomography (CT) scans are commonly performed in the assessment of CSLD. If PCD specific signs on CT were identified it may help clinicians in deciding when to refer for specialist testing. One potential PCD specific sign we have observed is sub segmental lobar collapse in excess of changes in other lung areas. We aimed to assess if complete collapse of a lobe is commonly found in CT of PCD patients, and its eventual association with respiratory tract infections causative germs.

Methods

Fifty-eight CT scans from 42 PCD patients were analyzed, looking for the presence of this feature and its association to other signs commonly seen in PCD and CSLD: bronchiectasis, atelectasis, bronchial wall thickening, air trapping and mucous plugging. A sub segmental lobar collapse was noted as present or absent. The five other features were marked independently and for each lobe as absent, moderate if their extent was <50% of the lobe or severe if it was >50%. A subanalysis of CT from children only is planned, as well as a study of concurrent airway microbiology.

Results

A completely collapsed lobe was found in 25/58 CT scans. The associated changes were mild in 21/25 and severe in 4/25.

Conclusion

A completely collapsed lobe is found in 43% PCD CT scans, and frequently in patients with minimal or moderate associated changes. This data shows this sign is commonly found in PCD and future work will determine if it is a PCD specific sign by assessing whether it is also found in other CSLD processes, as well as analysing more scans from children with PCD to determine how early this sign develops, and evaluating a possible association with respiratory tract infections.

Poster Walk**PW 35.****Paediatric Enteric Fever in Brussels: a case series over 14 years**

V. Selimaj, P Lepage, Y. Miendje Deyi, W. Mattheus, M. Hainaut

*Clinique Saint Jean, ULB-HUDERF, UZ Brussel, Sciensano, ULB-CHU Saint Pierre*Introduction

Enteric fever (EF) is a major public health problem and a witness of the global health disparities. It is caused by *Salmonella enterica* serovar Typhi (*S.typhi*) and *Salmonella enterica* serovar Paratyphi A,B,C (*S.paratyphi*) and is estimated to infect 12-26 million persons per year, claiming up to 216.000 lives annually. The majority of the persons affected, are children.

Paediatric data on enteric fever in Europe are missing. In this context, a case series of enteric fever was analysed to describe the clinical, biological and microbiological characteristics as well as the preventative and diagnostic challenges identified in a paediatric population living in Brussels.

Methods

We carried out a retrospective observational study of all lab-confirmed cases of typhoid and paratyphoid fever in children aged 0-15 years at the Centre Hospitaliere Universitaire Saint Pierre and Hôpital Universitaire des Enfants Reine Fabiola, between January 2005 and December 2018.

We looked at variables such as age, gender, history of travel, number of consultations prior to diagnosis, length of hospitalisation; clinical features such as fever, cough, headache, gastrointestinal symptoms; and biological findings such as Haemoglobin, C-Reactive Protein, White Blood Cell counts, Neutrophils, Lymphocytes, Eosinophils and Platelet counts, sodium and Liver enzymes concentrations.

Results

There were 30 positive isolates of *S.typhi* and *S.paratyphi*: 27 patients had positive blood culture, 1 patient had positive bone drainage, 2 patients had positive stool culture (one patient had missing data, and was excluded). More than half the patients were females 17/29. The median age was 3,5 years (range 5 months to 14 years). Half of our patients had recently travelled to endemic areas. For 80% of the patients there was a delayed diagnosis. Eosinopenia was present in 93% of the cohort. None of the patients had received any EF preventive education or vaccination.

Conclusions

Enteric fever poses a diagnostic challenge to clinicians working in non-endemic areas. Clinicians must keep it high in the list of the differentials at the returning traveller. The presence of eosinopenia in a febrile patient coming from the tropics or subtropics, should raise suspicion of enteric fever. Travellers to endemic areas, particularly the visiting relatives and friends should be better educated and vaccinated for enteric fever.

P 42.

A clinical case study of the Sleep Apnea Syndrome in infants

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CHU de Liège, U Liège

Introduction

Sleep apnea syndrome (SAS) in infants is a sleep related breathing disorder. It involves reductions and pauses in breathing that occur during sleep. Partial reductions in breathing are called “hypopneas.” Complete pauses in breathing are called “apneas.” Apneas may be of central, obstructive or mixed origin. If left untreated SAS may have life threatening impacts on infants.

Clinical Case

Patient of 1 month and 21 days of life, presented at the emergency room for nasal congestion and cough associated with fever. Clinical examination revealed weight and height below the 50th percentile, axial hypotonia and Pierre Robin like features with retrognathia and ogival palate. Complementary ENT examinations, included a nasopharyngeal endoscopy which revealed a local edema and erythema of the arytenoid with an appropriate mobility of the epiglottic plane. During the period of observation and monitoring in the pediatric ward, an episode of tonico-clonic movement of the left lower limb and nocturnal episodes of hypoxemia were observed. A polysomnography was conducted that revealed an apnea to hypopnea index which was moderate (index of 25/hr). Further investigations included the coupling of polysomnography study to that of transcutaneous carbon dioxide emissions and as a result obstructive sleep apnea syndrome (OSAS) was the final diagnosis. A treatment by continuous positive airway pressure (CPAP) was indicated. The infant showed improved axial tonus and -nocturnal oxygen saturation.

Conclusion

Polysomnography combined with the study of transcutaneous carbon dioxide emissions is the most appropriate method for the diagnosis of SAS. Polysomnography provides an Apnea to Hypopnea Index that helps to categorize the types of SAS. Moreover, the use of the endoscopic visualization of the airway can determine the etiology of OSAS. CPAP remains the gold standard for the treatment of OSAS where surgery should be considered as an alternative if CPAP is ineffective, and if the clinical state of the child is adequate.

Posters**P 43.****Bacterial Meningitis: Atypical presentation.**

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CHU de Liège, U Liège

Pneumococcal meningitis is a serious illness that still occur despite vaccination campaigns, its incidence is estimated at 0,13cases/100 000. Despite established and effective antibiotic strategies, its mortality is estimated to be 10% and the complications rate 30%.

A 11-year-old boy presents to emergency room for frontal headache, vomiting and sudden decrease in hearing since that night. He has a loss of appetite since 48h. No notion of fever.

His familial history is uneventful as well as his personal medical history. No chronic treatment.

At admission: his cardio-respiratory parameters are stable, he has no fever.

At clinical examination: he has spatial and temporal disorientation, difficulty to communicate, and seems to have serious hearing loss. No meningeal signs. The rest of neurological examination is difficult to perform. Examination of the other systems is normal.

The blood test showed an increase of leukocyte ($31,06 \cdot 10^3/\text{mm}^3$) and CRP (110,8 mg/L).

The toxicology analyses were negative.

The brain scan showed pansinusitis but nothing which explained the symptomatology.

The Lumbar puncture comes back positive: leukocyte $3860/\text{mm}^3$, proteins 4286 mg/L, glucose 9 mg/dL.

The microbiological culture showed a *Streptococcus pneumoniae* (capsular serotype 12B).

Antibiotics by Claforan 200mg/kg/day and Vancomycine 40 mg/kg/day was started. We also added corticoide for hearing. Vancomycine was stopped after 4 days and Claforan was stopped after 10 days of treatment.

The first audiometry shows: hearing threshold: 115dB for right side, 93dB for the left side.

The RMN were reassuring without abscess or osteitis.

With the treatment, he partially recovered the hearing on the left side. A cochlear implant was placed on the right side.

Diagnosis of meningitis can be difficult. For young children the symptomatology is recognized as being often not specific, while in older children it's more obvious and more typical. For them, fever and deterioration of the general state are the main symptoms which precede the other more typical symptoms such as vomiting, headache, photophobia and stiff neck. In our case, the initial symptomatology was unspecific. This highlights the importance of remaining careful when confronted with acute neurologic defect, especially hearing loss.

In addition, our case illustrates the need for rapid treatment to avoid complications. The main neurologic complications are: seizure, sensorineural hearing loss, hydrocephalus and motor problems

P 44.

Incomplete Kawasaki disease, a diagnostic challenge : about a toddler with fever and pyuria

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Introduction

Kawasaki disease is a well known disease but, in young children, symptoms could be atypical leading to incomplete Kawasaki disease. The atypical presentation makes the diagnostic more challenging so that the treatment could be delayed with potential serious consequences.

Presentation

We report here the case of a 20-month old child sent to the pediatric emergencies by the general practitioner for fever and jaundice. She presented fever for five days (despite treatment by Amoxicillin for an acute otitis media), slight rash spontaneously disappearing, cheilitis and dry conjunctivitis. Criteria for Kawasaki disease were not fully met, a fourth criteria out of five being necessary. So we looked after the criteria for incomplete Kawasaki disease following the algorithm of the American Heart Association (AHA). The child met the criteria : fever for 5 days, CRP > 3mg/dL, anemia, elevated ALT level and urine > 10 WBC/hpf. Waiting for the urine culture, we started the antibiotics for a presumed acute pyelonephritis. On the next day, the urine culture proved sterile. Treatment started with intravenous immunoglobulins and acetylsalicylic acid in anti-inflammatory doses. The girl becomes afebrile 30 hours after the perfusion and the biology was really improved with a marked decrease of the inflammatory syndrome. The echocardiography was within normal limits. The acetylsalicylic acid treatment was reduced to anti-aggregating doses for six weeks (stopped thanks to a normal second echocardiography) and a regular echocardiographic monitoring was set up at 3, 6 and 12 months of evolution.

Discussion and conclusion

Incomplete Kawasaki disease diagnosis remains complicated and difficult to establish given the absence of a sensible and specific test. The AHA established a diagnostic algorithm in 2017. Using this algorithm could be really useful if we don't have all the criteria for typical Kawasaki disease.

Posters**P 45.****Case report: An 11 year old boy with atypical meningitis**

Z. Casier, S. Van Molhem, A. Verschelde, L. De Keyzer

AZ Damiaan, AZ Sint Jan

An 11 year old boy, with no pertinent medical history, is seen in the emergency department with persistent fever for 7 days, worsening headache and photophobia. Other symptoms over the past few weeks were general malaise, conjunctivitis and pharyngitis. He was seen by his own pediatrician 5 days prior to this visit with fever and general malaise, with no abnormalities on clinical exam. A blood draw showed negative inflammatory parameters (white blood cells (WBC) 5400/ μ L, C-Reactive Protein (CRP) 17.5 mg/L). Infectious serology was negative for *Mycoplasma Pneumoniae*, Ebstein Barr Virus and Cytomegalovirus. On clinical exam we saw a drowsy but alert boy who obeys to commands. Vital parameters are normal except for tachycardia (HF >120bpm, sat 100%, BP 101/74 mmHg). He has severe neck stiffness and photophobia. Internal exam shows diffuse crackles on the right lung without respiratory distress. A new blood draw reveals leukocytosis (13000 WBC/ μ L) without elevated CRP (11.7 mg/L). Given the prominent neck stiffness a CT-scan and lumbar puncture were done. The CT showed no abnormalities and no evidence for raised intracranial pressure. Examination of the cerebrospinal fluid (CSF) showed 930 WBC with 68% neutrophils, low glucose (54 mg/dL) in relation to the blood glucose level (101 mg/dL) and high protein levels (141 mg/dL).

A chest-XR revealed a pneumonia of the right lower lobe. He was admitted to the pediatric ward where he was started on IV ceftriaxone and azithromycin. Doxycycline was also associated. In the following 48 hours the nasal swab was positive for *Mycoplasma Pneumoniae*. The PCR on CSF could not detect *Mycoplasma Pneumoniae*, nor could it detect any other causative agents. However, there was clear seroconversion for *Mycoplasma Pneumoniae* in the serology. Within 72 hours after admission, he was afebrile and his symptoms disappeared. He was discharged on day 8 of hospitalization, with no neurological sequelae.

Mycoplasma Pneumoniae is a rare causative agent of aseptic meningitis. In only 0.1% of the *Mycoplasma Pneumoniae* infections, the central nervous system appears to be involved, with encephalitis, aseptic meningitis and myelitis as possible neurological presentations. Central neural involvement may be self-limiting, however early administration of adequate antibiotic therapy may be crucial, considering the capacity of antibiotics to cross the blood-brain barrier.

P 46.

Post-infectious bronchiolitis obliterans: a rare complication of viral or bacterial lung infections

D. Quiévreux, E. Gueulette

CHU UCL Namur Saint-Elisabeth

Introduction

Lung infections are common in children, both viral and bacterial. If the majority resolve without sequelae, complications can occur which should be screened for.

Clinical case

We report the case of a 10-month old infant. In November 2019, he presented with a picture of mixed high and low viral respiratory infection, compounding into recurrent multifocal pneumonia. He will initially be treated with antibiotics, oxygen therapy and bronchodilators. Unfortunately, he was quickly rehospitalized for RSV co-infection. Clinically, the child was feverish, dyspneic, spastic and oxygen dependent each time for an unusually prolonged period. This will lead to look for a pulmonary complication, by means of a thoracic CT-scanner. In view of the images, the diagnosis of post-infectious bronchiolitis obliterans is suggested.

Discussion

Bronchiolitis obliterans is a chronic lung disease characterized by circumferential and transmural inflammatory involvement of the bronchioli. Its incidence is 0.5 to 17.8 /100,000. It is defined by elements of a triad: clinical (cough, wheezing dyspnea, recurrent infections), radiological (mosaic appearance) and functional (fixed obstructive ventilatory disorder). The most frequently found causative agent is adenovirus, but any viral or bacterial lung infection can be complicated by such lesions. Management will be based on anti-inflammatory treatment (systemic corticosteroids, azithromycin), symptomatic treatment (inhaled corticosteroids, bronchodilators, anti-leukotrienes, respiratory physiotherapy), treatment of intercurrent infections (antibiotic therapy) and comorbidities (gastro- esophageal reflux) as well as preventive vaccinations (influenza, pneumococcus).

The infant in our clinical case was treated according to these protocols. After 6 months of follow-up, he is currently on treatment consisting of inhaled corticosteroids, azithromycin, omeprazole and respiratory physiotherapy. After 1 year of follow-up, the radiological images were normalized.

Conclusion

Repeated pulmonary infections in children, of viral or bacterial origin, should lead to a complementary assessment in search of complications or underlying causes. Although still little known because rare, bronchiolitis obliterans is part of the differential diagnosis. Its management must be the subject of consensus and guide-lines.

Posters**P 47.****Erythema nodosum associated with cat scratch disease : a case report.**

H. Warnier, K. Nyamugabo, M-C Seghaye

*CHU de Liège, U Liège*Clinical case

A 9-year-old girl presented a long lasting painful submandibular lymphadenopathy and recurrent erythema nodosum on the lower limbs. History revealed that the girl owned a young cat. Lab tests revealed a mild inflammatory syndrome and positive serologic tests for Bartonella Hensae, and ultrasonography showed abscessed adenitis. Treatment for cat scratch disease, consisted in azithromycin given orally for a total duration of 3 weeks. Suppurative adenitis was treated by flucloxacilline (initially given parenterally and then orally, for a total of 14 days) and necessitated several punctures of the collection. Evolution of adenitis was slowly favourable (resolving within 2 months), and erythema nodosum finally resolved spontaneously.

Discussion

Cat Scratch Disease (CSD) is an infectious disease caused by Bartonella Hensae, usually occurring after a cat scratch or bite. The main manifestation of the disease is a localized lymphadenopathy, but it can also cause systemic disease (reaching nervous central system, eye, liver or spleen for example) or cutaneous lesions such as our patient. Erythema nodosum (EN) is a rare form of skin manifestation of CSD. EN is characterized by erythematous tender nodules, mainly localized on the lower limbs. It's a rare condition in children, that may be associated with various underlying conditions : first pediatric causes are infectious diseases (Group A Streptococcus, Mycoplasma, Bartonella and various other microbiological agents), but it can also result from inflammatory bowel diseases, Sarcoidosis, vasculitis or drug exposure.

Diagnosis is made based on physical examination and history, and biopsy is generally not necessary but if performed, it reveals aspecific panniculitis. It's a self-limiting condition that generally doesn't need any treatment itself, but the underlying cause must be identified and treated.

P 48.

Case report: Congenital pulmonary airway malformation in a 17 month-old girl

S. Goemanne, J-C Beghin

HUDERF-ULB

Background

Congenital Pulmonary Malformations are a group of rare diseases caused by aberrant embryological lung development, which occurs at different stages of intrauterine life. Several structures including the airways, lung parenchyma and vasculature may be affected.

Case report

We describe the case of a 17 month-old African girl born at full term after normal pregnancy with no relevant medical history. She was admitted to the emergency department for fever up to 40°C since 5 days, cough and respiratory distress. Clinically, the patient had grunting, tachypnea, chest retractions, wheezing and crackles in the right lung base. She was suspected to have bronchiolitis complicated by bacterial surinfection. Chest X-ray confirmed the presence of a round opacity but also revealed multiple air-filled cystic lesions in the right lower lobe. The child was admitted for intravenous antibiotic therapy and complementary investigations. Chest CT-scanner was suggestive of a large Congenital Pulmonary Airway Malformation (CPAM) that involved the entire right lower lobe. The patient underwent a right lower lobectomy by thoracotomy due to massive inflammation. Pathology confirmed the diagnosis of CPAM type 1.

Discussion

CPAM is an adenomatoid proliferation of bronchioles that form cysts during embryogenesis. It usually occurs in a single lobe and despite its very low incidence (1 per 10.000 live births) it is the most common congenital thoracic lesion. CPAM is classified in 5 types based on clinical, macroscopic and microscopic criteria. Type 1 is the most frequent (65%). Two third of the cases are detected during pregnancy or neonatal period because of respiratory distress. Among the remaining patients, 86% will become symptomatic before 13 years of age (median 2 years of age). Symptoms include recurrent pneumonia, respiratory distress and spontaneous pneumothorax. In the absence of hydrops, CPAM has a good prognosis, with more than 95% live birth. Management of symptomatic CPAM includes prompt surgery. For asymptomatic cases, increased rate of infection over time renders the surgery more difficult after months or years of evolution and pushes for recommendation of early elective surgery.

Conclusion

Despite the high quality quality of fetal imaging in our countries, CPAMs are undiagnosed in 33% of cases during pregnancy or neonatal period. The disease should be suspected at any age in the presence of recurrent pneumonia, pneumothorax or suggestive chest X-ray.

Posters**P 49.****Lymphocytoma and neuroborreliosis in a five-year-old child in the Hainaut: a rare presentation of Lyme disease.**

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*Centre Hospitalier epiCURA, ULB, CHU Saint-Pierre*Background

Neuroborreliosis represents the second most frequent manifestation of Lyme Disease in Europe. In children, facial paralysis is the most common and pathognomonic manifestation of Neuroborreliosis, especially if bilateral. Despite the fact that in Belgium Lyme disease is more common in the province of Flemish Brabant, Campine and in the Ardennes, we report a case of pediatric Lyme disease identified in the Hainaut and with an atypical presentation characterized by an initial lymphocytoma followed by low pathognomonic manifestations of neuroborreliosis.

Methods

We analyzed clinical and biological data of a 5-year-old girl who was hospitalized in our institution (EpiCURA Hornu, Belgium). Furthermore, we considered recent literature and publications about diagnostic criteria and new diagnostic tools for Neuroborreliosis in children.

Results

We report the case of a 5-year-old girl who consulted her family doctor in March 2019 for a left retro auricular induration without any other complaints. Two weeks later, the child consulted the pediatrician for an erythema migrans and a lymphocytoma behind the left earlobe. She also progressively developed the following clinical features: intermittent headache, asthenia, leg pain and memory disorders. The physical examination didn't highlight any neurological signs. A completed anamnesis revealed a recent tick bite behind the left earlobe before the first symptoms occurred. The tick was removed by friction. An Elisa and Western Blood Tests confirmed positive Borrelia serology. The cerebrospinal fluid analysis showed lymphocytosis, but we didn't find the presence of anti-Borrelia antibodies. The child received a 2 weeks course of ceftriaxone and we could observe a rapid complete resolution of symptoms. Three weeks later, she presented intermittent residual neurological symptoms such as headache, concentration disorders and leg pain. At the same time, we were informed about a positive Borrelia-specific PCR in the cerebrospinal fluid. We preferred to start again intravenous antibiotics for one week. Today, the child is doing better and has no complaints.

Conclusion

Better knowledge of the clinical symptoms and the biological diagnosis of this disease should allow earlier diagnosis and more appropriate treatment especially in the regions with the lowest incidence of the disease.

P 50.

Acquired torticollis in an infectious patient as a sign of otomastoiditis complicated by thrombophlebitis of jugular vein and intracranial thrombosis

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*GZA Sint-Vincentius, UZ Leuven*Background

A toddler of 1 year old with an uncomplicated medical history was admitted to our hospital with high spiking fever of 6 days and a toxic and uncomfortable appearance. The parents mentioned that he had been reaching out for his ears the days before. Clinically a hyperemic pharynx and bilaterally hyperemic eardrum were seen, without any sign of perforation or otorrhea. There was no drooling or stridor. On auscultation, transmitted rhonchi were heard. No protrusion of the pinna, no mass, erythema or tenderness over the mastoid was noted. Neurological examination was normal, without signs of meningism. Blood sample showed high inflammation markers with a CRP of 326 mg/dL. Chest x-ray and abdominal ultrasound showed no abnormalities. Shortly after admission, a torticollis with lateral flexion of the head to the right side combined with a rotational movement of the chin to the left side developed, accompanied by diffuse cervical pain on palpation.

Methods and results

Empirical antimicrobial therapy with a third generation cephalosporin were started. A CT of the neck and cervical spine showed a coalescent otomastoiditis with extension of the inflammation to the caudal region of the mastoid, starting to form Bezold abscesses, and further complicated with suppurative thrombophlebitis of the jugular vein. This is also known as Lemierre syndrome. An MRI showed thrombosis of the sinus venosus. A mastoidectomy was performed in a tertiary center, where microbiological samples were taken. Immunological screening was performed.

Conclusion

This case showed an atypical presentation with torticollis as a sign of otomastoiditis, complicated by sinus venosus thrombosis and thrombophlebitis of the jugular vein. Torticollis can be explained through irritation of the sternocleidomastoid muscle by thrombophlebitis of the jugular vein, or can be secondary to irritation of the muscle by Bezold abscesses, expanding caudal through mastoid bone. Complications of otomastoiditis occur in 15-30% of the patients and most often involve the intracranial region. Otomastoiditis is a clinical diagnosis, but imaging is indicated in atypical presentations and when complications are suspected.

Posters**P 51.****Streptococcus anginosus epidural abscess complicating a pansinusitis in a 9-year-old boy**

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*ULB-CHU Saint Pierre*Introduction

Sinusitis is a very common infection in children and most patients fully recover. However, persistent infection can lead to dangerous intracranial complications representing diagnostic, management and therapeutic challenges.

Clinical case

A previously healthy 9-year-old boy presented to the emergency department with a 5-day history of fever and headache. The day before presentation he developed a swelling of the forehead and since the morning a left eyelid swelling. On physical examination, the patient presented left periorbital edema and a fluctuant left-sided forehead mass. Ocular motion and neurologic examination were unremarkable.

Laboratory results revealed moderate inflammation. CT-scan showed a pansinusitis including the frontal cells with overlying soft tissue edema. No intracranial findings or bone destruction were found. The patient was immediately treated with ceftriaxone and oral corticoids. The fever regressed rapidly and the patient felt better. However, forehead and left eyelid swellings did not improve. After 3 days, a new CT-scan was performed that showed pansinusitis persistence and abscesses on either side of the left frontal bone without bone destruction. Ornidazole was added to the antimicrobial treatment and the patient underwent surgically drainage of the affected sinuses and of the left eyelid. It was decided not to drain the epidural abscess due to its small size.

Intraoperative cultures grew *Streptococcus anginosus* group susceptible to penicillin, a commensal of the oral flora, which is among the most commonly isolated pathogen in intracranial complications of sinusitis.

After a short postoperative improvement, the left eyelid swelling increased again. MRI revealed an expansion of the epidural abscess associated with a compression of the superior sagittal sinus. The patient underwent a second surgery to drain the subcutaneous abscess. Despite broad spectrum intravenous antibiotics and repeated sinus surgery, the intracranial abscess continued to grow in the following days and cerebritis and osteitis of the frontal bone developed. After drainage of the intracranial collection, the patient had a favorable outcome with prolonged intravenous antibiotics.

Conclusions

Imaging is the key to diagnose the rare complications of sinusitis. Broad spectrum intravenous antibiotics and drainage of the sinus are mandatory in case of intracranial complications. However, there is less consensus on when to perform neurosurgical drainage.

P 52.

Hair depigmentation due to cystic fibrosis

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Background/Aims

Hair depigmentation (hypochromotrichia) is a very rare presentation of cystic fibrosis (CF) with only four cases described in literature. Poor nutritional status due to pancreatic insufficiency and deficient intake is the main cause. We want to raise awareness about this clinical picture as early treatment of nutritional deficiencies is important to prevent life threatening complications.

Methods

The clinical, biochemical and radiographic data of a 5 month-old girl who presented with failure to thrive and hypochromotrichia as signs of CF are presented.

Results

A 5-month-old girl presented with failure to thrive. Clinical exam revealed proximal hair depigmentation. She was diagnosed with CF after two consecutive positive sweat chloride tests with chloride concentrations of 90.0 and 76.0 mmol/L (normal below 30). Gene analysis showed compound heterozygosity for CF mutations c.2551C>T p.(Arg851*) and c.2755delT p.(Tyr919Thrfs*4). Laboratory tests revealed normal electrolytes, normocytic anemia, hypoalbuminemia, slightly elevated transaminases, and low vitamin A and E and zinc. Pancreatic elastase was below detection limit, reflecting pancreatic insufficiency. Chest X-ray showed air trapping. Abdominal ultrasound revealed liver steatosis and no hepatomegaly. Throat swab culture grew Haemophilus influenzae. Treatment consisted of pancreatic enzyme replacement therapy (PERT), fat soluble vitamin supplementation (ADEK), oral antibiotics, inhalation therapy with hypertonic saline and regular chest physiotherapy. After initiation of treatment there was a significant catch up growth and weight gain and good respiratory status. Clinical exam also revealed reversal of the hair depigmentation.

A combination of nutritional deficiencies as seen in CF due to pancreatic insufficiency and deficient nutrient intake is the main cause of hair depigmentation. The lack of zinc, protein, essential fatty acids and the amino acid tyrosine seem to have a role in skin and hair changes in these patients. Protein-energy malnutrition, defined by anemia, hypoalbuminemia and edema is seen in 5 to 13% of children with CF. A reversal of the hair depigmentation can be expected after adequate treatment and improvement of nutritional status, as seen in our patient.

Conclusion

Hair depigmentation is a very rare presentation of CF. This case report shows this clinical finding associated with poor nutritional status and reversal after treatment.

Posters**P 53.****About a case of spectacular evolution of a severe dermo-respiratory syndrome under dupilumab**

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A twenty year caucasian old boy is followed since the age of four months for atopic dermatitis and respiratory symptoms at our outpatient consultation.

His pulmonary evolution was marked by a first episode of bronchiolitis at the age of 4 months with evolution in a few years to difficult to treat asthma. He was often hospitalized for oxygenotherapy and received systemic corticosteroids on 15 occasions. The treatment was supplemented by high dosis inhaled corticosteroids (ICS) with long acting beta2 agonists, beta2agonists as needed, antileukotrienes and an attempt of allergenic avoidance. A first outbreak of atopic dermatitis was described at the age of eleven months. He was treated with local corticosteroids since the onset of symptoms with ciclosporin during more than ten years. He presented and is still presenting a concomitant alopecia areata.

Complementary investigations revealed a clear and severe allergic status (biology and skin allergy tests). Pulmonary functions showed an (often) reversible obstructive syndrome since the age of three. The exhaled nitric oxide (eNO) reached a peak of 107 ppm (normal under 25 ppm). Bronchoalveolar lavage was performed and showed 240000 cells/ml including 3% of neutrophils (N: 0-2) and 4% of eosinophils (PNE) (N: 0-1). At the same time, chest x-ray and tomodensitometry were normal. The immune balance (cellular and humoral) was also normal; the blood level of eosinophils (PNE) was noted at 680/ μ l (N < 400). An 24- hour oesophageal pH monitoring and a sweat test were normal. The integumentary system was assessed by SCORAD (SCORing Atopic Dermatitis) with a result between 75 and 90/100.

A treatment with dupilumab (Duxipent) was started in May 2019 considering the failure of optimal management with partial control of the disease.(allergic avoidance was incomplete and difficult). Dupilumab is a monoclonal antibody that inhibits IL-4 and IL-13 signaling. The federal drug administration (FDA - USA) recommends it if the blood level of PNE is higher than 150/ μ l, the eNO > 20 ppm and a failure of first line treatments.

The patient reported only a transient local reaction at the injection site and conjunctivitis already described with this treatment.

After seven months of treatment, we noted a decrease of the dosis of ICS without oral corticosteroids, e-NO at 20 ppm, Scorad at 26/100 and even an improvement of the alopecia areata with tolerance of allergens. Quality of life reported by the patient was also improved.

P 54.

Case report: septic shock in a 12-year-old boy in the beginning of the covid-19 outbreak: a diagnostic challenge

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A 12-year-old boy was transferred to the emergency ward after he was found on the kitchen floor, vomiting bile. Three days earlier, the patient was presented at the emergency ward because of the impossibility to lean on his left ankle due to swelling and pain, caused by an inversion trauma that happened three days earlier. X-ray of the left ankle was negative. The patient was sent home with analgesics, support bandage and crutches. However, the patient was presented again that same day because of persistent pain without any changes clinically. Ice, elevation of the feet and rest was advised.

Two days later, the pain persisted. A CT-scan showed a fulminant swelling of the soft tissue. The patient was sent home with plaster bandage and a consultation within one week. The day after, the patient was found on the kitchen floor while vomiting bile. The patient was vomiting for one day, had diarrhea for four days and had a minimal food intake for two days. By arrival at the emergency ward, a deep hypoglycemia was diagnosed.

After adequate glucose administration, clinical exam still showed a somnolent patient with a poor peripheral circulation. The patient was in sinus tachycardia and hypotension and became progressively tachypnea. Oxygen was started. Arterial blood gas showed a normal glycemia, hyponatremia and high lactate. Blood sample indicated acute renal failure, beginning liver failure with aberrant coagulation and nascent sepsis, CRP 340mg/l.

Lung X-ray showed diffuse bilateral infiltrates. Lung CT showed multiple infiltrates without any ground-glass opacities, excluding COVID-19. Differential diagnosis included tuberculosis, septic emboli due to peripheral infection and bacterial consolidations.

After removing the cast, a hard, cold, swollen and painful lower leg was revealed. The patient was transferred to a university hospital after starting with broad-spectrum-antibiotics. A duplex ultrasound of the left ankle revealed a deep venous thrombosis(DVT). Eventually, the patient was diagnosed with a septic shock due to septic emboli on a DVT, caused by a septic arthritis. The diagnosis of inversion trauma, the absence of fever, warmth and redness of the joint and the lack of a complete anamnesis at first, made the diagnosis of a septic arthritis in this case very difficult. This case should remind the clinician to always bear septic arthritis in mind. It can present without fever and it is often preceded by trauma or fall.

Posters**P 55.****Case Report: Prolonged fever, splenomegaly and pancytopenia in a 4-year-old child**

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*ULB - HUDERF*Background

Visceral leishmaniasis (VL) is a systemic protozoan disease whose global incidence has increased in recent years due to increased international mobility, and concomitant factors that increase susceptibility, such as infection with human immunodeficiency virus (HIV) and malnutrition. Clinical features of VL mimic several other common diseases and varies depending on patient's immune status and the species of *Leishmania* provoking the infection. The spectrum of illness ranges from asymptomatic infection to fulminant, life-threatening infection. The diagnosis requires demonstration of the parasite. Early detection and proper management are crucial for control of this disease.

Methods

We report the case of a four-year-old boy who was admitted to our hospital with a two-months history of fever, abdominal distention and weight stagnation after a travel to Morocco. His physical examination revealed massive splenomegaly, while laboratory findings yielded pancytopenia.

Results

A bone marrow aspiration revealed a hypocellular marrow with no evidence of malignancy and no parasite identification at macroscopic bone marrow smear examination. Serology showed positivity for *Leishmania donovani* antibodies at indirect immunofluorescence test (IFA) and enzyme-linked immunosorbent assays (ELISA). *Leishmania* DNA was secondly detected at polymerase chain reaction (PCR) on bone marrow smear confirming the diagnosis of visceral Leishmaniasis and a treatment by amphotericin B was initiated. Visceral Leishmaniasis should be considered in the differential diagnosis of children with persistent fever, hepatosplenomegaly and pancytopenia with travel history to endemic areas. Definitive diagnosis requires demonstration of the parasite by either histopathology or culture of material obtained by needle aspiration or biopsy from affected organs. Serologic tests as IFA and ELISA has high sensitivity but are not specific for the stage of the disease. Molecular methods have remarkable sensitivity and specificity and allow species identification.

Conclusion

Visceral leishmaniasis must be suspected in children presenting the triad of fever, splenomegaly and pancytopenia coming from endemic counties. Multiple diagnostic approach is suggested for accurate diagnosis.

P 56.

When a pharyngitis in the Indian Ocean leads to cardiac surgery in Paris.

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Clinical case

Y.D is a 4 year old boy born in the Comoros. He presented to the emergency department of the Centre Hospitalier de Mayotte with dyspnea and cough in a febrile context. The clinical examination shows bilateral jugular vein distension, hepatomegaly, edema of both feet, labored breathing and a systolic hearth murmur. The chest radiography shows a cardiomegaly. A sinusal rythme with a prolonged PR interval is seen on the ECG. The ultrasound shows a mitral regurgitation. The blood sample showed a WBC of 16,3 G/L with a mild inflammatory syndrome. The culture of a throat swab shows *Streptococcus pyogenes*. ASLO and ASD are respectively 103 and 400 U/mL. According to the local epidemiology of Mayotte and the clinical presentation of severe mitral regurgitation, a rheumatic carditis was suspected. Next to the treatment of his cardiac failure, he received Amoxicilline and Prednisolone. Five months later he underwent mitral repair surgery in Paris.

Discussion

Acute Rheumatic Fever (ARF) is one of the non-suppurative complications of *S. pyogenes* pharyngitis. The incidence in Mayotte is 18,2/100 000 among people younger than 20 years old. Antigenic mimicry and an abnormal immune response of the host are the cornerstones of the pathophysiology of ARF in genetically predisposed children who were in contact with *S. pyogenes*. The diagnosis is made on a combination of clinical signs according to the revisited Jones Criteria. Carditis occurs in 50% of patients with and will present mostly as mitral valvulitis. Only 10% will present with cardiac failure. Currently the treatment is based on the eradication of *S. pyogenes* and anti-inflammatory treatment. Urgent surgery will be needed for severe valvulopathy. Valve repair will always be preferred. But if a replacement should be done, mechanical valves should be avoided because of the need of anticoagulant therapy and monitoring of the INR that would represent a challenge in developing countries. Acute rheumatic fever represent a real burden, causing 500 000 deaths per year. A screening of subclinical carditis in endemic regions could lead to early detection and early treatment. Unfortunately, there is a lack of human and material resources.

Conclusion

ARF is responsible for a high health burden in developing countries. The highest burden is related to cardiac involvement. Primary and secondary prevention are important to reduce ARF associated morbidity, but there is a lack of human and material resources.

Posters**P 57.****Chronic cough in a child with "severe asthma": may it be chronic pulmonary aspiration?**

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*U Liège, CHU de Liège, CHR Huy*Introduction

Asthma is a common cause of recurrent bronchitis in infants and young children, but the differential diagnosis are numerous. They should be explored when the child does not improve significantly despite high dose asthma medication.

We report the cases of two toddlers who were first diagnosed with asthma but who were found out to have chronic pulmonary aspiration (CPA).

Case presentation

Case 1: A 8-months-old girl presented with recurrent bronchitis and persistent cough and wheezing. She was diagnosed with asthma and gastroesophageal reflux disease. She was referred to the respirology clinic at 16 months. The respiratory symptoms were still present despite salmeterol 25 + fluticasone 150 (2 doses 2x/qd), Montelukast 4 mg and omeprazole 20 mg. The mother reported cough when swallowing liquids. Barium swallow study (BSS) and fiberoptic bronchoscopy (FiBr) were normal but a flexible nasal endoscopy (FNE) suspected a type 1 laryngeal cleft (LC T-I). A direct rigid laryngoscopy (DRL) confirmed the diagnosis. Appropriate treatment with thickened liquids and medical anti-reflux treatment resolved the respiratory symptoms.

Case 2: A 3-years-old boy was referred to the respirology clinic due to recurrent bronchitis and chronic wet cough starting at 7 months. He had a history of severe prematurity, bronchodysplasia and was fed by gastrostomy for orality disorders (with growth failure). The foster mother reported cough when eating solids and liquids. He was on high dose asthma medication (similar to Case 1 medication). Immune work up, sweat chloride test, FNE, and BSS, were normal. FiBr revealed a diffuse malacia and inflammation, no fistula. Finally, a second FNE identified a LC T-1 and a DRL ascertained the diagnosis. Chronic respiratory symptoms improved when oral liquids were stopped.

Conclusion

Chronic respiratory symptoms, particularly if not responding to asthma treatment, require careful evaluation to exclude underlying disease. CPA is often forgotten as a cause of chronic respiratory symptoms. Some clinical features should alert pediatricians: cough triggered by oral intake, feeding difficulty, chronic wet cough, recurrent respiratory infections. The main causes of CPA are anatomic abnormalities of the airways and the upper digestive tract, neuromuscular and neurological diseases and gastro-intestinal disorders. LC T-I/II diagnosis is difficult and usually requires a direct laryngoscopy, as feeding skills studies and FNE may be normal.

P 58.

A severe case of acute flaccid myelitis: clinical presentation, diagnosis and management

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Background

Acute flaccid myelitis (AFM) is the clinical syndrome caused by loss or dysfunction of anterior horn cells in the spinal cord as a result of infectious or inflammatory disease. This leads to flaccid weakness of the innervated limb. In the post-polio era, enterovirus (EV), EV-D68 and EV-A71 are the most well-known viruses to cause AFM. We present a case of AFM with severe clinical features caused by another EV.

Case

A 5 month-old boy was admitted to our Pediatric Intensive Care Unit (PICU) with AFM. Presenting with fever and anorexia since 24 hours, he progressively developed respiratory failure, bradycardia, dysphagia and weakness of the upper limbs with hypersensitivity of the lower limbs during the following 24 hours. He required mechanical ventilation, initially via endotracheal intubation, subsequently via tracheostomy. Extensive diagnostic workout was performed.

Magnetic Resonance Imaging showed extensive myelitis with pathological T2-hyperintense lesions in the gray matter of the spinal cord from medulla oblongata to the thoracic region Th2. Electromyography confirmed severe damage of the anterior horn of the upper limbs. Guillain-Barré syndrome was excluded and an (entero)viral infection was suspected and confirmed after repeated viral testing.

Corticoid treatment was initiated. After 1.5 months our patient progressively regained spontaneous motor activity of the head, lower limbs and right arm and partial respiratory recovery, requiring mechanical ventilation during sleep. After approximately 3 months he was discharged to a rehabilitation center. Upon discharge, there was still a significant axial and peripheral palsy with complete palsy of the left arm.

Results

Rhinovirus (RV) and human bocavirus were detected in the respiratory specimen. The National Reference Centre (NRC) confirmed RV-A in the respiratory sample and detected a coxsackievirus A9 (CV-A9) and bocavirus in the stool sample by next-generation sequencing. There were no pathogens detected in CSF. As CV-A9 has been described as a neurotropic virulent EV associated with AFM, we believe this to be the pathogenic agent.

There are no specific therapies for AFM available. Therapy must be focused on supportive care, limiting further damage to the spinal cord and early rehabilitation.

Conclusion

PICU referral is needed for patients with AFM and respiratory symptoms. This case report demonstrates the importance of analyzing multiple samples and the extensive search for viral causes.

Posters**P 59.****Varying Presentations of Multisystem Inflammatory Syndrome Temporarily Associated with COVID-19**

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*UZ Brussel*Background

A novel coronavirus identified in 2019 leads to a pandemic of severe acute respiratory distress syndrome with important morbidity and mortality. Initially, children seemed minimally affected, but there were reports of cases similar to (atypical) Kawasaki disease or toxic shock syndrome, and evidence emerges about a complication named paediatric inflammatory multisystem syndrome temporarily associated with SARS-CoV-2 (PIMS-TS) or multisystem inflammatory syndrome in children (MIS-C).

Methods

Two cases were compared and discussed demonstrating varying presentations, management, and evolution of MIS-C.

Results

Case 1: a black African 15 year old boy with idiopathic end-stage kidney disease on peritoneal dialysis developed severe signs of viral or inflammatory myocarditis after a Covid-19 infection.

Case 2: a black African 14 year old girl known with Lennox-Gastaut syndrome presented with symptoms similar to Kawasaki disease or toxic shock syndrome.

These cases are presented to increase awareness and familiarity among paediatricians and emergency physicians with the different clinical manifestations of this syndrome.

Conclusion

MIS-C is an uncommon entity and may occur with possible diverse clinical presentations. Early recognition and treatment are paramount for a beneficial outcome.

P 60.

The link between malnutrition, of allergic origin, and immune deficiency: about a case

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Background

A diet that is restricted by allergies may lead to nutritional deficiencies that in turn alter the body homeostasis. The potential resulting immune deficiency favors bacterial infections, which can themselves worsen malnutrition.

Clinical case

A 20-month-old patient with egg and cow milk protein allergy is admitted to the emergency room for febrile rhinopharyngitis. As cow's milk protein and egg allergy is severe and not followed, the patient's diet is restricted to a few foods. The patient has already presented pathological fractures with vitamin and phosphocalcic disorders supplemented recently. Clinical examination revealed paleness and bronchial secretions on auscultation with no sign of acute respiratory distress. Workup reveals a central pancytopenia of infectious origin with iron deficiency anemia, the presence of influenza B on a nasopharyngeal swab and a diffuse bronchopathy. A rachitic rosary is detected on chest radiography. Given the deterioration of the general condition, the patient is hospitalized for surveillance under cardio-respiratory monitoring and symptomatic treatment. After 2 days of hospitalization, in a context of acute respiratory failure, a left pulmonary secondary infection is highlighted. IV antibiotherapy is started but left bronchopneumonia worsens with a left pleural effusion, characterized as an empyema. A modification of the IV antibiotherapy and a decortication by thoracoscopy are necessary. When *s. pneumoniae* is detected on pleural fluids, the antibiotic spectrum is reduced. This treatment allowed a complete resolution of bacterial pneumonia. Substitutive treatment by vitamin D and calcium was also introduced.

Discussion

While most healthy children have natural defenses to fight off infections, those with deficient immune systems are at higher risk for diseases. Many factors can cause immune system deficiencies, and among these, undernutrition is one of the essential elements. Bacteria are therefore more resistant and the use of antibiotics with a wider spectrum is often necessary. In addition, any strong or prolonged immune stimulation is accompanied by hypercatabolism and thus an aggravation of the initial state of malnutrition.

Conclusion

Maintaining a varied diet, despite of food allergies is essential for the child's overall development, including the immune system homeostasis. Medical and dietary follow-up must be carried out to avoid nutritional deficiencies and thus the related complications.

Posters**P 61.****Subacute meningitis as presentation of neuroborreliosis in a seven year old child**

C. Genin, M. Creuven, P. Philippet

*CHC Mont Légia*Case report

We report on a case of neuroborreliosis in a 7 ½ year old girl with a clinical picture of isolated subacute meningitis. The patient has an 8 days history of marked asthenia without headache, fever or any neurological manifestation (especially nerve palsy). There was no history of tick bite or erythema migrans. Clinical examination shows nuchal rigidity with otherwise normal neurological examination. Standard blood tests (WBC, CRP) and cerebral CT-Scan were normal. Lumbar puncture showed lymphocytic pleiocytosis of cerebrospinal fluid (CSF) (505 WC/mm³ with 93% lymphocytes) with elevation of proteins level (1.224 g/L) and normal glucose level. Serologic tests revealed IgG antibodies to *B. Burgdorferii* in blood and CSF with intra-thecal synthesis. She was successfully treated by intravenous ceftriaxone (2g once daily) for 21 days.

Discussion

Facial nerve palsy is the most common manifestation of neuroborreliosis in children and can be associated with clinical (headache, nuchal rigidity) or biological (CSF pleiocytosis) signs of meningitis. Isolated meningitis without nerve involvement is less frequent (1% of pediatric Lyme diseases). Classic presentation of Lyme meningitis consist in headache with low or absent fever and subacute course of symptoms. Isolated asthenia is an unusual presentation. Diagnosis relied on typical CSF features (lymphocytic pleiocytosis, moderate elevation of proteins concentration, normal glucose level) and demonstration of intra-thecal synthesis of *Borrelia* antibodies (detection of IgG *Borrelia* antibodies in blood and CSF with positive CSF/blood index). Treatment recommendations for neuroborreliosis varies among different scientific authorities worldwide. Long-course antibiotic treatment (14 days for facial palsy, 21 days for meningitis) is commonly admitted. In children with meningitis, intravenous therapy with ceftriaxone is generally initiated until patient is stabilized. Oral regimen with doxycycline was proven efficient and is used as first line treatment in older children (> 8 years) for isolated facial palsy, or after initial IV therapy in meningitis. In younger children, complete parenteral course with ceftriaxone is recommended in Europe, when oral doxycycline is indicated in all age children by the American Center for Diseases Control.

P 62.

Trisomy 21, what about the lungs? – About 3 paediatric cases

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Cliniques Universitaires Saint Luc

Introduction

Non cystic fibrosis bronchiectasis is a rare but increasingly frequent diagnosis in paediatrics. We report the cases of 3 children with trisomy 21 who have developed bronchiectasis.

Case reports

A first trisomic girl presented with recurrent respiratory infections, including a severe right-sided pneumonia since the age of 14 months. Cough reflex was inadequate and chest CT at 2 years old demonstrated bronchiectasis of the middle lobe. Bronchoscopy showed collapse of the middle lobe's orifice and presence of purulent secretions. Bronchiectasis was attributed to severe respiratory infection on a background of hypotonia and inadequate cough.

A second trisomic girl with many co-morbidities (oesophageal and duodenal atresia, interventricular and interauricular communications) presented with recurrent bronchitis and pneumonia since the first months of life. She also had severe gastro-oesophageal reflux disease (GORD). Chest CT at 5 years old showed diffuse bronchiectasis of the middle and right lower lobes and tree in bud in the right superior lobe. Bronchoscopy showed bronchomalacia, an extra B6 bronchus and diffuse purulent secretions mainly in the lower lobes. The aetiology of bronchiectasis for this patient is complex: recurrent respiratory infections on a background of bronchomalacia, severe reflux due to oesophageal dysmotility and congenital heart disease.

The last patient is also a trisomic girl with multiple co-morbidities (duodenal atresia, atrioventricular canal). She presented recurrent respiratory tract infections since the third month of life, and was diagnosed with hypogammaglobulinemia. She also suffered from obstructive sleep apnoea (OSA), GORD and chronic aspiration. She received supplemental nocturnal oxygen therapy in addition to nocturnal CPAP treatment. This patient developed pulmonary hypertension. Chest CT at 4 years old showed bronchiectasis mostly in the right lower lobe. Multiple risk factors can explain the occurrence of bronchiectasis: GORD with chronic aspiration, immune deficiency, tracheomalacia, OSA, congenital heart disease.

Conclusion

Trisomic patients with chronic cough or recurrent respiratory infections are at risk of developing bronchiectasis for multiple reasons, including aspiration, immature cough reflex, hypotonia, immune deficiency amongst other co-morbidities. These case reports highlighted the importance of a regular follow-up at the respiratory clinic to avoid the occurrence of lung damage.

Posters**P 63.****Analysis of the characteristics and risk factors of complications in a pediatric population hospitalized for pneumonia**

P. Guillaume-Gentil , E. Rebuffat, C. Adler

*CHU Saint-Pierre, ULB*Background and aims

The incidence, risk of complications and sequelae, and mortality, associated with pneumonia, remains high in the global pediatric population, despite the introduction of conjugate vaccines against *Streptococcus pneumoniae* since the 2000s. The objective of this study is to analyze the incidence, the characteristics of pneumonia and their complications and to identify the risk factors for complicated pneumonia within the population of children hospitalized at the “Centre Hospitalier Universitaire de Saint-Pierre”, in order to understand the impact of this disease and its complications in a multicultural population of Brussels.

Materials and methods

It is a retrospective and descriptive study. The different data (demographic, clinical, microbiological, biological, radiological and therapeutic) were collected, analyzed and then compared among children with uncomplicated pneumonia and those who developed a complication.

Results

A total of 113 patients out of 2229 hospitalizations (5%) were admitted for pneumonia. A virus was isolated in 32 patients, a bacterium in 9, a virus and a bacterium in 2 and no germ in 70 children. The germs were identified by searching for viral antigens and/or viral culture of nasopharyngeal secretions, by blood cultures, by serology and by culturing pleural fluid. Eleven children (10%) developed a complication. Pleural effusion is the most common (5%). Elevation of c-reactive protein to admission biology is statistically significantly associated with a risk of developing a complication.

Conclusion

The search for new, more profitable and more sensitive means of identifying bacteria is necessary to better identify the children for whom antibiotic therapy is justified and necessary. A c-reactive protein greater than 100 mg/L on admission could constitute a biomarker encouraging close clinical monitoring, and also possibly carrying out a chest ultrasound to detect complications early.

P 64.

Assessment of the diagnostic value of eosinopenia in suspected bacterial infections in the pyretic child.

J. Vranckx, M. Vranckx

CHU Saint Pierre, Hôpital Civil Marie Curie

Aims

The diagnosis of bacterial infection in a febrile child is based on the association of the physical examination and the results of complementary exams. The main goal of this study is to assess the diagnostic value of eosinopenia in suspected bacterial infections in children presenting to the emergency room with fever. The second goal is to compare the diagnostic value of eosinopenia to the value of other biomarkers used more often, and to check their respective abilities to differentiate a bacterial etiology from a viral etiology to the fever.

Methods

We present a descriptive monocentric retrospective study based on the collection of laboratory values (C reactive protein, leucocytosis, relative neutrophilia and absolute eosinopenia) in children younger than 15 years old, that were admitted to the emergency department for fever between 07/01/2016 and 06/30/2017, and determination of their diagnostic value (sensitivity, specificity, positive and negative predictive values , likelihood ratio).

Results

232 patients were included, including 60 clinical bacterial infections (30 were documented) et 172 viral infections (84 were documented). Eosinopenia alone is not a good diagnostic marker (Se 46.67%, Sp 73.84%, NPV 76.12%, PPV 30.77%). The association of a C Reactive Protein value higher than 100mg/L with a neutrophilia higher than 85 % and absolute eosinopenia could assess of a bacterial etiology (p N.S. due to low number of patients). Among various combinations of markers, a normal C Reactive Protein value associated with eosinophilia has the higher power to exclude a bacterial infection (NPV 95%). C Reactive Protein and relative neutrophilia have the best areas under the Receiver Operating Characteristic area, respectively 0,8085 and 0,7710. Hyperleucocytosis is only usefull if very high (> 20,000/mm³).

Conclusions

In pyretic children admitted to the emergency department, eosinopenia is not a good marker of bacterial infection if used alone. On the other hand, used in association with other biomarkers, it potentiates their diagnostic values.

Posters**P 65.****Successful Systemic and Topical Treatment of Mycobacterium abscessus Otomastoiditis in Children.**

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*Radboud University MC*Background

Mycobacterium abscessus is an extensively drug-resistant opportunistic pathogen that can cause chronic otomastoiditis. There are no evidence-based treatment regimens for this severe infection.

Method

We treated four children with M. abscessus otomastoiditis with a structured regimen of topical imipenem and tigecycline, intravenous imipenem and tigecycline, and oral clofazimine and azithromycin and adjunctive surgery.

Results

All four patients attained culture negativity while on treatment, and no recurrences have been noted after a mean of 14 months of follow-up after treatment. The frequency and severity of adverse events is problematic. Intravenous tigecycline seems to be the main culprit, causing severe nausea and vomiting, as previously reported. The major adverse event was the social impact of the treatment, i.e. missing school for days or weeks and several admissions to hospital for either adverse events or surgery.

Conclusion

This structured approach to treat M. abscessus otomastoiditis led to cure in all four patients, with 1 year of follow-up after treatment. Adverse events were frequent, mostly caused by tigecycline.

P 66.

Septic arthritis of the temporomandibular joint: an uncommon but potentially severe complication of acute otitis media in children

I. de Brouhoven, M. Creuven, A. Joyeux, M. Lewin, P. Philippet

CHC Montlégia

Acute otitis media (AOM) is common in children and usually benign since the advent of antibiotics. It can however give rise to intratemporal and intracranial complications due to the multiple dissemination paths around the infected middle ear. Beside the more frequent mastoiditis, AOM can lead to septic arthritis of the temporomandibular joint (TMJ), a rare condition in the paediatric population and poorly described in the literature so far.

We report the case of a 3-year-old healthy child with otogenic septic arthritis of the right TMJ. The child presented with 4-days of isolated fever, unnoticed erythema and peri-auricular swelling of the right ear. Otoscopy confirmed an acute otitis media and biology revealed a highly elevated CRP with neutrophilic leukocytosis. The contrast-CT of the skull showed no filling of the right mastoid cells but an intra-articular effusion of the right TMJ and significant swelling of the adjacent soft tissues. After a bilateral myringotomie, intravenous empiric antibiotics were started with Cefotaxime. After 9 days of IV treatment and a persistent right otorrhea, a retro-auricular fluctuation appeared and confirmed to be a mastoiditis with a subcutaneous abscess on the repeated scan of the petrous bones. A transcutaneous puncture was performed and the antibiotherapy was expanded to Metronidazole. The evolution was finally satisfactory after 14 days of IV treatment. No bacteria was eventually found in the blood culture, mid-ear aspiration or abscess puncture.

Septic arthritis of the TMJ generally occurs by hematogenous dissemination or more rarely due to otogenic causes by contiguity with the surrounding anatomical structures.

Symptoms vary from fever, trismus, auricular pain or swelling to an asymptomatic presentation, making its clinical diagnosis difficult. Therefore the contrast-CT is essential to assess the TMJ, for acute diagnosis of septic arthritis as well as screening of long-term bone changes (fibrosis, joint ankylosis). CRP is mostly useful to monitor the response to treatment.

To date, no treatment consensus has been established for this medical emergency; a combination of IV antibiotics, joint decompression and physical therapy.

Septic arthritis of the TMJ should be thoroughly screened in case of complicated AOM, not responding to ongoing antibiotics. An early diagnosis, aggressive and multidisciplinary treatments can prevent the damaging sequelae on the maxillofacial growth and jaw mobility of the child.

Posters**P 67.****Chronic cough in children: don't forget post-infectious bronchiolitis obliterans.**

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*CHU de Liège, CHR Liege*Background

Chronic cough is one of the most common causes of medical consultation in children. Etiologies are numerous; asthma or prolonged cough after viral infection are common, but other rare and severe disease must be excluded, particularly if the child presents with other symptoms.

Post-infectious bronchiolitis obliterans (PIBO) is a rare, and severe complication of a respiratory infection. It is defined by a chronic occlusion of the small airways, probably due to an inflammatory response, and resulting in a peribronchial fibrosis. The exact prevalence is unknown.

A PIBO must be suspected in children with a typical bronchiolitis not improving after three weeks, particularly in case of a severe disease requiring mechanical ventilation. The gold standard diagnostic test for PIBO is lung biopsy, but a clinical scoring system (BO-score) was recently validated, including a typical clinical history, chronic hypoxemia, and a mosaic pattern on high-resolution computed tomography (HRCT).

The treatment is based on anti-inflammatory agents, but no recommendation exists concerning the ideal agent and treatment regimen.

Case report

We report the case of 4 years old girl, previously healthy, diagnosed with a PIBO at the age of four, seven months after a severe bronchiolitis. She presented with non-resolving chronic wet cough, shortness of breath at exercise, fatigue, growth faltering. The work up for cystic fibrosis, tuberculosis, immune defect were negative. Several nocturnal oximetry and daytime oxygen saturation measurements revealed a chronic hypoxiema, and a HRCT showing mosaic pattern areas confirmed the diagnosis (BO-score=8). The bronchoalveolar lavage confirmed an associated diagnosis of chronic bacterial bronchitis. The patient was treated with nocturnal oxygen therapy, a six-weeks antibiotics course, high-dose corticosteroids pulse therapy and chest physiotherapy.

Conclusion

Chronic cough is frequent in children, and often diagnosed as asthma. But a non-resolving chronic cough after a respiratory infection, particularly if associated with failure to thrive and hypoxemia, should prompt search for other rare and/or severe causes, such as PIBO. PIBO is a rare but severe obstructive lung disease. Rapid diagnosis and treatment may minimize bronchiolar fibrosis and improve the prognosis; once the disease is established, the treatment is only supportive.

P 68.

Subcutaneous emphysema and pneumomediastinum complicating high-flow nasal cannula therapy: a case report.

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UZ Brussel

Introduction

Humidified Heated High-Flow Nasal Cannula (HHFNC) is a rather novel non-invasive ventilation therapy that seems to be well tolerated and easily administered and air/oxygen is delivered at different flow rates $\geq 2\text{L}/\text{min}$. Evidence suggests that HHFNC provides positive airway pressure, which is variable and not predictable. Such an unpredictable rise in pressure may potentially cause air leak syndromes in children.

Case

A four year old girl, with a previous history of asthma, was admitted on the paediatric ward of a non-university hospital with respiratory distress and fever. Laboratory work-up showed no elevated inflammatory markers and the thorax radiograph revealed a pneumonia of the right lower lobe. Non-invasive ventilator support with high-flow nasal cannula was started at rate $30\text{L}/\text{min}$ ($=2\text{L}/\text{kg}/\text{min}$). Frequent inhalation of salbutamol and intravenous corticosteroids were administered for 24 hours and then interrupted. The respiratory distress and fever persisted, the repeated blood test revealed increasing signs of infection and the radiograph showed growing infiltrates for which intravenous antibiotics were started. In the following days she showed respiratory improvement so the flow and inhalation therapy could be reduced. The third day of hospitalisation she developed an increasing swelling of the neck. Thorax radiograph was repeated on day four and revealed extensive subcutaneous emphysema near the neck, thorax and left arm as well as a pneumopericardium for which a transfer to our university hospital was performed. Low flow oxygen therapy, corticosteroids, magnesium sulphate and more frequent inhalation therapy were used. A CT thorax confirmed subcutaneous emphysema, pneumomediastinum and -pericardium, intrapulmonary air leakage perivascular and pneumonia. Spontaneous resolution was observed in the following days.

Conclusion

We present a case of a girl with subcutaneous emphysema due to mediastinal pneumothorax due to bronchial obstruction and respiratory distress treated with HHFNC. The lack of studies using higher flow rates and the available case reports of serious air leakage in children treated with HHFNC indicate that caution must be exercised and the underlying lung disease, asthma or other, needs to be treated as well. Uncomplicated pneumomediastinum and subcutaneous emphysema can be managed conservatively with analgesia and avoidance of maneuvers that increase pulmonary pressure.

Posters**P 69.****A rare congenital lymphatic malformation presenting as chylothorax at the age of 13 months**

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UZ Gent

Chylothorax is a rare cause of pleural effusion in children, but the most common form of pleural effusion in neonates. It can cause significant respiratory morbidity, malnutrition and immunodeficiency. Therefore chylothorax requires timely diagnosis and treatment. We encountered a 13 month old girl with intrathoracic cystic lymphangioma as a cause of persistent bilateral chylothorax.

The girl presented with 1 week history of cough and fever. Chest X-ray showed white left hemithorax with large pneumatocele at the left upper lobe. She was treated for complicated pneumonia with chest drain and antibiotics, and eventually left upper lobectomy for persistent pleural effusion. After surgery, chyleous pleural fluid was noted and medium-chain triglyceride diet initiated.

Anatomopathology reported a thinwalled cyst and emphysematous changes which led to presumed diagnosis of congenital lobar emphysema with thoracic duct compression. The pleural effusion stabilized and she was discharged. Six weeks later, she presented with low grade fever and bilateral chylothorax. Bilateral chest drains were inserted which drained 800-1500ml fluid per day, despite stopping oral feeds and octreotide at maximal dose. First thoracoscopic thoracic duct clipping failed to decrease lymph drainage. Second thoracic duct clipping was combined with a right pleurectomy with slow decrease of chyle and transition to MCT diet. Revision of anatomopathology with CD31 staining showed cystic lymphangioma. No other localisations of lymphangioma were found. Six months after initial presentation, sirolimus was started because of persistent small bilateral pleural effusion and cystic lesion at the left upper mediastinum with failure to thrive. One year later, the cystic lesion is much smaller, the child thriving, and there have been no recurrences of large chylothorax.

Cystic lymphangioma is a rare benign cystic lesion composed of dilated lymphatic vessels. They usually present at birth or during the first years of life and most often occur in the head and neck. Cases of abdominal or intrathoracic lymphangioma are very rare (<50 in literature). Although rare, early differential diagnosis of chylothorax versus infectious pleural effusion is mandatory. Causative diagnosis requires a multi-disciplinary approach including surgery and anatomopathological examination with specific immunohistochemical staining. Cystic lymphangioma can successfully be treated with combined approach with surgery and sirolimus.

P 70.

Hepatic capillariasis in a refugee child with PICA

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UCL

Background

Hepatic capillariasis is a rare serious hepatic disease caused by the nematode *Capillaria hepatica*. *Capillaria hepatica* accidentally infects humans and is a parasite of mammalian liver primarily rodents. Infection occurs after ingestion of infective eggs.

We describe a 3 years old girl suffering from hepatic capillariasis. This case highlights the severity of the disease and the diagnosis workup.

Case report

A 3 year-old girl living in a refugee centre was referred for persistent high fever, weakness, PICA, abdominal pain, vomiting and hepatomegaly. The parents mention she is eating everything around her and in the nature (PICA). The blood test revealed microcytic anaemia (8,3g/dL), eosinophilia (12750/ μ L), hyperglobulinemia and elevated levels of ALT (360U/L) and AST (207U/L). Serologic tests were positive for *Fasciola Hepatica* and *Toxocara canis*. This result ended up being a false-positive due to a cross-reaction. Liver biopsy was done and revealed adult worms and characteristic eggs of *Capillaria hepatica*. The diagnosis was confirmed by a positive PCR for the same parasite on the liver tissue. At this time, our patient has been treated for 60 days by albendazole and corticosteroids with positive clinical and biological response.

Discussion

The clinical manifestations of capillariasis are nonspecific. The typical triad is fever, painful hepatomegaly and leucocytosis with eosinophilia.

The parasite and his eggs are undetectable in faeces. The larvae mature, mate and lay her eggs in the liver parenchyma and they cannot be excreted. Currently, there is no valid immunodiagnostic method for the diagnosis of hepatic capillariasis. In addition, serological cross-reactivity lead to misdiagnosis. There are false-positive tests results with other worms. Therefore, liver biopsy remains the cornerstone of diagnosis.

The deposition of eggs in the liver parenchyma causes granulomas and liver necrosis, which can lead to potentially fatal liver dysfunction. Thereby, the diagnostic must be quickly mentioned and the treatment initiates.

It consists in an association of anti-nematoid drugs and corticosteroids to reduce inflammation.

Conclusion

Hepatic capillariasis should be taken into consideration in a child with PICA presenting with persistent fever, hepatomegaly and eosinophilia. Liver biopsy remains the gold standard for diagnosis. Due to challenges in making the diagnosis, the prevalence of the disease and it's severity are underestimated.

Posters**P 71.****Listeria meningoenkephalitis in an immunocompetent child**

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*U Liège, UCL*Introduction

Listeria monocytogenes is a rare cause of bacterial meningitis (or meningoencephalitis) representing only 0,7% of all bacterial meningitis. It is known to affect vulnerable groups, such as neonates and immunocompromised people. Immunodeficiency is recognized as the main risk factor of developing neurolisteriosis. No other risk factor of developing neurolisteriosis has been well established. From 1996 to 2018, only 21 previously healthy children with neurolisteriosis have been recorded in literature. Case Presentation A 14-year-old boy was admitted to the pediatric emergency center with high fever, headache, recurrent vomiting and nuchal pain for 5 days. The patient also presented low limb paresis, auditory hallucinations and tongue paresthesia. The physical examination showed signs of meningitis. The initial blood test showed a light inflammatory syndrome (CRP 30mg/L), leukocytosis (10930/mm³) with predominance of lymphocytes and normal glycemia (112 mg/dL). Lumbar puncture showed a relative hypoglycorrhachia (44mg/dL), hyperproteinorrhachia (0.501g/L), and leucorrhachia (217/mm³ for 15 erythrocytes/mm³). The patient was hospitalized on suspicion of viral meningoencephalitis and a treatment by acyclovir was started. In the first 24 hours, the multiplex PCR revealed a positive result for *Listeria monocytogenes*. The EEG showed lightly disturbed trace. Antibiotherapy by high-dose amoxicillin and highdose gentamycin was then installed. The CSF bacterial culture confirmed results two days later (enriched environment). The patient showed clinical improvement after 48 hours of treatment. The duration of the intravenous treatment was 7 days for gentamycin and 21 days for amoxicillin. Screening tests for immunological deficit showed no specific results. Follow-up included auditory testing and cerebral MRI.

Discussion & Conclusion

Listeria meningitis (or meningoencephalitis) has a variety of symptoms ranging from viral to bacterial meningitis (with or without encephalitic component). Thus its diagnosis is rarely thought of in the first place when occurring in an immunocompetent patient delaying adequate treatment and leading to higher risk of severe sequels. Pediatricians should be aware that *Listeria* meningitis (or meningoencephalitis) can occur in healthy patients and that the use of PCR on the CSF can help to make the diagnosis.

P 72.

Refractory chylothorax in children : a case report

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Hôpital de Jolimont

Background/aims

Chylothorax in children is most frequently iatrogenic. It results from thoracic duct injury as a complication of cardiothoracic surgery. Conservative therapy generally consist of pleural drainage of chyle, dietary modification, parenteral nutrition and medications such as somatostatin analogs or glucocorticoids. Less common causes of chylothorax includes congenital/genetic vascular disorders that can be refractory to usual therapeutic measures. Targeted therapy could be the treatment of choice of these rare conditions.

We report this case to highlight a rare cause of chylothorax and its specific management.

Methods

Data were abstracted from medical records

Results

A three year old male child was referred for management of acute respiratory distress. 3 days earlier he had scarlet fever which rapidly resolved after well conducted amoxicillin therapy. Previous medical history was notable for scoliosis. There was a family history of psychiatric disorders on maternal side. Laboratory evaluation demonstrated normal C reactive protein level. Chest radiograph revealed bilateral pleural effusions. Blood cultures were drawn and the patient was given amoxicillin and clarithromycin dual therapy to overcome presumptive bacterial pleuropneumonia. Thoracentesis was performed and was consistent with a chylous effusion. He received bilateral continued pleural drainage for about 8 weeks. Usual therapeutic measures were ineffective. Finally instauration of rapamycin allowed to stop pleural effusion. The workup included lymphoscintigraphy which give unexpected images. Among multiple anomalies, we noted the absence of thoracic duct and the accumulation of nanocolloid in both hands. Pulmonary biopsy showed pleural lymphangiectasias. Genetic analyses revealed heterozygous state for a SHOC2 variant (c.74A>G; p.Glu25Gly).

Conclusion

Systemic congenital lymphangiomatosis is a rare disease characterized by benign, cystic, multifocal areas of lymphatic proliferation. Recently, use of rapamycin, an mTor inhibitor, have been shown to decrease lymphatic proliferation and associated pleural effusion. In our case, lymphoscintigraphy gave surprising results that was reminiscent of extremity lymphoedema seen in newborn with Noonan syndrome. Genetic analyses revealed a novel SHOC2 variant. SHOC2 mutations are known to be involved in a group of diseases called RASopathies. Significance of the discovered variant remains unclear. Further studies are ongoing.

Posters**P 73.****The prevalence of hypercapnia during an acute infection in children on chronic non-invasive ventilation: a retrospective study**

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Backgrounds/aims

The use of chronic non-invasive ventilation (NIV) in children has increased; this brings several challenges. Children with home ventilation are at risk for non-elective hospitalizations, with acute infections as the main cause. This study gained insight in the prevalence of hypercapnia in children on chronic ventilatory support during an acute admission.

Methods

In this retrospective cross-sectional study, we've included children aged between 0-18 years old, on regular use of bilevel positive airway pressure (BiPAP) or continuous positive airway pressure (CPAP) at home, with diagnosis of an acute infection, hospitalized at the pediatrics department or intensive pediatric care unit (PICU). Hospitalizations for non-infectious causes or scheduled admissions and records where data of measured pCO₂ were missing, were excluded. As secondary outcome parameters ventilator settings at time of admission, at 48 hours and at 14 days before admission were analyzed. Mann-Whitney test was used to compare continuous variables, while the Chi-square test or Fisher's exact test was used to compare proportions. For comparison of paired data the Wilcoxon signed-rank test was performed.

Results

There were 43 cases included, where the prevalence of hypercapnia was 23% (10/43) with a mean pCO₂ of 51,7 ± 6.4 mmHg. These children have lower oxygen saturation levels. All the hypercapnic patients had a lower airway infection and also in normocapnic patients this was the most frequent diagnosis. Indicators for the severity of disease were not statistically significantly different in patients with hypercapnia compared to patients with normal pCO₂ values. The data of 48 hours before admission showed that the respiratory rate was significantly higher in patients with hypercapnia.

Conclusion

The overall annual incidence rate for non-elective hospital admission because of an acute infection was 13%. There was a significant increase in respiratory rate in the hypercapnic patients compared to the normocapnic patients 48 hours before admission. Since tidal volumes in the patients were approximately 100 percent of the targeted tidal volume (TTV) before hospital admission, we suggest to increase these settings for TTV when signs of a lower respiratory infection appear.

P 74.

HIV-infected mothers who decide to breastfeed their Infants under close supervision in Belgium: about two cases

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UCL Saint-Luc, Cliniques Saint-Pierre

Background

In most industrialized countries, human immunodeficiency virus (HIV) infection remains a formal contraindication to breastfeeding. However, for the past 9 years, the World Health Organization (WHO) has recommended, for developing countries, that mothers infected with HIV and treated by combined antiretroviral therapy (cART) should breastfeed their infants. HIV-infected women coming from developing countries and living in industrialized settings are increasingly expressing their natural desire to breastfeed. To avoid uncontrolled breastfeeding practices and reduce the risk of mother-to-child transmission of the virus, there is an urgent need to consider the wishes of these women.

Methods

We report two cases in which specific guidelines were implemented in order to support the mothers' choice to breastfeed in Belgium.

Results

As a result of different prophylactic measures including antiretrovirals in mothers and infants and close follow-up, none of the infants were infected.

Conclusions

National or international recommendations for HIV-infected mothers who choose to breastfeed in industrialized countries remain unclear and discordant. There is an unmet need for experts to address this emerging issue and to develop an international consensus for the monitoring and prophylactic management of exposed-infants.

Posters**P 75.****Two cases of hemolytic uremic syndrome in the same childcare center in Brussels**

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CHU Saint-Pierre, Infection Prevention and Control, Common Community Commission (COCOM), PICU, UCL Saint-Luc

Introduction

Hemolytic uremic syndrome (HUS) is a rare disease characterized by the triad of hemolytic anemia, thrombocytopenia and acute renal failure (ARF). It is classically associated with Shiga toxin-producing *Escherichia coli* (STEC). HUS caused by STEC strains is a mandatory notifiable disease in the Brussels-Capital Region.

Cases report

A 2-year-old boy, with no particular medical history, presented to the emergency room with seizures, diarrhea and fever. The diagnosis of HUS was made on biology. PCR detection at the National Reference Center (NRC - UZ Brussels) revealed the presence of *E. Coli* O157: H7 in a stool sample. The child was transferred to a pediatric intensive care unit (PICU) for status epilepticus. He required mechanical ventilation for 12 days and benefited from peritoneal dialysis followed by continuous veno-venous hemofiltration for a total of 6 days. Hospitalization lasted 4 weeks.

This case was notified to the department of Infection Prevention and Control (COCOM) by the childcare physician. In order to put active surveillance in place, a prevention message was sent to the parents mentioning a case of HUS in the childcare and advising to consult a physician in case of signs or symptoms suggestive of gastroenteritis.

Three days later, a 2-year-old girl presented to the emergency room of another hospital with vomiting and a 5 days fever. Her mother brought a letter from the childcare center mentioning STEC infection case. Blood tests revealed the characteristic triad of HUS. The PCR-STEC was negative in both blood and stools samples. The patient was transferred to a PICU but did not require dialysis. She was discharged after 2 weeks.

On account of the contacts between the different sections of the nursery, it was decided to screen all 74 children and 27 adults. Stools were sent to the NRC. In addition to the two hospitalized children, 8 children were healthy carriers of the same strain of STEC; one child had only a brief episode of diarrhea. In order to avoid further contamination, these carriers were excluded from the childcare until two negative cultures were obtained from samples taken 24 hours apart.

Conclusion

This epidemic situation of STEC infection is rarely documented in childcare facilities. We report 2 cases of HUS in the same nursery care with 13 out of 76 children (13%) carriers. This result suggests human-to-human contamination and justifies close follow-up of children living together.

P 76.

Chlorhexidine anaphylaxis after topical use in a 13-year old girl: a case report.

L. Depoorter, J. Vanderkerckhove, M. Grosber, G. van Berlaer, Y. Vandenplas, E. De Wachter

UZ Brussel

Background

Anaphylaxis is a severe life-threatening systemic hypersensitivity reaction. It is a clinical diagnosis based on widely accepted criteria and is characterized by its rapid onset in skin and mucosal changes and/or airway, breathing and circulatory problems. Careful history taking to identify the causing trigger is essential. Evidence of IgE-sensitization on skin prick test or in vitro testing confirms the diagnosis.

Case

A 13-year old girl presented to the emergency department with a sudden anaphylactic event. Detailed medical history revealed that the event initiated immediately after eating her usual lunch consisting of cheese sandwiches, and a glass of cow's milk and 20 minutes after using a topical disinfectant spray Mercurochrome®, containing chlorhexidine digluconate, on a wound on her right knee after a fall earlier that day. She rapidly developed a generalized urticarial rash, dyspnea, throat swelling and nausea. After an antihistamine was administered without sufficient improvement, she presented at the hospital. She had no particular medical history and no history of atopic diseases. Physical examination revealed normal blood pressure, normal oxygen saturation, mild respiratory distress, urticarial skin rash and swelling of her throat. Anaphylaxis was diagnosed. Adrenaline 0.01 mg/kg was injected intramuscular with immediate clinical improvement. Intravenous glucocorticosteroids were administered to treat the lasting cutaneous reaction. Specific IgE-levels showed normal levels for wheat, milk, alpha-lactalbumin, beta-lactoglobulin, casein, cheese and omega-5 gliadin and a total IgE of 19.1 kU/L. Serum IgE-level of chlorhexidine was 4.0 kU/L, corresponding with a class 3 allergy. The diagnosis of a chlorhexidine allergy was confirmed.

Conclusion

Anaphylaxis is a clinical emergency and all healthcare professionals should be familiar with its acute and ongoing management. A careful history to identify triggers is key. Chlorhexidine is a widely used antiseptic and disinfectant. It is also used in many other products, including mouthwashes, eye drops and cosmetics. Delayed type IV allergic reactions such as contact dermatitis are most commonly observed. However, type I reactions such as anaphylaxis have been increasingly reported. Strict allergen avoidance and an anaphylaxis management plan with the use of adrenaline auto-injector and follow-up in an allergy clinic are essential

Posters**P 77.****Benefit of genetic sequencing in the treatment of a tuberculosis disease case.**

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*ULB-CHU Saint-Pierre, Sciensano*Background

Tuberculosis (TB) disease in childhood is difficult to confirm microbiologically due to paucibacillary nature of the disease leading to the lack of drug susceptibility testing (DST). In case of known index case (IC) contact, guidelines recommend to implement treatment based on IC *Mycobacterium tuberculosis* (Mtb) DST. We report a case of extrapulmonary TB (ETB) child presenting a different phenotype of Mtb than that of her linked IC.

Case report

A 47-month-old, presented to hospital with a 1 week history of pyrexia and some limping. Parents reported contact with a contagious TB case 3 weeks earlier. She did not receive BCG vaccine. Physical examination revealed a febrile alert girl, a tuberculin skin test of 17 mm induration. Limping was not objectified. Chest X-ray revealed normal. Laboratory test demonstrated a high inflammatory syndrome. Hepatic ultrasounds revealed micronodular pattern suggestive of miliary TB. As soon as diagnosis was suspected, a 4-drug classic anti-TB therapy was started. After 2 weeks, a quinolone was added to the treatment given result of phenotypic IC DST demonstrating isoniazide resistance (INH-R) of Mtb. Two months later, Mtb susceptible to all first-line drugs was cultured from child's gastric aspirates. Given paradoxical results and the fact no other IC source has been found, genotypic comparison of child's and IC's strains were performed. WGS confirmed transmission of IC's strain to the child and did not show any classic INH-R mutation on both strains. Evolution was favorable with treatment based on child's phenotypic results.

Discussion

WGS allows rapid detection of mutations known associate with resistance for anti-TB drug and to determine transmission events based on results of genotypic profiles obtained. Phenotypic DST (PDST) makes difficult to institute an optimal individualized treatment before several weeks. In our case, difference on phenotypes could be explained either by earlier contact with the IC than which reported by parents or by current infection of IC with two populations of Mtb showing different INH susceptibility. One of these population demonstrating INH-R which has not yet been identified genotypically.

Conclusion

Bacteriological confirmation of TB in childhood can be difficult but is necessary to obtain PDST and adapt treatment. PDST results may differ between child's and IC's strains of Mtb due to resistance mutations. WGS enables to confirm transmission in event of uncertainty.

P 78.

Cross-transmissions of *Pseudomonas aeruginosa* in a pediatric cohort with cystic fibrosis

J-C Beghin

ULB - HUDERF

Introduction

Pseudomonas aeruginosa (PA) infection is the most common pulmonary infection in patients with cystic fibrosis (CF). Chronic PA carriers defined as patients with more than 50% of the airway cultures positive for that germ during 1 year, present faster clinical deterioration. Molecular typing studies have confirmed the presence of PA cross-transmissions in unrelated patients. The objectives of this study were to highlight the occurrence of PA cross-transmission within our institution and to assess the current hygiene strategies.

Design

This prospective mono-centric study was conducted in 2019 during 4 months in a cohort of children with CF followed at the Hôpital Universitaire Des Enfants Reine Fabiola Cystic Fibrosis' Center (Université Libre de Bruxelles). Airway cultures positive for PA were obtained during routine consultation and submitted to molecular typing by pulsed field gel electrophoresis (PFGE). This technique compares different large bacterial DNAs fragments in order to detect similar strains suggesting cross infection. An epidemiological survey was also carried out to find the location of cross-transmission (hospitalization unit, physiotherapy center, school, family and extra-curricular activity). Finally, a questionnaire has been distributed to assess patients' and caregivers' knowledge of PA transmission modes.

Results

Among the 77 patients followed-up in the center, 34 patients were included in the study. Seven patients (20.59%), including 5 chronic carriers, presented PA in their respiratory sample. Among these 7 patients, 5 presented several PA strains (2 or 3). A total of 14 PA strains were genotyped by PFGE. Molecular typing results showed PA common strain in two 13 years old chronic carriers (28.57%). Cross-transmission was suspected and confirmed by descriptive epidemiological data (contacts in and outside the hospital). The PA strain identified (VIM-2 gene) highly suggested nosocomial acquisition. PA transmission modes were poorly understood in young children (7-11y) and adolescents (16-21y).

Conclusion

This study showed cross-transmission between 2 unrelated patients in a cohort of 7 PA infected patients. This proportion is comparable to Australian, New Zealand and English centers. These results encourage our center to improve hygiene strategies in its different care structures and to regularly assess the efficacy, feasibility, comprehension and psychosocial impact of these new cross-infection prevention measures.

Posters**P 79.****Invasive Haemophilus Influenzae infections in the pediatric patient a case series**

D. Vermeulen, P. Schelstraete

*UZ Gent*Background/Aims

Vaccination for Haemophilus Influenzae type b (Hib) is provided for free in the Belgian national immunization program since 2002 to prevent invasive infections, especially in children younger than 5 years. This had led to a sustained reduction in serotype b infections. However several studies have reported increasing trends in non-capsulated H. Influenzae, now the leading cause of invasive H. Influenzae disease in all age groups, particularly among groups more susceptible to infection.

Methods

Charts of pediatric patients, diagnosed with invasive H. Influenzae infection between January 2015 and December 2019 in the University hospital of Ghent, were checked for H. Influenzae serotype, vaccination status and comorbidities.

Results

We present 7 cases, all younger than 5 years of age, with an invasive H. Influenzae infection. Four of them presented with sepsis and three of them with meningitis. Five cases had a non-capsulated H. Influenzae infection, one case had H. Influenzae serotype F infection and in one case the serotype was not known. Only one patient was fully vaccinated for Hib. Three patients had different types of comorbidities.

Conclusion

Non-capsulated H. Influenzae was in our center the most frequent type causing invasive H. Influenzae infection since 2015 in pediatric patients. This results corresponds with epidemiological studies. Future surveillance of invasive H. Influenzae infections therefore should contain all serotypes.

P 80.

Homozygous nonsense mutation in RC3H1, which encodes Roquin-1, causes a novel immune dysregulation syndrome characterized by severe hyperinflammation

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UZ Gent, VUB and many others

Background

Roquin-1 is a posttranscriptional repressor protein of key immune-regulatory proteins, such as ICOS, OX40 and TNF α . In mice was shown previously that genetic mutations weakening or inactivating Roquin-1 or its paralog Roquin-2 result in a wide spectrum of immune-related diseases such as mucosal inflammation, lymphoproliferative interstitial lung disease and lupus-like autoimmune disease.

Case

The proband was followed for several years at another hospital because of mild mental retardation, sensorineural hearing loss, chronic diarrhea and petechia. At the age of 11 year, he presented with prolonged fever and liver failure. Neurometabolic work-up did not allow identification of an underlying molecular defect. He was found to fulfill all criteria for hemophagocytic lymphohistiocytosis (HLH). Using whole exome sequencing a homozygous novel stop codon mutation was detected in RC3H1 (c.2414G>A, p.688R/*) encoding roquin-1. Comparing the human R688* roquin-1 variant with a murine M199R variant revealed a striking phenotypic resemblance, both in immune cell activation and hypercytokinemia. Mechanistically, the mutated R688* roquin-1 did not localize to P-bodies and failed to interact with the CCR4-CNOT1 deadenylation complex, explaining how it was unable to control inflammatory cytokines such as TNF α , IL-2 and IL-17A.

Conclusion

The proband is the first patient reported with Roquin-1 defect. Our results show that inactivation of Roquin-1 provokes a hyperinflammatory syndrome by failure to control ongoing immune activation. However, if the mental retardation of the patient is a consequence of the immune dysregulation or a direct effect of posttranslational regulation of neuronal genes by RC3H1 remains to be investigated.

Posters**P 81.****Cluster of invasive meningococcal disease caused by a meningococcus serogroup B in a nursery school in Wallonia, Belgium in 2018**

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Sciensano, ULB-CHU Saint-Pierre, UCL Saint-Luc, CHC – Liège, CHU Liège, Agence pour une vie de qualité, infection prevention and control

Background

In Belgium, the estimated annual incidence of invasive meningococcal (IMD) is 1.14 cases/100.000 inhabitants. The majority of cases are of serogroup B with around 60 cases/year since 2012. Clusters in preschool settings are described, and time interval between index case and subsequent cases is generally inferior to 3 weeks. Here, we report a cluster of 3 IMD cases caused by a meningococcus B (MenB) occurring in a nursery school over a 9 month period.

Methods

Case-investigation and contact-management was coordinated by Agence pour une Vie de Qualité (AVIQ). Microbiology testing of strains was performed by the National Reference Center (NRC).

Results

The first cases of IMD (purpura fulminans and meningitis respectively) occurred in April and June 2018 in 2 previously healthy 4 year old boys attending the same pre-school classroom. Both times, following regional guidelines, ciprofloxacin chemoprophylaxis was given to close contacts including household contacts, toddlers in the same classroom and their teacher. The third case, a previously healthy 5 year old boy, developed sepsis and meningitis end of December 2018, 6 months after case 2. This child was not in the same classroom as case 1 and 2 but in the same age-group and in the same classroom as the twin of case 2.

All three cases recovered. Case 2 and 3 had received chemoprophylaxis prior to disease.

Microbiological analyses revealed a MenB with identical genotype for case 1 and 3 (cc269). The strains had identical antibiotic sensitivity profile, including absence of fluoroquinolone resistance. The second case was diagnosed by positive PCR (on cerebrospinal fluid) for MenB, performed in the hospital (no strain/sample sent to NRC).

Infectious Diseases Experts were contacted to discuss post-exposure management following case 3. Chemoprophylaxis (azithromycin) was once again given to contacts as defined above. Vaccination was recommended after final microbiological results predicted coverage by the 4CMenB vaccine. Forty children out of fifty have been vaccinated (2 doses).

In the 12 month follow-up after case 3, no additional cases were reported by the nursery school.

Conclusion

IMD clusters are difficult to manage and generate high public anxiety, particularly in case of an ongoing cluster, despite contact tracing and management. Recommendations on how to manage these situations are lacking. The use of 4CMenB in post-exposure prophylaxis is poorly documented and requires further investigation

P 82.

Phage therapy to allow liver transplantation in a toddler infected by an extensively-drug resistant *Pseudomonas aeruginosa*

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UCL Saint-Luc, Hôpital Militaire Reine Astrid

Background

One of the therapeutic strategies to tackle multidrug-resistance is the use of bacteriophages. The phage productions of the Queen Astrid Military Hospital (QAMH), Brussels, are in agreement with an adapted Belgian framework, set up in concertation with the Federal Agency Medicinal and Health Products. One septicemic case, infected with MDR *Pseudomonas aeruginosa*, was effectively treated with phage therapy (PT) intravenously (IV) for ten days (as monotherapy) in the QAMH in 2016.

Methods

To present a case on well-tolerated phage therapy in a transplanted child with uncontrolled liver infection due to XDR *Pseudomonas aeruginosa*

Results

A 14-month-old infant was first transplanted on 26/09/2018 for biliary atresia, using an ABO-incompatible living donor. After multiple complications (rejection, biliary digestive anastomosis perforation, cytomegalovirus infection and cholangitis with bacterial sepsis), a first carriage of MDR *Pseudomonas aeruginosa* (only sensitive to colimycin) was shown at day 20 post-transplant. One month later he developed an XDR *Pseudomonas* sepsis (colimycin R, aztreonam I and gentamycin S) treated with IV colimycin, aztreonam and gentamycin. Blood cultures remained positive and intrahepatic collections appeared on the abdominal ultrasound. Decision was taken on 23/11/2018 to drain one of the collections and to start with experimental IV and in situ PT after ethical committee approval. Twenty milliliters (2 mL/kg) of purified bacteriophage cocktail BFC1 (phage load of 107 pfu/mL) containing two sensitive bacteriophages active (defined in vitro in retrospect) against the patient's *P. aeruginosa* isolates, were administered as a 6-h intravenous infusion for 72 days once a day till two weeks after he received a new ABO compatible liver transplant on 03/02/2019 (so a total of 86 days of IV phage therapy). Intralesional injections with BFC1 were performed with 1 mL during 7 days and stopped due to abdominal swelling and discomfort. During liver transplantation 250 mL of BFC1 was used to rinse the abdominal cavity; the procedure was well tolerated.

Conclusions

To our knowledge, this is the first case of life-saving IV PT in an infant. The long IV PT treatment of 86 days was safe – no side effects could be related to IV PT. Ten months after retransplantation the 28-month-old child remains well on immunosuppressive medication.

Posters**P 83.****FINCA disease presenting as Severe Childhood onset Interstitial lung disease (chILD) with neurodevelopmental disorder**

I. Van Dijck, F. Vermeulen, M. Proesmans, M. Boon

*UZ Leuven*Background/Aims

FINCA disease should be considered as a possible genetic cause in children with chILD and neurodevelopmental disorders. The disease course can be variable.

Results

We present a 4-year old patient with psychomotor retardation, severe chronic lung disease with initial suspicion of childhood interstitial lung disease (chILD), severe course of respiratory symptoms needing recurrent admission to the intensive care unit and chronic oxygen treatment, nutritional problems and failure to thrive, dilatation of the aortic root and macrocytic anemia. Chest CT showed bilateral ground glass opacities with honey combing and air bronchograms with multiple subpleural cysts, mainly in the lower lobes. Lung biopsy showed interstitial fibrosis resembling non-specific interstitial pneumonia (NSIP) and type 2 alveolar cell hyperplasia.

Whole exome sequencing confirmed compound heterozygous mutations in the NHLRC2 gene, causing FINCA disease (fibrosis, neurodegeneration and cerebral angiomas) in this patient. This gene is expressed in multiple tissues and thought to have a role in apoptosis and tissue fibrosis. Only a few cases have been reported in the literature, all with rapid progressive symptoms and early death (before the age of 2 years). However, the respiratory status of our patient is currently stable at the age of 4 years, home oxygen was recently interrupted and oral nutrition was successfully restarted. This is only the 5th patient described, and the longest living patient.

Conclusion

New entity in pediatric pulmonology characterized by cerebropulmonary symptoms

LO 10.

Infliximab trough levels at the end of induction will predict endoscopic remission in paediatric patients with Inflammatory Bowel Disease

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UZ Leuven, KUL

Background

Although higher infliximab (IFX) trough levels (TL) have been associated with better outcomes, the ideal predictive sampling time and cut-points to achieve endoscopic remission remains unclear in children with inflammatory bowel disease (IBD). Therefore, we evaluated the pharmacokinetics of IFX during induction to predict outcome of IFX.

Methods

All children with Crohn's disease (CD) or ulcerative colitis (UC) starting IFX therapy (5 mg/kg at wk. 0-2-6-12) for active luminal disease from May 2017 till May 2019 were followed prospectively. IFX levels were measured by ELISA (TL at wk. 2-6-12, peak at wk. 0-2-6 and intermediate at wk. 1-4). IFX levels and cumulative drug exposure (AUC till wk. 12) were correlated with outcome at mo. 6. Clinical remission was defined as PUCAI/PCDAI <10, biochemical remission as CRP ≤5 mg/L + ESR ≤20 mm/h, endoscopic remission as SES-CD <3 or Mayo endoscopic sub-score =0 and deep remission if both clinical + endoscopic remission.

Results

A total of 252 serum induction levels were included from 32 patients (20 CD and 12 UC; 38% male; median age at start of IFX 13.8 years [11.3-14.9]). Clinical remission was achieved in 24 (75%) patients and 18 (56%) were in endoscopic remission (all in deep remission) at mo. 6. Endoscopic remission at mo. 6 was associated with significantly higher median IFX TL at wk. 4 (38.8 µg/mL [24.3–46.0] vs 23.5 µg/mL [10.5–36.6], p=0.017), at wk. 6 (19.9 µg/mL [10.1-26.3] vs 11.1 µg/mL [3.7–19.9], p=0.031), at wk. 12 (9.6 µg/mL [5.5–11.9] vs 3.5 µg/mL [2.7–7.2], p=0.004) and higher AUC wk. 0-12 (4574.7 µg*day/mL [3783.0–5160.8] vs 3722.9 µg*day/mL [3102.2–3991.9], p=0.008). Median IFX TL at wk. 12 were significantly higher in children with clinical remission (8.6 µg/mL [5.1–12.0] vs 4.3 µg/mL [3.1–5.9], p=0.033), but not for biological remission (6.7 µg/mL [4.0-12.0] vs 4.3 µg/mL [1.2–7.2], p=0.250) at mo. 6. ROC analysis identified an wk. 12 IFX TL ≥ 5.0 µg/mL and an AUC wk. 0-12 ≥ 4056.0 µg*day/mL as minimal target to achieve endoscopic remission at mo. 6 (AUROC: 0.796 [95%CI: 0.62-0.97] and AUROC: 0.778 [95%CI: 0.61-0.94] respectively).

Conclusion

Adequate IFX exposure during induction in paediatric IBD patients is associated with significantly better clinical, endoscopic and deep remission rates at mo. 6. Model-informed precision dosing can assist physicians to achieve optimal exposure during induction more precisely (and rapidly) what is essential for an optimal outcome.

Poster Walk**PW 18.****A systematic review on health care implications of a vegetarian diet in children**

L. Leclair, J. Toelen

*KUL*Objective

A vegetarian diet is a diet that excludes meat, while a vegan diet excludes all animal products (including dairy and eggs). Its health benefits and risks have mainly been described in adults, so that there is currently no consensus about implications of a plant-based diet in children. Vegetarianism and veganism is however gaining rapid popularity, which necessitates further research to reach evidence based guidelines.

Methods

This systematic review reports the currently known health benefits and harms of a vegetarian and vegan diet in children of 0-18 years old. This review was written according to the PRISMA-guidelines. On October 5, 2019, we searched the databases of PubMed, Embase, CINAHL, Web of Science, Cochrane Library and ProQuest. A set of predefined in- and exclusion criteria was used to screen eligible studies for inclusion.

Results

A total of 18 comparative cross-sectional studies were included in this review. The vast majority of studies were published in western countries after the year 2000. The median sample size of vegetarian children is 53 compared to 51,5 omnivorous children. In total, data was obtained from 1594 omnivores, 1159 vegetarians and 156 vegans. We extracted health outcomes from each study to be reviewed in one of the three different categories: anthropometrical, cardiovascular and biochemical.

Conclusion

There appear to be health benefits linked to a vegetarian diet in children, mainly coming from a lower incidence of modifiable cardiovascular risk factors. Although it is plausible that similar health benefits could be reached on a well-balanced diet that does not completely exclude meat. Parents who want to raise children on a vegetarian diet should be educated about the dietary sources of iron, vitamin B12 and calcium, since some studies observed a higher incidence of deficiency. Supplementation with vitamin D is recommended for all children regardless of diet. Data about a vegan diet was insufficient to draw conclusions about health benefits or harms. This review identified several knowledge gaps when it comes to the safety of feeding a plant-based diet to children.

PW 19.

Bowel Function in Children With Low-Type Anorectal Malformation After Surgical Repair

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UZ Gent

Aim

The aim of this study was to search for any association between demographic, clinical, and therapeutic characteristics of patients with a low-type anorectal malformation (ARM) and their long-term bowel function.

Methods

In this retrospective study, 108 patients were contacted, of which 80 patients (74%) were included. Demographic, clinical, and therapeutic information was obtained from the patients' medical records. The standardized Rintala questionnaire (resulting in a bowel function score (BFS)) was used to evaluate bowel function. Data was analyzed with SPSS 26 (significance level = 0.050).

Results

80 patients (21 boys (26.3%)) were included, 70 (87.5%) presented with a rectoperineal fistula, 9 (11.3%) with a rectovestibular fistula, and 1 (1.2%) with anus imperforatus. The Mann-Whitney U test showed a significant negative impact of trimming of the anorectum ($p = 0.003$) and the presence of a developmental disorder ($p = 0.013$) on bowel function. Patients with sacral/spinal anomalies also showed a negative trend on the BFS score ($p = 0.086$), but was not significant probably due to a small patient group. The other characteristics (sex, prematurity, time of diagnosis, preoperative interventions, dehiscence, time of operation, and ARM type) were not significant. Multiple linear regression confirmed these results.

Conclusions

Trimming and the presence of a developmental disorder have a significant negative impact on the bowel function of patients with a low-type ARM. A negative trend was observed in patients with sacral/spinal anomalies. These results may help the clinician in determining the prognosis for patients with low-type ARM, and, if confirmed by later studies, may result in a different surgical approach.

Poster Walk**PW 21.****Impact of Nutrition Support Team (NST) funding on the quality of care of Home Parenteral Nutrition (HPN) in children with benign diseases**

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ULB - ERASME, UCL Saint Luc, CHC Chrétien, KUL, UZ Leuven, UZ Gent, UZ Antwerpen

Background

Five Belgian academic hospital NST received from 2011 to 2017 a funding from the NHI for the follow-up of children HPN-patients with benign diseases. We reviewed the NHI requested quality of care annual reports aiming to analyse the impact of their funding on NST performance while comparing it with existing data.

Methods

Patient data were extracted from seven annual reports (2011-2017) of five Belgian NST academic units involving only children HPN-patients with benign diseases. Reported data are: Age, HPN-incidence and prevalence, HPN-related hospitalisation days/yr, catheter-related blood stream infection/1000 HPN days(CRBSI), number of catheter replacements/yr and patient autonomy. Statistical analysis involves linear regression.

Results

Mean children prevalence was 12,4/million/yr, range [10-14,8] and mean children incidence 2,6 patients/million/yr, range [0,8-4]. Mean age: 6,8yr range [7months-17yr]; gender ratio F/M-ratio 1,97. This represented 177 catheter yr over 7yr. The main HPN indication was short bowel syndrome 55%. Mean rate of CRBSI was 1,04/1000 HPN-days, range [0,6-1,6]. The mean number of catheter replacement/yr HPN was 0,47 range [0,40-0,56]. The mean number of HPN-related hospitalisation days/yr was 4,9 range [4,0-5,7]. Average 62% of children are able to go to school with parents continuing their working activities.

Conclusion

This study demonstrates that the funding of NST in academic hospitals with NST teams has allowed to assess and ultimately to control the number of new cases, to show lower HPN-complications than data, and to favour patient autonomy.

P 84.

Acute cholecystitis in a patient with normal imaging findings on initial presentation

B. Boon, K. Poschet, M. Claeys, A. Calders, A. Delmotte, F. De Meulder

GZA Sint-Vincentius

Background

A 3 year old toddler presented with vomiting, high fever and acute epigastric pain. The pain was colicky without radiating. Clinical examination revealed a diffusely tender abdomen on palpation. Vital signs were normal.

Methods

Laboratory investigations showed high signs of inflammation (CRP of 166 mg/dl, leukocytosis of 24000/mm³). Abdominal ultrasound was performed to evaluate for possible causes of acute abdominal pain and displayed unilateral mild hydronephrosis without any other abnormal imaging findings. Urinalysis was negative. A CT scan performed 2 days later, however, revealed significant gallbladder wall thickening with signs of necrosis and pericholecystic hepatic abscess formation. Repeated blood tests showed markedly reduced signs of inflammation with normal cholestatic liver enzymes. Since the clinical evolution, as well as the laboratory signs were favorable, a conservative approach was adopted. The patient received pain medication and made a full recovery.

Results

Repeated ultrasounds confirmed the pathologically thickened gallbladder wall with multiple hepatic abscesses. The gallbladder had a maximum thickness of 9 mm, where 3 or 3.5 mm are generally used as cut-off values for significant gallbladder wall thickening. Cholecystolithiasis or sludge was never visualised. Additional diagnostic testing to screen for underlying causes of pediatric acute cholecystitis was performed. In contrast to adult acute cholecystitis, acute cholecystitis in children is most often acalculous in origin. Fecal polymerase chain reaction (PCR) testing for *Cryptosporidium*, *Entamoeba histolytica* and *Giardia* was negative. No parasites were found on microscopic fecal investigation. The underlying cause of the acute cholecystitis remained uncertain.

Conclusion

Our patient presented with typical symptoms of acute cholecystitis, namely fever, acute abdominal pain and vomiting. Since acute pediatric cholecystitis is rare, it is often not included by health care providers on the initial differential diagnosis of a child with symptoms suggestive of acute cholecystitis. This frequently leads to substantial delays in diagnosis. The golden standard for diagnosis remains ultrasound of the gallbladder, with a thickened gallbladder wall as the most reliable finding. The normal imaging findings on initial evaluation in the presented case are highly unusual.

Posters**P 85.****Acholic stools and the diagnosis of a choledocholithiasis**

A. Lambiri, M. Maka

*EpiCURA Hôpital*Introduction

We describe a case of obstructive cholelithiasis with dilatation of the biliary tract in a 4-month-old male infant who presented to the emergency room with acholic stool and liver enzymes alterations

Case report

A 4 month old- male presented to the emergency room with pale stool. The work-up revealed an increase in the transaminases levels and a direct hyperbilirubinemia. The ultrasound showed an obstructive cholelithiasis with a secondary dilatation of the biliary tract

The patient was initially treated with endoscopic retrograde cholangiopancreatography (ERCP) and sphincterotomy followed by laparoscopic cholecystectomy. Shortly afterwards, he presented the same symptoms with an increase in the liver enzymes alterations. The ultrasound showed a choledocholithiasis. He was therefore taken for a secondary ERCP followed by biliary stenting. The intervention was successful, his transaminases and bilirubin levels declined and his initial symptoms disappeared.

Conclusion

There are many risk factors associated with the development of cholelithiasis in newborns and infants. However, it can also be idiopathic. The overall prognosis is favorable and the managerial approach is diverse.

P 86.

Seronegative autoimmune hepatitis in children: An atypical entity to mention

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CHU de Liège, U Liège

Introduction

Autoimmune hepatitis is a rare disease that most frequently affects young adults and children including infants. When no specific viral or autoimmune markers are identified in a child presenting with hepatitis, other causes of liver disease are searched for, including drug-induced or metabolic diseases, biliary diseases or malignant conditions infiltrating the liver. There are however instances when none of the known causes are found. Such condition is referring to as “seronegative autoimmune hepatitis”.

We report the case of a 13 years old girl with an acute liver failure and histological features pleading for an autoimmune hepatitis but with no specific markers and negative autoimmune serology. A positive response to immunosuppressive treatment was noticed with clear regression of transaminase after few weeks of treatment.

We describe the several similarities between seronegative hepatitis and autoantibody-positive hepatitis that supports the autoimmune etiology in common between them. The arguments in favors includes 1- histological criteria, 2- positive response under immunosuppressive therapy, 3- combination with other autoimmune diseases simultaneously or secondarily, 4-family medical history with autoimmune disease.

Conclusion

Autoimmune hepatitis is a chronic and progressive inflammatory disorder that's characterized by the presence of specific circulating autoantibodies and increased levels of immunoglobulin G. A seronegative form of AIH responsive to immunosuppressive therapy is also reported in several pediatric studies and must be included in the diagnoses as a distinct entity in childhood. Immunosuppressive treatment should be promptly started despite the lack of serological markers and even in children with normal or low levels of serum gamma globulins provided that all other underlying conditions have been excluded.

Posters**P 87.****Food Protein-Induced Enterocolitis Syndrome: an emergency diagnosis we must know!**

C. Micin, O. Guidi, M-C Seghayé, E. Bequet

*U Liège, CHU de Liège*Background

Food protein-induced enterocolitis syndrome (FPIES) is a non-immunoglobulin E mediated gastrointestinal food hypersensitivity. In acute setting, symptoms appear 1 to 4 hours after ingestion of the target protein by profuse vomiting, sometimes associated with diarrhea. It can lead to a critical condition, causing hypovolemic shock and involving the child's vital prognosis. Unfortunately, with its non-specific symptoms, the diagnosis of FPIES is often made after multiple symptomatic episodes.

Clinical case

We report the case of a 6-months-old girl who had several episodes of acute FPIES. Hospitalizations, with hemodynamic management, were necessary and the diagnosis was only suspected after 3 episodes. Fainting due to severe vomiting in a gastroenteritis context was the diagnosis made during the first two hospitalizations. Then, dietary diversification normally continued. Two months later, a new episode of lethargy appeared, leading to the diagnosis of FPIES induced by solid food. Suspected food triggers, such as potatoes and leeks, have been removed, but three new acute crises have occurred.

In this case, considering the difficulties in accurately identifying the proteins causing the syndrome, a strict diet with a cow's milk protein hydrolysate has been recommended.

Discussion

FPIES is difficult to diagnose. The clinical features are, most of the time, confused with a simple digestive infectious disease. In case of repetitive vomiting and lethargy occurring after a meal, every physician must evoke FPIES. The chronic form of FPIES is characterized by persistent vomiting, weight loss, failure to thrive...

The diagnosis of FPIES is based on one major criterion (vomiting within the following 1 to 4 h after ingestion of the suspect food) and at least 3 minor criteria.

FPIES is more frequently observed with cow's milk or soy. But in FPIES induced by solid food, acute episodes may be more severe and multiple food triggers may be involved. An early diagnosis is even more important.

Trigger proteins can be identified by using oral food challenge in-hospital.

Conclusion

The diagnosis of FPIES is delayed because the symptoms are wrongly attributed to a benign viral process. The treatment consists in a diet excluding trigger proteins. Usually, FPIES spontaneously resolves by the age of 2 to 6.

Given the serious acute condition and the specific hemodynamic management, FPIES is a disease to be known by every physician.

P 88.

When the bowel dances the twist: A rare case of sigmoid volvulus in a child

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UZ Brussel, VUB

Introduction

Acute abdominal pain in children is a frequent presentation in the emergency department (ED). Differentiating aetiologies is of major importance, as a minority of these children will need urgent care. We present a rare case of a sigmoid volvulus (SV) in a child presenting to ED with acute abdominal pain. Missing the diagnosis may result in life-threatening complications such as sigmoid gangrene/perforation, peritonitis, sepsis, and death.

Case

An 11-year old boy with a 2-year history of severe constipation, presented with acute abdominal pain. Clinical exam showed a distended, acute abdomen with clangor. Abdominal X-ray showed bowel obstruction with an extremely dilated colon. CT showed a dolichosigmoid with whirlpool sign of the mesentery and a secondary colonic dilatation suggestive of SV. Initial rectal tube placement deflated the abdomen and released lots of stool and gas. A conservative approach was maintained with nasogastric aspiration and nil by mouth. Major gastric discharge after 48 h warranted a control X-ray showing a possible new or persisting SV. Endoscopy could not confirm this, but showed a dilated and atone colon. Although oral intake had resumed successfully on day 5, clinical exam on day 8 revealed clangor again. Abdominal X-ray was again suggestive of SV, confirmed by contrast enema which could however not reduce the SV. Endoscopic detorsion was followed by a laparotomy for sigmoidectomy and placement of a terminal colostomy because of the dilated bowel. The resected part was sent to the anatomopathological analysis to exclude Hirschprung's disease. Also, blood was drawn to exclude genetic causes for intestinal obstruction. The boy recovered well afterwards.

Discussion

SV is rare in children. Diagnosis is based on history, clinical exam and imaging. Typical is a whirlpool sign on CT. Initial treatment consists of detorsion by endoscopic exsufflation or contrast enema with a risk of unsuccessful detorsion or perforation. If successful, elective sigmoidectomy is the gold standard, because of the high recurrence risk (>65%). If unsuccessful, emergency surgery should be performed. Constipation can be a cause of SV, but other associated conditions, such as Hirschprung's disease or intestinal pseudo-obstruction, are reported in 30% and should be ruled out.

Posters**P 89.****Rectal bleeding in infants: a clinical sign not to be neglected**

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*CHU de Liège, U Liège, CHR de la Citadelle*Background

Rectal bleeding is a regular cause of consultation in the pediatric out-patient clinic or in the emergency department. Its exact prevalence in children is unknown but some studies report it to be about 0,3%. Rectal bleeding has generally a benign cause and does not jeopardize hemodynamics. However, in rare situations, rectal bleeding may become a real vital emergency needing rapid recognition and treatment.

Case Report

We report on the case of a 10 months-old infant with rectal bleeding that led to the diagnosis of an ulcerated Meckel's Diverticulum. The anamnesis revealed a transitory and self-limiting episode of rectal bleeding two months earlier. At the time of presentation in our emergency department, the patient was in good general state with normal hemodynamic parameters, he did not show any sign of digestive discomfort or abdominal pain. The clinical abdominal and proctologic exams were reassuring. Laboratory examination didn't reveal any abnormalities apart of a discrete hypochromic microcytic anemia (Hb 10,5 g/dL). Coagulation tests were normal, the stool culture negative.

The possible diagnosis of a Meckel's diverticulum was hint at abdominal echography that showed the presence of a digestive formation with a diameter of 1,7 cm in the para-umbilical region. Therefore, the patient underwent an explorative laparoscopy during which the formation was removed. The anatomo-pathological analyzes confirmed the diagnosis of an ulcerated Meckel's Diverticulum. The post-operative evolution was uneventful.

Conclusion

This case points out the importance of a detailed anamnesis, a rigorous clinical examination and in particular the importance of complementary examinations that should always be considered when facing a rectal bleeding with no evident causal factor. The Meckel's Diverticulum is the most frequently encountered digestive malformation. It manifests often in an atypical way, staying asymptomatic for long periods of time. Clinical manifestations may be clinical signs of acute abdomen, of intestinal obstruction, of intussusception or digestive bleeding. With respect to the possible mortality still related to a complicated Meckel's Diverticulum, clinicians should always be aware about the danger to trivialize rectal bleeding that should therefore be explored.

P 90.

An interesting case “out of paper” with celiac disease leading to xylophagia: a case report

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UZ Leuven

Case report

A six year old girl without medical history has been seen at the consultation on the division of pediatric gastroenterology because of the ingestion of paper (in literature known as “pica”, more specific “xylophagia”) since six months. There was a merging failure of intake of normal nutrition and secondary weight loss, with also abdominal pain and fatigue.

Clinical examination did not expose something besides a tired-appearing young girl with clear weight loss of 2 kg. She was diagnosed with pica. Blood analysis showed an important iron deficiency anemia (hemoglobin 7,2 g/dL). This anemia could provoke pica, yet why would this girl have such an important iron deficiency? Iron supplementation was started, yet one week later more blood results were known and screening for celiac disease - given the vague clinical presentation - showed an immeasurably high value of anti-tissue transglutaminase antibodies (>128U/ml) with normal total IgA. Gastroscopic evaluation confirmed the diagnosis of celiac disease (chronic duodenitis with severe villous atrophy, Marsh classification 3C). The girl was put on a rigorous gluten free diet.

Clinical checkup one month later showed a child in good clinical condition, with a favorable weight gain and almost disappearance of xylophagia. Biochemically there was a slowly increasing hemoglobine with still low iron supplies. Anti-tissue transglutaminase antibodies remained immeasurably high, however showed - after dilution - a meaningful decrease (1405 U/ml before diet, 654 U/ml on gluten-free diet) in absolute values. In the following months, there was a biochemical recovery and total disappearance of the pica behavior.

Conclusion

In this case, the diagnosis of celiac disease with secondary deficient absorption of nutrients with the onset of iron deficiency anemia and presumably a subsequent presentation with xylophagia, was made within one month after presentation with a minimum of examinations. After confirmation of celiac disease with intestinal biopsies, the patient was put on a gluten free diet with good clinical evolution and resolution of pica. Take home message: this case underscores the need to weigh in mind the possibility of celiac disease in more indeterminate physical complaints.

Posters**P 91.****Constipation as the only symptom of ovarian teratoma: about a case.**

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*University Hospital Liège, University of Liège, Regional Hospital La Citadelle*Background

Constipation is very common in the pediatric population affecting 30% of children and adolescents. Most often, its origin is functional, but it is important to keep in mind that its causes can also be medical or surgical.

Clinical case

A 12 year old girl, with an uneventful medical history, is admitted to the pediatric emergency department for intermittent abdominal pain, constipation and abdominal bloating for 3 weeks, with a marked increase of the symptoms on the last week. She also complained about dysuria and small and frequent blood losses at menstruation for the last 2 months. The clinical examination shows a significant increase of the abdominal perimeter with a decreased peristaltic and a diffuse dullness to percussion, without pain on palpation. The rest of the clinical examination is normal. The abdominal ultrasound, supplemented by the abdominal CT, reveals a voluminous cystic partitioned abdominal mass with intra-lesional calcifications. The mass is 30 cm on its large axis, originates from the uterine appendages and is compatible with a mature teratoma. Given the volume of the mass, there is an intestinal compression, a right uretero-hydronephrosis and compression of the inferior vena cava. The surgery consisted of a right annexectomy and allowed the removal of the entire mass that weighted 2500g. Anatomico-pathology confirmed the diagnosis of mature, multi-cystic teratoma of the ovary with intact serosa. To date, no further treatment is required and rigorous medical monitoring has been implemented.

Discussion

Teratomas are congenital tumors that develop from primitive germ cells, most often in gonads. Among the different types, the most common is the mature teratoma (90% of cases). The latter is cystic, benign and generally composed of tissues from the 3 embryonic leaves (endoderm, mesoderm and ectoderm). The presence of fat in the cyst as seen at imaging is a pathognomonic sign of maturity. Mature teratomas have a slow growth (2mm/year) and are therefore generally asymptomatic until their size causes a mass effect. When the tumor is large, it can bend (15%), rupture (5%), become malignant (2%) or infected (1%).

Conclusion

Constipation, in healthy children, may simply be of functional origin but excluding medical and surgical causes is important. Constipation, accompanied by abdominal bloating, may be the only sign of an intra-abdominal compressive effect

P 92.

Acute abdominal pain in teenager

O. Leclercq, L. Lecomte

CH Jolimont

We present the case of a 14 year old who was admitted to the emergency room after being awakened by severe acute-onset periumbilical pain. He was equally nauseous, afebrile and presented no other symptoms. His past medical history consists of bilateral congenital deafness and thrombophilia due to a heterozygous G20210A mutation of the prothrombin gene. His acne is currently being treated by tetracycline and the dosage was doubled 3 weeks prior to presentation. Initial work-up showed mild inflammation, neutrophilia, hyperglycemia and elevated serum lipase (542 U/L for a normal range of 0-160U/L). Free intraperitoneal fluid was detected on an abdominal ultrasound and the subsequent abdominal Computed Tomography (CT) showed severe acute pancreatitis (AP) of Balthazar grade E with multiple areas of necrosis. He was transferred to an adult intensive care unit for analgesia and etiological work-up. He later developed a mild fever and the pain subsided after patient-controlled analgesia and several days of intravenous fluid and electrolyte supplementation. Bilateral pleural effusion and partial thrombosis of the splenic vein and a branch of the portal vein were identified on the day 4-CT and therapeutic low molecular weight heparin was administered. After complimentary work-up and ultrasound guided percutaneous biopsy of the fluid collection, no definitive etiology was identified. Oral intake was progressively reintroduced with a fat restricted diet and he was discharged after 9 days. Follow up will be essential because of the many possible complications ranging from pseudocysts to abscesses and fistulizations. Because of this, survivors of severe acute pancreatitis often have a reduced quality of life.

AP is a relatively uncommon diagnosis in pediatrics. The most common etiologies in adults being alcohol abuse and gallstones, children are often spared. Nonetheless, other potential causes can present in childhood and adolescence. The most frequent being anatomical abnormalities, infections, hereditary, metabolic, trauma or drugs. The pathogenesis between children and adults is similar so most of our knowledge of the pathology, treatment and outcome come from extrapolations of adult literature.

Tetracycline has been identified as a causative agent for AP on multiple occasions. Therefore, it is important to realize that something as “simple” as acne treatment can have potentially life-threatening adverse effects.

Posters**P 93.****Scurvy in a toddler - A case report**

Z. Ouchinsky

*ULB - HUDERF*Background

Nowadays plant-based milks are increasingly consumed by adults and children. Unfortunately most of them are unsuitable for the very special needs of young children and can be life-threatening in some cases.

Method

Description of a case-report.

Result

A 4-month-old girl presented at the emergency room with vomiting, asthenia, impaired general condition and weight loss.

Her parents are non-consanguineous Algerian and her older sister has a cow milk allergy (CMA). She was born at term by elective C-section with 2.735 kg and 55.5 cm. On her first days of life she received an hydrolyzed formula (Nutramigen®) for CMA suspicion.

At 2 months of age, because of digestive symptoms, a rice milk has been prescribed but the mother bought one in a supermarket, improper for toddlers (enriched with B12 and calcium but without any other micronutrients and poor in proteins and calories).

At admission, she weighed 3.7 kg (vs 4 kg 1 month earlier) and was 55.5 cm tall. Weight-for-height was 83% and height-for-age 88%.

She was pale, not in good condition, had brittle hair, limb edema, very dry skin, and cradles cap. We observed also a developmental delay with axial hypotonia.

Biological investigations showed anemia, hypoalbuminemia, and deficiencies in vitamin C, zinc and folic acid. Ionogram, ferritin, vitamin A, D and E were normal.

Radiographs of lower limbs were normal.

She drank very few and was sleeping a lot at the beginning of the stay. She was often very uncomfortable and had blood in her stools.

Management of her undernutrition started very carefully considering the risk of refeeding syndrome. She was fed by nasogastric tube because she was not able to drink enough. We decided to keep an hydrolyzed formula but chose one with medium chain triglycerides given the digestive symptoms (Alfaré®). She received vitamins and zinc supplementation.

After 3 weeks she was able to drink alone enough milk and her clinical state improved : healthier skin, no rectal bleeding, better tone, more active and comfortable baby. Discharge weight was 4.4 kg.

Eventually we concluded many of her symptoms could be explained by undernutrition but also by scurvy: rectal bleeding, asthenia, painful and swollen limbs, skin manifestations and poor appetite are often described in scurvy.

Conclusion

This case report emphasizes the danger of inappropriate plant-based diet in young children. Besides, it highlights scurvy has not disappeared, even if rare in developed countries.

P 94.

Case report of a rare cause of jaundice in children: autoimmune pancreatitis

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CHR Verviers, U Liège, CHR de la Citadelle, CHU de Liège

Introduction

Autoimmune pancreatitis (AIP) is a rare form of pancreatitis in children, combining specific imaging criteria, histopathological and biological features. There are some differences in clinical presentation and management between children and adults which complicates diagnosis.

Clinical case

We report the case of an 11-years-old boy who was presenting jaundice, fatigue, pruritus, peri-umbilical pain and diarrhea without fever. In the medical family history, a lupus was reported in his mother. Blood tests showed a cholestasis, an increase in lipase (>4x ULN) and transaminases (TGO: 2,8x ULN, TGP: 8,9x ULN).

IgG4 levels and autoimmunity testing were normal.

Transabdominal ultrasound showed a moderate dilatation of intra and extra hepatic biliary tract and magnetic resonance cholangiopancreatography an enlargement of the pancreatic head and irregularities in the main pancreatic duct without mass syndrom.

The echo-endoscopy highlighted stenosing cholangitis.

Spontaneous resolution of clinical and then biological features was observed in few weeks, without any treatment.

Discussion

AIP is a particular form of chronic pancreatitis classified into two subgroups in adults, based on HISORT criteria.

In pediatric patients, clinical presentation may include obstructive jaundice and/or abdominal pain. It is associated with specific imaging findings of pancreas in all reported patients.

Unlike adults, elevated IgG4 levels are uncommon and histopathological features of both types are often present. This suggests that children have distinct AIP pattern.

Pancreatic biopsy can confirm the diagnosis but there is a limited expertise in interpretation of pediatric pancreatic histopathology.

With corticotherapy, AIP remits at a high rate. However, it has significant adverse effects on the child's growth. Moreover, spontaneous resolution is not rare. The wait-and-see strategy must be considered on long-term studies.

Follow-up of AIP patients should be performed to exclude long-term complications like diabete or inflammatory bowel disease.

In our patient, several criteria were present to evoke AIP : clinical and biological presentations, imaging findings, history of lupus in mom and spontaneous resolution.

Conclusion

Pediatric AIP has distinct presentation compared to adults. In case of obstructive jaundice with an increase in lipase,AIP must be envisaged.Data in children are limited and long-term studies are necessary for a better understanding of that rare disease

Posters**P 95.****Mind the liver when Coombs comes positive in toddler.**

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*U Liège, CHC Liège*Background

Pediatric autoimmune hepatitis is a rare chronic inflammatory disease.

The clinical presentation is variable, from incidental discovery to fulminant hepatitis.

Without treatment, it progresses rapidly to liver failure and cirrhosis.

Currently, two types of autoimmune hepatitis are described:

- Type 1: smooth muscle (SMA) and / or antinuclear (ANA) antibody-positive
- Type 2: liver kidney microsomal type 1 (LKM1) and / or anti-liver cytosol type 1 (LC1) antibody-positive

Case report

1 year old boy referred for incidental discovery of raised hepatic aminotransferases in a context of virosis with prolonged fever.

The child had no medical or familial history except feeding problem.

Normal psycho-motor development, correct weight gain growth.

The clinical examination show only moderate hepatomegaly without splenomegaly or associated jaundice. The rest of the exam was normal.

Blood test :

- Increased liver test from one biology to another: GPT 1241 U / L, GOT 1427 U / L, GGT 50 U / L. Normal bilirubin.
- Normal liver function
- Viral serology negative for hepatitis A, B, C, E, EBV, CMV, Toxoplasmosis, Herpes, Parvovirus, Enterovirus, Adenovirus.
- Ceruloplasmine, copper normal and alpha 1-anti-trypsin : MM.
- No anemia.
- Immune: Hypergammaglobulinemia G, Coombs + IgG gliadin slightly elevated, IgA transglutaminases normal, HLA normal for coeliac disease.
- AAN, SMA, LKM-1, anti LC-1, anti-SLA, ANCA negative.
- Normal metabolic tests.
- Heterozygous TPMT

Abdominal ultrasound : normal.

Liver biopsy : presence of multinucleated hepatocytes, moderate inflammatory infiltrate, swollen hepatocytes, piecemeal necrosis, plasmocytes within interface hepatitis. No fibrosis.

Fibroscan : still showing a fibrosis score of 16.8 kPa with an IQR of 14%.

Treatment with Budesonide and Azathioprine was started with excellent biological progress under treatment.

Discussion

The diagnosis of autoimmune hepatitis is classically based on the presence of serum antibodies, hypergammaglobulinemia G, compatible histology, autoimmune family history and in the absence of other etiologies.

However, sometimes the patients present clinical history and histology compatible with autoimmune hepatitis without specific markers such as autoantibodies. Pediatric seronegative autoimmune hepatitis include a spectrum of disease. The presence of a hepatic cytolysis with positive test coombs and hypergammaglobulinemia must evoke the diagnosis in infant.

Posters**P 96.****Acute right iliac fossa pain, about a case**

E. Hoornaert, C. Koutny, G. Van De Poel

*Clinique Notre Dame de Grace Gosselies*Introduction

Acute abdominal pain is a common complaint in childhood and can be caused by a wide range of aetiologies, mostly benign. The challenge for a good physician is to identify those who require urgent treatment. Meticulous history taking and physical examination are essential to determine the cause of acute abdominal pain.

Results

We report the case of a 14-year-old girl sent to our emergency room for suspicion of appendicitis. She had abdominal pain in the right iliac fossa that appeared brutally the day before, was worsened while walking and was associated with nausea, without fever. Clinical examination showed a sensibility in the right iliac fossa. Initial lab tests were normal whereas abdominal ultrasound showed a large amount of ascites in the 4 quadrants of the abdomen and an ovarian haemorrhagic cyst of 6mm on the right side. B-HCG was negative. The mother reported us that there was a familial history of hereditary angioedema due to C1-inhibitor (C1INH) deficiency. Further lab tests showed a diminution in complement C4 and in C1INH. Diagnosis of abdominal angioedema due to C1INH deficiency was made. She was treated with tranexamic acid 1g 3x/day for 7 days with good clinical evolution.

Conclusion

Hereditary angioedema (HAE) is a rare genetic condition characterized by transitory and recurrent oedemas. Onset may occur at any age but often appears during childhood or adolescence. Triggering events may be any trauma (even the smallest: change of position), stress but also hormonal events such as puberty. Oedemas involve the skin or mucosae (e.g. laryngeal) and do not respond to usual treatment (antihistaminic, steroids, adrenaline). Intestinal track involvement induces acute abdominal pain that can mimic an intestinal occlusion or appendicitis sometimes associated with ascites and hypovolemic shock. Pathogenesis of HAE is a deficiency or dysfunction of C1INH which leads to an excessive production of bradykinin, causing increased vascular permeability and angioedema. Diagnosis relies on C4 and C1INH rates measurement. First line therapies include tranexamic acid, C1INH concentrate, recombinant human C1INH and Icatibant® (a synthetic bradykinin B2-receptor antagonist).

In patients with frequent or severe episodes, long-term prophylaxis is based on C1INH concentrate and tranexamic acid (better tolerated) rather than on attenuated androgens (Danazol®).

P 97.

Hydrogen and methane breath test in the diagnosis of lactose intolerance.

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UZ Brussel, VUB Brussel

Aims

The hydrogen (H₂) breath test is a non-invasive investigation with a high sensitivity and specificity to diagnose lactose intolerance. Patients with lactose intolerance also expire increased amounts of methane (CH₄) during a lactose challenge. The aim of this study is to evaluate the importance of CH₄ measurements.

Methods

We tested 59 children (5-15 years old) with symptoms suggesting a lactose intolerance with a lactose H₂ and CH₄ Breath Test. Lactose was administered orally (dose of 2 g/kg, maximum 50 g) diluted in a maximum of 250 ml of water. The expired air was collected in specific syringes with a capacity of 60 ml. One breath sample was taken before the intake of lactose and breath samples were collected after the ingestion of lactose every 15 min during 3 hours. The expired H₂ and CH₄ was measured with a specific analyzer (Microlyzer DP; Quintron Instruments, Milwaukee, Wis.). The result was considered positive when a H₂ peak exceeded 20 parts per million (ppm) over the baseline. CH₄ excretion was considered positive using if also reaching 20 ppm above baseline. A clinician, blinded for the results of the breath test, registered the symptoms of the patients during the test.

Results

26/59 (44%) were negative for both H₂ and CH₄. 25/59 (42 %) had a positive H₂ breath test result, 6/25 (24%) also positive for CH₄ and 19/25 (76%) negative for CH₄. 8/59 (14%) patients were only positive for CH₄. 7 out of these 8 patients reported a significant improvement of their symptoms on a lactose free diet. Since the intervention was open, and a blinded lactose challenge was not performed, a placebo effect cannot be excluded.

Conclusion

The results indicate that CH₄ measurement is possibly of additional value for the diagnosis of lactose intolerance. These results indicate that further research is needed before hard conclusions and recommendations can be proposed.

Posters**P 98.****First successful use of ustekinumab in an infant with very early onset inflammatory bowel disease: outcomes at 18 months follow-up**

C. De Geyter, B. Hauser, K. Huysentruyt, E. De Greef, G. Veereman, Y. Vandenplas

*UZ Brussel, The Hospital for Sick Children, University of Toronto*Background

Infantile onset IBD is a challenging disease within the group of very early onset inflammatory bowel disease (VEO-IBD), especially when it comes to treatment. While ustekinumab is an effective therapy approved for adult Crohn's disease, off-label use in the paediatric population is increasing. We describe the successful use of ustekinumab in an infant with VEO-IBD

Case presentation

A 5-months old, healthy born infant from allergic parents presented with bloody diarrhea. Treatment with an extensive protein hydrolysate and elemental formula resulted only in temporary improvement. Upper endoscopy and ileocolonoscopy showed mild antritis and duodenitis as well as some colonic aphteous lesions, an ulcer in the caecum and histologically mild ileitis and local cryptitis. The combination with mild anemia, inflammation and hypoalbuminemia, and negative coproculture, suggested the diagnosis of VEO-IBD, although immunologic and genetic work-up remained negative. Treatment with azathioprine, mesalazine and prednisolone was started, with temporary improvement. After a toxin-producing *Clostridium difficile* infection, eradicated with metronidazole and vancomycine consecutively, complaints remained and another ileocolonoscopy was performed. This showed diffuse aphteous lesions, deep and superficial ulcers with cryptitis, cryptabcesses and cryptdestruction, confirming diagnosis of VEO-IBD. Treatment was switched to infliximab (10 mg/kg week 0, 10 mg/kg week 2 and 15 mg/kg week 6) but failed to improve symptoms. Levels remained low and anti-TNF antibodies developed. Methotrexate was tried without effect. Because of transfusion dependent anemia and severe failure to thrive with unchanged lesions on ileocolonoscopy, ustekinumab in compassionate use was requested to Janssen.

Results

An induction dose of 65 mg ustekinumab IV at 19 months of age was administered followed by a maintenance dose of 45 mg SC/8 weeks up to now. Stool consistency and blood loss improved progressively and growth resumed. A colonoscopy after 14 months of treatment showed no more macroscopic, histopathological nor biochemical inflammation.

Conclusion

We report to the best of our knowledge the first patient with a VEO-IBD successfully treated with ustekinumab without side-effects and in complete remission after 1.5 year of treatment

P 99.

The search for information in media (websites and apps) by children with coeliac disease and their parents. A pilot study

E.I. Levy, T. Devreker, E. De Greef, Y. Vandenplas

*UZ Brussel*Background/aims

Coeliac disease (CD) is often difficult to diagnose because of the wide range of symptoms. Once the diagnose is confirmed, children and parents have to adjust their lifestyle. There is on the web a lot of information about CD. No study has evaluated so far the use of media (websites and apps) by CD children and parents.

Methods

This pilot survey was conducted at a pediatric department on the 25th and 28th of October 2019. We enrolled 51 children between the age of 0-18 years (including their parents) on these 2 coeliac days for their annual routine checkup. A multiple choice questionnaire (including also some open questions) was made with Monkeysurvey[®]. Children who used media by themselves participated on tablets when arriving at the event, if they didn't parents would answer for them.

Results

Of the 51 children, 23 answered the questionnaire by themselves. Children younger than 5 years (n:11) were not included in the media question part. The use of social media by children was very low (n:5) compared to parental (n:14) use (21.7% vs 82.2%). The use of media at the age of 6-12 years was 13.0% and 8.7 % in the age group 13-15 year. Children said they obtained enough information from the medical team (61.1%). Some (27.8%) had no interest in information from media. All media users, used the websites of the Flemish/French Belgian Coeliac Association websites and searched for information found by google-ing. Only 50 % of the parents that used internet was using apps too. YouTube (35.7%), forums (28.7%) and bakery workshops (0%) were not well known among parents. However, media were well used to search restaurants (web 70%, web/apps 30%) or holiday places/resorts (web 44.4% web/app 55.6%) which serve glutenfree meals. The "glutenfree" sign on food and cosmetic items was unknown to all children and only three parents know the correct answer (5.9%).

Conclusion

This pilot study showed that parents and children are interested and use information found on the world wide web. Parents and children should better be informed about media and on reliable and less reliable sources. This information should be provided by the medical teams.

Posters**P 100.****The inter-rater reliability of the Cow's Milk-related Symptom Score (CoMiSS) between parents and a trained healthcare professional**

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*UZ Brussel, LA FE Hospital*Background/aims

The Cow's Milk-related Symptom Score (CoMiSSTM) was developed as an awareness tool for primary healthcare providers (HCPs) to increase the awareness and knowledge of the most common manifestations of cow's milk protein allergy (CMPA) in infants. The aim of this study was to analyze the inter-rater reliability of the CoMiSS between parents and a trained HCP.

Methods

A pilot, cross-sectional, observational study was performed in 148 presumed healthy infants aged 0 to 6 months. An HCP determined the CoMiSS during a routine visit. Parents, blinded for the score of the HCP, were asked to fill in the same scoring template. Exclusion criteria included any detected health problem during the visit. The statistical analysis was performed with the Wilcoxon-test.

Results

At inclusion, mean age was 46 days. The overall mean \pm SD CoMiSS values scored by the HCP were 4.13 ± 3.54 . As for the parents, the mean \pm SD scores were 4.24 ± 3.61 . No statistically significant differences were observed for any item depending on the person performing the scoring. Symptoms with the highest mean scores regarding to both the HCP as the parents are faeces consistency and crying.

Conclusion

This study shows a statistically excellent inter-rater reliability between parents and a trained HCP of any item of the CoMiSS determined in a population of presumed healthy infants.

P 101.

Novel DGAT1 gene mutation in infant with protein-losing enteropathy (PLE)

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UZ Brussel

Background

We present a case of a Belgian female infant of consanguinous parents of Turkish origin with prolonged diarrhea, feeding difficulties and recurrent infections since the age of 2,5 months. She was born on term and after 2,5 uneventful months she presented in Turkey with an acute gastro-enteritis complicated by Klebsiella sepsis (probably originating from a necrotizing arthritis of the right hip, confirmed several weeks later on MRI), severe metabolic derangement and hypoalbuminemia. After stabilization and treatment she was transferred to Belgium where vomiting and diarrhea recurred every time formula feeding was restarted or increased. The infant was on several different regimen, initially hydrolysed whey protein and later amino-acid based formula. PLE with associated hypoalbuminemia, hypogammaglobulinemia and failure to thrive was thought to be secondary to septic arthritis, which was supported by findings on albumin scintigraphy. Cow's milk protein allergy was diagnosed but failed to sufficiently explain the clinical course. Genetic analysis showed a novel mutation in DGAT1 that supports a previously described aberrant gut epithelial lipid metabolism explaining all symptoms.

Methods

In Utrecht a gene panel analysis throughout targeted massively parallel/ next generation sequencing by the Illumina NGS sequencing system was performed. The congenital diarrhea panel contains 46 genes. This method shows a coverage of 98.5% and detects 95% of mutations in the panel.

Results

Genetic analysis showed a homozygous pathogenic mutation c.469-2A>G p.(?) in DGAT. Both parents showed a heterozygous state, confirming the result in their daughter. No other mutations were found in the examined genes.

Discussion

DGAT1 encodes for diacylglycerol acyltransferase type 1, a protein involved in triglyceride synthesis. Mutations in this gene are rare and most reported cases concern Turkish patients. Few known patients with a homozygous mutation present with severe congenital diarrhea and PLE. Although the mutation in our case was previously unknown, the phenotype is similar to other cases described. After enteral nutrition was replaced by parenteral lipid administration we saw a favorable evolution.

Conclusion

This rare case of feeding difficulties in an infant with a previously unknown DGAT1 mutation with favorable evolution after switch to parenteral lipids, supports the finding that DGAT1 deficiency causes PLE due to aberrant gut epithelial lipid metabolism.

Posters**P 102.****Diagnosis of vitamin B12 deficiency through newborn screening**

A. Empain, L. Marcélis, C. De Laet

*ULB - HUDERF*Background and aim

Vitamin B12 deficiency is well known to cause macrocytic anemia. Prolonged B12 deficiency during infancy has been associated with negative long-term health outcomes in children, including impaired cognitive function and developmental delay. Several studies show that B12 deficiency is prevalent in pregnant women leading to risk of deficiency in the newborn. Unrecognized neonatal B12 deficiency worsens if the infant is exclusively breastfed by a B12 deficient mother.

Cobalamin is a cofactor required by 2 enzymes, methionine synthase (homocystein remethylation), methylmalonyl-CoA mutase (valine, isoleucine, threonine catabolism and transformation into succinyl CoA). Abnormal urinary methylmalonic acid (MMA) and/or plasma tHomocysteine (tHcy) are sensitive markers of B12 deficiency. Methylmalonic aciduria (propionylcarnitine-C3- value) is part of the Belgian newborn screening program. Neonatal screening offers an opportunity to identify children with asymptomatic B12 deficiency.

The objective of this study is to assess which of increased C3 at neonatal screening are due to B12 deficiency.

Method

We have resumed all neonatal screenings showing a pathological elevation of C3 from January 2013 to August 2019. For each patient we looked at the metabolic parameters (MMA on dried blood spot and in the urine, tHcy, vitamin B12, folic acid, plasma amino acids). In case of newborn deficiency, a deficiency in the mother is investigated : causes and biochemical tests.

Results

From January 2013 to august 2019, 13 patients had elevated C3 on neonatal screening. Of these, 3 had a metabolic disorder (2 Cobalamin C deficiency and 1 methylmalonyl-coA mutase deficiency). 8 out of 13 patients were B12 deficient. The last 2 patients were born prematurely. Those had a spontaneous resolution of biological abnormalities. Among the 8 patients with B12 deficiency, 7 had an elevation of the urinary MMA and 6 had an elevation of tHcy. After two month of B12 supplementation, all the metabolic parameters were normalized. Regarding the maternal assessment, 4 out of 8 mothers had a history of bariatric surgery and the others were also vitamin B12 deficient.

Conclusion

Vitamin B12 deficiency can be very harmful to a child's development. It is important to diagnose it as soon as possible. Moreover, these diagnosis in children thanks to neonatal screening emphasizes the need for adequate maternal nutrition during pregnancy and lactation.

P 103.

Nasobiliary drainage prior to surgical biliary diversion in patients with progressive familial intrahepatic cholestasis type II

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Background

Progressive familial intrahepatic cholestasis (PFIC) can cause intense pruritus refractory to medical therapy. Surgical biliary diversion techniques, including partial internal biliary diversion (PIBD), have been developed over the years to relieve pruritus without requiring liver transplantation. Despite the efficacy and safety of PIBD procedure, some patients do not respond to surgery and there is currently no possibility to predict pruritus response according to genotype or other clinical parameters. Our aim was to evaluate nasobiliary drainage (NBD) as a method to predict pruritus response to PIBD, in order to avoid unnecessary surgery.

Methods

We present two PFIC 2 patients who underwent two and three transient endoscopic NBD prior to PIBD surgery, which was performed at the age of 25 and 28 years old respectively. Both patients suffered from invalidating pruritus refractory to medical therapy. Liver transplantation was not performed since both patients had normal liver function and normal liver histology (patient 1) or only very mild fibrosis (patient 2). Pruritus was assessed according to the following score: 0 = none, 1 = rubbing or mild scratching when undistracted, 2 = active scratching without evident skin abrasions, 3 = abrasions evident, 4 = cutaneous mutilation and scarring evident, impaired quality of life.

Results

Both patients repeatedly responded to NBD (pruritus score 4 to 0-1) and had complete pruritus resolution after subsequent PIBD (score 4 to 0). Serum total biliary acids (TBA) decreased from 282 $\mu\text{mol/L}$ and 314.4 $\mu\text{mol/L}$ before PIBD to 40.7 $\mu\text{mol/L}$ and 220.9 $\mu\text{mol/L}$ after PIBD respectively (normal TBA levels < 10 $\mu\text{mol/L}$). Follow-up duration after surgery was seven years for patient 1 and one month for patient 2. Pruritus did not recur after PIBD in our patients despite important variations in TBA levels during follow-up. Mild post-endoscopic biological pancreatitis occurred in 1/5 NBD procedures, and resolved spontaneously. The only adverse effect observed within 7 years post-PIBD was very mild transient osmotic diarrhoea, easily treated with cholestyramine.

Conclusion

Our data suggest that NBD is a safe and effective way to predict pruritus response before performing a permanent biliary diversion surgery in PFIC patients.

Posters**P 104.****Is shit in real life the same as on photo?**

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*UZ Brussel, The Hospital for Sick Children, University of Toronto*Background/Aims

The Brussels Infant and Toddler Stool Scale (BITSS) was developed to assess stool consistency in non-toilet trained children. In contemporary medicine we cannot deny the importance of the increasing use of photographic material, which is also the case for stool assessment. There is however no data if the use of photographic stool assessment is comparable to real time assessment. The goal of this study was to investigate the variability between real time vs. photographic assessment of stool consistency using the BITSS.

Methods

202 fresh stool samples of non-toilet trained children attending a day-care centre were rated according to the 4-point BITSS scale. Each stool sample was photographed and presented at a later time in a random order to the same observer. One observer rated all stools, the number of observers that rated each stool depended on the availability of the day-care staff. Observers that rated a minimum of 40 stools and their corresponding photos were included for further subanalyses. A Cohen's κ was calculated for the agreement in ratings between fresh sample and photo of each observer, a κ of 0.4-0.6 was seen as a moderate and 0.6-0.8 as substantial agreement. For the secondary goal we also computed inter-observer reliability, with the observer that has rated every stool serving as the reference observer.

Results

Overall, 576 ratings were performed, resulting in a substantial agreement with a global kappa of 0.63 (95% CI 0.56-0.70). Of the total of 576 ratings, 4% were hard stools, 53% formed, 34% loose and 9% watery stools. A substantial agreement between fresh stool samples and their photos was found ($\kappa = 0.70$, 95% CI 0.59 to 0.81 for observer 1, $\kappa = 0.73$, 95% CI 0.50 to 0.96 for observer 2 and $\kappa = 0.47$, 95% CI 0.17 to 0.77 for observer 3). The overall absolute agreement between observer 1 and 2 for the assessment of the fresh stool samples was 81.3% and 82.2% between observer 1 and 3, with a substantial agreement between all observers ($\kappa = 0.674$, 95% CI 0.43 to 0.91 for observer 1 vs 2 and $\kappa = 0.66$, 95% CI 0.40 to 0.93 for observer 1 vs 3).

Conclusion

The substantial correlation we found suggests that the BITSS is a good clinical tool to use for photographic material. The secondary outcomes confirm that the BITSS has a good inter-observer reliability and can be used in daily practise by both professional health care workers and people without medical background.

LO 1.**Interdisciplinary rehabilitation after cancer in children and adolescents**

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UZ Gent

Background/aims

Physical activity, psychosocial well-being and participation levels are often reduced in children and adolescents after cancer treatment. Increasing the healthy nutritional habits, physical fitness, psychological health and participation levels might decrease long-term (side) effects of the disease and improve quality of life (QoL). The current study determines the effects of a personalized interdisciplinary intervention program at the level of functioning, activities and participation.

Methods

A longitudinal cohort study was developed. Patients between 8 - 21 years and within 6 months up to 5 years post-acute cancer treatment were eligible for participation. Body composition, maximal oxygen consumption during cardiopulmonary exercise test, QoL via the PedsQL, and participation levels via the Children's Assessment of Participation and Enjoyment Scale of Questionnaire (CAPE) were assessed at baseline, after 4 months of intervention and at 12 months follow-up. Intervention consisted of an individualized physical program 3 times/week. Additionally, follow-up was executed on a monthly basis for 4 months concerning psychological well-being, guidance for nutritional habits and participation in terms of fatigue, energy management and hobbies.

Results

Twenty-three patients (mean age: 12.1 yrs \pm 3.2) were included. Preliminary results after 4 months treatment displayed increased VO₂peak ($1.30 \pm 0.35 > 1.38 \pm 0.33$ L/min; $p=.007$), load ($96.14 \pm 39.56 > 110.43 \pm 40.76$ Watt; $p<.001$), dry lean weight ($7.82 \pm 3.79 > 8.02 \pm 3.87$ kg; $p=.038$), and QoL-fatigue reported by parents ($66.73\% \pm 13.63 > 74.87\% \pm 14.82$; $p=.023$). Accordingly, heightened physical activity levels on the CAPE scale were reported, however, not significant ($p=.06$). The correlations of QoL with intensity of social activities were low to moderate ($.465 \leq r \leq .589$).

Conclusion

An individualized interdisciplinary intervention program of 4 months improved the physical fitness in children and adolescents after cancer treatment. The improvements in dry lean weight, QoL, social and physical participation and the reduction in fatigue directly after the program are promising. Nonetheless, these results should be confirmed in a larger sample size. Moreover, the outcomes after 12 months follow-up are needed to steer conclusions about long-term effects.

Short Oral Presentation**SO 14.****Is the preoperative preparation of Sickle Cell Disease patients optimal: Assessment of practices and post-operative complications.**

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Sickle cell disease (SCD) patients are at high risk for severe postoperative complications such as vaso-occlusive crisis (VOC) and acute chest syndromes (ACS). Systematic preoperative blood transfusion has been the most common preventive method used in Western countries. This attitude is based on the results of few old, randomized trials that demonstrated the benefits of preoperative transfusion. Nevertheless, this recommendation is now widely questioned.

The aims of this retrospective study were to assess the current quality of care, to identify the risk factors associated with postoperative complications and to provide useful data in order to improve SCD patient management.

Material and Methods

Medical records of SCD patients who underwent elective mild risk surgery at HUDERF between 2009 and 2019 were analyzed. All patients were managed for peri-operative care according to our institutional protocol which includes red blood cells (RBC) transfusion 2 to 5 days before scheduled surgery, specific pain management and avoidance of dehydration and hypothermia. Data on SCD severity (genotype, G6PD deficiency, complications and biology), preoperative preparation, operative parameters, post-surgical management and post-operative complications during the following year were described in details by statistical tests with a significant p value <0,005.

Results

Ninety-eight interventions were included among them 96 were statistically analyzed. Within the first 30 days, few complications developed after cholecystectomy (12,5%) and adenoïd/tonsillectomy (3%). Splenectomy was associated with high complication rates (75%) especially for ACS (42%). Prolonged operative time appeared to be the only independent predictive factor for vaso-occlusive complications (p=0.008 for ACS and p=0.015 for VOC). Adverse SCD effects seem negligible 3 months after surgery.

Discussion and conclusions

Perioperative management of our SCD patients should be adapted according to the observed results. For cholecystectomies and adenoïd/tonsillectomies, individualized and selective RBC transfusion protocols should be developed. More attention should be paid to splenectomy, given its high risk of complications. More specifically, multidisciplinary collaboration is required to reduce operative time and to assess the indication for preoperative RBC transfusion on a case by case basis.

SO 19.

Physical functioning in children surviving a brain tumour

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UZ Leuven

Background

Survival rates of children with brain tumours have largely increased over the past decades. Many long-term sequelae in these children are well known, but the physical functioning of these survivors is poorly described.

Methods

In this cross-sectional study on paediatric survivors of a brain tumour, physical functioning was assessed in 53 patients who were treated at the University Hospitals of Leuven and were at least 6 months out of treatment. Mean age at diagnosis was 6 years 2 months (range 0-14). Mean age at testing was 11 years 8 months with a mean follow-up time of 4 years 8 months. Pilocytic astrocytoma (44.23%) and medulloblastoma (17.31%) were the most common diagnosis. Overall motor function was evaluated using the Movement Assessment Battery for Children (MABC-2-NL) and/or Bruininks-Oseretsky Test of Motor Proficiency (BOT-2). Functional walking capacity was assessed with 6-minute walk test (6MWT).

Results

Mean scores of MABC-2-NL components, i.e. manual dexterity, aiming and catching, balance and total score were below 1SD of the norm values in respectively 68%, 74%, 65% and 75% of patients. Mean scores of fine manual control and body coordination of the BOT-2 were below 1SD in respectively 46% and 70% of patients. Sixty-six percent of patients scored normal on the 6MWT while 28% showed a mild decrease and 6% a moderate or severe decrease. Younger age at diagnosis (42%; <5 year) is significantly associated with lower scores on Body Coordination. Patients with adjuvant treatment (58%; either chemo and/or radiotherapy) showed significant worse scores for fine manual control of the BOT-2 ($p=0.024$) and Balance of MABC-2-NL ($p=0.036$), albeit these effect become insignificant after Bonferroni correction. Follow-up duration and localization (infra- vs supratentorial) revealed no statistical differences.

Conclusion

The majority of survivors of a childhood brain tumor exhibits long-term decreased fine motor control and balance. Adjuvant treatment further impacts their physical functioning. Functional walking capacity was slightly better preserved. These results indicate that all paediatric patients with a brain tumour need a rehabilitation program.

Poster Walk**PW 4.****The risk of rebound hypercalcaemia after denosumab treatment in paediatric oncology**

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UZ Leuven

Background

Central giant cell granulomas (CGCG) and aneurysmal bone cysts (ABC) are benign but destructive bone tumours that exhibit overexpression of the receptor-activator of nuclear factor $\kappa\beta$ ligand (RANKL). RANKL activates the RANK receptor on osteoclasts leading to bone resorption. Denosumab is a human monoclonal antibody that binds RANKL and inhibits this process making it an alternative to surgery. There is limited data available on the safety in children. We report two cases of rebound hypercalcaemia following denosumab treatment.

Findings

Surgery was deemed unsafe due to inevitable damage to surrounding tissues in the following cases. Subcutaneous denosumab (70mg/m²/dose) was given on day 1 of each 4-week cycle with loading doses in cycle 1 on day 1, 8 and 15.

Case 1

A 14-year-old boy with CGCG of the mandible was treated with 12 cycles of denosumab resulting in metabolic remission on PET-CT. Four months after cessation, he presented with vomiting, polyuria, hypercalcaemia and acute kidney injury. Hyperhydration and furosemide were started without improvement. Therefore zoledronate was given. This was complicated by symptomatic hypocalcaemia, hypomagnesemia and hypophosphatemia with prolonged QTc requiring intensive care. After correction of deficiencies and further hydration, kidney function and electrolyte imbalances normalised.

Case 2

A 7-year-old boy with a suprasellar ABC with bilateral compression of the optic canal presented with headache, vomiting and worsening vision. Thirteen cycles of denosumab were given resulting in improvement of the lesion. Four months after discontinuation he presented with vomiting, hypercalcaemia, and acute kidney injury due to dehydration and hypokalaemia. Hyperhydration and potassium supplementation were given. One dose of pamidronate resulted in increasing creatinine and hypophosphatemia. Intravenous fluids could be weaned off with improving renal function. He was discharged but presented two weeks later with the same clinical picture, again treated with hyperhydration and pamidronate.

Both patients remain asymptomatic, respectively 12 and 7 months after denosumab discontinuation, with normal kidney function and calcium levels.

Conclusion

Denosumab discontinuation can be complicated by severe rebound hypercalcaemia. Careful monitoring and patient education regarding hypercalcaemia symptoms after cessation is warranted. Preventive strategies, e.g. weaning course with decreasing doses, may be indicated.

PW 6.

Neonatal screening program for sickle cell disease : comparison of patients born 2000-2005 and 2010-2015, single center experience

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CHU Charleroi, ULB - HUDERF

Background

Universal Newborn screening (NS) for Sickle cell Disease (SCD) was implemented in all Brussels maternity wards since 2000 and has been shown to be effective in reducing early mortality and morbidity.

The aim of our study was to compare outcome of 2 NS cohort born at 10 years of interval.

Methods

This retrospective study included all children screened for SCD enrolled in the Belgian SCD database and born from 2000 and 2005 (Group 1) and from 2010 and 2015 (Group 2) and followed at Hôpital Universitaire des Enfants Reine Fabiola (Brussels, Belgium). Demographic data, baseline hematological values, clinical events, cerebro-vascular disease and disease-modifying treatment (DMT) were compared between both groups during the first 5 years of life.

Results

Among these 59 patients, most of our patients come from Democratic Republic of Congo and more than 80% had a severe genotype (HbS/HbB0). However, due to the recent changes in migratory flows, the number of patients from West Africa has increased considerably in recent years with more SC patients. Their number tends to reach the figure of the French cohort.

The number of patients who developed at least one SCD clinical event as well as the median age at the first event was the same between the 2 groups (nearly 80%).

The proportion of patients with splenic sequestration was significantly higher ($p=0.0083$) in group 2 (28% versus 74%). The number of patients with abnormal transcranial doppler velocities slightly decrease in Group 2 (14.3% vs 8.3%) with a higher rate of conditional velocities (5.7% vs 20.8%) but it was not statistically significant.

More patients in Group 2 were being treated with hydroxyurea (HU) even if it was not significant. However, higher doses were prescribed in Group 2.

The number of patients who presented a clinical event before the age of 5 is comparable to what is described in the literature. It seems to be lower in the second cohort, but it was not statistically significant. It could be due to a higher rate of prescription and dose of HU. In addition, the improvement of hematological values in the second cohort suggest also a better adherence to the treatment.

In conclusion, our data showed no significant change in the management of the complications presented by patients with sickle cell disease and their overall management. However, there is a trend towards increased hydroxyurea prescribing and dosing.

Posters**P 105.****A limping hiding a metastatic neuroblastoma**

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*CHU de Liège, CHR de la Citadelle, Notre Dame de Bruyères*Introduction

Neuroblastoma is a neuroendocrine tumor derived from sympathetic ganglion cells. This tumor mainly found in children and affecting boys more than girls. Its histopathology, presentation and evolution are heterogeneous. So, the treatment depends on its evolution

Case Report

A 6-year-old girl was referred to emergencies for pain in the right hip for a week leading to lameness. Initially considered as a hip synovitis, so, treated symptomatically. Facing worsening symptoms despite treatment, she was referred to us for further assessment. Clinically, she presented pain in the right hip hyperflexion that limited walking, no notion of fever or trauma and a child who otherwise had good clinical parameters and a normal clinical examination. Her medical history reports congenital hip dysplasia.

Thus we performed bilateral hip ultrasound and X-ray of right hip, no effusion or bone lesion was seen, blood biology revealed an inflammatory syndrome, the rest of the biology was otherwise without particularities.

So, an osteo-articular infection, was suspected and the patient was treated by antibiotics. 48 hours after the start of treatment, no clinical or biological improvement was noted. An abdominal ultrasound was therefore performed and revealed a lesion in the right adrenal gland and another between the ipsilateral kidney and the spine pushing back the inferior vena cava.

She was referred to our pediatric hemato-oncology unit where a biological assessment demonstrated, in addition to the inflammatory syndrome, an increased NSE, a bone biopsy revealed an infiltration of the marrow and confirming the diagnosis of neuroblastoma. A spinal MRI did not show intra-dural invasion but showed a metastatic carcinomatous of D10-L3, MIBG scintigraphy confirming the invasion of the dorsal-lumbar spine but also of the spine cervical, tumoral lesions visualized in pelvis, proximal femurs, shoulders, left internal subclavicular region... The lumbar puncture returned negative. Therefore, a stage IV metastatic neuroblastoma was diagnosed and chemotherapy treatment initiated. An ovariectomy for ovarian cryopreservation was also performed before the initiation of chemotherapy

Conclusion

Facing an osteoarticular pain, a metastasis is not the first diagnosis in the clinician's mind. Nevertheless after exclusion of other differential diagnoses or in the absence of clinical improvement, the clinician should not forget least common diagnosis, including a metastase as in your case

P 106.

A case report of Ileal Burkitt Lymphoma revealed by an intestinal invagination in a young child

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Introduction

Burkitt lymphoma is a rare entity that accounts for 50% of all childhood Non-Hodgkin Lymphomas. The abdominal presentation remains the most common form and can complicate to an intestinal invagination, which leads to an early diagnosis of the disease.

Clinical case

We described the case of a three-year-old boy who had been hospitalized for a week due to intestinal pain, vomiting and diarrhea. The abdominal ultrasound showed an intestinal invagination (typical 'target' sign). The patient was performed a laparotomy with ileocecal resection and terminal anastomosis. The irreducible nature of the lesion and the presence of mesocolic lymph nodes of pathological appearance could suggest an underlying lymphoma, which was confirmed on histopathology. The patient was referred to the pediatric oncology department for evaluation and initiation of the chemotherapy. The post-operative ultrasound and PETCT showed a progression of the mass over the week after surgery. According to St. Jude's staging system, the patient was classified as low LDH stage III Burkitt lymphoma and underwent five courses of chemotherapy.

Discussion

Abdominal imaging (ultrasound/CT-scan) played a decisive role in the management of the Burkitt Lymphoma. It enabled us to observe the tumor mass, the lymph nodes and the complications, as the invagination or the peritoneal effusion or others signs of intestinal pain that were a contraindication to reduction treatment by enema. Surgery had a limited role in the treatment of Burkitt's lymphoma (for localized stage I only) because surgery is rarely complete in stage II or III and the intervention could lead to complications and a delay of chemotherapy with a negative impact on overall survival. It is recommended to carry out a biopsy (ultrasound-guided puncture) and to start chemotherapy immediately, which may be sufficient to allow disinvagination by simple reduction of the mass. In the case of our patient, the start of chemotherapy led to the complete remission of the residual mass at day eight of evaluation.

Conclusion

The treatment of Burkitt's lymphoma is based primary on chemotherapy, which gives a very good response (OS 98% at 5 years for the abdominal low LDH stage III Burkitt lymphoma). Performant imagistic evaluation and initial management of patient is essential for the future outcome. Surgery should only be used in very limited cases.

Posters**P 107.****Case report : a painful pelvic mass in a fourteen years old girl.**

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*CHIREC, HUDERF-ULB*Background

Rhabdomyosarcoma (RMS) is the most common soft tissue sarcoma among children. Pelvic RMS are rare tumours with a poorer prognosis than the majority of RMS. We report hereafter a case of a teenager suffering from a pelvic RMS.

Case

A fourteen-year-old belgian girl was initially admitted to the emergency department for a suspicion of bartholinitis. She reported pelvic pain due to a mass evolving since two months. She did not report any vaginal bloody discharge or other symptoms. Physical examination only attested a painful indurated pelvic mass located between the labia majora and the anus, going through pelvic muscles. All ganglionic areas were free on palpation. There was no hepatosplenomegaly. She was treated by intravenous antibiotics for forty-eight hours. Blood test was normal. Tumoral markers, alpha fetoprotein and beta HCG, were negative. A pelvic ultrasound reported a strong suspicion of a malignant tumor with an eighty by forty millimeters fleshy high vascularized tumor with multilobulated contours. Magnetic resonance imaging confirmed a large pelvic mass located deeply in the right gluteal region and the perineum, infiltrating the external anal sphincter, the right side of the vagina, the tendinous centrum of the perineum and the puborectal muscle. The tumor was very suspicious of a rhabdoid origin. Further investigations such as PET scanner demonstrated a very captative tumor inoperable without mutilation as well as two inguinal lymph nodes. The biopsy confirmed the diagnosis of perineal alveolar RMS. Treatment was started following EpSSG RMS 2005 recommendations, chemotherapy followed by surgery, radiotherapy, adjuvant chemotherapy and maintenance therapy for a total duration of two years.

Discussion

Pelvic RMS prognosis is poor because it is often locally extensive and large at diagnosis, lymph node invasion is frequent and the alveolar histological subtype is particularly frequent. Current strategies focus on multimodal treatments involving surgery, chemotherapy and radiation but it is difficult to achieve successful treatment at this site. Because surgery can be very mutilating others techniques like brachytherapy and proton beam therapy are studied in order to preserve organs and their function.

Conclusion

Prognosis of pelvic RMS remain poor despite technical progress. Treatment approaches are studied to be as less invasive as possible and should be chosen following the benefits-risks balance for the patient.

P 108.

A rare case of concomitant Hodgkin Lymphoma and Langerhans cell hyperplasia in an 8-year-old child

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Background

Langerhans cell histiocytosis (LCH) is a rare clonal proliferative disorder of abnormal Langerhans cells (LCs). Association between a variety of malignant disorders and LCH has been previously reported. The most common being malignant lymphoma, including classical Hodgkin Lymphoma (cHL). However, abnormal proliferations of LCs (LC hyperplasia), histologically indistinguishable from true LCH, may rarely occur in association with a malignancy and are often incorrectly labeled as LCH. We report a case of a child with a cHL in whom an excess of LCs has been incidentally identified on a standard biopsy.

Case Report

An otherwise healthy 8-year-old girl presented with a one-week history of a left supraclavicular lymphadenopathy and fatigue. Medical history did not reveal any signs of disease. Physical examination revealed a palpable, firm and sensitive left supraclavicular mass measuring approximately 3 x 4 cm and multiple smaller right inferior cervical ganglions. The biological analyzes showed hyperleukocytosis with elevated levels of neutrophils, monocytes and eosinophils. The erythrocyte sedimentation rate was increased at 120 mm/h. Serological test excluded any recent viral infection. An ultrasound and cervico-thoracic CT scan were performed, showing a large mediastinal mass, bilateral cervical, supraclavicular and left axillary lymph nodes, all hypermetabolic on FDG-PET scan. A biopsy of the left supraclavicular lymph node showed extensive nodal architectural effacement by a nodular proliferation containing Reed-Sternberg cells (CD30+ and CD15 (partial)+, and CD3-, CD20-) intermingled with numerous eosinophils and multiple large cells with abundant eosinophilic cytoplasm, which were positive for CD1a, confirming to be LCs. A diagnosis of nodular sclerosis cHL with excess of LCs was rendered. Identification of a BRAF mutation is ongoing. Bone marrow aspiration and biopsy were normal. The patient was treated as stage IIA (ESR > 30mm/h) cHL with chemotherapy after inclusion in EURONET-PHL-C2-TL2 group.

Conclusion

The relationship between LC hyperplasia and concurrent lymphoma remains unknown and questions remain as to whether the presence of LCs represents a true clonal neoplasm or an exaggerated reactive phenomenon. Current data demonstrated the polyclonal nature of the LCs, which suggests that the excess of LCs may be driven by chemokine/cytokine or other stimuli produced by the associated neoplasm, without any prognostic repercussion.

Posters**P 109.****Favism – A yellow boy and his beans**

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*AZ Damiaan, AZ Sint-Jan*Introduction

Favism is a haemolytic response to the consumption of fava beans. The reaction manifests within 6 to 24 hours of the bean meal, with exhaustion, jaundice and dark urine. The high concentration of certain glucosides in the fava beans triggers destruction of red blood cells who are glucose 6-phosphate dehydrogenase (G6PD) deficient. Therefore favism only occurs in people with this X-linked defect.

Case Report

We present the case of an 18 month-old boy, presenting at the emergency department with fatigue, jaundice and dark urine. Symptoms started two days before presentation. Tachycardia (>120 bpm) and hypertension (124/60 mmHg) were observed. There was no relevant medical or family history. There was no access to medications or other toxic agents. The child is of Syrian descent.

Blood test showed a severe normocytic haemolytic anaemia: haemoglobin 5.2 g/dL, mean corpuscular volume 85.2 fL, haptoglobin 0.06 g/L, lactate dehydrogenase 535 U/L, indirect bilirubin 7.4 mg/dL, reticulocytosis 6.8%. Peripheral blood smear was normal. Direct and indirect Coombs were negative. Osmotic fragility, G6PD-deficiency and haemoglobin electrophoresis tests were sent for further analysis. Urine sample showed haemoglobinuria. Ultrasound of the abdomen showed no hepato- or splenomegaly. The patient was admitted and given a packed cell transfusion on which his vital parameters stabilized. Extensive case history revealed that the child had eaten fava beans two days prior to presentation, which made the diagnosis of G6PD-deficiency the most plausible.

Conclusion

This case illustrates a less known condition in Western countries. G6PD deficiency is particularly common in certain parts of Africa, Asia, the Mediterranean and the Middle East. Through global migration and immigration ethnic groups with the highest incidence of this inherited genetic disorder will enter into the Western healthcare system. A raised awareness for the potential of G6PD-deficiency will lead to proper screening by clinicians and to education of affected patients.

P 110.

Neonatal leukemia : Report of a case with Blueberry Muffin Syndrome

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Case report

A full-term newborn baby, born to parents without any particular medical history, presented at birth with some purple cutaneous nodules and papules. These lesions were widespread over the whole body but predominated on the trunk and scalp. This rash was consistent with a Blueberry Muffin Syndrome. The clinical examination was otherwise unremarkable.

We started an assessment. The baby had no thoracic or abdominal tumor such as neuroblastoma and wasn't affected by a congenital infection. We also excluded hemolytic disease of the newborn and hereditary spherocytosis. Skin biopsy showed some myeloid blasts. Finally, marrow aspiration led to the diagnosis of neonatal acute myeloid leukemia (AML). A genetic assessment ruled out trisomy 21 and Noonan syndrome.

At this stage, given the high toxicity of chemotherapy in newborns, we decided to delay chemotherapy as long as the baby remained clinically and biologically stable. The aim was to allow organ maturation and weight gain. We followed the newborn closely. After 6 weeks, he developed a severe neutropenia, therefore we decided to start chemotherapy. The evolution was favorable except for some breech skin complications.

Discussion

Blueberry muffin syndrome corresponds to cutaneous papules or nodules, often red or purple, presented at birth. These lesions predominate on the face, neck and trunk and correspond to a dermal erythropoiesis which is pathological after birth. The main etiologies are: congenital infection such as toxoplasmosis, rubella or cytomegalovirus, hemolytic disease of the newborn (ABO or Rh incompatibility), hereditary spherocytosis and congenital malignancies, including histiocytosis, neuroblastoma and leukemia. A skin biopsy is often realized to help diagnosis.

A neonatal leukemia is a leukemia that develops within the first 28 days of life. It affects one newborn in five million. 2 out of 3 neonatal leukemia are AMLs. They are frequently associated with some genetics syndrome such as Noonan syndrome and trisomy 21. The most common clinical signs are hepatomegaly, splenomegaly and leukemia cutis. The diagnosis is made by bone marrow aspiration. Treatment consists in chemotherapy with age appropriated doses.

Conclusion

Clinicians should not trivialize the presence of cutaneous papules or nodules (blueberry muffin rash) discovered during routine newborn examination, as these could lead to a potentially lethal diagnosis such as a congenital leukemia.

P 111.**Pulmonary embolism, an unusual presentation of Fanconi anemia**

P. François, P. Philippet, S. Schiffers, L. Rouffiange, S. Eiras da Silva, M. Thimmesch, C. F. Chantrain

CHC Liège

Fanconi anemia (FA) is a rare inherited bone marrow failure syndrome characterized by pancytopenia, congenital abnormalities and predisposition to malignancy. Pulmonary embolism (PE) is uncommon in children and has never been described in FA. We report the case of an adolescent who experienced massive PE that led to a diagnosis of FA.

A 15-year-old boy presented with right lower lobar pneumonia and pleurisy. Blood sample showed macrocytic anemia and thrombocytopenia. Two months later, he was admitted with chest pain, dyspnea and haemoptysis leading to the diagnosis of massive bilateral PE. He had thrombocytopenia at 40000/ μ L and macrocytosis. He was treated by surgical thrombectomy and with anticoagulant. Later, thrombocytopenia and macrocytosis persisted while anemia and moderate leukopenia progressively occurred. Thrombophilia evaluation showed a moderate antithrombin III (ATIII) deficiency at 50% and a double heterozygous mutation in the MTHFR gene. Several signs suggesting a congenital disorder were noticed: small size, triangular face, hypotelorism, discreet hypotrophy of thenar eminence and increased foetal haemoglobin. Medullary puncture showed moderate hypoplasia. The genetic check revealed a rise in chromosomal breakages suggesting a diagnosis of FA. Molecular biology confirmed two heterozygous deletions in the FANCA gene.

The association between FA and PE has not been reported. However, a recent study showed an increased incidence of thromboembolic events in patients suffering from aplastic anemia (AA). Pro-inflammatory cytokines and endothelial dysfunction could promote a state of hypercoagulability. Moreover, several cases of PE have also been reported in children with Blackfan-Diamond anemia. Factors, not related to FA, may have fostered the development of PE in our patient. First, the thrombotic assessment revealed ATIII deficiency and a double MTHFR mutation which is a controversial risk factor for thromboembolic phenomena. Secondly, the PE occurred after a four-hour coach trip that may have induced venous stasis and promoted PE, although there was no deep vein thrombosis. Lastly, the pneumonia may also have contributed as the infection increased coagulability.

In conclusion, we cannot affirm a real relationship between FA and PE even if an increased risk of thromboembolism in patients with AA has been reported. However, this case underlines the importance of considering congenital AA in cases of unexplained persistent cytopenia.

P 112.

Infantile hemangioma : watch out for the hidden part of the iceberg...

A. Fohn, S. Cao, M-C Seghaye, J. Longton

CHU de Liège, U Liège, CHR de la Citadelle

Aim

Infantile hemangioma (IH) is the most common benign vascular tumour in children (5-10% of infants under one year of age). The natural evolution comprises 3 phases: a proliferation phase (during the first months), a latency phase (towards the end of the first year) and a spontaneous regression phase (from 4 to 7 years). Therefore most of IH do not require any treatment. However, depending on the localization, there may be a risk of complication and treatment with a Beta-blocker is indicated in order to accelerate the involution of the lesion. Our presentation reports the case of a child with a bearded segmental IH. They are more aggressive and more at risk of complications (laryngeal involvement) and may be associated with syndromes such as PHACE(S) syndrome. This one includes abnormalities of the posterior fossa, facial IH, abnormalities in the anatomy of the cerebral arteries, cardiac malformations, ocular and sternal abnormalities. In the case of bearded hemangioma, fibroscopy and a thorough ENT examination should always be performed to rule out laryngeal involvement.

Method

This presentation is based on a clinical case study of a newborn with a bearded hemangioma with an initially unfavourable course of this one but that gradually regresses with treatment by Propanolol.

Result

We report the case of an infant born at term and from a vaginal delivery. At a few days of life, capillary lesions appeared on the face associated with subcutaneous swelling. The lesions were rapidly progressive. There was no respiratory distress but painful necrosis of the lower lip, preventing proper feeding. Given the facial localization and the segmental aspect of the infantile hemangioma, a check-up was performed to rule out a PHACE(S) syndrome because significant stenosis of the intracerebral vessels could contraindicate treatment with Beta-blockers. The results were negative. Propanolol was started and the IH gradually regressed, allowing a correct dietary recovery and the absence of laryngeal complications.

Conclusion

This clinical case reminds us of the importance of an early evaluation of the risk of complications of an IH in order to introduce a Beta-Blocker treatment as soon as possible (indeed the introduction of the treatment after the growth phase reduces its effectiveness), and of the importance of carrying out a complementary assessment given the risk of association with other malformations in the context of a segmental hemangioma on the face (PHACES syndrome).

Posters**P 113.****A successful management of generalized kaposiform lymphangiomatosis manifested by chylothorax in an 11-month-old boy: a case report**

F. Lebrun, P. Schlessler, J. Khamis, P. Philippet

*CHC Liège-Belgium, University of Liège*Introduction

Lymphangiomatosis is a rare congenital disease characterized by an abnormal proliferation of lymphatic vessels. The damage can be multi-systemic except in the brain which does not contain lymphatic vessels. The kaposiform presentation is a generalized and very aggressive form, which is distinguished by his histopathology and the presenting symptoms.

Case report

Accidental discovery of bilateral pleural effusion on cardiac ultrasound in an 11-month-old boy during a follow-up visit of an interventricular communication. A chest X-ray and chest CT confirmed the presence of bilateral pleural effusion, larger on the right. A pleural drain was placed at this side. Analysis of pleural fluid revealed a chylothorax (triglycerides at 720 mg/dl, WBC at 9700 with 98% of lymphocytes). Cardiac ultrasound also showed an associated moderate pericardial effusion. Clinically, the patient presents a greater fatigability, a relative anorexia and a polypnea since few days.

A full-body MRI confirms the presence of intrathoracic effusions and also shows significant jugulo-carotid lymphadenopathy, vertebral and splenic lesions and peri-aorto-cave centimetric lymphadenopathy. A lymph node biopsy will confirm the diagnosis of Kaposiform lymphangiomatosis. Initial management justified the installation of a thoracic drain, the prescription of an exclusive parenteral diet and the initiation of octreotide. Clinical evolution was initially favorable but we were then faced with two new degradations justifying drainings and a transient phase of mechanical ventilation. There was no significant biological disorders initially. In the course of the hospitalization several biological disorders appeared transiently, mainly because of the protein loss related to pleural drainages. After the diagnosis of lymphangiomatosis was confirmed by genetic analysis, the patient was treated with sirolimus. Subsequently, the clinical evolution was clearly favorable.

Discussion

Kaposiform lymphangiomatosis is a rare condition. Management is not yet formally validated. The treatments are generally supportive. Pleurodesis, pleurocentesis and ligation of thoracic duct were not considered given the mixed benefit reported in the literature. In addition to measures to limit the formation of chyle, several molecules are currently undergoing clinical studies to try to act directly on the formation of lymphatic, dysfunctional process responsible for the disease. In this context, the Sirolimus seems to occupy a prominent place.

P 114.

Severe phototoxicity associated with concomitant use of methotrexate and voriconazole, an overlooked drug-drug interaction

D.J. Bogaert, L. Verlinden, E. Vandecruys, G. Laureys, E. Verhaeghe, T. Bauters

UZ Gent

Background

We report a patient with T-cell lymphoblastic lymphoma who developed severe phototoxicity under concomitant treatment of methotrexate (MTX) and voriconazole. Voriconazole, an antifungal agent, is generally well tolerated. However, its use can be associated with various cutaneous reactions such as phototoxicity involving skin and/or lips. Toxic skin reactions caused by MTX are less frequent. MTX has been rarely associated with ultraviolet (UV) reactivation, an inflammatory skin characterized by the recurrence of previous sunburn.

Although concomitant use of MTX and voriconazole is not uncommon in an oncology setting, we suspect that the associated phototoxicity is often under- and/or misdiagnosed. Thus far, only two reports describing this phenomenon have been published.

Methods + results

The patient is a 17-year-old Caucasian male treated for T-cell lymphoblastic lymphoma (EORTC-CLG 58081). He was kept on secondary prophylaxis with voriconazole following pulmonary aspergillosis. The first interval phase of his chemotherapy protocol, encompassing four cycles of high-dose (HD) MTX, was given during the summer period. After the second HD-MTX cycle, he presented with erythema, swelling and yellow-black crusts of the lips. This was diagnosed as herpes simplex infection and acyclovir was started. PCR for herpes simplex virus was negative. Meanwhile, he got sunburned causing a temporary flare-up of the lesions. When the lips were healed, chemotherapy was restarted. After the third HD-MTX cycle, he developed severe erythema and edema at all sun-exposed areas (face, lips, neckline, forearms, hands, legs). There were no mucosal lesions. A skin biopsy confirmed the diagnosis of phototoxicity, which was considered as a drug-drug interaction due to the simultaneous use of MTX and voriconazole. Antifungal prophylaxis was switched to daily IV caspofungin. The last HD-MTX cycle was cancelled and continuation of his chemotherapy protocol needed to be postponed. Under local corticosteroid treatment and strict shielding from sunlight, it took two months for the phototoxic lesions to completely resolve.

Conclusion

The enhanced risk of phototoxicity under concomitant use of MTX and voriconazole is not widely known and was initially misdiagnosed in this patient. The diagnostic delay resulted in more extensive lesions, which had a considerable impact on his quality of life and required reduction of his chemotherapy.

Posters**P 115.****Atypical presentation of a precursor B-cell Lymphoblastic Lymphoma**

T. Alliet, P. Van der Speeten, Y. Eelen, L. De Somer, S. Pans, M. Renard, H. Segers

*UZ Leuven*Background

A 9-year-old Nepalese girl presented to the emergency unit with atraumatic pain, redness and swelling of alternating joints (left and right ankle, left elbow) since 2 months. There were no B-symptoms. She had already been treated with intravenous flucloxacillin for suspicion of septic arthritis without improvement. Non-steroidal anti-inflammatory drugs were initiated for a possible diagnosis of juvenile idiopathic arthritis, although the rapid alternation between affected joints was rather atypical. She was admitted because of persisting symptoms for further diagnostic work-up. Magnetic resonance imaging (MRI) revealed multiple bone lesions in both lower limbs. Positron emission tomography-computed tomography (PET-CT) showed multiple hypermetabolic bone lesions in both lower and upper limbs, but no primary tumour. Bone marrow aspiration showed no aberrations on morphology but flow cytometry showed a small population (0.02%) of leukemic B-lymphoblasts (strong CD10 expression, TdT+, CD19+, CD20-, CD34+, CD45+). This suggested the presence of a lymphoma, however the low lymphoblast count didn't meet diagnostic criteria. Eventually, final diagnosis of a precursor B-cell lymphoblastic lymphoma (PB-LBL) was made by biopsy of a lesion.

Methods

A literature search was conducted in the MEDLINE database for similar paediatric presentations of extranodal precursor B-cell lymphoblastic lymphoma (PB-LBL). Articles were selected based on title and abstract.

Results

Lymphomas account for approximately 7% of paediatric malignancies and are classified as Hodgkin's disease or non-Hodgkin's lymphoma (NHL). Lymphoblastic lymphoma accounts for 30% of the childhood NHL cases and is differentiated from leukaemia based on the marrow blasts count (<25% in lymphoma). Further categorisation into precursor B-cell or T-cell is based on immunohistochemical criteria of which PB-LBL comprises only 10% of the cases.

We found several paediatric case reports of extranodal presentations of B-LBL such as the concha, atrium, skin (5 cases), breast (2 cases), soft tissue (3 cases), pleura (2 cases), omentum. We only found two other cases of multifocal bone involvement (age 3 and 15).

Conclusion

PB-LBL is a rare entity that often has an extranodal presentation. The multifocal bone involvement, as seen in our case, is very rare and can be misdiagnosed as an infectious or rheumatologic process. Further investigation is warranted if there is insufficient response to treatment

P 116.

Recurrent cholesteatoma in a child with Fanconi anemia

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CHC Liège, U Liège, CHU-UCL Namur

Background

Cholesteatoma is a rare disorder consisting of abnormal growth of keratinizing squamous epithelium behind the tympanic membrane. It results either from an entrapment of epithelial rest during embryogenesis (congenital cholesteatoma) or from an abnormal retraction or migration through tympanic perforation (acquired cholesteatoma). Fanconi anemia is an inherited cause of bone marrow failure typically associated with physical abnormalities and increased risk cancer. Ten to 20 % of Fanconi patients presented with otologic manifestations such as hearing loss, thin tympanic membrane, narrow stenosis of external auditory canal, aberrant tympanic bony island, malformed malleus and abnormal course of chorda tympani. We report the case of a child with Fanconi anemia who developed recurrent cholesteatoma.

Case report

A 7-year-old boy was referred for mild neutropenia (700/uL) and thrombocytopenia (30000/uL). He also had several congenital abnormalities including short stature, microcephaly, ectopic left kidney, café-au-lait spots, dysmorphic face and stenosis of the right ear canal. In early childhood he had recurrent otitis, pharyngitis and a parapharyngeal abscess. At the age of 6 years, he developed a progressive hearing loss related to right cholesteatoma and was treated by surgical tympanoplasty. The diagnosis of Fanconi anemia was supported by abnormal chromosomal breakage testing and confirmed by the finding of homozygous FANCA mutation. After 2 years of evolution without significant infection, the child presented with a new hearing deterioration related to cholesteatoma relapse. He underwent surgical mastoidectomy and was treated later by related stem cell transplantation.

Discussion

This case is the first report of cholesteatoma in patient with Fanconi anemia. Several factors can potentially support this association: in animal models, genes of the FANC family are expressed in branchial arches from which outer and middle ear derive; anatomic abnormalities modifying pharyngeal dimensions, eustachian tube and middle ear morphology as observed in Fanconi patients are associated to an increased risk of cholesteatoma; immune defects and in particular progressive neutropenia favor chronic or recurrent otitis that predispose to cholesteatoma.

Conclusion

This report underlines the need of otorhinolaryngologic screening and follow-up in Fanconi patients.

Long Oral Presentation**LO 5.****Clinical and biochemical phenotype of 11 patients with bi-allelic mutations in TANGO2**

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UZ Gent, Queen Silvias Child Hospital, UZ Brussel

Background

TANGO2 (transport and golgi organisation 2) related disease is a recently described cause of recurrent crises with rhabdomyolysis and arrhythmias that can also present with developmental delay and (subclinical) hypothyroidism. The exact function of TANGO2 is unknown, but a link with mitochondrial energy metabolism has been suggested. Here we describe the clinical and biochemical characteristics of 11 patients (5 Belgian and 6 Swedish) from 7 families with bi-allelic mutations in TANGO2.

Results

The clinical phenotype was characterised by developmental delay and/or progressive spastic paraparesis with language delay. The majority of patients (8/11) had metabolic crises associated with catabolism causing decreased consciousness, ataxia and weakness with subsequent development of rhabdomyolysis. Interestingly three patients, diagnosed because of an affected sibling, never had documented rhabdomyolysis. The age of the first rhabdomyolysis ranged from 6 months to 18 years. Brain MRI was performed in 9 patients and showed, apart from mild white matter loss in 2 cases, no significant abnormalities.

Cardiac arrhythmias (prolonged QTc, torsades de pointes and VT) were seen in 5/11 patients. The arrhythmias were lethal in 4 patients (aged 9 months to 27 years). Echocardiography was performed in 8 patients and did not show structural defects.

Hypothyroidism was documented in 8/11 patients, and preceded rhabdomyolysis in one patient.

Abnormal acylcarnitine profile or dicarboxylic aciduria was observed in 7/11 patients during crises. Palmitate oxidation in fibroblasts from 2 patients was normal. Muscle biopsy was performed in six patients and did not show abnormalities on histology. Spectrophotometry of OXPHOS was performed in 4 patients and showed a slight decrease in complex II in 3 patients.

Six patients were homozygous for a large intragenic deletion, two were homozygous for a nonsense mutation, three were compound heterozygous.

Conclusion

These 11 patients demonstrate the wide phenotype of TANGO2 related disorders. Probably the incidence of rhabdomyolysis is overestimated in this report, as in all families this was the clinical clue to make the diagnosis. Moreover, because of the high prevalence of intragenic deletions, WES cannot be readily used to exclude this disease.

LO 6.

New FDG-PET analysis confirm previous evidence for the epileptic network of Lennox-Gastaut Syndrome

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Lennox-Gastaut syndrome (LGS) is a severe form of childhood-onset epilepsy characterised by generalized tonic seizures, characteristic electroencephalogram patterns, and cognitive impairment. The electroclinical phenotype arises from diverse aetiologies. Fluorodeoxyglucose Positron Emission Tomography (FDG-PET) during the interictal state often shows reduced brain metabolism in brain networks involved in the epileptic process. We aimed to show the cortical regions involved in the epileptic process of LGS by comparing brain metabolism in a group of patients with LGS to a group of epilepsy controls.

We compared FDG-PET metabolism in 17 patients with LGS to the uninvolved hemispheres in 18 age- and sex-matched control patients with left and right temporal lobe epilepsy (TLE, 10 with L-TLE and 8 with R-TLE). Each subject's FDG-PET scan was first split into left and right brain hemispheres. Each hemisphere was then intensity normalised to the mean signal within cortical white matter; co-registered to the patient's T1-weighted MRI; spatially warped and re-sampled to Montreal Neurological Institute space (2mm³ voxels); and spatially smoothed at 10mm. We flipped all the left hemispheres of R-TLE on their right orientation to gain statistical power and compare them to non-lesional LGS hemispheres. Non-parametric, permutation-based, two-sample t-tests were used to compare the hemispheres of the LGS patients to the "unaffected" hemispheres of the TLE control patients.

We found significant hypo-metabolism (LGS relative to TLE controls; $p < 0.025$, corrected) in midline and lateral fronto-parietal cortex, including middle and superior frontal gyri, posterior cingulate, precuneus, and angular gyri. These cortical areas correspond to the location of the Executive Control Network (dorso-lateral prefrontal, inferior parietal cortex).

Patients with LGS have areas of FDG-PET hypo-metabolism that closely resemble the areas of cortical activation we have previously reported using simultaneous EEG-functional MRI of interictal generalised paroxysmal fast activity and Single Photon Emission Computed Tomography of tonic seizures. Key areas of hypo-metabolism correspond to brain regions of known major cognition-related networks, particularly the fronto-parietally situated 'executive-control network'. Our results provide convergent clues for a key role of this network in the epileptic process of LGS and therefore point a new therapeutic target.

Long Oral Presentation**LO 7.****First epileptic EEG activity in infants with Tuberous Sclerosis Complex is associated with neurodevelopmental outcome at the age of 2 years**

J. De Ridder, B. Verhelle, J. Vervisch, K. Lemmens, K. Kotulska, R. Moavero, P. Curatolo, B. Weschke, K. Riney, M. Feucht, P. Krsek, R. Nabbout, A.C. Jansen, D. Domanska-Pakieła, M. Kaczorowska-Frontczak, C. Hertzberg, H.M. Hulshof, S. Samuelli, B. Benova, E. Aronica, D.J. Kwiatkowski, F.E. Jansen, S. Jóźwiak, L. Lagae

KUL, The Children's Memorial Health Institute, Tor Vergata University, Bambino Gesù Children's Hospital, Charité University Medicine Berlin, Queensland Children's Hospital, Medical University Vienna, Motal University Hospital, Necker- Enfants Malades Hospital, UZ Brussel, Vivantes Klinikum Neuköln, University Medical Centre Utrecht, University of Amsterdam, Stichting Epilepsie Instellingen Nederland, Harvard Medical School, Warsaw Medical University

Purpose

To study the association between the first epileptic EEG activity (interictal epileptiform discharges (IED) or seizures) in infants with Tuberous Sclerosis Complex (TSC) and neurodevelopmental outcome at 24 months of age.

Methods

In this study we included 83/94 TSC infants from the international prospective EPISTOP trial. The characteristics of the first EEG with epileptic activity (age at first epileptic EEG, presence of IED or seizures) were correlated with neurodevelopmental outcome (results Bayley Scales of Infant and Toddler Development III and autism spectrum disorder (ASD)) at 24 months using non-parametric test statistics.

Results

79/83 patients developed epileptic EEG activity by the age of 24 months. The median age at the first epileptic EEG was 77 days (IQR [23-111]). At the age of 4 months, 78% had at least 1 EEG with epileptic activity; at 5 months 83%; at 6 months 87%; at 12 months 93% and at 18 months 95%. In 63% epileptic EEG activity was multifocal. In 15% a seizure was recorded.

A younger age at first EEG with epileptic activity was significantly associated with lower cognitive (p-value 0.001), language (p-value 0.001) and motor (p-value 0.002) developmental quotients (DQs). The presence of seizures on this EEG was significantly associated with lower cognitive (p-value 0.047), and language (p-value 0.030) DQs, but not with motor (p-value 0.058) DQs. No significant associations were found between the presence of multifocal epileptic activity and the DQs. No significant associations were shown between a younger age at first epileptic EEG activity, presence of IED or seizures and a higher probability of ASD at 24 months.

Conclusion

In this TSC cohort, a younger age at first EEG with epileptic activity was significantly associated with a worse cognitive outcome. Multivariable analyses are needed to include the contribution of seizures, mutation status, MRI characteristics and treatment.

SO 1.

Executive function assessment in 2-year-olds born preterm.

A. Van den Brande

*UZ Leuven*Background

During the past decade there has been increasing interest in aspects of the broad construct of executive function (EF) in childhood. EF refers to higher order, self-regulatory, cognitive processes including working memory, inhibitory control and cognitive flexibility. Current literature has shown that children born preterm perform poorer than term-born peers on the three core EFs. These EF deficits have also been shown to be key to the behavioural and academic problems observed in this group. To our knowledge there are no studies assessing EF in preterm born children at the age of 24 months. Therefore, we performed a battery of three age-appropriate tasks assessing the three core EFs in this cohort. The aims of the study were to assess EFs in 2-year-olds born preterm and to examine the relationship between EFs and developmental outcome at 24 months corrected age.

Methods

A prospective cohort study was designed to study EF in extreme (EPTB), very (VPTB) and moderate (MPTB) preterm born children. The preterm born infants were followed from birth to 24 months corrected age. Multisearch multilocation (MSML), Reversed categorization (RevCat) and snack delay task (SDT) was applied to assess the EF of infants. Developmental outcomes were measured by using the Bayley Scale of Infant Development (BSID-III).

Results

At 24 months corrected age 99 children (16,2% EPTB, 49,5% VPTB, 34,3% MPTB) completed the EF assessment battery. Success rates were highest in MSML (85,9%), followed by SDT (66,7%) and RevCat (26,3%). More EPTB children were unable to inhibit their behaviour on SDT compared to VPTB and MPTB children ($p=0,013$) and MSML was positively correlated with postmenstrual age ($p=0,002$). Total task scores for MSML, RevCat and Snack delay were all positively correlated with mental developmental index (mean: 104,78, SD: 13,28) and psychomotor developmental index (mean: 105,72, SD: 15,27) on BSID-III ($p<0,001$).

Conclusions

This study provides evidence that it is possible to assess EF in 2-year-olds born preterm. Since EF scores are directly correlated with developmental outcome at 24 months, EF may be a valuable target for early identification of at-risk children and subsequent intervention.

Poster Walk**PW 12.****Pediatric neuroborreliosis**

J. Vanbekbergen

UZ Leuven

Diagnosing Lyme neuroborreliosis in the pediatric population can be challenging, especially in absence of erythema chronicum migrans or explicit neurological symptoms. Consensus on the diagnostic process in children is lacking. This retrospective study gives an overview of most frequent presenting symptoms, work up and management at the University Hospitals Leuven, Belgium. A retrospective analysis was performed of 20 pediatric patients with Lyme neuroborreliosis treated at the University Hospitals of Leuven from 2014 until 2019. Medical records were reviewed and data, including peripheral blood and cerebrospinal fluid results, imaging reports and treatment methods were collected.

All patients presented during spring or summer time. In only a minority of patients (25%) a history of a tick bite was reported, and erythema chronicum migrans lesions were never even noted. Clinical presentation varied but facial nerve palsy was the main presenting symptom (75%). Only fifty percent of these children had a positive immunoblot in peripheral blood, while eighty percent had intrathecal synthesis of *Borrelia* antibodies. All patients were treated with intravenous antibiotics, of which seventy percent had complete clinical resolution. Fifteen percent of the patients had only minimal sequelae, other data were lacking.

Pediatric Lyme neuroborreliosis can be difficult to diagnose. Serological testing alone is not sufficient for the diagnosis. A lumbar puncture, although invasive, is necessary for confirmation of the diagnosis by detection of intrathecal synthesis of *Borrelia* antibodies. Overall, prognosis is good after adequate antibiotic treatment.

PW 14.**A first case of acute flaccid myelitis associated with enterovirus D68 in Belgium**

I. Delpire, M. Rodesch , F. Vermeulen , F. Christiaens

ULB - ERASME

Introduction

Acute flaccid myelitis (AFM) is an acquired spinal cord disorder mostly affecting young children characterized by the acute-onset of flaccid weakness in one or more limbs combined with a spinal cord lesion restricted to the gray matter on magnetic resonance imaging (MRI). Recent publications outline the outbreaks relation between enterovirus D68 (EV-D68) infections and AFM.

We describe the first case of an AFM in a child associated with EV-D68 in Belgium.

Case report

A 4 years old girl (previously healthy and properly vaccinated including the polio vaccine) presented with paralysis of the right upper limb one week after an upper respiratory tract infection. The normal head-CT and a pleiocytosis in cerebrospinal fluid evoked acute disseminated encephalomyelitis, neuromyelitis optica spectrum disorders, AFM, transverse myelitis or Guillain-Barré syndrome. The MRI revealed spinal gray cord matter lesion from C2 to C7 and extending to the brainstem.

Treatment with high-dose corticosteroids showed no clinical improvement. A second line treatment with immunoglobulins intravenously (Ivlg) improved the patient clinical state. MRI showed a clear regression of the lesions. Naso-pharyngeal aspirate revealed an EV-D68; enterovirus A71 was found in the stools. The patient returned home with physiotherapy treatment and recovers some of her mobility but still has weakness in the lower right arm.

Discussion

The definition of AFM was proposed by the CDC in 2014 after a large outbreak of EV-D68 in the United States (US). Since then, there has been an increase of AFM cases associated with EV-D68 worldwide. In Belgium, 26 cases of EV-D68 were isolated from the respiratory tract in 2018 with no associated AFM cases.

MRI with a T2 hyperintensity in spinal cord grey matter with or without hyperintensity at dorsal brainstem and detection of EV-D68 specific nucleic acids in a respiratory specimen using a validated PCR assay can confirm this diagnosis. Ivlg is recommended in the acute phase and steroids could be not effective.

Conclusion

This is the first case of AFM associated with enterovirus D68 in Belgium. The prognosis of this pathology well described in outbreaks in the US is often not favorable. Proper recognizing and documenting of AFM is important to avoid under-diagnose so the pathogenesis can be better understood in order to find an effective treatment.

Poster Walk**PW 36.****Interest of Positron Emission Tomography in small vessel primary angiitis of the central nervous system**

M. Belcour, P. Dontaine, A. Monier, L. Lebrun, I. Salmon, A. Van Hecke, C. Fricx, O. De Witte, G. Boitsios, S. Goldman, X. De Tiège, A. Aeby

ULB - HUDERF, ULB - ERASME

Introduction

Primary angiitis of the central nervous system (PACNS) is an inflammatory brain disease affecting the medium and small vessels of the CNS. The diagnosis that requires confirmation by brain biopsy remains challenging due to unspecific clinical presentation and low specificity of the complementary exams. Instead of large and medium vessels vasculitis, the brain positron emission tomography (PET) is less well established in small vessel (sv) PACNS as there are no systematic studies in the literature. The goal of this presentation is to report two biopsy-proven pediatric cases of sv PACNS with PET investigation and discuss the utility of PET in the diagnosis of this rare condition.

Case Report

Patient 1 was a previously healthy twelve-years-old girl, who presented with paresthesia and focal seizures followed by the development few days later of a bilateral optic neuritis. The extensive autoantibody search was negative and we found no evidence of infection. The cerebrospinal fluid (CSF) revealed mild pleocytosis. Brain MRI showed bilateral high signal intensities in T2-weighted images and FLAIR images in the pre-central sulcus showing enhancement on the T1-weighted post contrast images and vasogenic edema on diffusion. EEG showed bilateral frontal spike and waves. Whole body Fluoro-deoxy-glucose (FDG) PET was compatible with an inflammatory process in both frontal lobes and no extra CNS inflammation. PACNS was suspected and a cerebral biopsy was performed, guided by MRI and FDG-PET, showing a lymphocytic predominant sv PACNS. Her symptoms progressively resolved under a treatment with levetiracetam, steroids and monthly cures of cyclophosphamide. Patient 2 has already been reported. A 9-years-old boy presented with headaches and focal seizures. Examination and blood tests were unremarkable. Brain MRI revealed a left occipital gadolinium-enhancing tumor-like lesion. The cerebrospinal fluid showed pleocytosis and elevated proteins. PET showed an increase in L-[methyl-11C]methionine (MET) uptake by the lesion, and no extra CNS inflammation. A left temporal stereotaxic biopsy guided by MRI and MET-PET showed a sv PACNS. Patient was treated with a combination of corticosteroids slowly tapered, and mycophenolate mofetil. He became clinically and radiologically stable 38 months after presentation.

Conclusion

This case-series demonstrate that both brain FDG and MET-PET are valuable tools for the diagnosis of sv PACNS and efficacious to guide brain biopsy.

P 117.

A rare presentation of congenital spinal dermal sinus

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Background

A spinal dermal sinus is a regularly occurring type of spinal dysrafism, with an incidence of 1 out of 2500 live births. A spinal dermal sinus is a tract into the intrathecal space, and predominantly localized in the lumbosacral region. Early detection is essential to prevent complications, e.g. infections. This anomaly is also associated with tethering of the spinal cord, which may lead to neurological complications.

Method

Case report

Results

We present a neonate with a skin defect near the left scapula, which was seen shortly after birth. MR-whole spine demonstrated a low cervical spinal dermal sinus, dorsal deviation of the dural sac and tethering of the myelum. The low cervical and paravertebral position is a very unusual location. The patient was referred to a pediatric neurosurgical center for surgical correction. The whole tract was removed and tethering was released, confirmed by postoperative MR. There were no complications.

Conclusion

Spinal dermal sinuses can develop at all spinal levels. Therefore, the whole spine must be thoroughly examined for its presence. Early diagnosis of spinal dermal sinus is crucial as earlier surgery is more successful in preventing irreversible neurological damage.

Posters**P 118.****Obstructive hydrocephalus leading to the diagnosis of diencephalic syndrome**

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U Liège, CHU de Liège

Diencephalic syndrome (DS) in children may be a neglected condition with unspecific symptoms such as emaciation and failure to thrive despite normal caloric intake often due to a hypothalamic tumor. Hyperalertness, euphoria, nystagmus and hyperkinesia are classically also described. DS is a rare disease the prevalence of which remains unknown. Diagnosis is facilitated if a hydrocephalus is present.

We report the case of a 2-year-old boy referred to the emergency room for headache, vomiting, sleepiness and weakness. The child was known to be thin since birth, therefore secondary failure to thrive took time to be noticed and remained first unexplored. Increased Head Circumference and nystagmus had been objectivated by his pediatrician but exploration had not been taken place yet.

Due to his clinical presentation of Intracranial Hypertension (ICH), a CT-scan was performed, that showed a mass syndrome focused on the diencephalon, inducing a noncommunicating hydrocephalus. Thus, an external ventricular drainage was placed to release his ICH, that allowed immediate improvement of the child's condition. The mass ablation was undertaken, the result of the anatomopathology revealed a pilocytic astrocytoma and the child was programmed for chemotherapy.

Conclusion

In our case, diagnosis of DS due to a tumour was made lately in the course of the disease at the stage of severe hydrocephalus despite symptoms existing several weeks before.

DS is an uncommon disease that should be considered in face of emaciation and failure to thrive in a young child.

P 119.

Sylvian stroke in a 12-year-old patient secondary to varicella arteriopathy

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Background

Strokes are rare in children and their management in this age group is not well codified. Given its severity, early recognition is mandatory for adequate treatment initiation.

Case report

A 12-year-old girl with a history of B lymphoblastic leukemia in remission without treatment for 7 years was admitted for a sudden loss of strength in her right upper limb and a facial paralysis followed by Broca's aphasia along with 4 episodes of vomiting. Biology, non-injected cerebral CT-scanner and carotid Doppler were normal. The electroencephalogram showed a slow dysrhythmia in the linguistic area.

The angio-CT scanner revealed a slender left internal carotid with a focal stenosis of the left M1 segment with ischemia of the left sylvian territory. Percutaneous partial arterial re-permeabilization was achieved before transfer to the PICU.

Medical treatment consisted of administration of noradrenaline to maintain supranormal blood pressure and of aspirin for anti-aggregation. Initial outcome was characterized by progressive improvement of aphasia in the first 24 hours but secondary neurological deterioration occurred after 48 hours. Cerebral magnetic resonance imaging (CMRI) revealed new subacute ischaemic foci in the left sylvian territory. Clinically however, the patient recovered progressively with a normal neurological examination at day six.

Extensive infectious-, autoimmune-, tumoral-, inflammatory- and coagulation testings were carried out and revealed no abnormality apart from positive VZV IgG. The child had had varicella four months earlier.

The control CSMRI performed one week later showed signs of blood brain barrier incompetence, reappearance of flow in the proximal part of left M1 and persistence of a discrete parietal irregularity of the latter compatible with sequelae of vasculitis.

The patient was discharged in good general and neurological condition under aspirin (1-2 mg/kg/d) with scheduled regular CSMRI checkups.

Discussion

This case of a post-varicella arteriopathy leading to a stroke in a young girl points out the absolute necessity to have a rapid diagnosis and treatment of stroke in children. As it is the case in adults, children should benefit from a clear procedure of care including emergency CSMRI, early arteriography and fibrinolysis, if indicated. Anti-aggregation by aspirin is recommended.

Conclusion

An established protocol for stroke treatment in children should allow rapid management and may improve prognosis.

Posters**P 120.****A case report of dominant COL4A2 mutation leading to progressive cerebellar atrophy, porencephaly and leukoencephalopathy.**

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*CHU-UCL Namur*Background

COL4A1 and *COL4A2* code for procollagen IV alpha-1 and alpha-2 chains, assembling in a heterotrimeric helix in the basement membrane of many tissues. Mutations in *COL4A1* and *COL4A2* lead to a systemic disease. Lesions frequently observed are ; cerebrovascular lesions causing infantile hemorrhage and porencephaly, renal lesions resulting in nephrosis and hematuria, ophthalmological signs as cataract, microphthalmia and blindness, cardiac signs causing arrhythmia and muscular lesions leading to weakness and myoglobinuria.

Case report

We report the case of a 6-year old girl presenting with developmental delay and language disorder. She is the only girl of non-consanguineous healthy parents. There was no history of alcohol, medication or drugs during pregnancy. Neonatal history revealed a short and twisted umbilical cord and polyhydramnios. The antenatal ultrasounds were normal and the posterior fossa showed no abnormalities. At birth, she had transient acute respiratory distress requiring oxygen therapy. She said about ten words but did not associate them. She walked at 18 months. She had behavioral disorder, microscopic hematuria and tricuspidian insufficiency. Clinical examination at 5 years showed mild ataxia, axial and peripheral hypotonia, hyperlaxity, bilateral ptosis, hypertelorism and a large mouth. Brain MRI performed the same year showed cerebellar abnormalities with a complete but depleted vermis, right cerebellar hemisphere atrophy, a left frontal porencephalic cavity and hypersignal of the periventricular white matter bilaterally. The angio-MRI was normal. Genetic analysis revealed a de novo heterozygous mutation in *COL4A2* (c.2821G>A, p. (Gly941Arg) in exon 32).

Discussion

Cerebrovascular lesions seen in *COL4A1* and *COL4A2* mutations are mainly infantile hemorrhage, porencephaly and periventricular leukoencephalopathy. In the literature, more common neurological features associated with these lesions are seizures (43%), hemiparesis (29%), tetraparesis (14%) and developmental delay (38%)¹. No clear genotype-phenotype correlation has been described yet². In the case of our patient, she presented developmental delay associated with aspecific motor abnormalities, language and behavioral disorder. Furthermore, brain lesions were progressive with initial normal ultrasound during the antenatal period. The key for diagnosis was the lesions seen on the brain MRI which oriented our genetic analysis. Nevertheless, diagnostic can still be complicated due to the broad severity range from small-vessel disease to massive hemorrhage, progressive lesions with a variable time of onset and the high rate of de novo mutation.

Conclusion

Mutations in *COL4A1* and *COL4A2* genes are a frequent cause of porencephaly and childhood cerebral hemorrhage with various phenotypic presentations and high rate of de novo mutations. Awareness amongst clinical signs should lead to the realization of a brain MRI to provide an early diagnosis and appropriate follow-up.

P 121.

Life as it is, from a different angle: Congenital laterocollis caused by trochlear nerve paresis.

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UZ Leuven, AZ Groeninge

Congenital torticollis is usually caused by muscular fibrosis in the sternocleidomastoid muscle. It is rarely associated with underlying neurological disorder or bone abnormalities.

Congenital laterocollis, on the other hand, is often caused by ocular abnormalities. We saw, at the pediatric neurology consultation, a 10 year old boy with congenital laterocollis on the left side. He was referred by the orthopedics department for torticollis with no apparent fibrosis in the sternocleidomastoid muscle. Additional imaging ruled out bone defects at the cervical vertebral column or an intracranial process. Consultation at the ear, nose, and throat-department couldn't indicate an underlying cause either. Neurological examination showed a laterocollis with limited flexion of the head to the left, no limitation in mobility of the head and neck and normal active and passive lateroflexion. When testing the eye tracking movements in lateroflexion to the right (Bielschowsky test), when looking to the left, the right eye turned to a more medial and cranial position. These findings are consistent with a right trochlear nerve paresis.

This case presentation draws attention to the need to distinguish between congenital torticollis and congenital laterocollis. A complete clinical neurological examination, including a Bielschowsky test, is essential. In this way, unnecessary investigations and even pointless surgical procedures can be avoided.

Posters**P 122.****Startling cause of abdominal pain in an 11 year-old child**

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Abdominal pain is common in children and can occur in many conditions. The pathologies also differ according to age groups. The diagnosis can sometimes be difficult to make.

While most abdominal pains are caused by minor conditions, the challenge for clinicians is not to miss severe cases.

Clinical case

11-year-old girl presenting at the emergency room for abdominal pain for 5 days not responding to analgesics, without other complaint or symptom.

Her medical background includes scoliosis, asymptomatic von Willebrand disease and a maternal history of neurofibromatosis type 1 (NF1). Genetic testing performed on the patient at the age of 6 suggested the presence of a microduplication of unknown pathological significance inherited from her mother.

Clinical examination reveals 11 non-progressive café-au-lait macules and a tenderness in the left flank, left iliac fossa and suprapubic area without abdominal guarding or palpable mass. The vital parameters and remaining physical examination are normal.

Differential diagnoses are suggested: mesenteric adenitis, renal colic, non palpable abdominal mass. Suspicion of NF1 supports the latter hypothesis (e.g. renal angiomyolipoma, plexiform neurofibroma, gastrointestinal stromal tumor).

An initial assessment includes a normal general blood test, an urinalysis with a macroscopic hematuria resolving spontaneously the next day, and an abdominal ultrasound imaging showing a non-significant grouping of the small intestine's loops in the lower pelvis without requiring additional exploration.

During the hospitalisation, and due to an increasing pain despite proper medications, an abdominal CT scan is performed, revealing a large right retroperitoneal homogeneous mass without calcification invading the sacrum with a maximal diameter of 9,2cm.

The MRI shows an extensive tumor from the sacral plexus within the pelvic cavity and the greater right sciatic foramen, suggesting a benign plexiform neurofibroma.

In this context, a biopsy is performed. The histological analysis is compatible with a neurofibroma, which cannot be removed given its localisation.

The proposed management consists of regular follow-ups and level I-II analgesic medications.

Conclusion

The suspicion of NF1 referred to in our case by the medical history and clinical examination widens the differential diagnosis range of abdominal pain and should rush complementary imaging when faced with persistent abdominal pain without obvious etiology.

P 123.

Sudden speech impairment: the case of a child with arterial ischemic stroke associated with focal cerebral arteritis

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CHC MontLegia, KUL

Case definition

A 9-year-old girl is brought to the emergency room for sudden speech impairment. The neurological examination was normal, except for slowed speech, trouble finding words and finishing sentences. An immediate head computed tomography came back normal and was followed by a magnetic resonance angiography (angio-MRI) within 24 hours. This showed focal lesions with diffusion restriction in the left frontal lobe, compatible with a subacute ischemic stroke, and an appearance of vasculitis in one branch of the middle cerebral artery. Aspirin (3 mg/kg/j) was therefore started and the patient's symptoms quickly resolved.

There is no evidence of varicella-zoster (VZV) infection in the previous year. There is a history of Ehlers-Danlos syndrome in the mother's family.

Extensive etiological investigation was carried out and showed a normal cardiac function without evidence of patent foramen ovale. Ultrasonography of the cervical and renal arteries and a thorough investigation for hypercoagulable status revealed no abnormalities. There was no evidence of autoimmune disease or active infectious processes in the blood or cerebrospinal fluid, including research for VZV. Hemoglobin electrophoresis was normal. The electroencephalogram showed slow focusing in the left temporo-parieto-occipital lobes.

Discussion

Cerebral arteriopathy is the most common cause of arterial ischemic stroke in children, occurring in almost half of the cases. It is a strong predictor of recurrence. Causes of arteriopathy include moyamoya, vasculitis, arterial dissection, sickle cell disease, postvaricella angiopathy and focal cerebral arteriopathy; the latter being the most common type and frequently associated with an infectious trigger.

Imaging of the arterial wall with angio-MRI in order to highlight irregularities is therefore crucial in the evaluation of newly diagnosed ischemic strokes and their follow-up, given the frequency of arteriopathies as a cause of ischemic stroke in children and their high risk of recurrence.

Treatment will be initiated with Aspirin and continued for at least 2 years, the period during which the risk of recurrence is highest.

Conclusion

In presenting this case of arterial ischemic stroke associated with transient focal arteritis, we point out that symptoms are often fleeting in pediatrics, making the diagnosis challenging. We also recall that precise etiological diagnosis is important due to the high risk of recurrence in some cases.

Posters**P 124.****An unexplained cause of status epilepticus associated with anti-TPO antibodies: could it be an Hashimoto's encephalopathy ?**

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*ULB - HUDERF*Background

Hashimoto's encephalopathy (HE) also known as steroid responsive encephalopathy associated with autoimmune thyroiditis is a rare entity in teenagers. We report the case of an atypical status epilepticus.

Case report

A 14-years-old girl was admitted to our PICU for status epilepticus. Induced barbiturate coma was needed to control seizures after failure of high doses of benzodiazepines. The patient suffers from Hashimoto's thyroiditis since she was 10 years old and takes levo-thyroxine's supplement.

The medical workup highlighted normal blood values and negative toxicology screening. The brain imaging was normal. However the protein level in the cerebrospinal fluid (CSF) was elevated (1,38 g/L) without elevation of leucocytes. The treatment included Ceftriaxone and Acyclovir until the bacterial culture and the herpes PCR came back negative. Levetiracetam was added to prevent further seizures. Withdrawal of benzodiazepines and barbiturates was started 24 hours after seizure control.

After excluding central nervous system infection, stroke, tumor, toxics and trauma, in the context of Hashimoto's thyroiditis (plasma anti-TPO antibodies:177 kUI/L), we suspected an Hashimoto's encephalopathy . The search of auto-antibodies in CFS including anti-TPO antibodies were negative. The low sensibility and specificity of these tests could not confirm or exclude this diagnosis. Due to severe clinical presentation, we started an empirical intravenous corticosteroid therapy at 2mg/kg/j on the third day. The clinical recovery was complete one week later and the control brain MRI, EEG and Evoked Potentials were normal. Yet the efficiency of the corticosteroid therapy is uncertain because of concomitant treatment with midazolam and thiopental.

Discussion

Hashimoto's encephalopathy is an uncommon syndrome and is considered as an exclusion diagnosis. Refractory status epilepticus is a common onset and isolated elevation of CSF protein a common finding. The first-line therapy is systemic corticosteroid. There are no recognized and well established diagnosis criteria but the diagnosis is highly suggestive in presence of anti-TPO antibodies in serum associated with the improvement of neurological state by corticosteroid therapy after excluding other causes.

Conclusion

A patient suffering from Hashimoto's thyroiditis with atypical status epilepticus could be suggestive of an autoimmune encephalopathy and the corticosteroid therapy should be started immediately.

P 125.

A recurrence of Giant Bilateral Inguinal Hernias in an Infant with Pallister-Killian Syndrome (PKS).

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Background

Pallister-Killian is a rare genetic condition caused by mosaic tetrasomy of the short arm of chromosome 12(12p), due to the presence of an isochromosome 12p (consisting of two 12p arms). It is a multisystem disorder characterized with significant hypotonia, intellectual disability, characteristic facial features, sparse hair and multiple congenital malformations.

Methods

We report a case of a male patient of a G8P7Ab1 42- year- old mother, who was diagnosed with PKS in the first month of life. He was born at 39 6/7 weeks gestation (birth weight:3860gr) to consanguineous Moroccan parents. He was noted antenatally to have an unilateral ventriculomegaly and polyhydramnios. He was born with a significant hypotonia and multiple congenital anomalies including a coarse face with a high forehead with temporo-frontal balding, sparse eyebrows and lashes, a broad nasal bridge, hypertelorism, shallow supraorbital ridges, upslanting palpebral fissures, low-set ears, a wide mouth with a thin upper lip, macroglossia and a high-arched palate. Feeding difficulties were present: GERD, swallow difficulties, constipation, and micro-and macro-aspirations. Echocardiography revealed a small ASD type 2. At the age of 2 months he developed massive bilateral inguinal hernias. Surgical reconstruction: right herniotomy and left orchidopexia. At the age of 11 months there was a bilateral recurrence of the inguinal hernias. Laparoscopic reconstruction was done. Afterwards, a non-contrast magnetic resonance imaging showed a striking cortical and subcortical cerebral atrophy, corpus callosum atrophy, subarachnoidal cyst formation and a thin aspect of the optic nerve bilaterally up to the chiasm.

Results

The SNF array showed a pathogenic multiplication of the entire arm of chromosome 12p. Additional FISH analysis confirmed the presence of a mosaic isochromosome 12p.

Conclusion

Pallister Killian Syndrome (PKS) (OMIM#601803) is a rare chromosomal aneuploidy. At the moment, the mechanism by which the isochromosome 12p arises remains largely unclear but the primary error is suggested to be prezygotic due to a meiosis II nondysjunction and often of maternal origin (in the majority of cases the maternal age is >35 years). Hernias are frequently seen in this syndrome. Congenital diaphragmatic hernia was present at birth in about 40% of the reported cases. Umbilical hernias are also frequent. Blyth et al. reported inguinal hernias later in life in 12% of the reported cases. To our knowledge this is the first case of PKS with recurrence of giant inguinal hernias. A laparoscopic approach for the recurrent hernia is the best therapeutic choice in this anatomical challenging syndrome.

Posters**P 126.****A case of GLUT1 deficiency syndrome – benefits of genetic testing in childhood epilepsy**

S. Neuens, A. Van Hecke, J. Soblet, C. Vilain

HUDERF-ULB

Epilepsy is one of the most common neurologic conditions in children. Genetic testing has gained importance in the workup of childhood epilepsy. This case illustrates the role of genetic testing in the diagnosis of childhood epilepsy and how it can effect therapeutic decision-making.

A 20 month old boy was referred to our institution for recurring status epilepticus. The first seizure occurred at the age of 18 months and was characterized by tonico-clonic movements, eye revulsion, hypersialorrhea and loss of consciousness for 30 minutes. Seizures resolved spontaneously. Clinical examination was normal. Initial EEG and cranial MRI were normal. Treatment with Valproic acid at 20µg/kg was started following this episode.

At the age of 21 months he presented a limping gait without prior trauma. Hip MRI was normal, and physiotherapy didn't alleviate his symptoms.

At the age of 2 and a half years he presented a second similar episode of convulsions lasting for over an hour. Seizure control was achieved only after administration of continuous IV midazolam. Daily Valproic acid doses were increased to 30µg/kg. Following this episode a worsening of his gait disturbances was observed, with the apparition of an ataxic gait. At this moment neurologic examination showed diminished right patellar reflexes. Cranial and spinal MRI, as well as long-term EEG were normal. His mother described less severe gait disturbances when valproic acid doses were omitted, leading to its replacement by Levetiracetam.

In trio clinical exome sequencing revealed a de novo R51P variant in the SLC2A1 gene, encoding Glucose transporter 1 (GLUT1). The similar R51H variant has been previously described in a child with dystonic movement disorders. Glycorachia was confirmed to be low at 38.3 mg/dL. Following this discovery, anti-epileptic treatment was stopped and a ketogenic diet was started.

SLC2A1 mutations are associated with GLUT1 deficiency syndrome, characterized by a wide clinical spectrum encompassing seizures, movement disorders, and cognitive impairment due to decreased glucose concentration in the central nervous system. Worsening of the child's gait disturbances under valproic acid therapy might be explained by its effect on GLUT1 transporter activity.

In this case genetic testing provided a diagnosis, valuable information on disease prognosis, and directly influenced therapeutic decision-making, leading to the discontinuation of seizure medications and instauration of ketogenic diet.

P 127.

Gut and liver dysfunction: a surprising diagnostic clue for metabolic disease

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UZ Gent

Introduction

Elevated transaminases are frequently found in children, and are usually benign. Nevertheless, if abnormalities persist or if clinical signs of underlying liver disease are present, further investigation is warranted.

Case description

A Caucasian boy was referred to the department of pediatric gastroenterology at the age of 15 months because of persistently elevated transaminase levels. Clinical history revealed intermittent episodes of severe abdominal cramps, diarrhea and hypoglycemia requiring hospital admission. On clinical examination, hepatosplenomegaly was noted. Otherwise the boy had a good general appearance without dysmorphism. Blood test showed moderately elevated liver enzymes (AST 251 U/L, ALT 428 U/L, GGT 41 U/L, LDH 512 U/L), elevated lactate (41.5 mg/dL) and decreased coagulation factors. Ultrasound showed mild hepatosplenomegaly, and strikingly diffuse enlarged hyperechoic kidneys. Because of a strong suspicion of an underlying metabolic disorder, blood and urinary metabolic tests were initiated. Transferrin isoelectric focusing (IEF) showed a type I pattern, which is associated with congenital disorders of glycosylation (CDG). Mutational screening revealed compound heterozygous missense variants in the mannosephosphate isomerase (MPI) gene within the RmIC-like cupin domain, confirming the diagnosis of MPI-CDG, previously known as CDG Ib. Mannose treatment was started at 21 months of age. After one month, this led to a clinical improvement, normalization of the biochemical abnormalities, coagulation factors and an almost normalization of the transferrin IEF.

Discussion

MPI-CDG is a very rare disorder, caused by a mutation in the MPI gene on chromosome 15q24. It leads to a reduced conversion of fructose-6-phosphate to mannose-6-phosphate, which serves as a crucial substrate for proper N-glycosylation. The predominant symptoms are chronic diarrhea with failure to thrive, protein-losing enteropathy, coagulopathy and hepatic dysfunction. MPI-CDG is distinct from other CDGs by the lack of neurologic symptoms and can be treated effectively with oral mannose supplementation. If untreated, it can be fatal. Only twenty-five patients have been described worldwide and the infrequency with which it is encountered makes MPI-CDG a diagnostic challenge.

Conclusion

MPI-CDG should be considered in the differential diagnosis of patients with unexplained chronic diarrhea, liver disease and coagulopathy to allow early diagnosis and effect

Short Oral Presentation**SO 2.****Recovery kinetics of gas exchange parameters and heart rate after maximal exercise in children with repaired Tetralogy of Fallot compared to controls**

I. Coomans, S. De Kinder, H. Van Belleghem, D. De Wolf, K. De Groote, J. Panzer, L. Muino Mosquera, H. De Wilde, T. Bové, K. François, J. Boone, K. Vandekerckhove

UZ Gent

Background

It has been well demonstrated that children with repaired Tetralogy of Fallot (rToF) have a diminished exercise tolerance. The recovery phase after maximal exercise has been less investigated. In this study we evaluate the recovery kinetics of children with rToF in comparison to healthy peers.

Methods

A group of 46 children with rToF was compared to a control group of 46 healthy children, matched for gender, age, weight and length. 20 patients had received a pulmonary valve replacement (PVR) over time. All participants performed a maximal incremental cardio-pulmonary exercise test (CPET). Oxygen uptake (VO₂), carbon dioxide output (VCO₂) and heart rate (HR) were measured during 6 min of recovery after CPET. Changes were calculated per minute and expressed as percentage drop from maximum value. Data was fitted to a mono-exponential model providing half-life time T_{1/2} to describe the decline.

Results

Exercise performance was reduced in the rToF group (VO_{2peak}: 35,33 vs. 43,64 ml/min/kg; p<0,001 and maximal load: 110,5 vs. 136,5 W; p<0,001). Maximal HR (171 vs 193 bpm; p<0,001) was lower in the rToF patients, due to chronotropic incompetence. A slower decrease of VO₂ and VCO₂ was observed in the rToF group at each minute of the recovery phase (all p<0,05). This was confirmed by a larger T_{1/2} in the VO₂ (50,97 vs. 44,55 s; p<0.05) and VCO₂ (66,38 vs 61,37 s; p<0.05) recovery kinetics in children with rToF. HR kinetics during recovery using both methods showed no difference between the two groups. No difference in recovery kinetics was found in the PVR group compared to the non PVR group. There was no correlation between the recovery parameters and the degree of RV dilatation on echocardiography.

Conclusion

Besides a lower exercise tolerance, rToF patients have prolonged recovery of VO₂ and VCO₂. This points towards an impaired oxygen debt repayment after exercise, and appears to be independent of RV dilatation or PVR. Since there is an equal recovery of HR after maximal exercise, inadequate stroke volume adaptation might explain the slower VO₂ recovery. More research is needed to further elucidate the underlying mechanism of recovery kinetics in rToF patients.

SO 7.

The long term fate of subaortic stenosis in childhood

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VUB, UZ Gent

Introduction

Subaortic stenosis (SAS) is a common cause of left ventricular outflow tract (LVOT) obstruction in children. It can be divided into a discrete form consisting of a circular membrane or fibromuscular ridge and a long and narrow tunnel-type. The ideal timing of intervention remains controversial. While immediate surgical results are satisfying, reoperation rate amounts up to 20 % and surgery does not seem to protect the aortic valve.

Methods

We conducted a retrospective study of 84 children (0-18y) diagnosed with SAS between 1992 and 2017. General characteristics, hemodynamic and surgical data were reviewed.

Results

Seventy-one patients underwent surgery. Median age at diagnosis was 2.3 (IQR 5.3) years and peak gradient $31.8 (\pm 24.6)$ mmHg. Discrete SAS was present in 89 % (63/71) and tunnel-shaped SAS in 11 % (8/71). Median age was 3.4 (IQR 7.3) years at intervention and peak gradient was 53.0 ± 31.3 mmHg. Reoperation rate was 20 % in case of discrete SAS and even 75% in patients with tunnel-shaped SAS ($p = 0.003$). Reoperation was associated with higher residual peak gradient immediately after surgery ($p = 0.003$), irrespective of the type of surgery (resection +/- myectomy) or any other investigated potentially predictive factor. Aortic insufficiency (AI) was present in 41 % of the patients before intervention and in 37% after. Prevalence of AI at latest follow-up was 61%. Thirteen patients had only conservative follow-up. Their median age at diagnosis was 3.8 (IQR 8.0) years and peak gradient at diagnosis was 11.6 ± 12.3 mmHg. Median follow-up time was 9.3 (IQR 6.2) years with a peak gradient of 18.5 ± 12.5 mmHg at latest follow-up. Peak gradients were significantly lower ($p = 0.001$) and median age was significantly higher at diagnosis in this group ($p = 0.04$). AI was present in 15% of patients at diagnosis and in 54% at latest follow-up.

Conclusion

SAS remains a complex disease. Reoperation rate remains high, especially in tunnel-shaped SAS and depends on the early postoperative gradient not on the type of operation. Low initial gradient and late diagnosis are predictors for a milder disease that doesn't require intervention during childhood. AI is frequently associated and is often progressive, despite surgery.

Short Oral Presentation**SO 8.****Oxygen uptake kinetics and local muscle oxygenation during submaximal exercise in children after the Fontan procedure compared to healthy peers**

Y. Bagdasarian, I. Coomans, D. De Wolf, K. De Groote, J. Panzer, L. Muino, H. De Wilde, T. Bové, K. François, J. Boone, K. Vandekerckhove

UZ Gent

Aims

Oxygen consumption and muscle oxygenation during submaximal exercise in children with univentricular heart (UVH) has been poorly investigated. This study compares the oxygen uptake kinetics and the local tissue oxygenation after the Fontan procedure compared to healthy peers during a submaximal constant load test (CLT).

Methods

18 UVH and 16 healthy control children performed a CLT and a maximal incremental ramp exercise test (CPET). 13 patients had dominant RV, 5 patients dominant LV. CLT was performed at 30% of the maximal exercise capacity during 6 minutes using cycle ergometry. Near infrared spectroscopy (NIRS) was used at m. Vastus Lateralis to measure local tissue oxygenation index (TOI) in both tests. The oxygen uptake kinetics (VO₂) was measured during CLT and tau (time to double VO₂) was calculated in both groups. Correlations between VO₂ and TOI and other parameters were investigated.

Results

UVH patients had lower VO₂peak (29±8 vs. 46±12 ml/min/kg, P <0.01), peakload (72±19 vs. 133±67 W, P<0.001) and maximal heart rate (HR) (168±13 vs. 193±12 bpm, P<0.001) compared with the controls. In CLT, HR was higher from 2 mins onwards in UVH patients. Higher tau-value and thus slower VO₂-kinetics was found in the patient group (31.3±6.3 vs. 25.1±5.7, P<0.01). The TOI was lower at the onset of exercise (59.9±4.26 vs. 67.6±5.5s, P< 0.001) in UVH. The TOI decreased steeply in both groups immediately after start of the exercise, but even more steep in the patient group. TOI remained lower in UVH children. Slower VO₂ kinetics in CLT was correlated with lower VO₂peak, lower rest TOI and lower TOI correlated with lower VO₂peak.

Conclusion

During submaximal exercise at 30% of VO₂peak, slower VO₂-kinetics, higher HR-response and different local TOI-patterns are determined. They correlated with a decreased maximal exercise performance in UVH children. This reflects a mismatch between O₂-delivery to O₂-demand of the working muscle from the onset of a low-level exercise. A good correlation was found between VO₂peak and TOI and between VO₂peak and VO₂-kinetics of CLT, this means that the constant load test is useful whenever patients are not able to perform maximal CPET or for inbetween evaluation.

SO 29.

Endocarditis prophylaxis in the "real life" of the general pediatrician and/or dentist

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Endocarditis is a potentially life-threatening disease in children with CHD and correct prophylaxis (EP) is of utmost importance. EP guidelines are well known by pediatric cardiologists. However, the knowledge and correct application of these guidelines by other stakeholders of children's health is less certain.

We conducted a survey among pediatricians and dentists in Flanders, Belgium. The survey consisted of a part assessing the knowledge about EP guidelines and included a few test cases. The survey was completed by 910 dentists (16.2% response rate for 5604 dentists), 100 pediatricians (17.5%) and 16 congenital cardiologists (59%).

65% of the dentists did not know any guideline. The majority of dentists would look for information on the internet or from the child's physician. In the pediatrician's group 47% did not know any of the guidelines and the majority of pediatricians would contact the child's pediatric cardiologist.

In the dentist group we focused primarily on the knowledge of high and low risk treatments and the identification of patients at risk. 87% identified correctly low risk treatments, but only 64% identified correctly all high risk procedures. Of the dentists knowing EP guidelines, 83% asked for the presence of CHD and allergy to antibiotics. Pediatricians correctly defined most defects at high risk, but scored lower for the correct identification of lower risk CHD. Most pediatricians were able to identify high risk procedures, but failed to identify many procedures correctly as low risk. Pediatricians were well aware of dental hygiene measures.

The practical cases were different for dentists and pediatricians. Dentists asked more advice of the patient's physician, but would wrongly withhold treatments in high risk patients (29%). Pediatricians had difficulties with the identification of low risk procedures and the correct classification of valvar diseases.

Conclusion

The knowledge of Flemish dentists and pediatricians of the EP guidelines is low. Too many children receive unnecessary antibiotics, and some children are unduly deprived of necessary dental procedures. EP guidelines should not only be spread by the cardiology scientific publications, but also by the dentistry and pediatric scientific societies and journals in order to improve their knowledge of the EP guidelines.

Poster Walk**PW 16.****Melody valve in mitral position in very young children with atrio-ventricular septum defect and severe mitral valve dysfunction**

J. Hubrechts, B. Cools, R. Heying, B. Eyskens, R. Rega, B. Meyns, M. Gewillig

UZ Leuven

Introduction

The availability of prosthetic valves in infants with atrioventricular valve annulus <15mm is very limited. Off-label use for surgical implantation of a folded or trimmed Melody valve is an option for mitral valve dysfunction. Moreover, the Melody is expandable by percutaneous catheter balloon dilatation.

Technique

The Melody is implanted via left atriotomy. The native valve is excised; the annulus measured with Hegar stifts. The stent is folded at both extremities to reduce final length. A pericardial strip is attached in the middle or at the edge of the stent as suture ring; the valve is shrunk manually and the band is sutured in the mitral annulus; stay suture in LV if ventricular protrusion. Then the valve is dilated with a balloon. Finally the stent is fixed on the interatrial septum. During growth balloon dilatation was performed through the interatrial septum; stable position of guide wire was assured by apical snare from retrograde transaortic access.

Patients, results

We describe 3 similar AVSD patients, in which the implantation of a Melody valve was life-saving as they developed all severe mitral dysfunction (stenosis MS or regurgitation MR) after prior AVSD surgery with valvuloplasty. None of them had known genetic syndromes or major comorbidities.

P1, 16 m old, 9.5kg, MR, pericardial strip in the middle of the valve, inner size expansion 16mm, complicated with 3th degree AV-block and pacemaker implantation. Balloon dilatation at 17 m to 20mm, follow-up (FU) until 66 m of age.

P2, 5 m old, 6.3kg, MR, pericardial strip in the middle of the valve, inner size expansion 10mm, no complication. Balloon dilatation at 10 m to 14mm, FU until 30 m of age.

P3, 4 m old, 4.1kg, MS, pericardial strip at the ventricular end of the valve, inner size expansion 14mm, complicated with 3th degree AV-block and pacemaker implantation. No balloon dilatation, FU until 6 m of age.

Our results show 100% of mechanical success, no mortality nor early valve replacement; no endocarditis, LVOT obstruction or inflow obstruction. At last check-up, none of them had MR, MS mean gradients 7, 12 and 5mmHg, without retrograde PHT.

Conclusion

Melody valve in mitral position in infants functions well down to 10mm of mitral annulus. LVOT obstruction can be avoided by positioning the stent completely in the atrium, without obstruction of pulmonary venous flow. Expansion up to 14-16mm in small infants is associated with higher incidence of AVB.

PW 17.

Direct paratracheal lymphosclerosis for plastic bronchitis after Fontan: percutaneous versus endoscopic transtracheal technique

J. Hubrechts, B. Eyskens, C. Dooms, G. Maleux, B. Cools, R. Heying, D. Boshoff, M. Gewillig

UZ Leuven

Background

Plastic bronchitis (PB) after Fontan palliation results from abnormal mediastinal lacteals leaking into the bronchial tree. Itkin et al. showed the possibility to embolize these abnormal lymphatic vessels via cannulation of the ductus thoracicus, which is a complex and demanding technique, especially when absent/hypoplastic/stenotic. We report on a new technique comparing two possible accesses: percutaneous and endoscopic .

Objective

To assess efficacy of embolizing abnormal mediastinal lymphatic vessels and leaks in Fontan patients with PB by direct access to the pathological region, shortcutting the ductus thoracicus. Experience in 2 patients.

Technique and results

Mediastinal lymphatic anatomy and function was investigated by inguinal intranodal gadolinium based MR lymphangiography.

In patient 1, paratracheal dilated lymph vessels were punctured directly percutaneously with a 22G Chiba needle through the intercostal space under fluoroscopic and cone-beam computerized tomographical (XperGuide®, Philips) and ultrasound guidance. Good intralymphatic position was ascertained by injecting water soluble contrast with drainage to abnormal lacteals; after flushing with glucose 5%, occlusion was obtained by injection of 5-10 cc of a mixture of lipiodol/n-BCA N-butyl cyanoacrylate (Histoacryl®) 4/1 under fluoroscopic guidance (DAB 25.34 Gy/cm²). There was a total remission of PB, now 15 months of follow-up.

In patient 2, all 4 abnormal hypertrophic mediastinal lymph nodes (paratracheal, but also bronchial and subcarinal) were punctured with a 22G ViziShot EBUS-TBNA needle under direct guidance and visualization of an EBUS EndoBronchial UltraSound-guided bronchoscope. After flushing with 0.5 ml glucose 5%, occlusion was obtained by injecting 10 cc of a mixture of lipiodol/n-BCA 5/1 under fluoroscopic control (DAB 15.2 Gy/cm²). There was a total remission of PB, now 2 months of follow-up.

Conclusion

Direct paratracheal lymphocclusion is effective and obviates the need for transductal access. When comparing both direct puncture techniques, we find the trans-tracheal EBUS puncture more elegant having easier and safer access, and a more complete occlusion with less radiation than the percutaneous puncture.

Poster Walk**PW 37.****Outcome of strategy in Pulmonary Atresia, ventricular Septal Defect and Major Aortopulmonary Collateral Arteries (PA, VSD, MAPCA's)**

B. Eyskens, B. Cools, R. Heying, J. Hubrechts, D. Boshoff, F. Rega, B. Meyns, M. Gewillig

*UZ Leuven*Background

To improve counseling for patients with PA, VSD, MAPCA's we evaluated the outcome in a consecutive patient population over the last 20 years. Treatment options are combinations of reconstruction of the pulmonary vascular bed by unifocalising the collateral vessels, rehabilitation of native pulmonary arteries (PA) and ultimate "repair" with closure of the VSD and valvulation of the right ventricular outflowtract (RVOT).

Methods

Retrospective review of 33 consecutive patients born between 1/ 2000 and 6/2019, directed to therapeutic strategy with the aim of final surgical repair.

Results

11/33 patients were diagnosed prenatally at 24 (20-32) weeks GA, 22 patients postnatally a mean age of 2,1 (0,1-28) months. Mean birth weight was 3045 (2088-4120 g). 8 patients were diagnosed to have 22q11 del. Anatomical features: hypoplastic central PA n = 28, absence of central PA n = 3, hemitruncus arteriosus n = 1, left arterial duct n = 1. Primary one-stage repair with unifocalisation of MAPCA's could be performed in 3 patients (mean age 6m, range 0,5-13,8m). 18 Patients could be successfully repaired (mean age of 44,5mths, 8,3-108,4 mths) after a staged approach of surgical and/or percutaneous interventions: aortopulmonary (Ao-Pu) shunt n =8 , RVOT-PA patch/conduit n =9 , unifocalisation of MAPCA's n =3 , balloondilatation and or stenting PA or MAPCA n = 15. In total 21/33 (64%) patients were successfully repaired. After a mean follow-up of 6,1 yrs, mean age 10,2 yrs, they have a good functional condition NYHA 1-2 and acceptable right ventricular systolic pressures of 65mmHg (32-90 mmHg). 3 Patients needed surgical reintervention after full repair: pulmonary valve replacement n =2, tricuspid valve plasty n = 1. 8/33 (24%) Patients died: 2 because of severe hypoxia before any intervention (day 2 and day 85), 4 after palliative intervention (RVOT patch, 2 Ao-Pu shunt, unifocalisation), 1 after staged repair. 4 Patients remain palliated (3 Sano shunt, 1 RVOT Contegra conduit).

Conclusion

Although PA/VSD/MAPCA's is considered as a disease with poor prognosis, 64 % of our patient population could achieve a successful repair, even with very small native pulmonary arteries. A negative prognostic factor is severe cyanosis at birth.

P 129.

Unilateral lung whiteout during neonatal period, about a case

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Introduction

Respiratory distress is a common occurrence during the neonatal period. Prompt investigation to identify the underlying cause and appropriate management is essential to improve outcomes. Many causes of respiratory distress in a new born are unique to this age group. Amongst them there are respiratory causes, but there are also various non-respiratory aetiologies such as heart failure due to congenital heart disease

Methods

We report the case of a girl born at 37 weeks of gestation. Pregnancy was normal, and the new born left the maternity with a normal oxygen saturation of 95%. At 14 days of life, she returned to our emergency room in respiratory distress that appeared one hour beforehand, with no fever. Initially, the girl was pale, with tachypnoea, tachycardia, was poorly perfused, presented major respiratory distress, and had a hepatomegaly of 5 cm. Initial oxygen saturation was measured at 75%, oxygen was administrated by nasal cannula and CPAP. As oxygen saturation was not improving, she was intubated. She also received vascular fillings, inotrope support by adrenalin, and prostaglandin to reopen her ductus arteriosus. Large spectrum antibiotics were also administered. Chest X-ray showed a right lung whiteout and blood tests were normal. Cardiac ultrasound showed dilatation of the right chambers and a supradiaphragmatic obstructed total anomalous pulmonary venous connection (TAPVC). The following day, surgical intervention indicated that there was a mixed TAPVC with the right pulmonary veins converging into an infra-diaphragmatic confluence which was obstructed, and the left pulmonary veins joined to form a non-obstructed separate venous connection

Conclusion

TAPVC is a cyanotic congenital heart disease in which the pulmonary veins fail to connect to the left atrium and drain into the systemic venous circulation. There are four anatomic variants: supracardiac, infracardiac, cardiac, and mixed. Those with obstructed TAPVC often present signs of shock, cyanosis or respiratory distress. Diagnosis is made by cardiac ultrasound, but in obstructed TAPVC chest X-ray can also show an enlargement of the cardiac silhouette, pulmonary vein congestion, or pulmonary oedema, which can be asymmetric, as found in our patient. In case of obstructed forms, treatment is based first on stabilization of the patient by administration of oxygen, ventilation, inotropic drugs and in some cases prostaglandin therapy. Urgent surgical repair is recommended

Posters**P 130.****Kingella Kingae endocarditis with vegetation and mitral valve perforation in a 7 month old infant.**

S. Verbeeck, S. Van Nuijs, F. Slap, B. Suys

*GZA Sint Augustinus*Background

This is a case of a 7 month old infant presenting with an upper airway infection, 3 days of high fever and diarrhoea. Laboratory studies showed a leucocytosis (23.500/mm³) and elevated CRP (353mg/L). Urine and stool samples were negative but lumbar puncture showed 846 WBC/ μ l. Hemoculture was obtained and antibiotics were started.

Evolution

At the moment hemoculture showed growth of *Kingella Kingae*, we planned a bone scintigraphy and echocardiography.

Echocardiography showed a fluttering vegetation of 10mm on the mitral valve and a mitral insufficiency. Cardiac surgery was performed to remove the vegetation and to repair the underlying perforation of the mitral valve.

Head CT was performed to exclude septic emboli (because of leucocytosis on lumbar puncture) and the patient was treated with IV antibiotics for a 4 week period following the surgery.

Conclusion

This is a valuable case for a poster presentation because of its interesting images and the important message to always remember the possibility of endocarditis when obtaining a positive hemoculture.

P 131.

How to detect incomplete forms of Kawasaki disease?

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Kawasaki disease is a systemic inflammatory disease affecting the medium-size arteries. It affects 4 to 25 in 100,000 children in Europe. The diagnosis is based on clinical criteria and blood tests but can be delayed due to the existence of incomplete forms and non-specific clinical signs especially in infants. Indeed, irritability and persistent fever can be the only symptoms of the disease. Incomplete forms are associated with an increased incidence of coronary aneurysms due to delayed diagnosis. An algorithm for incomplete forms has been created by the American Heart Association to help diagnosis and provide prompt management.

Here, we report the case of a 5-month-old boy suffering from an incomplete form of Kawasaki disease. He was admitted in the emergency unit with fever, intestinal symptoms and rhinitis. Biological tests showed a neutrophilia, thrombocytosis, high inflammatory biomarkers and slightly elevated troponin I levels. No specific infectious etiology was found. Repeated COVID-19 antigenic test was negative. Due to the persistence of clinical symptoms with high inflammatory biology pattern of unknown origin, we performed an echocardiography leading to the diagnosis of Kawasaki disease. It revealed multiple medium-sized coronary artery dilatations. The maximum diameter found was 6 mm on the right coronary- and 7,7 mm on the left coronary artery. Treatment with intravenous immunoglobulins and high-dose aspirin was started immediately. The patient responded very well to the treatment. The 9 months follow-up shows stabilization of the aneurysms without any sign of coronary insufficiency at rest.

In conclusion, Kawasaki disease with coronary artery aneurysms might occur without specific symptoms, which delays diagnosis. Therefore, diagnosis should always be considered in face of prolonged hyperthermia of unknown origin with compatible blood test pattern and echocardiography should be done to exclude the presence of coronary artery dilation. Prompt recognition of this disease and adequate treatment prevents the development of coronary aneurysms with thrombi deposition and myocardial ischemia.

P 132.**Long QT interval in patients with Turner syndrome**

J. Dauby, A-S Crochelet, M-C Lebrethon, J. Harvengt, S. Bulk, M-C Seghaye

*U Liège*Introduction

Turner syndrome (TS) is well known to be associated with cardiovascular malformations. In addition, current knowledge points out the presence of electrophysiological anomalies associated with malign cardiac arrhythmia, in particular, the presence of long QT. We report on the case of 2 patients with TS in whom prolonged QT was diagnosed.

Cases presentation

A 16-y-old teenager with a TS (XO mosaicism) was diagnosed at 15 because of primary amenorrhea and growth retardation. Since diagnosis, the girl was treated with growth hormone and substitute estrogen. During a follow-up consultation, the resting ECG revealed for the first time a prolonged corrected QT interval (Bazett) of 460 msec. The stress ECG showed a fast increase of the heart rate without repolarization or rhythm disorders during or after physical stress and also confirmed the prolongation of the QT interval 4 minutes after stopping the effort (Schwartz criteria). A 24-h-Holter record was realized and showed several ventricular extra-systoles that disappear during physical stress. In this context, a β -blocker therapy was introduced. The patient is currently stable.

A 26-y-old girl, diagnosed at birth with TS (XO), has been treated with growth hormone until the age of 15. She was also treated for hypothyroidism. The cardiac check-up showed a sinus tachycardia, an intermittent prolonged QT interval of 450 msec and a dilatation of aortic sinus of Valsalva. Therefore, a β -blocker therapy was introduced at 18 to prevent both arrhythmia due to QT interval prolongation and an increase of the aortic bulb dilatation. Since then, the patient has regular cardiac follow-up that reveal a favorable evolution under β -blocker treatment.

Discussion & Conclusion

Prolonged QT interval is more frequent in Turner patients than in the general population and explains a higher rate of cardiac death independently of the presence of a structural malformation in the former patients. The cardiovascular screening of Turner patients should include repeated standard ECG, stress ECG with QT measurement 4 minutes after the end of stress and Holter records. Genetic counseling should also be considered with respect to the identification of long QT-related ion channel mutations and prophylactic treatment with β -blocker to avoid ventricular arrhythmias and sudden cardiac death introduced as soon as possible.

P 133.

Convulsive seizures in LQTS : think about malignant tachyarrhythmias

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U Liège

Introduction

Congenital long QT syndrome (LQTS) is an inherited condition characterized by QT segment prolongation at standard ECG due to a genetic defect of an ionic channel. Various genetic mutations are known to be responsible for this syndrome. Clinical features can include generalized seizures that are secondary to malignant ventricular tachyarrhythmias, sometimes misinterpreted as epilepsy. Genetic analysis is of central importance for diagnosis and treatment.

Case report

We report the case of a young 13-year-old girl who was admitted in the emergency room for convulsive seizure occurring at rest. This patient is known since birth to have a LQTS associated with a congenital bilateral deafness (Jervell and Lange-Nielsen syndrome). The QTc varies between 450 and 500 msec. The genetic work-up showed mutations on the KCNQ1 and SCN5A genes. Her mother, one sister and one step-brother carry a LQTS mutation without deafness. The patient is treated by beta-blockers since birth but showed several generalized seizures leading to the suspicion of associated epilepsy. However, pathologic EEG has never been recorded. Conversely, malignant tachyarrhythmias have never been documented. At time of the last seizure, treatment consisted of nadolol 1,25mg/kg/day. In order to ensure the diagnosis of tachyarrhythmia, a Medtronic Reveal LINQ Recorder has been implanted. Nadolol dosis has been adapted to weight (1,7mg/kg/j). Since Reveal recorder implantation four months ago, no adverse event has been reported.

Discussion

This case shows a classical presentation of LQTS associated to central deafness in the context of a Jervell and Lange-Nielsen syndrome. The seizures are more likely to be due to ventricular tachyarrhythmia causing low cardiac output than to epilepsy. Indeed, our patient carries two risk factors for arrhythmogenic events: duration of QTc >500 msec. and the presence of two genetic ion channel mutations. Beta-blocker therapy, in particular nadolol offers a satisfactory but not complete prevention against ventricular tachyarrhythmia. For that reason, the indication for internal defibrillator implantation or in specific cases, a left sympathetic denervation must be considered and based on the records of the Reveal device.

Conclusion

The congenital long QT syndrome is a rare condition that may lead to malignant ventricular tachyarrhythmia. This latter manifest by cerebral seizures often misinterpreted as epilepsy.

P 134.**When respiratory symptoms hide a double aortic arch**

S. Finocchiaro, N. Farhat, M-C. Seghaye

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Anomalies of the aortic arches represent 1% of congenital heart diseases. Double aortic arch accounts for 30%-50% of cases. We report the case of a patient in whom the abnormality, despite causing her significant respiratory symptoms, was diagnosed as late as in her 10th year.

Case presentation

The child is referred to a pulmonologist at the age of 4 months because of post-meal choking episodes and stridor. Tracheomalacia was diagnosed and the presence of an arteria lusoria was suspected in a swallowing study and at angio-CT. The child showed signs of severe gastroesophageal reflux disease and received a Nissen fundoplication at the age of one year which only very slightly relieved the symptoms.

During her childhood, the girl suffered from repeated respiratory infections. She experienced more than 5 laryngo-tracheitis per year and at least as many wheezing bronchitis. The patient presented a permanent asthmatic symptomatology that did not respond to treatment. However, these symptoms decreased over time.

At the age of 10, during a cardiac assessment for lipothymic discomfort, a new CT angiography was performed to precise the aortic arch anatomy. The latter revealed a complete co-dominant double aortic arch, responsible for a tracheal compression of 50% of the lower part of the trachea. The child underwent surgical treatment consisting of a section of the ligamentum arteriosum, of the distal left aortic arch with dorsal pexy of the descending aorta and pexy of the left subclavian artery at the lateral thoracic wall. There were no post-operative complications.

Discussion

Double aortic arch is often symptomatic already in young infants. The most frequently reported symptoms are those of tracheal compression including recurrent bronchopulmonary infections, stridor, respiratory distress, apnea and choke. These signs tend to improve over time, surely due to the development of the trachea which becomes firmer. Signs of esophageal compression such as dysphagia or regurgitations may falsely be interpreted as gastroesophageal reflux.

Conclusion

The diagnosis of a double aortic arch, and, in a broader sense of an anomaly of the aortic arch, should be suspected in case of neonatal respiratory distress without obvious etiology, persistent stridor in an infant, severe asthma resistant to treatment or recurrent respiratory infections associated or not with a gastroesophageal reflux, dysphagia or choking episodes related to solid food ingestion.

P 135.

Hypertrophic cardiomyopathy : A Breathtaking disease

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Background

Hypertrophic cardiomyopathy (HCM) is the most common genetic heart disorder and accounts for 40% of all five cardiomyopathies in pediatrics. With a rate of five per million, HCM is more likely diagnosed in late adolescence but can occur in any child regardless of age. Most are asymptomatic, but some develop symptoms such as fatigue or dyspnea, especially when an effort is required. We report the case of an adolescent with severe HCM, whose symptoms were attenuated due to her physical activities restriction during the COVID-19 lockdown.

Case presentation

A 14-year-old girl presented with a history of nocturnal dyspnea eased by emesis and associated with cold extremities. Before the COVID-19 lockdown, as a sportsperson, she noticed unusual fatigue and shortness of breath while training, and then also a gradual onset while walking. We highlighted an orthopnea as she reports sleeping with two pillows. One year ago, she was diagnosed with asthma that did not respond to treatment. Physical examination showed a systolic murmur and a thready pulse. We explored her condition with an electrocardiogram that showed global hypertrophic signs. Echocardiography revealed overall hypertrophic cardiomyopathy with a ventricular septum thickening of 30 mm (Z score: 6,75) and an impaired diastolic function. A magnetic resonance imaging defined the thickness and revealed areas with fibrosis. During her hospitalization, she received beta-blockers as supportive treatment to reduce the workload of her heart. An internal defibrillator was also implanted to prevent sudden death due to malignant arrhythmias. A discussion will soon take place with her parents regarding a heart transplant.

Discussion

People with HCM may present non-specific symptoms such as dyspnea, which can be more apparent in sportspeople. When they cease physical activities, it can mask their symptoms and prevent them from seeking medical advice, but paradoxically it can prevent them from having severe consequences as malignant arrhythmias. In the end, we are facing patients whose symptoms are being minimized while the disease is progressing with an increase in the probability of sudden death.

Conclusion

HCM is a rare disorder in pediatrics and has an uncertain evolution from one child to another. This disease includes stages, ranging from the asymptomatic patient to the very symptomatic one with an irreversible form that will require heavier supportive treatment and sometimes heart transplantation.

P 136. E

Establishment of a predictive model of VO₂ in patients with total cavo-pulmonary anastomosis (FONTAN)

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Background

The Fontan procedure, the last of a series of cardiac surgeries in patients with single ventricle physiology, offers improved exercise capacity, although reduced when compared to a healthy population. We hypothesized that we can establish a predictive model of late cardiovascular performances based on the pulmonary artery (PA) diameters.

Methods

We studied 175 patients with single ventricle physiology followed between 1960-2020. Descriptive analysis from the cardiac catheterizations and exercise testing was performed to evaluate the potential of a stepwise regression analysis to predict VO₂.

Results

148 (84%) patients underwent a Glenn surgery and 139 (79,4%) a Fontan palliation. Pre-Glenn PA diameters were 8,4±4mm (RPA) and 8,6±3,9mm (RPA) (NAKATA index 271± 100mm²/m², age 3,9 +/- 0,5 years old); pre-Fontan PA diameters were 9,4±2,2mm and 10,2±4,6mm for RPA and LPA respectively (NAKATA index 209±41,9mm²/m², age 3,9+/-0,9 years old). 83 patients (47,3%, age 20,14+/-17 years old) performed an exercise test with a mean peak VO₂ of 24,7±12,1 ml/kg/min (percentage of predictive value 59,7±26,5%). The stepwise regression predicted the VO₂ as Constant + Coefficient x PAm, a statistically significant correlation with LPA pre-Glenn (p 0,0355) and RPA pre-Fontan (p 0,0429).

Conclusion

Our model based on the preoperative PA diameters allowed to predict VO₂ in patients with single ventricle palliation.

LO 11.**Dietary fiber intake and gut-derived uraemic toxins in a Belgian paediatric CKD cohort**

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UZ Gent

Introduction

Chronic kidney disease (CKD) in children, is a pro-inflammatory and invalidating systemic condition that leads to an unacceptably high morbidity and mortality. Central is the accumulation of organic metabolic waste products, coined as uraemic toxins (UTs). Several of these UTs are protein-bound and gut-derived. Accruing adult evidence points out distinctly dysbiotic gut microbiota in CKD, resulting in a state of increased proteolytic fermentation, that might be counteracted by dietary fiber. Several attempts to alter UT generation with pre-, pro- and synbiotics yielded contradictory results. This is unexplored in the paediatric population. Therefore, we aimed to define the relationship between dietary fiber intake and protein-fiber ratio (PFR) versus UT concentrations.

Methods

In this 2-year prospective observational longitudinal study, we included 262 visits of 47 predialysis CKD 2-5 or transplant patients [9.1 (1.0-17.0) years]. Total and free levels of 6 protein-bound UTs (indoxyl sulfate (IxS), p-cresyl sulfate (pCS), p-cresylglucuronide (pCG), hippuric acid (HA), indole-acetic acid (IAA) and 3-carboxy-4-methyl-5-propyl-furanpropionic acid (CMPF), coupled to in-depth diet histories were collected. Linear mixed models were used to assess the relationship between the UTs versus dietary fiber and PFR, considering eGFR.

Results

Total dietary fiber intake was low, especially in advanced CKD: 13.3 g/day/BSA (7.9-19.8) in CKD1-3 versus 8.8 (1.7-11.6) in CKD 4-5 ($p = 0.018$). While no association between fiber intake and UT concentrations was found, a significant association between PFR with total serum IxS [$e = 0.039$ (0.0014 – 0.076) $p = 0.042$] was seen.

Conclusion

Our data show that fiber intake in paediatric CKD is low, and that a higher PFR is associated with higher levels of IxS. The neglect of current CKD nutrition guidelines for dietary fiber intake thus seems unjustified. They further illustrate the complexity in restoring eubiosis and suggest a multifactorial approach, in which the interaction between fiber and protein intake might be a piece of the puzzle.

Long Oral Presentation**LO 12.****The impact of chronic kidney disease on the quality of life and psychosocial functioning of children and their family**

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UZ Leuven

Background/aims

In the frame of RIZIV/INAMI convention of pediatric nephrology, tertiary centers of pediatric nephrology provide multidisciplinary care for children with chronic kidney disease (CKD). This multidisciplinary care is essential, as studies have shown that CKD has an impact on the psychosocial development and Quality of Life (QoL) of these children and their families. In addition, systematic follow-up is very important as psychological interventions can benefit from these results. Many studies have focused on the QoL of children with CKD. However, less is known about the illness-related stress of the parents and about the association between illness-related stress of parents and psychosocial outcomes of their children. Therefore the aims of the present study are to investigate (1) illness-related stress in parents of children with CKD and compare it to parents of children with cancer and (2) explore the associations between illness-related parental stress and child psychosocial parameters.

Method

All children with CKD included in the convention of UZ Leuven were approached. QoL was assessed by means of PedsQL Core 4.0 and illness-related parental stress by means of Pediatric Inventory for Parents (PIP). To explore stress in the parents of children with CKD, we compared their scores on the PIP with previously published scores in a sample of pediatric cancer.

Results

In total we included 49 children (30 boys; M age= 10.06, SD = 5.05) and their parents. The average parental stress related to the frequency of stressful events in our parents of children with CKD did not significantly differ from parents of children with cancer ($t(171) = 1.85, p > .05$). However, stress due to the perceived difficulty of medical stressors in our population significantly differed from a pediatric cancer sample ($t(171) = -4.02, p < .001$), with lower perceived difficulty of medical stressors. Finally, all scales of the PIP and PedsQL are significantly negative correlated ($p < 0.01$), showing that more parental stress is associated with a lower QoL in the child.

Conclusions

Our study show that CKD also affects the parents and that parental stress is comparable to parental stress due pediatric cancer. Moreover, more parental stress is associated with a lower QoL in the child. These results show that health care services should keep providing a multidisciplinary team and focus on the entire family of children with CKD, in order to optimize their wellbeing.

SO 3.

Kinetic modeling as guide for dialysis prescription in acute neonatal hyperammonaemia: an example using CarpeDiem and Fresenius 4008 machine

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UZ Gent

Background

Acute neonatal hyperammonaemia is associated with poor neurological outcomes and high mortality. As these outcomes are inversely related to the duration of the hyperammonaemic coma, prompt management that guarantees a fast decline in serum ammonia is crucial. Using our experience with different haemodialysis machines, we developed a kinetic model for acute neonatal hyperammonaemia to draft a dialysis prescription protocol that can ensure a fast decline in serum ammonia (goal: <400µmol/L in <4hours).

Methods

From all dialysis sessions performed in 2020 at Ghent University Hospital (Belgium) in newborns with hyperammonaemia, dialyzer clearance and extraction ratio were calculated using intradialytic ammonia concentration-time curves. A single compartment kinetic model with a distribution volume of 60%-80% of body weight was assumed, and generation was derived from the interdialytic concentration increase. The calibrated single compartmental model was further used to simulate serum ammonia decline in infants of 2-5kg for different ammonia start concentrations (3000, 1500, 800, 400, 200µmol/L), dialysis machines/dialyzers and settings (blood flow QB 30-50mL/min).

Results

Four patients (3.24±0.40 kg) underwent 13 dialyses: 5 with the 4008 machine and FXPaed dialyzer (Fresenius Medical Care, Germany); and 8 with the CarpeDiem machine (50% with 0.15m² respectively 0.25m² dialyzer) (Medtronic, USA). QB was 30-35mL/min (4008-FXPaed), 22-35mL/min (CarpeDiem 0.15), and 30-34mL/min (CarpeDiem 0.25). Extraction ratios were 38±5% for 4008-FXPaed, 10±3% and 13±3% in the CarpeDiem 0.15m² and 0.25m² dialyzer. Generation was 0.40±0.25µmol/min, with no observed impact on dialyzer clearance and extraction ratio. Using the 4008-FXPaed, the time to decrease start concentrations of 3000µmol/L (3kg) to <400µmol/L was 315 min for a QB of 30mL/min and 190min for a QB of 50mL/min, while it was 205 and 125min for a start concentration of 1500µmol/L, and 110 and 65min for a start concentration of 800µmol/L. In general, for start concentrations >800µmol/L in 3kg child, the CarpeDiem machine was found inadequate to decrease serum ammonia in <4h. Increasing body weight (5kg) resulted in longer time intervals to reach target.

Conclusion

Kinetic models can guide our management decisions and treatment protocols by predicting which treatment goals can be reached in a given time period with a particular dialysis prescription, available resources and/or dialysis modality.

Short Oral Presentation**SO 4.****Perinatal determinants of renal function and blood pressure in former extremely low birthweight infants in late childhood**

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Background

Extremely Low Birthweight (ELBW) infants suffer from adverse renal and cardiovascular outcomes in later life, including lower estimated glomerular filtration rate (eGFR) and elevated blood pressures. Less is known regarding the perinatal risk factors for these outcomes.

Methods

Within the "PREMATurity as predictor of Cardiovascular-renal Health" (PREMATCH) study, renal function and blood pressure in 93 ELBW children and 87 controls were evaluated around age 11 during a visit to the study's research center. We compared eGFR and systolic/diastolic blood pressure (SBP/DBP) between cases and controls. An adverse outcome was defined according to chronic kidney disease (CKD) stage 2 (eGFR <90 ml/1.73m²/min), whereas elevated DBP/SBP were defined as an average SBP/DBP ≥90th percentile. We investigated perinatal risk factors for adverse outcome amongst ELBW children. We developed a proof-of-concept model predicting the risk of CKD stage 2 at 11 years wherein points were assigned for sex (male), ventilation therapy (>10 days) and intraventricular hemorrhage (IVH; any).

Results

We replicated the finding that ELBW children suffer from lower eGFR (94.1 vs. 106.5 ml/1.73m²/min, p<0.001) and higher SBP/DBP (75th vs. 47th percentile, p<0.001; 68th vs. 54th percentile, p<0.001). No direct correlation between SBP/DBP and eGFR was observed for our cohort of ELBW children. Male ELBW children developed CKD stage 2 more frequently (OR=3.33, p=0.055). ELBW children who developed CKD stage 2 received significantly longer ventilation therapy (17 vs. 9 days, p=0.006) and suffered more frequently from IVH (40% vs. 15.8%, p=0.056). Risk of CKD stage 2 increased linearly from 0% to 80% with regard to the points scored in the prediction model. No ELBW children developed microalbuminuria by age 11. No perinatal factors associated with elevated SBP at age 11. ELBW children who developed elevated DBP had a significantly higher gestational age (29.5 vs. 27.2 weeks, p=0.001), received a shorter treatment with oxygen (18 vs. 40 days, p=0.09) and were discharged at a significantly lower weight (1786 vs. 2240 g, p=0.01).

Conclusion

At age 11, ELBW children have poorer renal function and higher blood pressures, however early mechanisms appear independent. Renal function in ELBW children is influenced by identifiable perinatal factors that may allow for those at increased risk to receive a more intensive follow-up. Blood pressure is not adversely influenced by perinatal factors.

SO 13.

FCGG Renal Biopsy Network: first epidemiological report on pediatric renal diseases

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UZ Gent, NBVN, UZ Leuven, UZ Antwerpen, UZ Brussel, UZ Leuven, AZ Nikolaas

Objective

The Flemish Collaborative Glomerulonephritis Group, FCGG, was founded in 2016 as a collaboration between renal pathologists and nephrologists, to collect all native kidney biopsies within the NBVN organization, in order to standardize diagnosis and therapy. Uniform renal biopsy request and renal biopsy report forms, and a new comprehensive list of renal pathology diagnoses for coding were introduced. The 2017-2019 epidemiological data of the pediatric patients (age= 0-17 years) are presented.

Methods

Following informed consent and in compliance with GDPR, basic patient and categorical renal data, semi-structured medical information of renal histopathology and the clinical renal disease are collected.

Results

In 2017-2019, 127 renal biopsies in pediatric patients were reported in the Flemish region [3.3 per 100,000 pediatric inhabitants per year] – 6% of all stored biopsies; more boys N=73 than girls N=54. Three “clinical” patterns were equally represented: only proteinuria >1g/day; only hematuria; and combination of proteinuria and hematuria. Acute or chronic renal failure were present in 25%. In each age/gender group (0-5; 6-11; 12-17), glomerulopathy was most common. The glomerulopathy differed among age/gender groups. In the youngest age group (0-5 years; N=33) minimal change disease / nephrotic syndrome of childhood predominated (50%), followed by Henoch-Schönlein nephritis and Alport’s disease. The middle age group (6-11 years; N=47) mainly presented with diseases with hematuria (47%): IgA nephropathy, Henoch-Schönlein nephritis and Alport’s disease; followed by minimal change disease (28%). Greater impact of gender was noted in the highest age group (12-18 years; N=47): IgA- and GBM mediated nephritis was present in 50% of the boys, whereas a more diverse palette of kidney diseases was present in female teenagers. Children of Western-European descent presented with hematuric renal diseases (50%) and with nephrotic diseases (25%), while the reverse was noted in the group without a Western-European descent.

Conclusion

The FCGG network provides a structured format for cross-talk between renal pathologists and nephrologists. Reliable estimates of pediatric renal diseases based on histology are now available in our region. Due to the diverse renal spectrum of the teenager group, a renal biopsy is indispensable.

Short Oral Presentation**SO 15.****Genotype – phenotype correlation in a pediatric autosomal dominant polycystic kidney disease (ADPKD) cohort**

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Objective

The correlation between genotype and phenotype is well described in ADPKD adults. *PKD2* is milder than *PKD1* disease, with end stage kidney disease occurring on average 20 years later, and patients with *PKD1* truncating mutations having a more severe outcome than *PKD1* non-truncating mutations. Still, large differences in outcome occur even within families carrying the same gene variation. Only a few cases series reported the genetic profile of severely affected ADPKD children and suggest an additional effect of hypomorphic genes. We therefore aim to analyse the geno-phenotype profile in a well characterized pediatric ADPKD cohort.

Methods

Clinical, familial, biological and imaging data were collected longitudinally in children diagnosed with ADPKD. Genotypic analysis was done using a custom Agilent SureSelect gene panel containing 136 ciliopathy-associated genes, including *PKD1* and *PKD2*. Mutations and/or variants identified were individually evaluated for pathogenicity.

Results

57 ADPKD children from 44 families were diagnosed at a mean (\pm SD) age of 4.1 (\pm 4.9) years. ADPKD diagnosis was made in 32 children (56%) because of asymptomatic screening as requested by the family; 7 (12%) due to presenting symptoms (6 due to urinary tract infection and 1 due to post-traumatic macroscopic hematuria); 9 (16%) due to a coincidental finding of renal cysts on US and in 9 cases (16%) a prenatal diagnosis was performed. Twenty-nine children (51%) met the definition of very-early onset (VEO) disease. We identified pathogenic mutations in 100% of our patients, in which the prevalence of *PKD1* truncating, *PKD1* non-truncating, *PKD2* and *GANAB* mutations was 75.4%, 19.3%, 3.5%, and 1.8%, respectively. Four cases (7%) were due to a de novo mutation. Interestingly, in 29 patients (51%) the germline mutation was the only identified mutation. However, in the rest of the subjects additional variants were identified in other ciliopathy-associated genes. In 12 cases (21%) the additional identified variants found in either the *PKD*, *PMM2*, *HNF1B*, *DNAJC1*, *CEP290*, *NEK1*, *MKKS*, *NPHP4* or *PKHD1* genes were scored to have a potential phenotypic effect, which will be evaluated by continued follow-up of this cohort.

Conclusion

We report the first large cohort of genotyped ADPKD children, including an extensive panel of ciliopathy genes next to the PKD genes. Interestingly, we found a high prevalence of additional and potentially modifying variants in this young population.

SO 17.

Comparative analysis from 2005 to 2020 of outcome parameters in Belgian children with a kidney allograft

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UZ Leuven

Introduction

Kidney transplantation is the treatment of choice for children with end stage kidney disease because it has important advantages over other methods of kidney replacement therapy (hemodialysis, peritoneal dialysis) in terms of morbidity and mortality. To meet the request of RIZIV/INAMI in the current analysis we provide an overview of hard outcome parameters (graft function, growth parameters, blood pressure) in a cross-sectional cohort of children with kidney transplantation followed in Belgian University Hospitals in 2020 and compared this to the cohort of Belgian pediatric kidney transplant recipients in 2005.

Patients and methods

Retrospective data were collected from the cross-sectional cohort of children followed in the University Hospitals of Ghent, Leuven, Brussels (HUDERF, UCL St Luc) and Liege. Clinical and functional characteristics were expressed as mean + SD or as a standard deviation scores (SDS) and were compared with the normal values for age and sex. Statistical analysis was performed by statistical program SPSS for Mac.

Results

In 2020 the cohort consisted of 125 children aged 3-19 years old with a mean age of 13.5 + 4.3 years (78 male / 47 female). In 2005 the cohort of patients consisted of 127 children aged 4-18 years old, with a mean of 14.0 year + 4.6 years (66 male / 61 female). Mean eGFR in 2020 and 2005 was similar (64 + 26 ml/min/1.73 m² vs. 68.1 + 17 ml/min/1.73 m², respectively). Although stunting is still common, mean height SDS was considerably better in 2020: -1,2 SDS + 1.4 SD than in 2005: -1.9 SDS + 1.5 SD (p<0.01). In addition, mean systolic and diastolic blood pressure were lower in children followed in 2020 compared to 2005 (+0.7 versus +1.0 SDS (p<0.01) for systolic and +0.6 versus +0.8 SDS (p<0,05) for diastolic blood pressure.

Discussion

This cross-sectional analysis of the national data shows that the current cohort of children in follow-up after renal transplantation in Belgium have better standardized outcome parameters for height, systolic and diastolic blood pressure compared to a historic cohort. The mean estimated GFR remains mildly compromised (CKD stage 2) while growth and cardiovascular parameters show a positive trend. This is most likely the result of excellent multi-disciplinary care provided by pediatric teams including highly specialized pediatric nephrologists and nurses, psychologists, dieticians and social workers.

Short Oral Presentation

SO 20.

ADPedKD: A global online platform to explore the childhood phenotype of Autosomal Dominant Polycystic Kidney Disease

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Introduction

Autosomal Dominant Polycystic Kidney Disease (ADPKD) is the 4th common cause of renal replacement therapy worldwide. As the disorder has been historically considered an adult-onset disease, there is a lack of longitudinal data from large pediatric cohorts. However, evidence is growing that first manifestations of ADPKD may be detected in childhood and children represent a specific target population for future treatment, allowing a better chance of preserving long term kidney function. To better define the pediatric spectrum of the disease, a global multicenter observational study on childhood-diagnosed ADPKD was launched in 2017.

Methods

The ADPedKD registry is a worldwide web-based database, including both retrospective and prospective longitudinal data from young ADPKD patients (≤ 19 years). Australia, North-America and the United Kingdom joined the initiative with their source databases, namely the KidGen Collaborative (KidGen), NIH-funded Hepato-Renal Fibrocystic Disease (HRFD) and National Registry of Rare Kidney Diseases (RaDaR). Under informed consent, de-identified patient data, including genetics, radiological and laboratory findings, treatments and follow-up were enrolled in the database accessible via <https://www.ADPedKd.org/>.

Results

1019 ADPKD children (from 89 centers and 33 countries) are enrolled in the registry of which 167 patients from RaDaR, 17 from KidGen, 11 from HRFD and 824 from ADPedKD (401 male/ 423 female) with a mean (\pm SD) age at diagnosis of 6.3 ± 5.2 years. 81 children (9.8%) were diagnosed prenatally at a mean gestational age of 26.8 ± 7.8 weeks. Reasons for initial visit were: family screening in 325 (39.4%), postnatal incidental finding in 223 (27.0%), presenting features (such as hematuria, hypertension, urinary tract infections and flank or back pain) in 150 (18.2%) or unknown/not available in 126 (15.3%). Genetic testing was performed in 42.8% of the population, with the following results: PKD1 mutation (85.4%), PKD2 mutation (11.7%) and others (6.0%).

Conclusions

The ADPedKD registry is a unique source of clinical observational data that will provide deep phenotyping of children with ADPKD and will allow to define unified diagnostic, treatment and follow-up recommendations.

SO 21.

Dietary fibre is associated with serum levels of uraemic toxins in children with chronic kidney disease

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Background

Imbalanced colonic microbial metabolism plays a pivotal role in generating protein-bound uraemic toxins (PBUTs), accumulating with deteriorating kidney function and contributing to the uraemic burden of children with chronic kidney disease (CKD). Dietary choices impact the gut microbiome and metabolism. The aim of this study was to investigate the relation between dietary fibre and gut-derived PBUTs in paediatric CKD.

Methods

Sixty-one (44 male) CKD children (9±5 years) were prospectively followed at 3-month intervals for 2 years. Dietary fibre and protein intake was evaluated by either 24-h recalls (73%) or 3-day food records (27%) and coupled to measurements of total and free serum levels of different PBUTs using liquid chromatography. We used linear mixed models to assess associations between fibre intake and PBUT levels.

Results

We found an inverse association between increase in fibre consumption (g/day) and serum concentrations of free indoxyl sulfate (-3.1% [-5.9%; -0.3%] (p = 0.035)), free p-cresyl sulfate (-2.5% [-4.7%; -0.3%] (p = 0.034)), total indole acetic acid (IAA) (-1.6% [-3.0%; -0.3%] (p = 0.020)), free IAA (-6.6% [-9.3%; -3.7%] (p < 0.001)), total serum p-cresyl glucuronide (pCG) (-3.0% [-5.6%; -0.5%] (p = 0.021)) and free pCG levels (-3.3% [-5.8%; -0.8%] (p = 0.010)).

Conclusion

The observed associations between dietary fibre intake and the investigated PBUTs highlight potential benefits of fibre intake in the paediatric CKD population. The present observational findings should inform and guide Adaptations of dietary prescriptions in children with CKD.

Short Oral Presentation

SO 30.

Reaffirmation of empiric antimicrobial treatment of febrile urinary tract infections in infants and children in Tournai-Belgium.

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*CHwapi Tournai*Background

From July 2014 until July 2017, we conducted a prospective study about empiric antibiotherapy of febrile urinary tract infections (UTI). We carried on the study for another three-year-period in order to enroll more patients and to compare the evolution of susceptibility/resistance. The final goal was the validation of our previous recommendations.

Methods

From 30th July 2014 to 29th July 2020, we collected prospectively data of 415 patients aged 0 to 15 years (1/3 boys, 2/3 girls) with general symptoms compatible with UTI and confirmed by suprapubic aspiration or bladder catheterization in children <3 years and by clean void or bladder catheterization in children ≥ 3 years of age. Results were analysed regarding age-groups (0-2 months, 2 months-urinary continence and after urinary continence), known uropathy at the time of diagnosis and type of episodes (first or recurrence). The resistance pattern of E Coli (the most common pathogen isolated in 87.58%) and other pathogens was also reported.

Results

In the group with no known uropathy at the time of UTI diagnosis, 285 patients had a first episode of UTI. Resistance in E Coli is 48.42% to ampicillin, 22.81% to amoxiclav and cotrimoxazole, 3.86% to cefuroxime. With recurrence (83 patients), the resistance increases to 55.42% to ampicillin, 26.51% to amoxiclav, 10.84% to cefuroxime but is only 3.61% to ciprofloxacin and 1.20% to amikacin. These data analysed in the 3 age-categories have the same trends.

In the group with a known uropathy, 11 patients had a first episode of UTI. Resistance is 54.55% to ampicillin, 9% to cefuroxime, 18.18% to temocillin and 0% to ciproxine and amikacin. In case of recurrence (36 patients), the rate of resistance increases to 69.44% to ampicillin, 16.67% to cefuroxime, 33.33% to cotrimoxazole, 11.11% to ciproxine, 5.56% to temocillin and 2.78% to amikacin.

If we compare the 2 three-year-periods, the results were globally identical except for resistance to amoxiclav that increased in the second period.

Conclusion

In case of a first UTI episode with no known uropathy, cefuroxime is the first choice for empiric antibiotic treatment. Ampicillin and amoxiclav are not suitable because of emergence of a high resistance rate to amoxiclav in the second period.

In case of a first episode of UTI with a known uropathy, and in case of UTI recurrence with or without uropathy, resistance tends to increase and amikacin becomes our first choice as an empiric treatment.

PW 2.**The choice between deceased and living donor renal transplantation in children: a multicentric cross-sectional study**

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Introduction

The aim of our study was to prospectively evaluate factors influencing the choice between a deceased (DDKT) and living donor for kidney transplantation (LDKT) from the perspective of parents and physicians.

Methods

We included patients with CKD stage 4 and 5 at the University Hospitals of Ghent, Leuven and Antwerp between February and December 2019. Questionnaires were distributed among parents and physicians in order to evaluate the potential differences between the medical recommendation and parental choice.

Results

Twenty-eight patients (mean age 10.48 yr, range 2-19 yr), 10 girls and 18 boys were included. Six patients had a history of previous kidney transplantation. Parents of 13 children preferred DDKT and 13 LDKT, in 2 cases there was no preference. Physicians recommended DDKT in 14 cases and LDKT in 14 cases. The factors precluding LDKT were multifactorial for both parents as physicians. In 7 cases the parents had (a) medical reason(s), in one case there were both medical and social reasons and in 5 cases no reason was given. The medical reasons were as follows; parents found unsuitable as a donor after medical screening (n=5), child too small for a LDKT (n=4), necessity of a combined liver- and kidney transplantation (n=1). In one case self-employment was a reason for preferring DDKT. The reasons the nephrologist advised against LDKT included; medical (n=6), social (n=4), combination of both (n=3) or lack of motivation (n=1). The medical reasons were as follows; parents found unsuitable as a donor after medical screening (n= 6) or a child too small for a LDKT (n=1).

Conclusion

In this study parents and physicians preferred future LDKT in respectively 46% and 50% of cases, while 35% of the children underwent LDKT in Belgium in 2018-2019. From the physicians perspective social factors play an important role in not actively promoting LDKT, although in only one case parents reported a social reason for preferring DDKT. Refusal to donate without specifying a reason is a potentially modifiable factor and strategies aimed at education and guidance of the families might contribute to a higher incidence of living donation in our setting.

Poster Walk**PW 15. E****Effects of fecal microbiota transplantation for recurrent clostridium difficile in children with renal replacement therapy**

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UZ Leuven

Background/aims

Clostridium difficile infection (CDI) is one of the most common causes of healthcare-associated infections. If recurrent CDI (rCDI) doesn't respond to antibiotics, treatment with a fecal microbiota transplant (FMT) may be a solution. The first results of FMT within immunocompromised children or with a solid organ transplantation are promising but until now, there are no data on FMT for children with rCDI and end stage renal disease who receive replacement therapy.

The microbiome plays an important role in growth, the development of the immune system and nutrition. Changes in the microbiome can cause dysbiosis which increases the chance of developing CDI. When CDI is treated with FMT, the goal is to implant healthy bacteria and to restore the symbiotic environment in the gut.

One of the functions of the kidney is to metabolize and excrete uremic retention molecules (URM). In patients with chronic kidney disease (CKD), this function cannot be fully executed and will lead to an accumulation of URM. Furthermore, the composition and the metabolic activities of the intestinal microflora change in patients with CKD. One of the functions of the healthy microbioma is to produce harmful metabolites. In a dysbiotic environment, the production of these metabolites will change and contribute to the accumulation of the URM.

We explored the effect of FMT on the level of URM as a potential future intervention for the modification of systemic exposure to these toxins in patients with CKD.

Results

All three patients have had no recurrences and did not develop any complication. The dysbiosis of the microbiome in all three patients with CKD and Clostridium hasn't resolved after FMT despite eradication of CDI. Both the number of bacteria and the species richness did not statistically differ between patients nor between time-points so the dysbiosis remains. The amount of URM depends on the CKD stage. In case of a high CKD stage and thus low renal clearance, URM will be higher. Three months after FMT took place, 64.28% of the URM are reduced so we can assume that FMT has an effect on the production and accumulation of URM. This may be explained by the leaky gut but needs more research.

Conclusion

FMT seems to be a safe and effective method to eradicate recurrent CDI in this specific population. Despite CDI was resolved, the dysbiosis persisted after the transplantation. However, most of the URM decreased after FMT.

PW 20.

Effects of faecal microbiota transplantation for recurrent clostridium difficile infection in children with chronic kidney disease

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UZ Leuven, KU Leuven

Background/aims

Recurrent *Clostridium difficile* infection (CDI) is a growing problem in children with different manifestations of chronic disease, including chronic kidney disease (CKD). In case of multiple recurrences despite antibiotics, faecal microbiota transplantation (FMT) can be considered, although the experience in children is limited. In addition to the effect on CDI, FMT could have a positive effect on the intestinal dysbiosis and the accumulation of uremic retention molecules (URM) associated with CKD. In this case series we want to investigate the clinical efficacy of FMT in children under renal replacement therapy (dialysis/transplantation) and its effect on the intestinal dysbiosis and URM.

Methods

We describe 2 children with a kidney transplantation and 1 child on haemodialysis. All suffered from recurrent CDI despite multiple courses of metronidazole and vancomycin. Donor feces was collected from their parents. Next to the clinical efficacy of FMT we analyzed stool before and after FMT for the microbiome composition and compared to healthy controls. CRP and faecal calprotectin were analysed as parameters for the gut and systemic inflammation. A panel of URM were analysed before and after FMT to determine the level of URM before and after the treatment.

Results

CDI resolved after FMT in all three patients without any adverse event. One patient had a single recurrence after 3 months for which he received a second FMT, afterwards no new recurrences were seen. We could not show any statistical difference for CRP and calprotectin over time after FMT. We observed a reduced richness (ten times less in comparison with healthy controls) and bacterial diversity, which did not improve after FMT. The patient that received hemodialysis demonstrated higher concentrations of URM in comparison with the other two children. Three months after FMT took place, 64.28% of the URM are reduced.

Conclusion

FMT is an effective treatment for recurrent CDI in our patients with chronic kidney disease without serious adverse events. Additional analysis of the microbiome showed an important intestinal dysbiosis in our patients, although we did not see a significant improvement 3 months after FMT. Nevertheless a reduction was seen of the most URM after FMT.

Poster Walk**PW 24.****The effect of a multidisciplinary weight loss program on renal circadian rhythm in obese adolescents**

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Background/Aim

Adolescent obesity is a serious health problem associated with many comorbidities. Obesity-related alterations in circadian rhythm have been described for nocturnal blood pressure and for metabolic functions. We believe renal circadian rhythm is also disrupted in obesity, though this has not yet been investigated. This study aimed to examine renal circadian rhythm in obese adolescents before and after weight loss.

Methods

In 34 obese adolescents (median age 15.7 years) participating in a residential weight loss program, renal function profiles and blood samples were collected at baseline, after 7 months, and again after 12 months of therapy. The program consisted of dietary restriction, increased physical activity, and psychological support.

Results

The program led to a median weight loss of 24 kg and a reduction in blood pressure. Initially, lower diurnal free water clearance (-1.08 (-1.40--0.79) mL/min) was noticed compared with nocturnal values (0.75 (-0.89--0.64) mL/min). After weight loss, normalization of this inverse rhythm was observed (day -1.24 (-1.44-1.05) mL/min and night -0.98 (-1.09--0.83) mL/min). A clear circadian rhythm in diuresis rate and in renal clearance of creatinine, solutes, sodium, and potassium was seen at all time points. Furthermore, we observed a significant increase in sodium clearance. Before weight loss, daytime sodium clearance was 0.72 mL/min (0.59-0.77) and nighttime clearance was 0.46 mL/min (0.41-0.51). After weight loss, daytime clearance increased to 0.99 mL/min (0.85-1.17) and nighttime clearance increased to 0.78 mL/min (0.64-0.93).

Conclusion

In obese adolescents, an inverse circadian rhythm of free water clearance was observed, with higher nighttime free water clearance compared with daytime values. Weight loss led to a normalization of this inverse rhythm, suggesting a recovery of the anti-diuretic hormone activity. Circadian rhythm in diuresis rate and in the renal clearance of creatinine, solutes, sodium, and potassium was preserved in obese adolescents and did not change after weight loss.

PW 38.

Growth after pediatric kidney transplantation: the effect of pre-transplant recombinant growth hormone and post-transplant corticosteroids

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Background

Catch-up growth after pediatric kidney transplantation (kTx) is usually insufficient to reach normal adult height. We aimed to analyze the effect of pre-transplant use of recombinant human growth hormone (rhGH) and corticoid withdrawal on longitudinal growth in the 1st year after kTx.

Methods

Patients who underwent kTx between 1996 and 2018 in 5 Belgian and Dutch centers before the age of 18 yrs old were included in this retrospective study. The height standard deviation score (SDS) at 1 year after kTx and the difference between height SDS at kTx and 1 year post kTx (Δ height SDS) in children who were and were not on corticosteroids at 1 year (CS+ and CS-) and who were and were not treated with rhGH before kTx (rGH+ and rGH-) were evaluated using a Kruskal-Wallis test and post-hoc testing. Linear regression models were fitted to identify factors associated with height SDS and post-transplant catch-up growth.

Results

188 patients were included, with median age at kTx 9.2 yrs. The numbers of patients in the CS+/rGH+, CS+/rGH-, CS-/rGH+ and CS-/rGH- groups were 76, 59, 23 and 29. The respective median height SDS before kTx and at 1 year after kTx in these groups were -1.37/-1.60, -1.00/-0.70, -1.40/-1.39 and -1.75/-0.99. Catch-up growth varied significantly between the groups ($p=0.002$). 45 and 46 children had a height < 2 SDS at kTx and at 1 year after kTx, respectively. Pubertal stage was associated with height SDS at 1 year after kTx, whereas donor source, graft function, rhGH administration of rhGH before kTx as well as continuation of steroid therapy 12 months post kTx were associated with the difference between height SDS at kTx and 1 year after kTx.

Conclusion

Catch-up growth at 1 year after kTx was highest in children without corticosteroids and rhGH and lowest in those on corticosteroids at 1 year and who had received rhGH pre-kTx. The use of corticosteroids at 1 year post kTx is the most important factor associated with catch-up growth in children after kTx.

PW 40.**Cytopenia in pediatric patients with Autosomal Dominant Polycystic Kidney Disease**

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*KU Leuven, UZ Leuven, CHU*Background

Cytopenia has emerged as a new extra-renal feature in Autosomal Dominant Polycystic Kidney Disease (ADPKD). In adult studies, ADPKD is associated with lymphopenia, leukopenia and thrombopenia through all chronic kidney disease (CKD) stages. The underlying mechanism is unknown. A decreased polycystins expression is associated with decreased proliferation in immortalized lymphoblastoid cell lines. Sequestration of white blood cells in the cystic organ and the uremic inflammatory environment in CKD could also be involved.

Methods

Cross-sectional study including: <19 years, ADPKD diagnosis (more than 1 renal cyst with a positive family history and/or confirmed genetic test), with normal renal function. For each patient, detailed questionnaire on the frequency of antibiotic use and infections in the month prior to inclusion. Blood samples collected and biological data on leukocyte, lymphocyte and thrombocyte counts compared between ADPKD versus sex and age-matched healthy controls (HC). Leukocyte and lymphocyte counts adjusted for age according to local laboratory values. Neutropenia: neutrophil count <2.0 x10⁹cells/L (mild neutropenia: <1.5 x10⁹/L, moderate neutropenia: <1.0 x10⁹/L, severe neutropenia: <0.5 x10⁹/L). Thrombopenia: < 150x10⁹/L.

Results

43 ADPKD patients (21 boys) and 43 HC (20 boys) with mean age and respective Standard Deviation (SD) of 11.2 4.5 and 11.1 4.3 years. 5 patients with co-medication (ACE inhibitor (N=3), Methylphenidate (N=2)). No patient under immunosuppressive therapy. No patient had infection in the month prior to inclusion. No differences between the 2 groups except for the estimated GFR (ADPKD: 122.1±23.9 ml/min/1.73m²; HC: 109.4±20.2 ml/min/1.73m²; p=0.009). Significant reduction in total leukocyte count (ADPKD: 6.2x10⁹/L (±2.6 SD); HC: 7.3x10⁹/L (±2.0 SD); p=0.007) and neutrophil count (ADPKD: 2.9x10⁹/L (±1.0 SD); HC: 3.6x10⁹/L (±1.6 SD); p=0.01) in ADPKD. Significant differences in the monocytes count (ADPKD: 0.4x10⁹/L (±0.2 DS); HC: 0.6x10⁹/L (±0.2 DS); p<0.001) and thrombocytes count (ADPKD: 257.5x10⁹/L (±55.4 DS); HC: 298.0x10⁹/L (±67.9 DS); p=0.002) between the two groups but no differences in the eosinophils and lymphocytes count.

Conclusion

In the pediatric patients, ADPKD is associated with lower leukocyte, neutrophil, monocyte and thrombocyte count. Even without phenotype associated with neutropenia, these results suggest a role of polycystins in the lymphoid and myeloid lines.

PW 41.

Pharmacokinetics and blood pressure effects of ACE-inhibitors in children with hypertension

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Universiteit Gent, UZ Gent

Background/Aims

Angiotensin-converting enzyme inhibitors (ACE-inhibitors) are the most frequently prescribed drugs in the paediatric population for primary and secondary hypertension. Hogg et al. (2007) have conducted a clinical study to evaluate the pharmacokinetics (PK) of lisinopril in children older than 6 years of age with a normal kidney function and no comorbidities. This group however entails less than 10% of the paediatric population using ACE-inhibitors. Therefore, a new study was set up to analyze the PK and the blood pressure effects of ACE-inhibitors in children and adolescents with hypertension and comorbidities, which represents the majority of the paediatric population.

Method

The study included 13 children between 2 and 18 years old with hypertension who received the ACE-inhibitor lisinopril. The lisinopril dosage was between 0.0125 mg/kg to 0.05 mg/kg per day according to the glomerular filtration rate (GFR). Moreover, a dose-titration of 0.1 mg/kg to 0.2 mg/kg was performed in accordance with the clinical and biochemical evolution. Just before and 4 hours after drug-intake in the morning, blood was drawn at 10 occasions. The data were analyzed using population PK in Monolix®.

Results

The plasma concentrations of lisinopril were best described with a one-compartment model with 1st order absorption and 1st order elimination, parameterized in terms of clearance and volume. The average absorption rate constant (k_a) was estimated at 0.079 1/hr, the volume of distribution (V/F) at 15.1 L and the clearance (CL/F) at 16.2 L/h. These results match the PK parameters of lisinopril in adults as obtained by Thomson et al. (1989) when scaled for body weight. Based on the preliminary PK model, an average 24-hour concentration between 4.3 (0.1 mg/kg) and 8.5 ng/ml (0.2 mg/kg) was predicted for a body weight between 9.6 kg and 97kg. No significant covariates could be identified, possibly due to the small group size and limited GFR range. Moreover, in the systolic and diastolic blood pressure groups, a ≥ 6 mmHg reduction was measured of 95% and 62% respectively. Further analyses are required to identify the optimal dose regimen of lisinopril in CKD population, by also accounting for the PD-effects on blood pressure. No serious adverse events were reported.

Poster Walk**PW 42.****Characteristics of nycthemeral rhythm of urinary water and solute excretion in children with enuresis**

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*UZ Gent, Universiteit Gent*Background

Nocturnal enuresis (NE) is caused by a mismatch of the nocturnal urine production and the functional bladder capacity. The International Children's Continence Society (ICCS) defined nocturnal polyuria (NP) as nocturnal diuresis (ND) exceeding the Expected Bladder Capacity (EBC) by 130%. NP is well documented as pathogenetic mechanism in monosymptomatic, but little is known of its incidence/characteristics in non monosymptomatic NE. NP, when associated with increased water diuresis, is attributed to abnormal circadian rhythm of vasopressin (AVP). This is why NE is treated with desmopressin, an AVP agonist. However 40-60% of the patients show insufficient response to it, suggesting that other factors like nutrition and circadian rhythm of other renal functions may be involved

Methods

The aim was to study the circadian rhythm of the kidney on water and solute excretion in fractionated urine samples over 24h (4 day and 4 night samples); a retrospective analysis of data from 402 enuretic children by whom a 24h urine concentration profile at a home setting was performed. The children were divided according to ND into 3 subgroups based on the ICCS definition a) low-normal ND if ND was <90% of EBC (113 cases) b) high-normal ND if ND was 90-130% of EBC (103 cases) c) NP if ND was >130% of EBC (91 cases)

Results

Increased diuresis rate and free water clearance overnight is present as well in the NP as well in the high-normal ND group.

Moreover both groups demonstrate not only an abnormal circadian rhythm of diuresis ($p=0,001$), but also of osmolar excretion ($p<0,001$).

The predominant findings are observed in both groups in the two first nighttime samples. Children with NP produced more urine not only at night but also during the daytime and in a 24h period ($p<0,003$). They also excreted more osmols in these three periods ($p<0,001$) comparing to the other groups

Conclusion

Where the clinical indication for desmopressin is often restricted to children with NP (>130%EBC), our data suggest that:

- NP should better be defined as $ND>100\%$ EBC and that this population might be equal desmopressin responsive
- overall pathogenesis is more complex than an abnormal circadian rhythm of AVP; in up to 60% of patients abnormalities of other circadian rhythms of renal functions might be involved, as well as increased nutritional intake
- NP is the highest and Uosmolality the lowest in the early night collections; thus treatment should target fast rather than long term action

P 137.

Neonate with transient pseudohypoaldosteronism.

S. Verbeeck, E. Levtchenko, K. Casteels, D. Mekahli, N. Knops

UZ Leuven

Background

A 3 month old infant presented with failure to thrive and severe dehydration on clinical examination. Blood sample showed a hyponatremia of 125mmol/L, severe metabolic acidosis with a bicarbonate of 7 mmol/L and potassium of 5.5mmol/L. There were no elevated infectious parameters and no fever.

Diagnosis

Further examinations showed no clinical signs of congenital adrenal hyperplasia and normal levels of cortisol and 17-OH progesteron. A sweat test was normal, there was no history of extrarenal losses. Urine sample showed signs of urinary tract infection and ultrasound revealed left sided hydroureteronephrosis. There were no signs of reflux on MCUG. Very high aldosterone and renin levels confirmed diagnosis of transient pseudohypoaldosteronism triggered by UTI. Transient pseudohypoaldosteronism is seen solely in young infants and is triggered by a UTI and/or an obstructive uropathy. It is caused by a decreased sensitivity of the mineralocorticoid receptor to aldosterone. This sensitivity is already low in infants and decreases even more as a reaction to urinary tract infection or obstruction.

Treatment

Administration of antibiotics and optimisation of fluid and salt intake had a good result with normalization of blood electrolytes within days and a spectacular recovery of the weight curve.

Conclusion

- In infants with failure to thrive, even in the absence of infectious parameters or fever, remember to check a urine sample.
- In children with hyponatremia, hyperkalemia and metabolic acidosis, consider the possibility of transient pseudohypoaldosteronism.

P 138.**Fortuitous discovery of severe hyponatremia and primary hyperoxaluria of an infant**

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*Vivalia, Centre Hospitalier de l'Ardenne (Libramont), U Liège*Case report

Our patient was 3 months old. She was brought by her parents to the emergency department for a palpebral oedema that appeared a few days ago. She was born at term with a low birth weight and has difficulties to gain weight. But in the last few weeks, the child had gained a lot of weight. On clinical examination, an oedema of the lower limbs was also noted. Blood biology showed an anemia with a hemoglobin at 6,2 g/dl, severe hyponatremia at 100 mmol/L and a major renal failure.

Additional exams and a transfer to intensive care were organized.

At first, we suspected a congenital nephrotic syndrome. After further examinations, including a genetic testing, a primary oxaluria type 1 was found. Thus far, the patient is still hospitalized and dialyzed pending a double transplant (liver-kidney).

Discussion

Primary hyperoxaluria (PH) is characterized by an oxalate overproduction. PH is caused by a mutation in one of the three enzymes involved in glyoxylate metabolism.

PH type 1 is the most common form (about 80% of the cases). In this case, the deficient enzyme is the glyoxylate aminotransferase (AGXT) involved in the transformation of glyoxylate to glycine and present in the liver.

The first symptoms appear within the first year of life. As the excess of oxalate is mainly excreted by kidneys, they are the prime target for oxalate deposition and the first manifestations are renal.

Measuring this excretion of oxalate confirms the hypothesis but the definitive diagnosis relies on a molecular testing. If genetic testing isn't available, liver biopsy can be considered only if there is a strong clinical suspicion for PH. Prenatal diagnosis is possible.

The efficacy of the treatment is depending on early diagnosis. Indeed, if introduced soon enough, a good medical management can preserve kidney function. The treatment must also be symptomatic. For example, in this patient, we faced a severe hyponatremia that we had to correct. As this condition was probably chronic, it was dangerous to correct it too quickly. Indeed, in those patients with chronic hyponatremia, cerebral adaptation can be observed and they may be less symptomatic. Therefore, treatment should only be provided to symptomatic patients and correction of hyponatremia should not exceed 6 to 8 mEq/L per day.

Furthermore, dialysis may be used to reduce oxalate deposition. But the definitive cure of PH type 1 is the liver transplantation as it corrects the underlying enzymatic defect due to mutation of the AGXT gene. If there is a concomitant renal failure, it should be combined with a kidney transplant.

Conclusion

Primary hyperoxaluria is a rare perinatal diagnosis. Early diagnosis and a regular follow-up can improve the prognosis and quality of life of these patients, but liver transplantation is the only curative treatment.

P 139.

Hyperkalemia in a patient with nephrotic syndrome

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ZNA Koningin Paola Kinderziekenhuis Antwerpen

Background

With the exception of dilutional hyponatremia, electrolyte abnormalities are uncommon in children with nephrotic syndrome. Abnormalities in serum potassium are not reported to occur if glomerular filtration is preserved.

Methods

We report on a 14-year old girl who presented with a first episode of nephrotic syndrome. She developed facial oedema since a couple of weeks. Diagnosis of nephrotic syndrome was delayed because of suspicion of and unsuccessful treatment of allergy. On presentation she had gained 10 kg and had severe edema. Her blood results showed a creatinine of 0.8 mg/dl, sodium of 136 mmol/l, potassium of 6.2 mmol/l and albumine of 18.7 g/l. Urine results showed nephrotic range proteinuria (9 g/g creatinine), sodium of less than 12 mmol/l and potassium of 58 mmol/l. Screening for secondary causes of nephrotic syndrome was normal.

Result

She was started on high dose prednisolone (60 mg/m²). Albumine infusion in combination with furosemide was given by which potassium and creatinine normalised. After correction of the fluid balance albumines were stopped, furosemide was to treat the severe edema. After 3 weeks of prednisolone she went into remission.

Conclusion

The hypothesis is that, in spite of secondary hyperaldosteronism because of volume depletion, sodium delivery to the distal tubule is decreased and therefore potassium secretion is impaired which leads to hyperkalemia.

Posters**P 140.****Urinary tract infection, even though urinalysis and culture are negative: a case report of a 5-year-old girl.**

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*AZ Groeninge, KU Leuven*Background

Urine samples are not always easy to interpret. There aren't any diagnostic tests of which the sensitivity and/or specificity is 100%. Traditionally, the primary diagnosis of a urinary tract infection is based on the presence of pyuria and bacteriuria on flow cytometry. However, the sensitivity and specificity of pyuria are respectively 63-100% and 40-98% and those of bacteriuria are respectively 73-95% and 44-100%. Firstly, we'll discuss a case of a 5-year-old girl. Secondly, we'll review the value of some diagnostic tests.

Methods

Case presentation: A 5-year-old girl presented to the hospital with fever and a rigid abdomen. Blood sample showed leukocytosis and a high C-reactive protein. Urinalysis was only positive for bacteria. Thoracic radiograph and abdominal ultrasound couldn't find a cause for the symptoms. To exclude an intra-abdominal abscess, computed tomography was performed and showed nephritis of the right kidney. Blood culture and urine culture remained sterile. Alpha 1-microglobulin in urine turned positive after a few days.

Results

Literature review: To diagnose a urinary tract infection; Leukocyte esterase has a high sensitivity, but a lower specificity. Nitrite has a lower sensitivity, but a high specificity. Pyuria has comparable results on manual and automated microscopy. However, bacteriuria has better results on manual microscopy. Urine culture has a sensitivity of 56-100% and a specificity of 71-100%. DMSA-scan is a good standard, but is rarely used in practice for the diagnosis. Alpha 1-microglobulin, a marker of tubular pathology, and procalcitonin, a marker of bacterial infections, could be useful to diagnose or exclude acute pyelonephritis.

Conclusion

Negative urinalysis, doesn't always exclude urinary tract infection. A combination of all diagnostic tests and especially the clinical presentation should be considered to exclude urinary tract infection with high probability.

P 141.

Post-transplant focal segmental glomerulosclerosis recurrences associated with bowel obstruction: a case report.

E. Surgun, K. Ismaili, E. Hennaut, N. Tram, B. Chiodini, X. Lolin, B. Adams

HUDERF-ULB

Introduction

Idiopathic nephrotic syndrome affects 1–3 per 100,000 children per year. Approximately 85% of cases show complete remission of proteinuria following steroid treatment, however the absence of complete remission within 4–6 weeks defines steroid-resistant nephrotic syndrome (SRNS). One of the principal histopathological entities encountered in SRNS is focal and segmental glomerulosclerosis (FSGS) which often leads to end-stage renal disease (ESRD). Post-transplant FSGS recurrence is frequent with variable rates from 6 to 59% and are challenging to treat. Therapeutic plasma exchange and Rituximab are two of the standards treatments validating the hypothesis of a circulating plasma factor associated to the high risk of recurrence in these patients. Suggested risk factors for recurrence include the age at onset of the disease, a rapid progression to ESRD and a history of previous relapses. The remission rate of pediatric recurrent FSGS has been reported to be as high as 70%, therefore FSGS diagnosis should not be a cause for postponing kidney transplantation.

Case report

A 12 years old female presented with abdominal pain and vomiting. She is known for SRNS with FSGS who progressed to terminal renal disease in 3 years and led to a kidney transplant from cadaveric donor. Six months after the transplant, the patient had a relapse of nephrotic syndrome concurrently with a bowel obstruction, treated laparoscopically. Therapeutic plasma exchange sessions and Rituximab injections led to the complete remission of the proteinuria.

On admission the patient showed persistent abdominal pain, lack of appetite and post-prandial vomiting. The radiological assessment was compatible with an acute ileus secondary to abdominal adhesions that was treated by laparoscopic surgery. Concomitantly, the patient showed an increased proteinuria indicating a relapse of nephrotic syndrome. This was the second FSGS recurrence on the transplanted kidney, also associated with a bowel obstruction that needed surgical treatment. The remission of the nephrotic syndrome was obtained with plasmapheresis and anti-CD20 treatment.

Conclusion

We described a patient who presented two post-transplant FSGS relapses, both associated to bowel obstruction and both episodes responded well to therapeutic plasma exchange and Rituximab injections. Although FSGS recurrences after renal transplant are well reported, the association with bowel obstruction has not been yet described to our knowledge.

P 142.**6-year-old boy with an unusual renal infection**

K. Van den Hende A-S Cornelis, E. Snauwaert, J. Vande Walle, A. Raes

*Universiteit Gent*Background

Acute focal bacterial nephritis (AFBN), also known as acute lobar nephritis/nephronia (ALN), is a localized bacterial infection of the kidney without renal tissue necrosis. It's a rare form of interstitial bacterial nephritis and involves typically one or more lobes. AFBN can be situated somewhere in the middle of the spectrum of urinary tract infections, varying from uncomplicated pyelonephritis to intrarenal abscess. There is still limited knowledge in children, but most patients with AFBN present with nonspecific conditions, such as fever, abdominal pain and vomiting. This lack of specific symptoms, in combination with aspecific laboratory findings, makes it difficult to recognize the disease.

Methods

A 6-year-old boy, with no relevant medical history, presented with high fever (> 40°C) since 1 day, abdominal pain and vomiting. He had no diarrhea or dysuria. On examination he had a sick appearance. The abdomen was soft, but painful in the right flank. Blood results showed increased inflammatory indexes with a leucocytosis with high neutrophils and an elevated CRP (195 mg/L). Urine sediment was negative, as were the chest X-ray and the ultrasound of the abdomen. IV Ceftriaxon and fluids were started. As there was no clinical improvement a CT scan was performed. This showed a bilateral pyelonephritis with incipient abscessation in the right kidney, and important subcapsular effusion. A few days later the urine culture showed an *E. faecalis* > 100.000/ml. Additional anamnesis learned that the patient is frequently constipated, drinks less than 500 ml a day and has holding maneuvers.

Results

The patient was treated further with IV Ceftriaxon for 10 days in total. After a couple of days there was a good clinical improvement. The IV antibiotic therapy was followed by oral Amoxicillin-clavulanic acid for another 2 weeks and Movicol. On imaging there was a normalization of the kidneys, without scars on short term.

Conclusion

This case illustrates a typical case of AFBN: A young child without urinary tract infections in the past, difficult to diagnose due to the absence of specific signs and symptoms, and with negative urinalysis (no WBC). In absence of imaging, the disease is probably underdiagnosed. It should be considered in children with fever and septic features (such as sick appearance, chills, vomiting...), increased inflammatory findings and/or abdominal pain, because this suggests a deep bacterial infection.

P 143.

Diagnostic challenges in a boy with tubulointerstitial nephritis and uveitis (TINU) syndrome: A case report and review of literature.

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Imeldaziekenhuis Bonheiden, UZ Leuven

We present a case report of an adolescent with TINU syndrome and a quick review of literature. Tubulointerstitial nephritis and uveitis syndrome is characterised by an association of uveitis and tubulointerstitial nephritis. It is an uncommon cause of acute kidney injury in children: only roughly 250 cases have been reported.

The condition is possibly underdiagnosed in children due to diagnostic challenges. Patients often present with nonspecific symptoms and the timing of eye symptoms varies in relation to the interstitial nephritis. The aetiology and pathogenesis of TINU remain largely unknown. No standard therapeutic protocols have been established to date.

We describe a case of an atypical presentation of TINU syndrome (TINU) at our clinic: A 15-year-old Armenian male presented to the Emergency Department with a one week history of left-sided abdominal pain. Laboratory findings were notable for elevated inflammatory markers and elevated serum creatinine. Urine analysis showed proteinuria and pyuria. A computed tomography of the abdomen was performed and showed signs of acute right sided pyelonephritis. Empiric antimicrobial therapy was initiated. Urine culture remained sterile and repeated blood analysis did not show any improvement of the inflammatory markers or renal function. This prompted an additional diagnostic work-up including a renal biopsy. The diagnosis of an interstitial nephritis was made. Upon follow-up, he presented with a unilateral red eye with blurred vision and photophobia. On ophthalmological exam, he had a right sided anterior uveitis. The diagnosis of TINU was made. Treatment including topical and oral corticosteroids was initiated.

This report illustrates that abnormal urinalysis or renal function impairment should always raise suspicion, but the diagnosis of TINU can be challenging if renal and ocular manifestations do not occur simultaneously.

P 144.**A puzzling case of recurrent upper urinary tract infections with urine leakage in a newborn**

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*Hôpitaux Iris Sud, ULB, HUDERF-ULB*Introduction

Congenital anomalies of the kidneys and urinary tract (CAKUT) are amongst the most common congenital anomalies, occurring in around 1% of births. Ureteral duplications, a common CAKUT, can come in both asymptomatic and symptomatic forms depending on whether the implantation of the ureter is atopic or not. It affects females more than males with a sex ratio of 2/1. We report here the case of a 3.5-month-old girl with an atopic ureter implantation who presented with 4 separate bacterial upper urinary tract infections (UTIs) in the space of 2 months.

Case

The girl is the first daughter of a non-consanguineous Vietnamese couple and was delivered via vaginal birth at term without incident. She had 4 pyelonephritis incidents from the age of 1 month and 11 days to the age of 3.5 months despite prompt, appropriate, treatment and ongoing antibioprophylaxis. The pathogens successively highlighted were multisensitive *Escherichia Coli* (2X), *Pseudomonas aeruginosa* resistant to trimethoprim and *Staphylococcus aureus* resistant to penicillin. Urine sampling for culture was obtained by catheterization at first, followed by urine bags when the urinary tract defect was diagnosed. The symptoms presented in each episode were fever, intense crying and a lack of appetite associated with urine leakage for the last 2 UTIs. Physical examination was unremarkable except for severe irritability. The blood test performed revealed a major inflammatory syndrome (WBC up to 28,680/µL and CRP level up to 200,24 mg/L) during each infection and the radiological assessment, including ultrasonography, voiding cystourethrography, dimercaptosuccinic acid renal cortical scintigraphy and magnetic resonance urography showed a right renal duplication with signs of obstructive uropathy of the upper pyelon drained by a megaureter ending in the vagina. No concomitant vesicoureteral reflux was observed. She responded well to the intravenous antibiotic therapy administered for each UTI and finally underwent surgery at the age of 3.5 months (upper right polar uretero-nephrectomy by laparoscopic route) with a favorable follow-up at 2 months post-op.

Discussion

According to the literature, duplication anomalies of the urinary collecting system are a common anatomical defect and are usually uncomplicated. In certain forms of major malformations, such as ectopic ureteral implantation, it can be associated with obstructive uropathy and with impaired renal function if untreated. One of the po

P 145.

Gastrointestinal protein loss in a child with polycystic kidney disease and SARS-CoV2 infection

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UZ Ghent

Background

Gastrointestinal (GI) involvement appears to be common in children with SARS-CoV-2 infection. GI protein loss is a very rare presentation. It is characterized by an excessive protein loss due to impaired integrity of either the intestinal or gastric mucosa. Until now, the main known infectious agents involved in protein losing enteropathy (PLE) are Salmonella, Shigella, Campylobacter, Giardia and Rotavirus and CMV for protein losing gastropathy (Ménétrier).

Case report

A 4-month old boy with polycystic kidney disease was admitted because of 1-day history of high fever and diarrhea. Physical examination revealed a sick child with tachypnea, distended abdomen with enlarged kidneys. In combination with high inflammatory parameters, broad spectrum antibiotics were started empirically and he was transferred to the paediatric intensive care unit (PICU). Additional ultrasonography and computed tomography excluded severe intraabdominal pathology, except for the polycystic kidneys. Blood cultures showed Staphylococcus Aureus. Other cultures were negative. Nasopharyngeal swab turned out to be positive for COVID-19. Although infectious parameters gradually normalized, the child developed a significant amount of ascites with respiratory decompensation, for which drainage was necessary during 5 days. Meanwhile, the boy had persistent mucous diarrhea. Ultrasonography showed no evidence for portal hypertension or thickened gastric fold. Notable laboratory findings included serious hypoalbuminemia and hypogammaglobulinemia. Urinalysis excluded proteinuria, liver function tests were normal. Although alpha-1-antitrypsin was not measured on stool, the absence of urinary losses with normal liver function and protein intake are suggestive for a GI protein loss. Endoscopy was considered too invasive and wouldn't induce therapeutic consequences. Supportive therapy, consisting of multiple albumin transfusions and intravenous immunoglobulins was started. Dietary modification with high protein content was recommended. He recovered gradually, with disappearance of the GI symptoms.

Conclusion

Our case describes a child with severe GI symptoms and significant hypoalbuminemia suggesting GI protein loss. COVID-19 infection is an evolving pediatric challenge and is still surprising clinicians with its varied systemic presentations. More epidemiological studies will be needed to reveal the role of SARS-CoV2 in PLE or protein losing gastropathy.

P 146.**Acute interstitial nephritis in an adolescent girl: to COVID-19 or not to COVID-19**

L. Peremans, A. De Jaeger, L. Dossche, J. Vande Walle, E. Snauwaert, A. Prytula, J. Dehoorne, A. Raes

*Universiteit Gent*Background/aims

Acute tubulointerstitial nephritis (AIN) is a rare cause of acute kidney injury (AKI) in children. Underlying causes are hypersensitivity to medication, infection-mediated AIN and auto-immune disorders. AIN may be a presentation of the single or multi-organ dysfunction associated with pediatric inflammatory multisystem syndrome (PIMS) that some children develop secondary to a COVID-19 infection.

Methods

Case description.

Results

A Caucasian girl of 15 years old with blank medical history was treated with Azithromycin and Non-steroidal anti-inflammatory drugs (NSAID) for a tonsillitis. PCR-test for COVID-19 was negative. Despite treatment the fever persisted, and her general condition deteriorated for which she was transferred to our pediatric intensive care unit. On admission she had fever (40°C), tachycardia (100bpm) and hypotension (99/43mmHg). Neurologically she appeared slow, but with normal neurological examination. Beside supportive therapy, IVIG was administered because of the tentative diagnosis of PIMS, based on the combination of persisting fever, hypotension, hyperinflammation (normal leukocytes with lymphopenia (470/ μ L), elevated CRP (262mg/dL), renal insufficiency (creatinine 3.20 mg/dL, urea 151 mg/dL) and a history of high-risk exposure to COVID-19. Acute interstitial nephritis was suggested to be the cause of AKI because of the presence of hematuria (876.4/ μ L), non-nephrotic proteinuria (0.59g/g creatinine), elevated alpha-1-microglobulin (33.06 mg/L) together with the finding of enlarged kidneys with a hypodense aspect and loss of corticomedullar differentiation on ultrasound.

A confirmatory kidney biopsy was not performed because of thrombocytopenia (58 x 103/ μ l). Early administration of steroids resulted in rapid improvement of the renal function and clinical condition. Results of the COVID-19 PCR-test and serology remained negative, and other microbial causes were excluded. Anti-streptolysin O titre, C3 and C4 levels and autoimmune screening were negative. Additional MRI of the brain turned out to be normal.

Conclusion

We present a case of an adolescent girl who presented with acute interstitial nephritis and a hyperinflammatory state mimicking PIMS. The etiology of AIN is probably multifactorial but will remain uncertain. Early recognition and prompt treatment are crucial to achieve optimal recovery. An international survey would be beneficial to document identical cases.

P 147.

Recurrent acute kidney injury with multifactorial aetiology in a 10 year old girl with a complex urogenital malformation.

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AZ Sint-Jan Brugge-Oostende, UZ Gent

Background

The causes of acute kidney injury (AKI) are commonly categorised in prerenal, renal and postrenal. Here we present a case of recurrent AKI with multifactorial aetiology.

Clinical case

A 10 year old girl with a complex congenital cloaca malformation presented with severe bladder-boweldysfunction and CKD (chronic kidney disease) stage 2 based on a dysplastic solitary right kidney. She previously received multiple reconstructive surgeries (recto-, ano- and vaginoplasty). Cystomanometry demonstrated a neurogenic bladder and a high grade vesico-ureteral reflux. Bowel irrigations and laxation (macrogol) together with clean intermittent catheterisation (CIC) and anticholinergic therapy resulted in improvement of the faecal and urinary incontinence, as well as the kidney function (CKD1). Despite correct CIC techniques she developed recurrent febrile urinary tract infections (UTI's), for which a ureterreimplantation was performed.

In the follow up, she presented with recurrent (4 times/5 months) severe postrenal AKI with VUJ obstruction (Creatinine Episode E1: 3.7 mg/dL; E2: 3.76 mg/dL; E3: 3.06mg/dL; E4: 5.3 mg/dL). The first 3 episodes were completely reversible with surgical release of obstruction (nephrostomy/double J stents). Additional MRI showed a small impression on the ureter on the level of the a iliaca communis and a bilateral hydrosalpinx (conservative follow-up). Although a cystoscopic incision of the right ureter ostium was performed, she developed a relapse of AKI with an important urinary bladder retention despite CIC, based on (nocturnal) polyuria. Indwelling catheterisation overnight in addition to CIC and anticholinergics resulted in a partial recovery of the kidney function (CKD3).

Discussion

This case represents recurrent AKI with multifactorial aetiology 1) VUJ obstruction 2) intermittent ureter obstruction by crossover of the a iliaca communis and bilateral hydrosalpinx 3) pre-existing CKD based on renal dysplasia, associated with (nocturnal) polyuria, secondary to deficient concentration capacity 4) high pressure neurogenic bladder, despite treatment with CIC and anticholinergics, and small bladder volume compared with diuresis-volumes 5) decompensation of renal function by severe constipation and/or UTI's.

Conclusions

Children with complex urogenital malformations and pre-existing grades of CKD, are at high risk of developing episodes of AKI, that can lead to severe CKD. A multidisciplinary specialised follow up is necessary.

P 148.**10-year follow-up of the teratogenic effects and neurocognitive development after prenatal ACE-Inhibitor exposure: a case report**

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*UGent, UZ Gent, AZ Sint-Jan Campus Brugge*Background/Aims

Angiotensin-converting enzyme inhibitors (ACE-inhibitors) are among the most frequently prescribed antihypertensive drugs. Ingestion during pregnancy has a known increased risk of fetopathy, with well-described congenital malformations. Until now, little is known about the long-term outcome and the impact on a child's life. The objective of this case report is to analyze long term outcome following prenatal exposure to ACE-inhibitors and describe the subsequent impact.

Case report

A 10-year-old child has been in follow-up for almost a decade. During his fetal period, his mother suffered malignant hypertension and had to continue her prescribed ACE-inhibitors despite known teratogenic effects. The mother gave birth to a 33-week-old infant through a semi-urgent caesarean section. During the neonatal phase, a severe kidney impairment due to bilateral hypodysplasia was observed. After initial need for dialysis, renal function recovered partially, but caused the need for dialysis and kidney transplantation at the age of 4-years. Aside from renal impairment, the child was born with hypocalvaria (i.e. incompletely formed skull bones) as well as severe abnormalities of the central nervous system, resulting in motor impairment (including a right-sided paresis) as well as cognitive retardation.

Conclusion

Renal hypoplasia is well known in newborns prenatally exposed to ACE-inhibition, but there is a gap in knowledge on other organs and long-term prognosis. This case documents that other organs are equally involved, and that long-term neurocognitive development is compromised.

P 149.

Mystery diagnosis: Bartter syndrome as a rare cause of failure to thrive

A. Matthys, L. Gheuens, S. Karamaria, A. Prytuła, E. Snauwaert, L. Dossche, J. Vande Walle, J. Dehoorne, A. Raes

UZ Gent

Bartter syndrome is a rare autosomal recessive tubulopathy. There are 4 types differing in severity and age of onset. Type I and II are the most severe presenting with polyhydramnios, prematurity and neonatal with hypokalemia, metabolic alkalosis, polyuria and hypercalciuria.

Case presentation

A 9 month old girl was transferred to our hospital because of hypokalemia and hypernatremia.

She was born vaginally after a 34 weeks pregnancy complicated by polyhydramnios with multiple evacuation need. She had good Apgar scores and a birth weight of 1750 g (SD-5.7). She was admitted to neonatology for prematurity and dysmaturity. She received nasal CPAP for one day and phototherapy. No further problems occurred and she was discharged after 36 days.

During the first months of life she was drinking well and followed her own growth. From the age of 4 months old feeding difficulties occurred and failure to thrive manifested. Her development was conforming her age. Initial examinations showed no abnormalities.

At 9 months she was admitted because of excessive vomiting. She was mildly dehydrated and her weight stagnated on 5 kg for 4 months. There was no history of polyuria. Due to hypernatremia (156 mmol/L) and hypokalemia (2.58 mmol/L) not responsive to IV fluid, she was transferred to our department.

We observed a girl in good clinical condition with pronounced frontal bossing, small hands and a wide nose bridge. The combination of polyhydramnios, hypokalemic alkalosis (K 2mmol/L), nephrocalcinosis and polyuria after rehydration pointed us to a tubulopathy, most likely type Bartter. Genetic analysis was initiated.

Under potassium supplementation, indomethacin and compensation of fluid losses the electrolyte disturbance were normalized. She was discharged with hypercaloric diet and additional fluid through gastrostomy. She remains in good condition and continues to gain weight.

Conclusion

Bartter types I and II are usually severe disorders that can be caused by various genetic defects, reducing the activity of one of several electrolyte transporters in the thick ascending limb. Even when not all clinical symptoms are present, it should be considered by failure to thrive with a history of polyhydramnios.

Posters**P 150.****Leptospirosis and Hantavirus induced acute tubulointerstitial nephritis in children: case series**

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UZ Gent, Universiteit Gent

Background

We present two children with acute tubulointerstitial nephritis (ATIN) triggered by leptospirosis and Hanta virus infection. The role of corticosteroids in the management of ATIN is controversial.

Case series

A 12 year old boy and 10 year old girl were admitted with elevated serum creatinine, thrombocytopenia and jaundice. There was no oliguria or hypertension. Urine analysis revealed tubular proteinuria. Kidney biopsy was performed in one patient and showed tubulointerstitial inflammation with mild mesangial proliferation. As per our institutional protocol systemic corticosteroids were administered in view of deteriorating renal function with respective serum creatinine values of 5.2 and 4.1 mg/dl. Both children exhibited an excellent clinical and biochemical response to treatment. Further investigations showed positive IgG titer against *Leptospira* in the first and positive IgM and IgG titer against Hanta virus in the second case.

Discussion

Due to the low incidence, leptospirosis and Hanta virus (20/year and 10-100/year in Belgium) may be easily overlooked.

Conclusion

Leptospirosis and Hanta virus should be considered as potential causes of ATIN in children. Both our cases showed an excellent response to corticoid treatment, suggesting an underlying common immunological mechanism.

P 151.

Frequently Relapsing Nephrotic Syndrome in an -11-year old male with heterozygosity for two variants in NPHS1 gene.

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Background

The NPHS1 gene encodes nephrin, a component protein for the renal glomerulus slit diaphragm, which prevents protein from being passed in the urine. Mutations in the NPHS1 gene are found in CNS (-Finnish type) and later in SRNS.

Methods

We report on an -11- year old Surinamese-Creole male patient who presented with steroid dependent frequently relapsing nephrotic syndrome. He was the 3rd child of non-consanguineous parents. Both parents and other 4 siblings have no nephrological symptoms. Onset of the nephrotic syndrome at the age of 2 y 11/12. He was first treated with human albumin infusions, diuretics and steroids (60 mg.m² QD for 6 weeks, 40 mg/m² for 4 weeks followed by taper). After a steroid free period of 3 months , he developed 4 flares after a common viral infection over a 6-month period, for which he was started on a cyclosporine. He remained without relapses for 2 years with cyclosporine and a low dose methylprednisone (5 mg AD), but started back with frequent relapses on tapering off this medication. Kidney biopsy: Minimal Change Disease (MCD). Few years after the initial diagnosis of nephrotic syndrome, rituximab was started (375 mg/m² 4 files with an interval of 1 week). Remission for 14 months. Afterwards, 2 flare-ups occurred after an infection. Patient was treated with steroids and cyclosporine was restarted. There was a complete remission for 6 months. A second rituximab treatment was given at the age of 10 years with subsequent tapering of steroids and retention of cyclosporine. Currently the patient is in remission. GFR: (Schwartz formula: 101,9 ml/min/1.73 m²). Lab: No hematuria, C3,C4: normal.

Results

A heterozygosity was found for 2 variants in NPHS1: c.2734G>A (p.(Ala912Thr)) and c.1747G>A (p.(Gluc583Lys)). Conclusion: The associations of these two rare variants has been previously reported as novel and potentially pathogenic by Bierzynska A. et al. Kidney International (2017);91,937-947. Patient 36 in their nephrotic syndrome cohort was a male of mixed ethnicity (black&white) whose nephrotic syndrome started at 0.14 years old. No consanguinity, sporadic case steroid sensitive but frequently relapsing, first biopsy mesangial proliferative changes (MPC), CKD1, no transplantation, no extra-renal phenotype and follow-up of 17.73 years. Points of comparison are the mixed ethnicity, frequently relapsing steroid sensitive, CKD1,male and 8 years follow-up. Learning point: Repeat kidney biopsy for possible detection of MPC.

P 152.**A neonate with an abdominal mass**

S. Karamaria, A. Raes, J. Dehoorne, A. Prytula, J. Vande Walle

*UZ Gent*Introduction

Neonatal adrenal hemorrhage (NAH) is a relatively uncommon condition (0.2-0.5%). The most common predisposing factors are perinatal asphyxia/hypoxia, birth trauma, prolonged labor, sepsis, macrosomia, hemorrhagic disorders. However spontaneous occurrence in otherwise normal neonates is also described. The clinical presentation may vary between asymptomatic, flank mass, anemia, persistent jaundice, scrotal hematoma, adrenal insufficiency and shock.

Case report

A premature boy was born after an uncomplicated twin pregnancy of 34+4weeks to a gravida 1 para 1 woman. He was vaginally delivered with a birth weight of 2100g and had Apgar scores of 8 and 9 at 1 and 5 minutes respectively. The amniotic water contained meconium but no extra measures of resuscitation were needed.

The first clinical examination revealed an abdominal mass in the left flank and therefore he was transferred to the neonatal intensive care unit for further investigations.

Intravenous antibiotics because of slight elevated CRP and phototherapy for indirect hyperbilirubinemia were started. During the first 24h the neonate developed hematuria. Creatinine was also slightly elevated during the first 72h but was thereafter normalized.

An ultrasound (U/S) with doppler showed an enlarged left kidney, with no dilation of the pyelum and a normal corticomedullary differentiation; in the center a vascular knot suggestive for arteriovenous malformation or fistula was visualized (no high or pulse flow). The renal vessels and the right kidney were normal. Additional exploration with MRI on day 5 in order to exclude malignancy was performed and showed a structure filled with fluid above the left kidney, subsequent to adrenal bleeding and fluid retention in the calices. The neonate was released after 7 days of observation and was further followed up in the outpatient clinic.

U/S performed on a regular basis revealed initially a dilatation of the calyces of the left kidney, followed by gradually shrinking of the kidney. At the age of 1 month a DMSA scan showed a completely non-functional left kidney. At the age of 5 months the left kidney was not visualized on U/S any more. The right kidney was enlarged to compensate.

Conclusion

NAH should be suspected in neonates even in the absence of important risk factors. Its clinical manifestations may vary widely. Once malignancy is excluded Doppler U/S is reliable for follow up. Conservative management is sufficient in most of the cases.

P 153.

Acceptance & Commitment Therapy with parents of children with chronic kidney disease

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UZ Gent, AZ Sint-Jan, Universiteit Gent

Background/Aims

The psychological impact of chronic kidney disease (CKD) goes beyond the child and affects the entire family. Compared to parents of healthy children, a higher presence of stress, anxiety and depression symptoms can be seen. The aim of this study is to set up an Acceptance and Commitment Therapy (ACT) group intervention with parents of CKD patients and evaluate the effectiveness.

Methods

Sixteen parents joined our ACT intervention which was organized during one whole day. ACT is a third generation behavioural therapy program and is known to be effective in the treatment of depressive and anxiety symptoms in parents of children with chronic diseases in general. To evaluate the impact of the intervention on psychological wellbeing and ACT constructs, parents completed 4 questionnaires before and after the intervention: Hospital Anxiety and Depression Scale (HADS) for anxiety and depression symptoms, Flexibility Index (FIT60) and Acceptance and Action Questionnaire-II (AAQ-II) for psychological flexibility and acceptance, and World Health Organization Quality of Life Questionnaire-BRIEF (WHOQoL-BRIEF) for quality of life. Additionally parents completed a questionnaire regarding their socio-demographic situation.

Results

Because the ACT intervention was only recently organized, results are not available yet. However, preliminary analysis indicate a benefit of the intervention. Secondary analysis of full data will be available at the meeting in march 2020.

Conclusion

Cf. Results

Posters**P 154.****Clinical characteristics of Galloway-Mowat syndrome and mutations in the TPRKB gene**

L. Peremans, B. Callewaert, A. Raes, J. Vande Walle, J. Dehoorne, L. Dossche, E. Snauwaert, A. Prytula

*Universiteit Gent*Background/aims

Galloway-Mowat syndrome (GAMOS) is a rare genetic entity characterized by nephrotic syndrome with microcephaly and neurological involvement. Counseling parents concerning clinical course and prognosis is difficult because of the heterogeneity. We describe the relatively mild nephrological course in two patients with mutations in the TPRKB gene.

Methods

Case description and literature review.

Results

Patient 1 is a male born from Syrian consanguineous parents. Nephrotic syndrome was diagnosed at the age of 6 years. He had a history of developmental delay and oculomotor dyspraxia. He had a short stature and microcephaly. Neurologic evaluation revealed a mixed spastic dystonic disorder. Serum creatinine was within reference range, but there was hypoalbuminemia and nephrotic range proteinuria. Genetic study showed a heterozygous missense variant in exon 3/5 of TPRKB gene.

Patient 2 is a male born from Caucasian consanguineous parents. He was known with developmental delay, and nephrotic syndrome was first diagnosed at the age of 4 years. Clinically he had mild peripheral oedema and microcephaly. Neurologic evaluation showed a mixed spastic dystonic disorder. Kidney biopsy showed no abnormalities, but genetic study identified a homozygous c.446A-G transition in exon 5 of the TPRKB gene.

Both patients were treated with lisinopril and supportive care, in view of resistance to corticosteroids and calcineurin inhibitors. They did not require albumin infusions. Both have a stable kidney function until now.

Conclusion

In both patients we found a mutation in the TPRKB gene – one heterozygous and one homozygous – encoding a subunit of the KEOPS complex.

Individuals with mutations in the KEOPS genes usually present with nephrotic syndrome of early onset, resulting in early end-stage renal failure and death. Only one other patient has been reported with a mutation in the TPRKB gene, and this patient developed end-stage renal disease and died at the age of 6.8 years. Our two patients presented with steroid- and calcineurin inhibitor resistant nephrotic syndrome, only at the age of 4 and 6 years, with a preserved kidney function until now. Neurologically we saw a mixed spastic dystonic disorder with periventricular leukomalacia on imaging.

Our case series shows that some children with GAMOS display a relatively mild nephrological course. Further research is necessary to establish genotype-phenotype associations.

P 155.

Robotic-assisted kidney transplantation in children: initial experience in a tertiary centre

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UZ Gent

Introduction

Kidney transplantation (KT) is the gold-standard treatment for end-stage renal disease (ESRD) in children. Robot-assisted kidney transplantation (RAKT) in adults is becoming increasingly common with potentially improved morbidity compared to open KT. Our objective was to evaluate feasibility and outcomes of RAKT in children.

Patients & Methods

An 8-years-old boy with ESRD received a kidney transplant from his mother. Simultaneously in two operation theatres, the boy underwent single-port (GelPOINT®) right laparoscopic nephroureterectomy (LNU) and his mother underwent robot-assisted left donor nephrectomy (RADN). Two full surgical teams were operating at the same time. Subsequently, the boy underwent RAKT, introducing the graft through the GelPOINT®.

Results

Total operative time for LNU, RADN and RAKT was 180, 140 and 195 min respectively, with warm, cold and rewarming ischemia times 1.5, 200 and 47 min respectively. Blood loss was 300, 20 and 50 cc respectively. No intraoperative complications were noted. Convalescence of both donor and recipient was uneventful, with good kidney function at one-year follow-up.

Conclusion

RAKT in children is technically feasible and safe, resulting in excellent graft function. Concomitant nephrectomy can be done laparoscopically through the single-site GelPOINT®. An experienced RAKT team with the full support of pediatric nephrologists is mandatory.

Posters**P 156.****Initial screening for bedwetting: the use of questionnaires and voiding diaries. First results from a National Belgian study**

S. Karamaria, N. Ranguelov, P. Hansen, V. De Boe, P. Verleyen, J. Vande Walle, L. Dossche, A. Bael

*UZ Gent, Universiteit Gent, Cliniques Universitaires St-Luc, KUL, CHU Tivoli, UZ Brussel, AZ Groeninge, ZNA Koningin Paola Kinderziekenhuis, Universiteit Antwerpen*Background

International guidelines have a consensus that stratification of nocturnal enuresis (NE) into non-monosymptomatic (NMNE) and monosymptomatic (MNE) is mandatory at intake to optimize therapeutic approach. This stratification is based on clinical parameters (presence or absence of Lower Urinary Tract Symptoms (LUTS) respectively). To identify clinical parameters a checklist (Clinical Management Tool (CMT)) and/or voiding diaries based on home recordings can be used. However, these recordings can be time consuming and difficult for the family. Moreover, the added value to the CMT, especially in treatment naïve patients, is rather expert opinion than evidence based

Methods

The aim of this study run in 7 Belgian Hospitals, was to document in treatment naïve NE patients >5 years: 1) The prevalence of MNE vs NMNE 2) the added value and correlation of CMT and/or diary in differentiating NE

Two study visits were scheduled: At visit 1 CMT was obtained, after a thorough medical history and basic assessments. If daytime incontinence and/or LUTS were identified, the diagnosis was NMNE. After the 1st study visit, a 2day voiding diary (fluid intake, voiding volumes, incontinence) was registered at home. During the second study visit, this diary was evaluated; if the micturition frequency was >8 or <3 and/or there was daytime incontinence, the diagnosis was NMNE

Results

In total 109 children were included, of which 19 were lost in follow up. Mean age was 7,7 (± 2); 62 were boys (68,9%) and 27 were girls (30%). 68 (75, 6%) were included at a non-University center. Based on the CMT 18 children were diagnosed as MNE (20%) and 72 children as NMNE (80%). Based on the diary 32 children were diagnosed as MNE (35,6%) and 58 children as NMNE (64,4%). 66 children (73%) had the same diagnosis with both methods. From the parameters present in both methods incontinence was significantly different ($p=0,016$). Statistical significant differences of MNE vs NMNE based on the diary were daytime incontinence, frequency, urge, interrupted stream and drinking at night; based on the diary were urge, daytime incontinence, maximum and average voided volume

Conclusion

NMNE is more frequent than MNE in treatment naïve patients. CMT alone versus CMT + diary had a different sensitivity and specificity of identifying LUTS : in absence of validation of the importance by a therapeutic trial outcome we state that we can only consider patients as MNE when and CMT and diary do not demonstrate LUTS

P 157. P**Prevalence and resistance pattern of microorganisms in pediatric UTI in the period 2000-2019. A retrospective analysis in UZLeuven**

Y. Dejonckheere, N. Knops

*UZ Leuven*Background

Urinary tract infections (UTI) are common bacterial infections in children with an incidence of 19 episodes per 1000 children per year. If untreated, UTI's can cause severe long-term complications like hypertension and renal scarring. Because of potential changing trends in microbial characteristics of UTIs in children and increasing antimicrobial resistance against the currently used antibiotics, a retrospective analysis of the prevalence and resistance patterns of uropathogens in our hospital was performed to get more insights into whether our current protocol is still sufficient.

Methods

Retrospective data were collected from admissions for pediatric UTI from 2000 to 2019 with a query search in the digital hospital information system (KWS). Search terms were: "urinary tract infection", "UTI", "urosepsis", "pyelonephritis", "cystitis". Statistical analysis was performed by statistical program SPSS for Windows.

Results

There were 1010 admissions with a boy-girl ratio 1:1,55. Median age at diagnosis was 1.1 years. A secondary underlying condition (VUR, neurogenic bladder) was present in 31% of patients. E. Coli was the most prominent microorganism, namely in 76% of cases. However, there is a trend toward increased presence of atypical organisms like Pseudomonas and Enterococci (last 5 years 37% non-E. Coli versus 23.6% between 2000 and 2004 ($p=0.006$)). In our population, the presence of ESBL is 5.9% and increasing during the last 8 years. The last 5 years, 33% of pathogens proved resistant to Amoxicillin Clavulanic acid, in contrast to 8.7% between 2000 and 2004 ($p<0.0001$).

Conclusion

This retrospective study of a large population of children admitted with UTI in a center for tertiary care shows that E. Coli still is the most important cause of UTI, but we have to take into account an increasing risks for non E. Coli pathogens together with an increasing prevalence of antibiotic resistance, especially ESBL producing microorganisms. All these factors lead to the implication that we have to be extra careful in our choice of antibiotics and that Amoxicillin Clavulanic acid may risk being an inferior choice of empiric therapy in the future.

P 189.**Combined hyponatremia with hyperkalemia in young infants: a diagnostic challenge.**

B. Meertens, A. Raes

UZ Gent

Hyponatremia is the most common electrolyte disorder in hospitalized children. The underlying mechanisms are often poorly understood, and this complicates its management, particularly in neonates. In this presentation I will discuss the diagnostic challenge of this electrolyte disorder with a specific focus on combined hyponatremia with hyperkalemia in young infants. The aim is to provide a clear overview of the diagnostic steps needed to define the specific origin of the electrolyte disturbance in order to optimize treatment.

The results from an extensive literature study of the etiology of combined hyponatremia and hyperkalemia in young children that were hospitalized will serve as background to a clinical case, which will be presented in detail.

It will be shown that the etiology of hyponatremia in young infants is diverse and often multifactorial. Neonates are especially vulnerable to hyponatremia due to immaturity of the renal tubules, which causes natriuresis. As a consequence of relative aldosterone resistance in early life, the collecting duct cannot compensate these losses. When there is a specific trigger that increases aldosterone resistance (e.g. pyelonephritis or obstructive uropathy) or decreases aldosterone production, this further destabilizes the fragile (ad)renal equilibrium and causes major electrolyte disturbances. Hyperkalemia and hyponatremia itself can be life-threatening but most deaths are related to the severe underlying condition that leads to an electrolyte disturbance. Depending on the report, there is a 12-14% mortality rate in literature. Hyponatremia is also used as one of the factors to determine the Score for Neonatal Acute Physiology-Perinatal Extension (SNAP-PE).

By way of a conclusion, it will be argued that hyponatremia can be easily overlooked in lab reports. But especially when combined with hyperkalemia, it is an anomaly that always requires further investigation and careful monitoring to assess whether or not treatment is necessary.

LO 3.**Four novel variants in the MKRN3 gene causing central precocious puberty**

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Hôpital Universitaire des Enfants Reine Fabiola, Université Libre de Bruxelles, Centre Hospitalier Universitaire du Sart-Tilman, Université de Liège

Background

Idiopathic central precocious puberty (iCPP) is defined by the premature re-activation of the hypothalamic-pituitary-gonadal axis with normal MRI of the central nervous system, causing the development of secondary sex characteristics prior to the age of 8 years in girls and 9 years in boys. In 2013, Abreu et al identified loss-of function mutation in the MKRN3 gene of fifteen patients from five families with iCPP, highlighting the implication of this maternally imprinted gene in this still poorly understood condition. Since this study, other mutations have been described and now represent the most common genetic cause of iCPP.

Objective

The objective of the study was to document the clinical course of puberty in four unrelated girls harbouring pathogenic MKRN3 variants.

Design and participants: This is an observational case series study of patients with iCPP followed in our center at the Hôpital Universitaire des Enfants Reine Fabiola.

Results

We identified six different variants in the MKRN3 gene of five unrelated girls with CPP. One was inherited from the mother and classified as probably non-pathogenic, and the other five were predicted to be deleterious by in silico analysis. Four of the five pathogenic variants were novel: two of them were missense mutations, one was a nonsense mutation and one was a frameshift mutation. Segregation analysis was done in the three girls with a family history of CPP. In all cases, the mutation was inherited from the father. The two young brothers of one of the patients were also carriers but asymptomatic given their young age. The index cases all had a very frank peak LH response to the GnRH test and a very rapid pubertal development.

Conclusion

We have identified four novel MKRN3 mutations in children with CPP. One of them had no known family history of CPP but a very early and rapid pubertal onset. An MKRN3 defect should be considered in all patient with CPP at a young age or with a lack of information regarding pubertal timing in relatives and no maternal history. The family study of a patient also allowed us to detect a MKRN3 mutation in her two young brothers, highlighting the value of screening asymptomatic patients and provide regular clinical follow-up with early treatment if necessary.

Long Oral Presentation**LO 4.****Partial CRISPR/Cas9 IL1R1 & IFNGR1 Knock-Down Improves β -cell Survival And Function Under Cytokine-Induced Inflammation**C. Daems, J. Vanderroost, E. Sokal, P. Lysy*UCL Saint-Luc*Background/Aims

Type 1 diabetes (T1D) is a disease characterized by the autoimmune destruction of pancreatic β cells. This destruction is mediated by lymphocytes T helper and cytotoxic, and by the action of the pro-inflammatory cytokines IL1 β and IFN γ inside the islets of Langerhans. We propose a new approach to alleviate islet inflammation by targeting pro-inflammatory cytokine receptors. Our hypothesis is that the downregulation of inflammatory pathways may improve β cell survival in the context of inflammation after T1D onset.

Methods

We knocked-down IL1R1 or IFNGR1 receptors in the MIN6 β -cell line by using the CRISPR/Cas9 gene editing system.

Results

The knockdown efficiency was evaluated by immunostaining and ranged from 12 to 40%. Naive MIN6 or CRISPR-knockeddown MIN6 cells were treated with IL1 β or IFN γ during 48 h at various concentrations (5 or 10 ng/mL). Cell viability of CRISPR-IFNGR1 and CRISPR-IL1R1 cell lines was improved after cytokine exposure compared to naive MIN6 (117 ± 16 vs $84 \pm 19\%$; $p=0.015$ and 134 ± 20 vs $71 \pm 4\%$; $p=0.016$). The assessment of insulin secretion capacities of CRISPR-IFNGR1 and CRISPR-IL1R1 cells showed higher secretion rates (1.24 ± 0.21 vs 0.35 ± 0.14 I.A.; $p=0.006$ and 0.91 ± 0.22 vs 0.32 ± 0.09 ; $p=0.014$), after cytokine treatment, as compared to naive MIN6. Gene expression of the pro-apoptotic receptor Fas was decreased inside the CRISPR-MIN6 cell lines and the expression of the pro-inflammatory cytokine Il6 gene was decreased inside the CRISPR-IL1R1 cell line, as compared to MIN6 controls. Similarly, gene expression of ER stress markers Atf4 and Chop decreased inside the CRISPR-IL1R1 and CRISPR-IFNGR1 cell lines, respectively, as compared to controls. Our results show that the targeting of IL1R1 or IFNGR1 could protect pancreatic β cells from the inflammatory attack found in T1D by decreasing apoptosis, inflammation and ER stress.

Conclusion

Our results show the feasibility of the CRISPR technique to protect β cells and require further in vivo analyses to fully address the potential of this system to be translated into clinical research protocols. The possibility of a translational perspective of our knockdown system is suggested by the ongoing clinical trial using the CRISPR/Cas9 system to evaluate the safety of PD-1 knockout engineered T cells in treating metastatic non-small cell lung cancer (NCT02793856).

SO 18.

Long-term adiposity outcomes of children born after maternal bariatric surgery

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UZ Brussel – VUB, UZ Leuven, Universiteit Antwerpen, Danone Research

Background

Bariatric surgery before pregnancy results in improved neonatal outcomes for the mother and child. However, long-term data on the consequences at childhood age are currently lacking.

Methods

EFFECTOR is a cross-sectional, long-term follow-up study of children (aged 4 to 11 years) born from mothers that underwent bariatric surgery (BS) before pregnancy (n= 36), overweight/obese controls (OW/OB) matched on pre-pregnancy BMI (n=36) and normal weight controls (NL) (n=35). We performed prospective collection of adiposity data.

Results

Neonates born after bariatric surgery had the lowest weight SD score at birth (-0.26 vs. 0.34 in OW/OB and -0.09 in NL; p=0.04) and smallest height SD score at birth (-0.18 vs. 0.36 in OW/OB and 0.04 in NL; p=0.04). At childhood age, the children after bariatric surgery presented with the highest weight and BMI SD scores (0.70 vs. 0.14 in OW/OB and -0.09 in NL; p=0.006 and 0.47 vs. -0.02 in OW/OB and -0.42 in NL; p=0.01). A similar difference in excess fat percentage and waist SD score was seen (5.7 vs. 1.4 in OW/OB and -0.1 in NL; p<0.001 and 0.61 vs. 0.16 in OW/OB and -0.15 in NL; p=0.04).

Conclusions

Maternal bariatric surgery does not appear to protect the offspring for childhood overweight and obesity.

Short Oral Presentation**SO 22.****Long-term endocrine and reproductive outcome of adolescent and young adult men born appropriate for gestational age with non-syndromic hypospadias**

L. Tack, A-F Spinoit, S. Riedl, A. Springer, U. Tonnhofer, M. Hiess, J. Weninger, A. Mahmoud, K. Tilleman, E. Van Laecke, P. Hoebeke, A. Juul, J.C. Albrethsen, E. De Baere, H. Verdin, M. Cools

Universiteit Gent, UZ Gent, Medical University of Vienna, St Anna Children's Hospital, Medical University of Vienna, University of Copenhagen, Rigshospitalet

Background/Aims

Hypospadias affects approximately one in 200 newborn males. Based on the hypothesis of testicular dysgenesis syndrome, these boys are believed to be at risk of having testicular dysfunction. However, little evidence currently supports this hypothesis. In this study we aimed at identifying the endocrine and reproductive outcome of men born appropriate for gestational age and with non-syndromic hypospadias as compared to healthy male peers.

Methods

Men born AGA with hypospadias (n=167) and controls (n=50) aged 16-21 years, were recruited in Ghent University Hospital and Medical University Vienna for cross-sectional endocrine and reproductive screening. Assessments included genital examination, testicular ultrasound, morning blood sampling for hormone assays (gonadotropins, total and free testosterone, insulin-like factor 3 and inhibin B). In addition, targeted next generation sequencing for genes (n=241) involved in gonadal development and spermatogenesis was performed in those with a poor outcome.

Results

None of the participants had experienced problems regarding pubertal onset and progression. Stratification based on severity of undervirilisation revealed higher gonadotropin and lower inhibin B levels only in complex hypospadias, but not isolated hypospadias. No differences were seen in androgen levels. Insulin-like factor 3 levels were higher in hypospadias cases compared to controls ($p=0.004$). Oligo- or azoospermia (<15.106 spermatozoa/mL) was found in 19/149 (12.7%) cases and 2/50 (4%) controls. High sub- and infertility rates were found in complex hypospadias (30%). However, very few had inhibin B or FSH levels above the clinical thresholds. No (likely) pathogenic variants were found in investigated genes.

Conclusion

None of the participants encountered problems regarding pubertal onset and progression, nor in steroidogenesis. However, semen production is a point of concern in men with hypospadias, especially in complex hypospadias. As this exam is non-invasive and yields low costs, routine semen analysis should be routinely offered at the end of puberty to all boys born with hypospadias. Mono-genetic mechanisms are unlikely to cause hypospadias.

SO 23.

Eating habits of children born after maternal bariatric surgery

K. Van De Maele, C. De Geyter, Y. Vandenplas, R. Devlieger, I. Gies

UZ Brussel, UZ Leuven

Introduction

Mothers who underwent bariatric surgery before pregnancy have worrisome eating habits and a poor diet quality. Little is known about the eating habits of their offspring. Since parental eating behaviors have passed on to their children and the rates of obesity in these children are high, their eating habits might be as bad as their mother's.

Methods

EFFECTOR is a cross-sectional, long-term follow-up study of children (aged 4 to 11 years) born from mothers that underwent bariatric surgery (BS) before pregnancy (n= 36), overweight/obese controls (OW/OB) (n=71) and normal weight controls (NL) (n=35). Anthropometric and eating habits data (47-item Food Frequency Questionnaire (FFQ)) were collected prospectively.

Results

The prevalence of overweight and obesity is highest in the children of the BS group (38.9% vs 15.5% in OW/OB and 5.7% in NL; $p=0.004$). Overall breakfast skipping prevalence is 9.2% in the entire study population. Most of the children skipping meals (breakfast, lunch or dinner) belong to BS group. Children of the BS group consume most artificial sweetened beverages (low calorie zero and light beverages) (38.9% vs 8.6% in NL and 26.8% in OW/OB; $p=0.01$). Daily use of artificial sweetened beverages is noted in 19.4% of BS children (vs 13% in OW/OB and none in NL).

Conclusion

Poor maternal eating habits after bariatric surgery are passed on to their children. These children are particularly at risk to skip meals and consume large amounts of artificial sweetened beverages (low calorie zero and light beverages).

Short Oral Presentation**SO 25.****Individuals with NR5A1 (SF1) variants and typical or diverse sex development are at high risk of hyposplenism**

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UZ Gent, Universiteit Gent, Erasmus MC Rotterdam, Hadassah Hebrew University Medical Center

Background

Heterozygous mutations in Steroidogenic Factor1 (SF1, NR5A1) cause 46,XY and 46,XX disorders of sex development (DSD), azoospermia, and primary ovarian insufficiency. NR5A1 is also involved in embryonic spleen development, by transactivation of T-cell Leukemia Homeobox 1 (TLX1). Hypo- or asplenism have occasionally been observed in DSD patients with NR5A1 mutations.

Aims

We aimed to determine the prevalence and functional consequences of splenic defects associated with NR5A1 variation. We assessed spleen anatomy and function in 22 DSD patients (17 heterozygous, 5 homozygous; karyotypes: 46,XY in 17, 46,XX in 5) and 5 asymptomatic carriers from 18 families, harboring 14 different NR5A1 variants. Spleen function was investigated by total blood and thrombocyte counts, % pitted red blood cells (PRBC), Howell-Jolly Bodies (HJB) and relative counts of spleen-dependent non-switched memory B cells. Spleen anatomy was studied by ultrasound, the functional impact of 7/14 NR5A1 variants on TLX1 transactivation capacity by luciferase assays.

Results

None of the participants had severe bacterial infections in the past. 59% of patients and 40% of carriers had thrombocytosis. Increased % PRBC was not observed; HJB were present in 5/19 (26%) tested patients and 0/5 carriers. Preliminary analysis showed significantly decreased non-switched memory B-cells in NR5A1 patients and carriers as a group in comparison with controls ($p=0.006$), with no differences in non-switched memory B-cells between patients and carriers (ns). Anatomical spleen abnormalities were observed in 14/22 (63%) patients and 2/5 (40%) carriers, spleen hypoplasia was most frequent. In vitro transactivation of TLX1 by NR5A1 mutants relative to WT was significantly decreased for 4/7 tested variants, increased for 1 and not different from WT for 2 variants.

Conclusions

Anatomical and/or functional hypo- or asplenism is suggested by at least one test in 77% of patients and 60% of carriers with NR5A1 mutations. In vitro functional tests cannot accurately predict in vivo splenic function. None of the tests is by itself sufficiently sensitive or specific for functional hypo- or asplenism, but applying a combination of techniques is complex and expensive in the clinical setting. Given the functional and anatomical hyposplenism shown here and its known risks, we suggest splenic surveillance and possibly prophylactic measures in all patients with NR5A1 variants and their family member carriers.

PW 29.

Long-term endocrine and reproductive outcome of adolescent and young adult men born small for gestational age with non-syndromic hypospadias

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Universiteit Gent, UZ Gent, Medical University of Vienna, University of Copenhagen, Rigshospitalet

Background/Aims

Children born SGA are known to have an increased risk for multiple conditions which can arise at all ages. A strong association between low birth weight or length and undervirilisation, in particular hypospadias in males is well-known. However, few studies have focused on outcomes of this subgroup of boys born SGA and undervirilized. In this study we aimed at studying the testicular function with regards to steroidogenesis and spermatogenesis in adolescent and young adult boys born SGA with hypospadias and tried to identify the underlying genetic mechanisms associated with a poor outcome.

Methods

Cases (n=26) and controls (n=50) aged 16-21 years, were recruited in Ghent University Hospital and Medical University Vienna for cross-sectional endocrine and reproductive screening. Assessments included antropometric measurements, testicular ultrasound, morning blood sampling for hormone assays (gonadotropins, total and free testosterone, insulin-like factor 3 and inhibin B). In addition, targeted next generation sequencing for genes involved in gonadal development and spermatogenesis (n=241) and low birth weight / length and poor growth (n=233) were performed in all cases.

Results

None of the participants had experienced problems regarding pubertal onset and progression. SGA cases were on average 4.4cm shorter than controls (p=0.012). In addition, 35% had not reached their genetic height potential. This was most pronounced in those with complex hypospadias (71,4%).

Higher LH, FSH and insulin-like factor 3 levels were found in hypospadias cases (p=0.034, 0.004 and 0.001, respectively). No differences in androgen levels were found. Oligo- and azoospermia were found in 13/22 (59.1%) cases who provided a semen sample. No (likely) pathogenic variants were found in investigated genes.

Conclusion

Growth and fertility are major concerns in boys born SGA and undervirilized. Therefore, routine follow-up of growth during childhood is warranted with referral for semen analyses at the end of puberty. Steroidogenesis and pubertal development are unaffected. Further studies are needed to elucidate the underlying mechanisms of the frequent poor testicular function in SGA boys born undervirilized.

Poster Walk**PW 30.****Diagnostic dilemma of a suppressed serum TSH in a female teenager with an asymptomatic goiter.**

S. Ryckx, B. Mahieu, M. Piqueur, F. Van Acker, D. Klink, J. De Schepper

*ZNA Koningin Paola Kinderziekenhuis Antwerpen, ZNA Middelheim, UZ Brussel*Background/Aims

Suppressed serum TSH with normal thyroid hormone levels can be seen in subclinical hyperthyroidism or incipient central hypothyroidism. Differential diagnosis between these conditions can be complicated in case of a poor or absent symptomatology. In addition, non-thyroidal illness and laboratory interference need to be excluded. We want to highlight the differential diagnosis and diagnostic work-up of a suppressed serum TSH (0.04 mU/L) in a 14 -years-old female adolescent with a recent onset asymptomatic goiter, but without clinical signs of hyper-or hypothyroidism.

Methods

TSH, FT4, and FT3 were measured by ECLIA on the Roche Cobas 8000 e801 platform. Measurements of TSH were also performed after heterophilic blocking (HBT) and spiking with serum of another patient with a known stable TSH concentration (recovery experiment).

Results

Physical examination was normal (absence of exophthalmia, tremor and tachycardia). Repeated serum TSH was 0.04 mU/L (N : 0.51-4.30), while FT4 was 19.7 pmol/L (N : 12.6 - 21.0) and FT3 5.7 pmol/L (N 3.3 – 7.7). Anti-TSHR or anti-TPO antibodies were not present. Ultrasound of the thyroid showed a globally enlarged thyroid gland (lobe volumes 10 and 12 ml) with bumpy outlines, but a normal reflectivity and vascularization. A TRH test showed a small rise of TSH (from 0.12 to 1.09 mU/L) and a normal PRL response (from 11.1 to 45 µg/L). Other basal pituitary hormone levels were normal. Similar TSH results were obtained with another (not streptavidin/biotinylated binding based) automated immunoassay on Abbott Architect i2000, as well as after HBT and recovery experiments. A Technetium scintigraphy of thyroid showed an enlarged thyroid with a normal homogeneous uptake (2.2 %). Serum TSH normalized after 3 months. Final diagnosis was colloid goiter with transient idiopathic TSH suppression.

Conclusion

An unexplained transient TSH suppression for 3 months was observed in an asymptomatic adolescent with a colloid goiter. After exclusion of laboratory interference (absent heterophilic antibodies), non-thyroidal illness (normal FT3), toxic nodular thyroid disease or antibody-negative Graves' disease (normal thyroid scintigraphy) and insidious central hypothyroidism (normal TRH test), 3-monthly TSH monitoring is advocated as a transient TSH suppression may occur.

P 158.

Hypothalamic lipoma and growth hormone deficiency

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UZ Leuven

Background

Intracranial lipomas are rare, congenital lesions, most often located at the midline. Most hypothalamic lipomas are asymptomatic, but some cases have been associated with precocious puberty, hypothermia, headache and/or obesity.

Case presentation

A 7-year-old boy was referred for short stature and proved to be partially growth-hormone deficient. Magnetic resonance imaging (MRI) revealed a lipoma in the paramedian hypothalamus. Growth hormone treatment resulted in swift and uncomplicated catch-up growth.

Conclusions

The present case appears to be the first to link hypothalamic lipoma to GH deficiency. The neuro-endocrine pathophysiology underpinning this link remains to be explored.

Posters**P 159.****Osteogenesis Imperfecta: pre-natal diagnosis and post-natal management**

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ULB - HUDERF, CHU Brugmann

Background

Osteogenesis imperfecta (OI) is a rare connective tissue disorder, with varying genotypes and prognosis. Pre-natal genetic diagnosis could help parents decide the future of pregnancy, even if there is no clear phenotype-genotype correlation. Reducing obstetrical trauma and avoiding perinatal fractures is a major challenge pediatricians and obstetricians have to face when delivering an affected child.

Our aim is to report two cases of OI harboring the same mutation, but diagnosed in different conditions.

Case presentation 1

A 18 years old woman, G2 P0, was referred for skeleton anomalies at 22 weeks of amenorrhea.

Ultrasounds showed abnormal bones morphologies : short (<percentile1) and bowed long bones with fractures and soft skull with large sutures evoking a severe skeletal dysplasia. Amniocentesis revealed a mutation COL1A2 (c.3034G>A) and confirmed OI.

After parents decided to continue the pregnancy, counseling explained the diagnosis, the difficulties of perinatal management (for the mother and the newborn) and the prognosis with caution.

Case presentation 2

A 2870g and 50cm male infant was born at 39 weeks after an uneventful pregnancy. The delivery was uncomplicated by vaginal route. At day 1, after a positive Ortolani test, multiple femur fractures occurred. Hypomineralisation, wormian bones and old rib fractures were present on a whole body radiography. A severe OI was suspected and confirmed by mutation detection of COL1A2 (c.3034G>A). A treatment with biphosphonates was started at 2 weeks of life, once a month combined with orthopedic management. New humerus, radius and femur fractures occurred without major trauma in the first 4 weeks of life.

Discussion- conclusion

Prenatal diagnosis of OI can be suspected during pregnancy, as soon as 16 weeks, depending on severity but it has to be confirmed with genetic sampling (amniocentesis, choriocentesis).

Neonatal care has significantly improved in the past decade in the management of OI, there was also solid progress in fetal intervention.

Post-natal management consists of increasing the bone density with bisphosphonate. Physical rehabilitation can be useful. In the case of bone fracture, long periods of immobilization should be avoided in order to spare bone density.

These two cases are a good representation of the different clinical outcomes in the presence of the same causative collagen mutation.

P 160.

Primary polydipsia in a 14-year-old girl: what to expect?

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UZ Brussel

Background

Polydipsia-polyuria can be caused by primary polydipsia (PP) or diabetes insipidus (DI), being divided in central diabetes insipidus (CDI) and nephrogenic diabetes insipidus (NDI). PP is rare in children, especially in adolescents and is often associated with a wrong drinking habit in infants or a behavioral/psychiatric disorder. The aim of this case report is to show that PP can present suddenly on adolescent age without any behavior problems and may resolve quickly after reduction of the aberrant drinking behavior.

Methods

A 16h water restriction test and MR imaging of the brain were performed in a tertiary center.

Results

A 14-year old girl presented with sudden onset of polyuria and polydipsia (drinking up to 10L of water a day) since 5 days. She had no prior medical personal/family history, no recent head trauma or neurological complaints. She was not under any stress, taking any medication or excessively exercising. Urine and blood analysis showed low serum sodium (133mmol/L), serum (261mOsm/k) and urine osmolality (47mOsm/kg), with normal serum glucose, electrolytes and kidney function. There was no glycosuria. MR imaging of brain showed no abnormalities. A water deprivation test led to an increase of urine osmolality up to 720mOsm/kg and serum osmolality up to 286mOsm/kg. Offering psychological support to stop her habitual drinking resulted in a normalization of the hyponatremia and disappearance of symptoms within one week.

Conclusion

Even after applying the water deprivation test, it can be rather difficult to differentiate PP from CDI. The water deprivation test permitted the diagnosis of PP in our case, the short duration of excessive drinking might have preserved the medullary concentration gradient. It is important to make the right diagnosis because if PP is mistaken for CDI and treated with desmopressin, severe hyponatremia may occur. Given the acute onset and the absence of underlying stress or psychiatric condition, a hypothalamic pathology affecting the thirst center, was to be excluded in our patient. The rapid normalization of her increased thirst is an argument against a hidden hypothalamic pathology. PP may present with a very sudden onset in adolescents without a psychiatric condition or stress. Fluid restriction in combination with psychiatric support will result in a quick resolution of the polydipsia-polyuria and hyponatremia and might help in excluding partial CDI if performed within the first week of onset.

Posters**P 161.****Normosmic Congenital Hypogonadotropic Hypogonadism (nCHH) due to GNRH1 mutation: a rare etiology**

C. Gernay, E. Boross, C. Vilain, H. Steyaert, C. Heinrichs, C. Brachet

ULB - HUDERF

Aim

Characterization of a rare cause of normosmic congenital hypogonadotropic hypogonadism (nCHH)

Methods

Case report. Clinical, endocrine and genetic evaluation in a patient

Results

The proband was referred at 8 months because of a relative small penis. He was the fifth child of consanguineous parents born in a small aramaic community (Idil, in Turkey). The father (160.6 cm) had delayed puberty, the mother (160.6 cm) has menarche at the age of 16 but got pregnant at 17. Physical examination showed 2.8 cm stretched penile length, distal hypospadias and high scrotal testes. After GnRH test, FSH increased from 0.7 to 6.0 and LH from < 0.2 to 1.2 mIU/ml (normal low response). Testosterone increased from <1 to 29 ng/dl, and dihydrotestosterone from < 2 to 17 ng/dl. InhibinB was at 89 ng/l. Karyotype was 46, XY. Kidneys were normal at ultrasound. He received testosterone (Sustanon 25 mg/month) for 3 months.

At the age of 2, he benefited from orchidopexy, hypospadias repair and circumcision.

At the age of 14 years, the testes were 3 ml bilaterally in the scrotum. His stretched penile length was 4 cm. Both his pubic and axillary hair were at stage 2. His height was 151.4 cm (-1.4 SD), his weight was 45,25 kg (- 0.5 SD). His bone age was 13. He had a normal sense of smell. Testosterone was undetectable. Inhibin B was very low at 20 ng/L. AMH level was also low (28.2 ng/ml). GnRH stimulation test increased FSH from 1 to 2.34 and LH from 0.3 to 2.68 mIU/ml (low response), respectively.

Clinical exome sequencing in trio revealed a homozygous mutation in the exon 1 of the GNRH1 gene (c.92G>A), the parents were heterozygous for the same mutation.

Conclusion

Congenital hypogonadotropic hypogonadism is a rare condition caused by GnRH deficiency, clinically manifested by micropenis, cryptorchidism and sexual infantilism. Sexual steroid are low with low to normal gonadotropin level, and otherwise normal pituitary function. It can be accompanied by anosmia (Kallmann Syndrome KS), or be normosmic (nCHH). Although more than 30 genes have been associated with this heterogeneous condition, mutations in GNRH1 were only described in four families, including another Turkish family reported with the same mutation than our patient. With the significant advances in the discovery of genes responsible for CHH, genetic testing is now an essential step in the investigation of this heterogeneous syndrome. Mutation in GNRH1 is a rare but known cause of nCHH.

P 162.

Message in a bottle: an infant with Cushing features.

Q. Jordens, K. Casteels, F. de Zegher

KUL

Background

Hypercortisolism is a condition resulting from prolonged exposure to elevated levels of glucocorticoids of either endogenous or exogenous origin. This may cause various, sometimes very subtle, physical and psychological changes.

Result

A three-month-old girl was investigated for inconsolable crying with sudden polydipsia and polyuria following an adenovirus gastroenteritis with high thrombocytosis. Extensive investigation ruled out diabetes mellitus and nephrogenic diabetes insipidus. Positive urinary CMV PCR detection suggested the possibility of CMV-mediated neuro-hypophysitis and concomitant central diabetes insipidus. Unexpectedly, the aforementioned noted symptoms resolved spontaneously after 7 days of hospitalisation. Two weeks after discharge, she presented with a Cushingoid appearance. The results of an ACTH-stimulation test pointed to adrenocortical suppression by exogenous glucocorticoids, and a renewed review of the medication disclosed the intake of a commercially available vitamin-D suspension and an omeprazole syrup made by the local pharmacist. There proved to be a temporal association between the intake of this syrup (also prior to the first admission) and the symptoms of hunger/thirst and Cushingoid appearance. Toxicologic analysis of the syrup revealed that it contained not only omeprazol, but also betamethasone-valerate. After halting the intake of this syrup, the girl returned to a healthy condition, including a swift normalization of cortisolemia.

Conclusion

Writing and delivering a prescription is a routine procedure for the doctor and pharmacist respectively. Dispensing errors (i.e. a discrepancy between the prescribed medication and the delivered medication) remain rare but are unlikely to ever be absent. To detect drug-related issues, such as in the present case of exogenous glucocorticoid excess, a repetitive and detailed review of all medication intake may be required. In order to resolve odd clinical cases, there is sometimes a need to think “outside the box” or “inside the bottle”

Posters**P 163.****Impaired Hypoglycemia Awareness in children and adolescents with type 1 diabetes**

A. Messaaoui, S. Tenoutasse, L. Hajsellova, L. Crenier

*ULB - HUDERF, ULB - ERASME*Background/Aims

Impaired Hypoglycemia Awareness (IHA) is associated with an increased risk of Severe Hypoglycemia (SH) in adults with diabetes but is less known in youth. The purpose of this study was to determine the prevalence of IHA and its clinical characteristics in a cohort of children and adolescents with type 1 diabetes.

Methods

This prospective observational study of two month duration included all eligible subjects with type 1 diabetes attending in the Diabetology Clinic at the University Children's Hospital Queen Fabiola (Brussels, Belgium), aged from 4 to 20 years. IHA was defined by a score ≥ 4 on a linear analogue scale, derived from the Gold method. For the aim of this study, SH was defined as an event leading to loss of consciousness. SH were collected by reviewing the patients' logbook and adjudicated by an endocrinologist. Quality of life (QoL) was determined by mean of the 3-level version of the EuroQol instrument. A logistic regression was performed to ascertain the effects of variables on IHA and SH.

Results

On the 404 subjects included, 128 (31.7%) were diagnosed with IHA. Among these, 21% experienced at least one SH vs 11% of patients without IHA during the study period ($p=0.009$). Subjects with IHA were younger (median [IQR]) (12.9 [10.4-16.3] vs 14.2 [11.7-16.9] years; $p=0.025$), younger at diabetes diagnosis (6.2 [3.5-9.3] vs 8.0 [5.0-11.2] years; $p=0.001$), more often c-peptide negative (78 vs 67%; $p=0.037$), female (57 vs 44%; $p=0.014$) and performed more glucose controls (7.0 [4.0-10.0] vs 6.0 [4.0-8.5] per day; $p=0.046$) than subjects without IHA. There were no differences in diabetes duration (5.3 [2.8-8.5] vs 6.2 [3.6-8.6] years), in HbA1c (7.6 [7.0-8.5] vs 7.7 [7.1-8.6]%) or in nocturnal occurrence of SH (8.7 vs 5.7%). Patients who experienced at least one SH were 3 times more likely to exhibit IHA ($p=0.008$). Decreasing QoL was associated with an increased likelihood of IHA. Subjects with IHA performed more glucose controls (7.0 [4.0-10.0] vs 6.0 [4.0-8.5] per day; $p=0.046$).

Conclusions

A significant proportion of youth with type 1 diabetes have IHA. Screening for IHA should be an important part of routine diabetes care in children, as IAH is associated with SH.

P 164.

Fetal goiter with congenital hypothyroidism and hearing loss: a case report

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CHIREC Delta, Hôpital Erasme, ULB - HUDERF, CHU Brugmann

Fetal goiter can cause many complications such as polyhydramnios, fetal death, preterm delivery, labor dystocia and neonatal asphyxia. In most cases, it is associated with hypothyroidism. It can be treated with intra-amniotic infusion of L-thyroxine. So far, there are no guidelines regarding dosage and frequency of administration to achieve disappearance of the goiter and euthyroid status in the newborn.

Congenital hypothyroidism is the most frequent congenital endocrine disorder and leads to cognitive disability if not treated in time. 15-20% is due to thyroid dysmorphogenesis which is caused by mutations inherited in an autosomal recessive pattern in genes encoding for key molecules involved in thyroid hormone synthesis.

The index case was born at term of non-consanguineous parents. Prenatal ultrasound had shown polyhydramnios due to a fetal obstructive goiter. The mother did not have a personal nor family history of thyroid disease. At 32 weeks, fetal TSH levels were at 370 mU/L and free T3 levels at 3,4 pmol/L while maternal thyroid function was completely normal. A schedule of five intra-amniotic L-thyroxine infusions between 32 and 39 weeks of gestation were given.

A blood sample at 1 hour of life showed elevated TSH (363 mU/L), low T4 (5.5 pmol/L) and extremely elevated thyroglobulin confirming congenital hypothyroidism. Ultrasound showed a moderate goiter. Treatment with oral L-thyroxine of 10µg/kg/day was initiated.

Brainstem Auditory Evoked Potentials (BAEP) were performed at 9 days of life and showed symmetrical sensorineural hearing loss reaching 60dB nHL. After one month, when TSH plasma levels were in the normal range, BAEP were repeated and showed improved thresholds at 35 dB nHL. At three months and a half, in a context of normal tympanograms, the thresholds ameliorated with another 10 and 5 dB.

With next generation sequencing, compound heterozygosity for two pathogenic variants in the thyroxine peroxidase (TPO) gene were detected: c. 209 C>T (novel mutation) and c.1184_1187dupGCCG (previously reported in congenital hypothyroidism).

In conclusion, we report a patient with congenital goitrous hypothyroidism due to a compound heterozygosity for TPO mutations. Intra-amniotic L-thyroxin treatment reduced fetal goiter but did not normalize TSH and T4 levels at birth. BAEP showed a sensorineural hearing loss with improvement over the first months of life with L-thyroxine treatment, which suggests hearing impairment is induced by hypothyroidism.

Posters**P 165.****Epidemiological and clinical factors influencing the presence of ketoacidosis in Children with new onset type 1 diabetes. Review of the last ten years**

A. Perchec, A. Messaaoui, L. Hajselova, S. Tenoutasse

*ULB - HUDERF*Background

Diabetic ketoacidosis (DKA) at diagnosis of type 1 diabetes (T1D) is a cause of high morbidity that can be preventable if diagnosed early. The aims of this study are to characterize the prevalence of DKA at diagnosis in children followed in the largest center for pediatric diabetology in Belgium and to highlight the risk and protective factors of DKA in our population.

Methods

A retrospective study including all children under 18 years of age with new onset T1D over 10 years, from 1 January 2009 to 31 December 2018, at the Queen Fabiola Children's University Hospital. Anamnestic, clinical and biological parameters were studied.

Results

On the 511 children, 39.9% met the DKA criteria. The average age at diagnosis was 9.1 (5.6-12.3) years. The classic symptomatology of hyperglycemia (polyuria, polydipsia, fatigue, weight loss) was indeed found in the anamnesis, and even more frequently in case of DKA. Children presenting with DKA had more nausea/vomiting (60 % with DKA versus 16 % without DKA, p-value=<0.001), abdominal pain (57 % with DKA versus 30 % without DKA, p-value=<0.001) and/or impaired consciousness (16 % with DKA versus 4 % without DKA, p-value=<0.001), with higher blood glucose (515 [407-681] mg/dl with DKA versus 412 [304-547] mg/dl without DKA, p=<0.001) and glycated hemoglobin levels (11.9 [10.9-13.5] % with DKA versus 11.3 [9.7-12.8] % without DKA, p=0.004). The factors protective of DKA were the presence of a family history of T1D (32 % without DKA versus 18 % with DKA, p-value=<0.001), a higher level of maternal education (24 % without DKA, 16 % with DKA, p-value=0.045) and regular monitoring by a pediatrician (12 % without DKA versus 6% with DKA, p-value=0.02). Conversely, being a younger girl coming from the Middle East and reporting directly to the emergency department were risk factors for DKA.

Conclusions

The prevalence of DKA at diagnosis of T1D in our study remained significant, nearly 40%. The early identification of clinical signs of hyperglycemia and the training of health care professionals remain a major challenges in the prevention of DKA.

P 166.

A rare cause of failure to thrive

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CHU Saint-Pierre

Aldosterone plays an essential role in fluid and electrolyte homeostasis through the renin-angiotensin pathway. The biosynthesis of this hormone requires 4 enzymes: cholesterol side-chain cleavage enzyme, 3- β -hydroxysteroid dehydrogenase, 21-hydroxylase and corticosterone methyl oxidase (CMO). Two types of deficiencies can be observed in relation to this fourth enzyme: CMO type I deficiency, with a low aldosterone blood level, and CMO type II deficiency, with a normal or low aldosterone level and an accumulation of its precursor 18-hydroxycorticosterone.

A 3-month-old baby girl was admitted to the Saint-Pierre Hospital (Brussels) for failure to thrive. She was born at 40 weeks of gestation from a non-consanguineous Albanian couple. While her birth weight was on the 50th percentile (3.740 Kg), it had dropped on the 3rd percentile at three months of life. Blood pressure was normal for her age. Physical examination revealed hypotonia and weakness but no other abnormality. First laboratory results revealed hyponatremia (128 mmol/L) and hyperkalemia (5.2 mmol/L) with normal renal and liver function. On the urinary spot, natriuresis was undetectable and urine osmolarity was 151 mOsm/Kg. Cardiac and abdominal ultrasounds, as well as the sweat test, were normal. Further blood investigations showed extremely high renin level (93,760 mIU/L), while aldosterone level was in the normal range (196 ng/L). Other hormonal tests (cortisol, IGF1, ACTH, 17-OH-progesterone, testosterone, DHEA, SHBG) were normal. We were unable to find a laboratory where multiteroid analysis by chromatography could be performed in Belgium. The increased renin level with normal aldosterone level suggested the diagnosis of CMO type II deficiency. Indeed, this diagnosis was confirmed by genetic analysis, which revealed a mutation on each allele of the CYP11B2 gene coding for the CMO (C554C>T and C541C>T). The baby was supplemented with Sodium Chloride and substitutive 9- α -fluorohydrocortisone. Electrolyte abnormalities were quickly corrected, and three weeks later weight gain was observed. The renin level decreased to 1,808 mIU/L after four months of treatment.

Electrolyte levels should be assessed in every child with an unexplained failure to thrive. The combination of increased renin and normal aldosterone levels is pathognomonic of CMO type II deficiency. To the best of our knowledge, this is the first case of CMO type II deficiency confirmed by genetic analysis reported in our country.

Posters**P 167.****Intraamniotic treatment of a new form of congenital hypothyroidism: a case report**

O. Pollé, P. Lysy, A. Lourtie, P. Bernard

*UCL Saint-Luc*Background

Fetal goiter (FG) is a rare condition that may lead to obstetrical and/or neonatal complications (e.g. polyhydramnios, fetal malposition, tracheal compression). Besides the exclusion of associated complications, the investigations focus on the evaluation of biological signs of hyper/hypothyroidism in the mother and the fetus, guiding the therapeutic decision. In fetal hypothyroidism, thyroid hormone substitution remains a challenge as maternofetal transfer of T4 is limited. We report the case of a FG in the context of congenital hypothyroidism and discuss the role of intra-amniotic infusions (IAI) of levothyroxine in this setting.

Case report

A 32-yo woman was referred to our clinic at 23 weeks of gestation for FG. Detailed echography of the fetus confirmed the goiter (4.28 cm³, >95th percentile) and showed no other abnormalities. The mother had normal thyroid function tests without positive autoantibodies (i.e. anti-TPO, -TSHR and -TG). Because of the significant volume of the goiter, a cordocentesis was performed and confirmed the diagnosis of FG in the context of nonimmune congenital hypothyroidism (TSH>100mU/L; fT4 5.5 pmol/L). The decision to initiate an oral treatment with L-thyroxine (50 µg/d; 1µg/kg.d) in the mother was taken and later increased to 75 µg/d (1.5µg/kg.d). As the goiter became clinically patent for the fetus (neck hyperextension and increase of amniotic fluid index (AFI)), a series of four IAIs (400 µg L-thyroxine per IAI) was started. After the second injection, a significant decrease of the goiter volume and of the AFI was observed. After the uneventful delivery, the endocrine assessment of the newborn revealed a persistent goiter, hypothyroidism (TSH >100mU/L, fT4 11pmol/L, undetectable thyroglobulin) and normal bone maturation (90th percentile for 40 weeks of gestational age). The infant was immediately started on 50 µg/d (170 µg/kg.d) of L-thyroxine. Genetic panel assessment revealed a new form of compound heterozygous mutations in the TG gene.

Conclusion

L-thyroxine IAIs are feasible and may be a useful therapeutic option to improve FG and reduce related obstetrical complications, as recently suggested by Ribault & coworkers. Despite the absence of thyroid status normalization at birth, L-thyroxine IAIs may have a positive metabolic impact on the fetus by improving hypothyroidism-related clinical features (e.g. bone age). Nevertheless, consensus on the standardization of the procedure is still required.

P 168.

Prepubertal gynecomastia: what to suspect first?

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UZ Brussel

Introduction

Most cases of prepubertal gynecomastia are classified as idiopathic. However, an exogenous or endogenous hyperestrogenism (from estrogen producing testis or adrenal tumors) has always to be excluded. We want to highlight the eventual hormonal and clinical correlates of hyperestrogenism in a prepubertal boy with bilateral gynecomastia.

Case report

A 9 year and 10-month-old boy was referred for non-prolactinoma related hyperprolactinemia. He presented himself with bilateral breast enlargement and slight pubic hair development, but without body odor or axillary hair development. He had no previous medical or surgical history, but he had taken commercial Perilla oil tablets for 6 months for concentration problems, which had been stopped one month before the onset of breast development. He denied the intake of other medication, food supplements, herbal products or use of cosmetic ointments since the start of gynecomastia. Physical examination disclosed bilateral mammary swelling with areolar pigmentation, but without galactorrhea. Tanner stage was A1P2G1. Testicular volume was 2 ml bilaterally.

Hormonal analysis showed an elevated serum prolactin, normal cortisol, undetectable gonadotrophins and estradiol, but a very high serum SHBG. MRI of the pituitary was normal. Bone age was 11.5 years. Ultrasound of the testes and adrenal were normal. During the GnRH stimulation test, no LH and FSH responses were observed. By further questioning, his mother confirmed the use of a nonformulary estrogen cream, obtained at an anti-aging clinic. Handwashing after cream application was not always performed and mother and son were sharing bath towels and blankets.

Discussion

The areolar hyperpigmentation together with the typical hormonal profile (suppressed gonadotrophins and elevated PRL and SHBG) made us suspect hyperestrogenism as an underlying cause for the gynecomastia. In addition, the contrasting finding of pubic hair and normal adrenal androgens supported our hypothesis, as pubic hair growth can be seen in patients exposed to estrogens. The use of estrogen cream was initially not disclosed by the mother but was afterwards confirmed by her and the prescribing physician.

Conclusion

In the evaluation of prepubertal gynecomastia, the use of cosmetics, herbal medicines or topical medications, including estrogens, should not only be questioned in the child, but also in the parents and close caregivers.

Posters**P 169.****Renal Pseudohypoaldosteronism type I in a 6-day-old neonate**

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Clinique Saint-Pierre Ottignies

Renal Pseudohypoaldosteronism (PHA1) is a hereditary disorder characterized by aldosterone resistance leading to renal salt loss. Symptoms appear during the neonatal period. Common laboratory findings include hyponatremia, hyperkalemia and metabolic acidosis, due to decreased activity of epithelial sodium channels (ENaC).

Case report

A 6-day-old male infant was admitted to our neonatal department for significant weight loss and feeding difficulties. Laboratory investigations revealed hyponatremia, hyperkalemia and metabolic acidosis. A few days after admission, high blood pressure was observed.

As first hypothesis, we suspected a congenital primary adrenal insufficiency. Initial management consisted in glucocorticoids, mineralocorticoids supplementation and correction of electrolytic disorders by external sodium intake. Determination of androgenesis enzymes returned normal.

Further investigations revealed an elevation of plasma renin activity and plasma aldosterone, which suggested the diagnosis of PHA1.

Sequence analysis of the NR3C2 gene demonstrated a heterozygous c.2839C>T mutation, confirming the diagnosis.

With salt supplementation, sodium level returned to normal range and body weight gain improved.

Discussion

The reduction of intravascular volume activates the renin-angiotensin-aldosterone system (RAAS), resulting in elevated plasma renin activity and plasma aldosterone.

Normally, the activation of ENaC by aldosterone increases renal Na⁺ reabsorption.

The renal form of PHA1 is caused by mutations in the gene NR3C2 which encodes the mineralocorticoid receptor (MR) in the distal renal tubule. The pattern of inheritance is autosomal dominant, but there are sporadic cases due to de novo mutations.

The mutations prevent binding of MR to aldosterone and decrease the expression of ENaC in the distal nephron.

The diagnosis is based on genetic analysis. It has a good prognosis. The extent of salt wasting improves over time and the treatment only consists in salt supplementation until early childhood.

Conclusion

In the context of persistent sodium and potassium imbalances in children who have normal adrenal function, the determination of plasma aldosterone and plasma renin are determinant elements to guide the diagnosis.

Of interest in our case, the strikingly arterial hypertension is rarely described. It could be explained by an over-activation of the RAAS. Thereby, normal or high blood pressure does not rule out salt loss syndrome.

P 170.

Rare cause of Cushing syndrome in a 12-year old girl with striae and weight gain

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UZ Leuven

Background and methods

Cushing's syndrome is rare in children. Its hallmark is glucocorticoid excess, which is most often caused by exogenous glucocorticoid administration, or by an endogenous excess of pituitary ACTH, referred to as Cushing's disease. Here, we report a girl with an extremely rare adrenal cause of Cushing's syndrome.

Findings

A 12-year old girl with an unremarkable medical history presented with generalised purple striae on the abdomen, back and limbs. For a few months, she had experienced excessive weight gain, besides menorrhagia, headaches and abnormal sleep. At clinical examination, she was hypertensive and had a moon facies with plethora, a buffalo hump and hirsutism on back and neck. Serum cortisol (36.5 µg/dL) and urinary free cortisol (1264.8 µg/day) were strikingly elevated, conferring also mineralocorticoid effects since hypokalaemia was consistently accompanied by hypernatraemia. Plasma ACTH was undetectably low, suggesting primary adrenal disease. MRI revealed a 9 cm large tumour mass with central necrosis in the left adrenal gland, with features of hypermetabolism on FDG-PET-CT. Pre-surgical reduction of the signs and symptoms of cortisol excess was achieved by administering a steroidogenesis inhibitor (ketoconazole) and a mineralocorticoid receptor antagonist (spironolactone). Open unilateral adrenalectomy, removing a mass of 9x9.8x6.5 cm, was followed by tertiary adrenal insufficiency. Hydrocortisone administration was started. Histopathologic analysis confirmed the presence of an adrenocortical carcinoma with a Weiss score of 6. Two months post-surgery, a low normal ACTH suggested incipient recovery of the adrenocorticotrophic axis, while cortisol levels were still low. Long-term prognosis, without any long-term treatment, is favourable.

Conclusion

Clinical signs and symptoms of Cushing's syndrome during childhood, unexplained by exogenous glucocorticoids, require further investigation. Hypercortisolism associated with low ACTH levels are indicative for a primary adrenal cause, possibly an adrenocortical carcinoma. Pre-surgery, dual treatment with ketoconazole and spironolactone may be helpful. Post-surgery, glucocorticoid substitution should be ensured before slow dose tapering into sub-substitutive range, guided by clinical and endocrine markers, until recovery of the full adrenocorticotrophic axis.

P 171.**Mediastinal immature teratoma leading to a supraphysiological serum testosterone concentration in an adolescent with Klinefelter syndrome**

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*UZ Gent*Background

Germ cell tumors (GCT), including teratomas, account for less than 5% of malignancies in childhood and adolescence and are mostly located in the testes in males. Mediastinal GCT usually manifest in adults, with the exception of Klinefelter syndrome (KS), where cases have been described in both prepubertal males and adolescents. Up to 31% of male patients with mediastinal GCT have KS, showing in general thorax-associated symptoms. We report the finding of an immature teratoma/seminoma in an adolescent KS male without symptoms, but presenting with a testosterone concentration above adult values at his yearly biochemical assessment.

Case report

The yearly blood analysis of a 15 year old with KS (47, XXY) male showed a very elevated serum testosterone (1412 ng/dL, normal range: 267.0 - 929.0 ng/dL) with suppressed LH and FSH (< 1U/L) levels. He had no general or specific complaints. Testis volume (12 and 12 ml) and pubertal staging (A2P4G4) had not changed during the last year. At physical examination a small left glandular breast swelling was palpated. Both alpha-fetoprotein (AFP) (59 µg/l) and β-hCG (2.16 IU/L) were elevated. Ultrasound of testes and adrenals were normal. CT thorax showed an anterior mediastinal hypodense rounded mass (49/34/53 mm) with heterogeneous contrast uptake and enhanced metabolic activity at a F-18 FDG PET-CT scan. Histologic examination of the surgically removed mass, which was attached to the thymus, showed an immature teratoma (90%) and a small component of seminoma.

Discussion

KS is a known risk condition for extragonadal GCT, although routine screening is not recommended given the overall small risk. The genetic origin of mediastinal GCT in KS is still unknown. The contact of the mass with the thymus in our patient suggest that the transformed cells may originate from primordial cells misplaced during embryogenesis.

Another particular finding in our patient was the absence of associated symptoms, except for gynecomastia, which is a typical finding in KS adolescents with mediastinal GCT. Elevated AFP was previously described in KS males with immature teratoma. Despite the high β-hCG level, no testicular growth was observed in our case.

Conclusion

In KS males anterior mediastinum is a preferential location of GCT, which does not always give thorax-associated complaints or testicular enlargement. Yearly testosterone and gonadotropins monitoring might be of help in early detection of GCT in KS adolescents.

P 172.

Are low-grade inflammation or testosterone aromatization in adolescent males with overweight or obesity associated with pubertal timing?

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UZ Brussel, Zeepreventorium

Background/Aims

An early, normal or delayed pubertal onset have been described in overweight/obese males. A greater degree of adiposity has been associated with a higher risk for delayed puberty, but the underlying mechanism has not been fully explored. We investigated whether a higher degree of low-grade inflammation or an increased testosterone aromatization might be more frequently present in overweight males with a delay in genital development.

Methods

Pubertal status assessment by Tanner staging and measurement of morning serum testosterone, estradiol, leptin, and high sensitive CRP (hsCRP) by standard laboratory methods were performed in 191 male adolescents, aged between 10 and 18.6 yr (median 12.84 yr) with overweight or obesity (BMI z-score >1.3), starting an ambulatory (n = 136) or a residential weight loss program (n = 55). Their median (range) BMI z-score was 2.32 (1.34 – 3.38). Delayed/slow and early/rapid genital development was defined by a Tanner genital stage respectively below the 10th or above the 90th percentile age distribution (national Flemish standards of 2004).

Results

In 3 males pubertal development was advanced, while in 34 it was delayed, in the remaining 156 adolescents genital stage was normally timed. Males with a delayed timing or progression of genital development were older (median(range) age: 14.84 (11.62 – 18.54) yr vs 12.31 (10.02 – 18.62) yr, p<0.005), shorter (height sds: -0.55 (-1.92 – 1.48) vs 0.49 (-3.03 – 3.79); p<0.005), but showed a similar BMI z-score (2.31 (1.44 – 3.30) vs 2.32 (1.34 – 3.22); p= 0.928) and waist z-score (2.06 (1.39 – 3.06) vs 2.21 (1.14 – 3.04); p =0.325) in comparison to males with a normally timed puberty. Median serum estradiol, leptin, and hsCRP concentrations did not differ significantly between those males with a normal or a delayed pubertal onset or progression.

Conclusion

Pubertal delay is more frequently observed than early puberty in males referred to obesity clinics. Neither an increased inflammation, nor an increased estradiol production appear to be associated with a later onset or slower progression of genital development in male obesity.

P 173.

Algorithmic adaptation of insulin therapy and carbohydrate consumption during exercise in children with type 1 diabetes: results of the CAR2DIAB study

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Evaluate the evolution of interstitial glucose during two sessions of aerobic exercise monitored in children or adolescents with type 1 diabetes after adjustment of insulin doses and carbohydrate intakes according to a combined algorithm.

Methods

Twelve patients with type 1 diabetes (15.1 ± 2 years, duration of diabetes: 9.5 ± 3.1 years) performed 2 sets of exercise sessions after cardiological evaluation. The first series (TE # 1) consisted of moderate-to-vigorous exercise with vigorous exercise coupled with maximal effort (total duration: 30 minutes). The second exercise series (TE # 2) was performed in the context of "real-life", during exercises categorized and monitored by connected watches. TE # 2 sessions were performed after insulin dose modifications using a decision algorithm [1], and fast carbohydrate diets were suggested using the algorithm proposed by Riddell in 2011 [2]. Patients were under CGM monitoring.

Results

Patients did not experience episodes of severe hypoglycemia, symptomatic hyperglycemia or ketosis. Analysis of CGM data (15h) in TE # 2 sessions revealed an overall improvement in mean glycemia [\pm standard deviation] (104 ± 14 mg / dL vs 122 ± 17 mg / dL during TE # 1; <0.001), associated with a decrease in the proportion of hyperglycaemia in periods ranging from 4h to 15h after exercise. The proportion of hypoglycaemia was not changed, except during the period TE # 2 + 4-8h, where a significant increase in hypoglycaemia <60 mg / dL was observed (25 vs 6.2%, $P = 0.04$), without associated complication.

Conclusion

In our pediatric series, the application of algorithmic adaptations of insulin doses and carbohydrate intakes globally allowed the improvement of glycemic control for 15h following the exercises performed in "real life" in children with type 1 diabetes.

10. Other

SO 31.

Impact of COVID-19 on Belgian paediatric trainees.

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Background and objectives

The COVID-19 pandemic has put strain on the activities and well-being of health care workers. We aimed to measure the direct and indirect impact on a personal and professional level for paediatric trainees in Flanders, Belgium.

Study design

Junior representatives of Flemish Society for Paediatrics (Jong VVK) conducted a follow-up cross-sectional study among fellow paediatric trainees. The impact of COVID-19 on daily tasks, education and emotional well-being for the first (March - April 2020) and second wave (October - November 2020) of the pandemic were studied.

Results

One hundred and nineteen surveys were completed in the first wave, consisting of 51% (119/233) of the total number of paediatric trainees in Flanders. Eighty surveys were completed in the second wave. Educational program changes occurred in 25% (30/119) of trainees and more than half (61%; 72/119) described the pandemic as an impediment for their educational progress. The perception of impaired education persisted for 30% of the responders (24/80) during the second wave. One out of three (30%; 35/119) felt their job was more exhausting than usual and 38% (45/119) perceived more stressed at work. These numbers were comparable at both timepoints. Increases in stress paralleled with increased irritability in daily life and poorer sleep quality. Increased stress at work correlated significantly with higher workload, having to do uncomfortable tasks, feelings of exhaustion and unsafety, and feeling less part of a team or less useful during the COVID-19 crisis.

Conclusions

COVID-19 had an important impact on the daily tasks, education and emotional well-being of the paediatric trainees. Medical training centres should be aware that there is a perception of impediment on the educational program of the paediatric. Policy makers and medical trainers should be cautious not to allocate all resources towards the specialities that were most prominently on the front lines of COVID-19 patient care.

Poster Walk**PW 8.****The virulence of a highly necrotizing clinical strain of *Streptococcus pyogenes***

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Background/Aims

Group A *Streptococcus* (Strep A) causes more than 500.000 deaths per year and is responsible for a broad spectrum of diseases ranging from pharyngitis to life-threatening invasive diseases such as necrotizing fasciitis. In 2008, a case of nosocomial transmission of Strep A occurred in Brussels between an infected child with necrotizing fasciitis and a nurse. After cutting herself with the scalpel used during child surgery, the nurse developed a finger necrosis. An identical Strep A strain was isolated from both cases. The strain, called L01, was genome sequenced to identify potential virulence genes explaining its hyper-necrotic phenotype.

We are characterizing the isolate recovered from this extremely severe clinical presentation to better understand the mechanisms used by the bacteria to invade host cells and escape from the immune system. We identify 4 enzymes called DNases (Sdn, Spd1, Spd3 and Spd4) in the L01 genome which could explain the phenotype of this clinical isolate. Our first goal is to investigate the specific role of DNases in Strep A virulence. In parallel, we also want to identify genes implicated in virulence by a comparative transcriptomic analysis of the L01 with a less pathogenic reference strain.

Methods

We have monitored the survival of our strain in whole blood and its fitness in the presence of exogenous DNA in comparison with a reference strain to see if an advantage of growth was observed. We have also performed RNA-seq analysis of the strain in different conditions and in comparison with the reference strain.

Results

We have shown that the presence of exogenous DNA gives a fitness advantage to the L01. Moreover, the L01 survives better than the reference strain in human whole blood. Furthermore, analysis of transcriptomic results in a rich growth medium during exponential phase highlights approximately a hundred of genes that were differentially expressed between the L01 and the reference strain in the tested condition.

Conclusion

This study will allow us to understand the mechanisms underlying the molecular basis of the necrotizing fasciitis and the specific pathways implicated in it. This could lead, in the future, to the development of diagnostic tests to detect this pathology quicker.

PW 28.

The European paediatric clinical trials network conect4children: activities of the Belgium National Network and its progression

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Background/Aims

Paediatric clinical trial networks have made many and unique advances over the last years through the possibility of a pan-European network conect4children (c4c) funded by the Innovative Medicines Initiative (IMI). With its start in 2018, it has achieved substantial deliverables that could redesign the landscape of paediatric drug development for a durable and efficient growth. The Belgium Paediatric Clinical Research Network (BCPRN) with its headquarters localized in Ghent, plays an essential role in these developments being involved in most of the work packages of c4c. One of the finalities of this project was the establishment of a paediatric clinical trial center, connected to the adult trial department at University Hospital Ghent. Furthermore, the European Paediatric Translational Research Infrastructure (EPTRI) has been started up to stimulate and accelerate preclinical paediatric drug development. To this date, little is published about this international consortium and how it could impact all future paediatric trial structures.

Method

This report describes an update on the progress of c4c over the past 3 years, including how the preliminary data of SAFEPEDRUG before 2018 is applied for pan-European clinical research.

Results

In the proof-of-viability clinical studies, three academic studies have started including patients over Europe, in which Belgium is the first to receive national approval for the KD-CAAP study. Aside from the four initial industry proof-of-viability clinical studies, four additional studies have been included. BPCRN together with the Health, Innovation and Research Institute of UZ Gent (HIRUZ) is involved in the work package of data-management and data coordination, which will form the basis of the first paediatrics-based CDISC approved data-standardization collection. In the work package for completing a sustainable business model, a complete SWOT analysis and legal structure has been developed for the future structure(s). BPCRN is also partnered with a collection of central institutions for expert opinions.

Conclusion

The SAFEPEDRUG preclinical research has evolved into a clinical setting within the pan-European network of c4c. Over the past 3 years, substantial developments and progress has been made with the Belgium network activity by BPCRN. Both are essential to achieve substantial improvement in medicines for children in EU and worldwide.

Poster Walk**PW 31.****Eczema herpeticum, a dermatological emergency**

J-C Beghin, S. Pohlen

*ULB - HUDERF*Background

In paediatrics, the frequency of visits to the emergency department (ED) for skin complaints varies between 5 and 17% of all ED visits. Most of these cases are not true emergencies; however, evaluation of the skin can reveal potentially fatal diseases.

Case report

We describe the case of a 9-month-old boy, born at full-term after a normal pregnancy. He had no relevant medical history except eczema on the elbows and knees treated with emollients. The infant was admitted to the ED after 24 hours of high fever (39.6°C) with a pruritic papulopustular eruption and yellow crusts on the face and the eczema lesions. Uncomplicated impetigo was diagnosed and he was discharged with oral Flucloxacilline and topical care. The next day, the infant returned to the ED with a deterioration of the general status, vesicles and black crusts on the cheeks, lower limbs, upper limbs and genital area. Biology showed a slight inflammatory syndrome. Rapid expansion of the vesicles and the presence of large necrotic crusts on a pre-existing eczema led to suspect eczema herpeticum (EH). The child was treated with acyclovir for 10 days of which 5 were intravenous (IV) and improved quickly.

Discussion

EH, or Kaposi Juliusberg syndrome, is an acute disseminated cutaneous infection induced by herpes simplex virus (HSV). It occurs in less than 3% of children with atopic dermatosis. The syndrome is defined by an extensive rash with vesicles, pustules and large or necrotic crusts. It is occasionally accompanied by pruritus, fever, malaise, vomiting, diarrhea and lymphadenitis. Herpetic keratitis and encephalitis are the most serious complications. Mortality can reach 75% if untreated and varies between 1 and 9% with antiviral therapy. EH diagnosis is mainly clinical. The most sensitive method to detect HSV is polymerase chain reaction (95%). The Tzanck test (cytological diagnosis) is a rapid and cheap identification method but sensitivity (40 to 80%) and specificity are poor, depending upon the examiner's expertise. Serological tests for HSV, bacterial and virological swabs are of little interest. Treatment with oral or IV aciclovir should be initiated promptly for at least 7 days to minimize morbidity and mortality.

Conclusion

EH is commonly misdiagnosed as eczema exacerbation or bacterial superinfection. In light of the severity of EH, rapid recognition of symptoms and fast initiation of antiviral treatment are crucial to reduce morbidity and mortality related to the disease.

PW 32.**Some trainees are more equal than others - the pediatric residency pay gap**

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Jong VVK, UZ Gent, UZ Brussel, VVK, Zeepreventorium, ZNA Koningin Paola Kinderziekenhuis

Background and aims

In Belgium, medical doctors in specialty training (residency years) are employed under a distinct and unique statute (*sui generis*). Although legal provisions exist, recent polls showed relevant heterogeneity in adherence to employment contracts, wages, and social benefits.

Methods

Junior representatives of the Flemish association for pediatrics (Jong VVK) conducted a descriptive cross-sectional study among trainees in pediatrics in Flanders in 2019, who are employed in both university and non-university training centers. By means of an anonymized online survey their wages, working conditions and fringe benefits were collected and studied.

Results

Fifty-four surveys were completed by 48 unique trainees. Data concerned employment regimens from 21 hospitals. The average gross and net monthly salary were, respectively, € 3182,63 (standard deviation € 221,86) and € 2424,28 (SD € 223,35). Noteworthy, the difference between the most wealthy incomes (P95) versus the least (P5) was € 713,79 net per month. Only minimal effect of seniority could be found. The average extra salary for all earnings besides the standard income (e.g. on on-call duties, overtime, transportation expenses,...) was € 305,87 per month, an additional income of barely 15%. The average net income per hour (€ 9,32) only just exceeds the nationally fixed minimum wage, undervaluing the trainee's past education, medical degree, experience and responsibility. Fringe benefits were infrequently provided. Besides 63% in the possibility of receiving a bike fee, 51% a hospitalization insurance and 45% an insurance concerning civil liability, other benefits were provided in less than a quarter of respondents with no-one receiving meal vouchers, 2% a vacation salary and 8% an end-year bonus.

Conclusion

The trainee in pediatrics in Flanders is being paid disproportionately and unfairly for his extensive and stressful professional activities. The roots for this inequity can be traced back to the outdated *sui generis* statute, an inadequate legal framework and a weak and non-vacant position of the resident on the labour market. There is a relevant heterogeneity in gross and net income and fringe benefits are unevenly provided. Moreover, it is striking that the individual trainee is not well informed about his or her employment conditions and transparency is lacking.

Poster Walk**PW 39.****Belgian CAP48 cohort of juvenile idiopathic arthritis (JIA): What is the role of the emergency department in the early management of JIA?**

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Juvenile idiopathic arthritis, although rare, is the most common rheumatic disease in children and can be an important cause of disability, of which the need for early treatment has been demonstrated by several studies. The aim of our study is to observe the patient's journey before their diagnosis, and more specifically in the emergency department since it seems to be the reference point at the time of the first symptoms.

Method

This is a retrospective study that included a questionnaire analysis and a review of medical files. We studied the CAP48 sub-cohort, which includes 61 children with juvenile idiopathic arthritis followed at the Queen Fabiola Children's University Hospital in the pediatric rheumatology department from 2013 to 2019.

Results

Forty-eight patients were finally recruited with 26 girls (54%). The average age of diagnosis is 5.1 years (SEM: 7.5 months). The first practitioner to be consulted mostly is the emergency physician (43.5%). The diagnosis is made in average after three physician's visits. The lapse of time between the first symptom and the diagnosis is 10.1 months on average (SEM: 3.2 months). In general it is the pediatrician who refers to the specialist in pediatric rheumatology (28%). In most cases, it is this specialist who first introduces the diagnosis.

Conclusions

The diagnosis of juvenile idiopathic arthritis remains a huge challenge for any practitioner. Therefore, emergency physicians have an important role since their clinical suspicion depends the speed of the care journey.

P 174.

Hair dancing to the rhythm of the beat

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UZ Leuven, Imeldaziekenhuis Bonheiden

A 4-year old boy presented with a longstanding history of rhythmic pulsation of the scalp hair. The physical examination was notable for an area of scalp hair that vibrated at frequency of about 90 beats per minute. There was no palpable mass and neurological examination was normal. Ultrasonography revealed a discrete vascular channel that had a transcranial extension and communicated with the sinus sagittalis superior. The diagnosis of a sinus pericranii was made. Sinus pericranii is a rare vascular malformation in which there is an abnormal connection between intracranial and extracranial venous structures. MRI imaging showed no other intracranial abnormalities. The patient was managed conservatively with regular follow-up.

P 175.**Recurrent vulvar ulcers in young girls**

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Vulvar ulcers are a painful condition needing a rationale for appropriate diagnosis. They have numerous causes and are usually self-limiting, but diagnosis-based treatment can reduce the duration of ulcers.

Case report

We report on an 11-year-old girl presenting for the third time in 7 months with oro-genital ulcerations and fever, without any other symptoms. She is nonsexually active and there was no evidence of trauma. Extensive ulcers of the labia minora and majora with loss of substance were the hallmark of the clinical examination. Behçet's disease was suspected and systemic corticoids quickly controlled the lesions.

Differential diagnosis

- Lipschutz's ulcers are large vulvar ulcers that mainly affect nonsexually active girls younger than 20 years and usually resolve within 3 weeks. They can occur with a flu-like illness, EBV or Mycoplasma. They appear as an acute condition, rarely relapsing, and are not associated with mouth ulcers.
- Sexually transmitted infections such as syphilis, chlamydia, HIV and HSV were ruled out.
- Cyclic neutropenia, agranulocytosis and vitamin B1, B2, B6, B12, folate, iron and zinc deficiencies were not found.
- Inflammatory bowel disease diagnosis proved irrelevant due to absence of gastrointestinal symptoms, normal abdominal ultrasound and gastroscopy, negative ASCA and ANCA, and an unremarkable histological analysis from a guided biopsy of an ulcerated area.
- Genital ulcers are common in Behçet's disease (BD), being the initial sign in about 18% of patients. However, diagnosis is solely based on clinical criteria. Our patient only presented with recurrent oro-genital ulcerations; she had no cutaneous lesions and her ophthalmological examination was normal. HLA-B51, often associated with BD, was negative.

Discussion

Although formal diagnosis couldn't be established, the recurrent and aggressive presentation of the genital ulcers with loss of substance as well as reported recurrent oral ulcers are strong clinical clues to suspect the first stage of BD. In addition to pain management, topical treatment with high potency corticosteroids and systemic corticosteroids progressively controlled the lesions. Colchicine was later started for a better long-term control.

Conclusion

Recurrent vulvar ulcers are a painful condition and primarily need appropriate pain management. Accurate diagnosis, though not always assured, enables to focus on best treatment to reduce duration of symptoms and risk of recurrence

P 176.

A rare case of subcutaneous emphysema after video-assisted thoracoscopic surgery for pleura empyema

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UZ Brussel, ZNA Koningin Paola Kinderziekenhuis

Introduction

About 0.6% of paediatric pneumoniae are complicated by empyema and more recent studies indicate that the incidence is increasing. Empyema thoracis is defined as a collection of pus in the pleural space. There are three stages described in the natural course of empyema: the exudative, fibrinopurulent, and organizing phase.

Because of advantages the video-assisted thoracoscopic surgery (VATS) has become mostly the standard of care for the treatment of empyema in children.

Multiple studies have shown that VATS decreases significantly the length of hospital stay, in comparison to thoracocentesis or chest tube alone in children.

Case

A 3 year old girl without any relevant history presented with fever during 1 week, dyspnea, signs of dehydration and of thoracic and abdominal pain. Pleura empyema of the right lung was confirmed by Chest X-ray and Thoracic Ultrasound. A treatment with intravenous broad spectrum antibiotic was administered (cefotaxim and flucloxacillin). Also oxygen therapy was started. A VATS was performed and purulent liquid was evacuated and a chest tube was left behind. A chest tube was placed too.

In Post-operatively a diffuse subcutaneous emphysema was seen as complication. It started at the thoracic and abdominal level and quickly ascended to the occipital and orbital regions. She was in severe pain for which continuous intravenous morphine was started. The insertion opening of the drain was enlarged but showed no significant improvement. Respiration was supported with low flow oxygen 2 l/min through a nasal cannula. two litres/min oxygen with nasal cannulae. In The emphysema was treated conservatively and resolved in about 5-6 days. Three days after VATS the chest tube was removed. Cultures of the blood and the pleura purulent liquid were negative. Antibiotics were given for 16 days intravenous and continued orally for five days.

Discussion

This is a case of subcutaneous emphysema after VATS procedure, which is a rare complication in children. The incident is not well known in children. The other significant complications are bleeding, infections, postoperative pain and recurrence at the port site. Further in adults there are reports of respiratory arrest and subsequent death in patients with subcutaneous emphysema.

Conclusion

A rare pediatric case with impressive subcutaneous emphysema as a rare complication after VATS procedure. Emphysema slowly absorbed and high doses of painkillers were given to keep

P 177.**Linear IgA bullous dermatosis**

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*CHM Mayotte, CHU Liège*Introduction

Linear IgA dermatosis is a autoimmune disease with an incidence of 0.5-1/1 000 000. There are 2 presentations, pediatric around 5 years and adult. Clinical lesions are typical. The physiopathology is poorly understood but it seems to be a genetic sensitivity (HLA Cw7). We describe a case encountered in Mayotte.

Case presentation

We report a case of a 3-year-old child with no pertinent past. The patient presents a pruritic bullous eruption for 10 days. Parents report an octopus meal 12 hours before the rash appears. He is treated by antihistaminic and antibiotic (AMOXICLAV) for 7 days. There is no improvement and the rash spreads all over the body. He is hospitalized in pediatric.

Physical examination reveals vesiculo-bullous lesions of different ages all over the body, including face but sparing mucous membranes. Lesions are circular, irregular with central erosion, peripheral elevation and, sometimes, vesicles around the lesion ("rosette"). There are some isolated bullous vesicles. The content of the vesicles is citrin. There is no other symptom.

Laboratory testing shows leukocytosis with lymphocytosis and eosinophilia without inflammatory syndrom. Liver function test results, renal function, are all within normal range.

Dermatological advice suggests autoimmune disease, especially lupus, dermatitis herpetiformis, linear immunoglobulin A dermatosis and juvenile bullous pemphigoid.

The workup autoimmun (nuclear autoantibody, extractable nuclear antigens autoantibody, immunoglobulin assay, transglutaminase IgA autoantibodies, endomysium IgA autoantibodies) was normal. The biopsy skin showed a microscopic appearance of bullous dermatosis by subepidermal detachment with the presence in direct immunofluorescence of linear deposits of IgA at the dermo-epidermal junction. This description is compatible with linear IgA dermatosis.

A treatment by DAPSONE is started with corticoid cream. Development is favorable thereafter.

Conclusion

Linear IgA dermatosis should be part of the differential diagnosis for infant with bullous vesicle eruption. The skin biopsy confirms diagnosis. The physiopathology is poorly understood but it seems to be a genetic sensitivity (HLA Cw7). The choice treatment is DAPSONE (diaminodiphénylsulfone) and topical corticotherapy. The side effects of DAPSONE (anemia, methemoglobinemia) require a control of activity in G6PD before its introduction. Disease appears around 5 years old and rarely persisting after puberty.

P 178.

Time to diagnosis : about a case of juvenile dermatomyositis

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Background

Juvenile dermatomyositis is an idiopathic disease characterized by non-infectious inflammation of muscles and skin. Although rare, it is the most common autoimmune myopathy in children and can be an important cause of disability. Early therapeutic care has a positive impact on the prognosis.

Case

We reported hereby the case of a 12-year-old boy who was admitted at the emergency department with muscle pain in both arms, severe fatigue, weight loss, recent twangy voice and a heliotrope rash. First symptoms started 6 months before treatment first administration and the child saw 4 different specialists (a general practitioner, a dermatologist, an orthopedist and several emergency physicians) before a pediatrician evokes the diagnosis and refers the patient to a pediatric rheumatologist.

Results

The diagnosis of dermatomyositis seems to remain a challenge for practitioner who are not involved in inflammatory diseases in children, in particular because of the gradual onset of symptoms. This case highlights the long patient's journey before diagnosis.

Conclusion

It is interesting to observe the delay to diagnose this pathology in our country and see if this time could be shorter with more specific training for first line physicians.

P 179.**Infantile hemangioma : Don't miss the PHACE syndrome**

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*CHC, U Liège*Background

Infantile hemangioma (IH) is the most common benign soft-tissue tumor of childhood with an incidence of 5 to 10%.

IH usually does not occur with systemic involvement but extensive and segmental IH, especially on the face, scalp, and cervical region, can be associated with malformations of the posterior fossa of the brain, arterial anomalies of the central nervous system, coarctation of the aorta, cardiac defects and ocular abnormalities. This association, known as PHACE syndrome, was first described in 1996. PHACE syndrome is observed in 2 to 3% of IH cases. The likelihood of a segmental or large IH on the face being associated with PHACE syndrome is 20 to 30%. Pathogenesis is still unknown but studies on genetics are still ongoing.

Case report

We report on a full-term, normal birthweight boy with a large, segmental telangiectatic lesion on the face. An extensive workup found an absence of the left internal carotid artery, an hypoplasia of the left common carotid artery, an asymmetric development of cerebellar hemispheres and a retro-orbital hemangioma. These findings and the expected natural evolution of the facial IH confirmed the initial hypothesis of PHACE syndrome.

Oral propranolol was carefully introduced at 6 weeks of age and slowly increased to the target dose of 1,5 mg/kg/day, clinically effective and currently well-tolerated dose.

Conclusion

A systematic screening for PHACE syndrome must be performed in all infants with a large (>5cm diameter) segmental IH located on the face or scalp, based on a physical examination, echocardiogram, MRI and MRA of the head, neck and cervical vessels.

Results of initial screening will guide multidisciplinary care and future monitoring of related morbidities.

Regarding the increased stroke risk in PHACE patients with arterial anomalies, oral betablockers will be introduced slower and at lower doses than in other IH requiring systemic treatment.

P 180.

Lipschütz ulcer: diagnosis and management. About an unusual case of recurrence and a rapidly favorable course with topical corticosteroid therapy.

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Introduction

Lipschütz ulcer is a rare clinical entity that usually occurs in young women. It manifests as acute ulcers. The diagnosis is based on history and clinical examination. Triggering factors can be encountered and relapse is unusual.

Case Report

We describe the case of a very young 9-year-old patient admitted to our clinic for an extremely painful acute vulvar ulceration with fever and influenza-like syndrome. The clinical examination showed a typical appearance of Lipschütz ulcer ("kissing lesions"). Herpes simplex virus and venereal bacterial infections were locally ruled out but the *Mycoplasma pneumoniae* serology was found positive. She presented an unusual recurrence after treatment with topical corticosteroid despite an initial rapidly favorable outcome with complete remission without scarring, with the same lesions and a same good response to topical corticosteroids. No relapse has been mentioned more than six months later.

Discussion

Lipschütz ulcer is an uncommon entity that is characterized by an acute painful necrotic vulvar ulcer. It typically occurs in young and sexually inactive women. The lesions are usually large and deep with a necrotic center. The bilateral symmetrical appearance known as "kissing lesions" is really characteristic. Lesions may be preceded by prodromal symptoms. The diagnosis is based on history and clinical examination. Additional investigations are useful to exclude other causes such as sexually transmitted infections or non-infectious diseases. Etiology can be idiopathic or associated with other infections as *Mycoplasma*. Healing occurs in two to six weeks, usually without scarring and without any recurrence. Treatment includes effective pain relief, wound care and topical corticosteroids or sometimes systemic corticosteroids can be considered.

After literature review, we propose to use the major and minor criteria defined by Farhi et al. to facilitate the diagnosis and we suggest the decision-making algorithm from Rosman et al. to determine the best approach.

Conclusion

Lipschütz ulcer is rare. Genital lesions are often typical ("kissing lesions"). Diagnosis is based on history and clinical examination. Lipschütz ulcer is a diagnosis of exclusion and other investigations are useful to exclude other causes. The treatment is first symptomatic with pain management but sometimes the use of corticosteroid is necessary. Recurrence is rare and healing is generally complete.

P 181.**Macrodystrophia Lipomatosa: a little bit “toe” much.**

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An otherwise healthy girl was born at a postmenstrual age of 38 weeks after an uncomplicated pregnancy with an enlargement of the first and second left toe. Clinical examination showed a bigger aspect of the left foot with an increased length and circumference of the first and second toe. No additional abnormalities were observed during clinical examination.

Plain radiography and MRI of the left foot revealed cortical thickening of and abundant fibrofatty tissue around the involved phalanges. Histology showed abundant fatty tissue infiltrating the dermal connective tissue, scattered in a fine, mesh-like fibrous tissue. Genetic testing identified a heterozygote c.1624G>A (p.Glu542Lys) missense mutation in the PIK3CA gene in 69% of the cells of the affected tissue (dermis/subcutis of the biopsy specimen), confirming the presumed clinical diagnosis.

The diagnosis of Macrodystrophia Lipomatosa (MDL), a rare congenital form of partial gigantism affecting all mesenchymal elements of the involved body part, was made. Recently an underlying gain-of-function mutation in the PI3K-AKT-MTOR pathway was identified causing this peculiar clinical, radiological and histological presentation.

P 182.

Infantile transient smooth muscle contraction

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UZ Leuven

Infantile transient smooth muscle contraction is a rather unknown disorder, which is not described in textbooks. In Pubmed only a few reports mention this entity. We would like to highlight the existence of this condition, in order to create awareness among fellow pediatricians and avoid unnecessary investigations.

We describe a healthy 3 month old boy, who presented to the paediatrics department after referral. Since birth, this child showed daily transient rippling of the lower limbs. This cellulite-like skin occurred mostly after temperature changes, such as cold air exposure or bathing, and gentle touching. It particularly involved both knees, although expansion to the lower legs was possible. No color change was noted. The infant did not experience pain or discomfort. It generally lasted a few minutes before the skin gradually returned back to normal. A beneficial effect of rubbing of the limbs was noted. At the moment of consultation the clinical examination did not show any abnormalities. There were no associated skin lesions. A duplex ultrasound was performed and could exclude the presence of a vascular malformation or lymphangioma. No biopsy was performed. The diagnosis of infantile transient smooth muscle contraction was made, based on limited reports in literature. The child was thereafter discharged, without further investigations or treatment. The rippling disappeared after 2 years.

Infantile transient smooth muscle contraction was first described in 2013 and is not widely known among pediatricians. This phenomenon is characterized by transient, localized rippling of the skin, mostly the limbs. It is suggested to be caused by intermittent contraction of the smooth muscle fibers, as a result of autonomic nervous system immaturity. The frequency and intensity of the rippling gradually decrease with age, after which it fully disappears. Pediatricians should keep the existence of this entity in mind when examining neonates, in order to avoid unnecessary testing or treatment, and thus prevent needless anxiety in parents.

P 183.**Ankyloblépharon filiforme adnatum : a case report.**

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Introduction

Ankyloblepharon Filiforme Adnatum (AFA) is a rare but potentially amblyogenic congenital palpebral malformation described for the first time in 1881 by Von Hasner. It is characterized by a single or multiple fine bands of an extensible tissue, joining the upper and the lower eyelids. The prevalence is unknown, with only few isolated cases described in the literature. AFA may be isolated but it is more frequently associated with others abnormalities

In 1980, Rosenman proposed a classification of AFA into 4 groups.

- Type I includes cases in which no associated defect appears.
- Type II includes cases associated with cardiac, digestive or central nervous system defects.
- Type III occurs in association with ectodermal syndromes.
- Type IV occurs in association with cleft lips and/or palates.

In 1993, Baccal added a type V which includes AFA association with chromosomal abnormalities

In 2007, Williams added a type VI which includes the same features as type I but where there is also a family history of isolated congenital AFA.

The treatment consists of a simple resection of the fibrous bands as soon as possible without the need of anesthesia or sedation. In addition to the treatment, a physical examination and other investigations must be carried out to search for associated malformations.

Methods

Case report

Results

The patient is a full-term female baby with an unremarkable gestation. She's the first baby of a couple without any health problems. There is no history of congenital abnormalities or consanguinity in the family. During the first hour of life, she was noted to have bands of fibrous tissue joining the upper and lower lids of her eyes.

A detailed clinical examination was realised and no further malformations were found. An ophtalmic examination diagnosed AFA. The karyotype and the brain and cardiac ultrason were normal.

During the second week of life, we noted a spontaneous resolution of the fibrous bands in the left eye. At the 20th day of life, the fibrous bands of the right eye were excised without anesthesia or sedation by the ophtalmologist

The patient was seen in consultation at 2 months of life with a favourable evolution.

Conclusion

AFA is a rare but potentially amblyogenic congenital abnormality of the eyelids. A simple treatment minimises any risk of amblyopia and enables a full examination of the eye. Its presence should alert the pediatrician to the possibility of associated congenital anomalies.

P 184.

A pediatric case of aerodigestive thermal burn after ingestion of hot milk

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ULB – HUDERF

Background

Thermal burns (TB) of the aerodigestive tract after ingestion of hot beverages are rare compared to caustic burns. However they can be serious and require specific considerations. This case illustrates the management of a young patient with oropharyngeal and esophageal TB.

Case report

A 2-year-old boy without any medical history was admitted to our PICU for the management of TB of the upper airways and esophagus. Four hours after swallowing a bottle of hot milk, his parents (absent at the time of the event) brought him in the Emergency Department and reported stridor, drooling and asthenia. Clinical examination confirmed breathing difficulties. To secure airways, intubation with a 3.5 mm diameter cuffed endotracheal tube was performed blindly by an experienced intensivist, because of inflammatory fibrinous magma at the glottic level. Nasofibroscopy showed significant edema and fibrinous deposit on the supra-glottic structures. Upper gastrointestinal endoscopy revealed friable esophageal mucosa of the upper two thirds, whitish membranes and circumferential ulcerations. High dose steroid therapy was administered for 3 days allowing extubation at day 5. Nasogastric tube was placed on arrival. Enteral feeding was started on day 3. Proton pump inhibitors were administered to prevent gastroesophageal reflux lesions. Antimicrobial prophylaxis was initiated for 7 days. The child left the ICU on day 6. Follow-up of lesions was carried out by endoscopy which demonstrated a rapidly favorable evolution.

Discussion

In the literature, few cases of oropharyngeal TB are described after ingestion of boiling beverages. There are currently no guidelines for the management of TB with oropharyngeal and upper digestive involvement. Our patient was therefore treated based on existing data concerning caustic esophageal burns (CEB) as well as on the clinical and endoscopic course of the lesions. No data exists regarding potential long-term complications such as stenosis or malignant transformation in these cases.

Conclusion

We present the case of a child affected by aerodigestive TB with hot neutral fluid. In absence of specific guidelines, management was based on CEB. Rapid favorable evolution under treatment was observed allowing early extubation and enteral nutrition without complications. Nevertheless, we recommend starting with maximalist supportive care and adapting treatment rapidly according to clinical and endoscopic evolution.

P 185.**The European paediatric clinical trials network beyond the Atlantic: the collaboration between I-ACT for children (US) and European network activity**

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Background/Aims

Paediatric clinical trial networks have made many advances over the last few years. In Europe, the Innovative Medicines Initiative (IMI)-funded conect4children (c4c) initiative has promoted innovation and standardization within all levels of paediatric drug development.

Method

This report describes a trans-Atlantic collaboration to conduct trial feasibility with I-ACT for Children (Institute for Advanced Clinical Trials for Children), a US-based non-profit organization for the advancement of paediatric clinical trials. An industry sponsored study was presented to European national paediatric network coordinators by I-ACT for Children. Results: An early engagement survey was conducted, followed by sites signing a Confidentiality Disclosure Agreement (CDA), receiving the protocol synopsis, and conducting a full site feasibility questionnaire. The CDA process was completed in 7 European countries within 12 days. The four-step process until full-feasibility report was completed in a total of over 25 sites Europe-wide within a 12-week mark.

Conclusion

This successful and efficient collaboration paves the way for a broader platform for paediatric drug development. Platforms such as c4c and i-ACT for Children are essential to achieve substantial improvement in medicines for children in EU, the US and worldwide.

P 186.

Linear IgA Bullous Dermatitis challenging recurrent bullous impetigo

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Background

Linear IgA Bullous Dermatitis (LABD) is a rare bullous disease, characterized by the linear deposit of IgA at the dermoepidermal junction. It is the most common autoimmune blistering disease in children.

Case Report

We report a case of a 14 month-girl, who first came into the Emergency Department with a skin rash and peri-oral vesicular and crusty lesions. Some other lesions were also found on the arms, thighs, and perineum. Despite positive lesion cultures, unsatisfactory lasting improvement was achieved after systemic and topical antistaphylococcal antibiotic. A negative PCR analysis of a bullae ruled out chickenpox. She presented a second burst with more severe and extensive tense bullae on the face and limbs associated with oozing and vesicular erythema in the napkin area. Pruritus was intense. Some lesions had an annular appearance like a string of pearls and suggested LABD. A skin biopsy was performed to confirm diagnosis. Standard histopathologic examination with direct immunofluorescence (DIF) demonstrated a subepidermal bullous dermatosis and linear IgA deposits along the basement membrane. After pre-therapeutic assessment, a Dapsone therapy was started and clinical improvement was noted within the first two weeks. Discussion: LABD is a rare cutaneous disorder but the most common autoimmune blistering disease in children. The complete pathogenesis is unclear, but the skin lesions may result from an antibody-induced local inflammatory response. It may be idiopathic or drug-induced. The childhood clinical presentation is characterized by tense vesicles and bullae with annular appearance (“string of pearls”) usually localized on the face, ears, and genitals. Diagnosis is based on histopathology and confirmed by DIF. Differential diagnosis includes more common bullous impetigo and other autoimmune bullous disorder like dermatitis herpetiformis or bullous pemphigoid. Dapsone is the first line treatment. It has to be administered carefully because of well-known adverse effects. Our case showed a typical presentation without any triggering factor and experienced a good response to treatment.

Conclusion

LABD is a rare cutaneous disorder but the most common autoimmune blistering disease in children. It can be easily mistaken for other common cutaneous lesions like bullous impetigo. The presence of more inflammatory recurrent bullous lesions and the “string of pearls” sign is suggestive. A skin biopsy is mandatory to confirm diagnosis.

P 187.**Multiple cutaneous abscesses due to *Pseudomonas aeruginosa***

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Pseudomonas aeruginosa (PA) is rarely encountered in skin lesions in children, except in ecthyma gangrenosum or bathtub folliculitis. To our knowledge, multiple cutaneous abscesses in a healthy infant have never been reported in the literature so far.

We report the case of a 6-month-old girl with a 3-day course of fever and bronchitis. She presented with three erythematous cutaneous nodules on the right arm and the left leg. A blood analysis showed a C-Reactive Protein at 177.1 mg/L with a hyperleukocytosis. Rapidly the nodules reached the number of eight and a systemic antibiotherapy was started with intramuscular ceftriaxone first due to lack of easy venous access and then intravenous amoxicilline plus clavulanic acid. A neutropenia appeared while the CRP level decreased. In the following days, the nodules evolved into abscesses. A biopsy of a nodule showed panniculitis with pus, leukocytes infiltrate and the culture grew full of PA. Surgical punctures of the other abscesses also drained off pus with cultured PA. The child went rapidly well. The plastic bathtub, the tap, the aerosol mask and connector, and the « Babycook » device were tested negative. Her twin brother and the family had no cutaneous lesion. Systemic antibiotics were discontinued and oral ciprofloxacin was started during two weeks. Repeated blood cultures remained negative. An extensive work-up did not reveal a deep infectious source (heart, abdomen, bone). The humoral immunity was normal. The child experienced a progressive decrease of the abscesses with and after antibiotics.

Nodular panniculitis due to PA is a rare condition. Differential diagnosis with erythema nodosum or other causes of panniculitis may be challenging before evolution to abscess. A positive bacteremia, an infected skin wound or contaminated liquid containing devices are often considered the origin of the cutaneous lesions.

This report aims to answer questions such as the source of the contamination, the usefulness of the antibiotics in the treatment, the need for surgical drainage, and the link between leukopenia and infection or immunodeficiency.

P 188.

Development of a rapid molecular diagnosis test for acute otitis media

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ULB – HUDERF

Background/Aims

Acute otitis media (AOM) is the main reason for antibiotic prescriptions in paediatrics. This infection could result in severe complications (mastoiditis, meningitis and hearing loss). The etiological agent in AOM can be viral or bacterial. An antibiotic treatment is required only in bacterial AOM as it reduces symptoms and limits complications. However, the etiologic diagnosis of AOM is difficult as the middle ear is usually inaccessible to microbiological sampling. Moreover, the 5 most common etiological agents of AOM (*Streptococcus pneumoniae*, *Streptococcus pyogenes*, *Haemophilus influenzae*, *Moraxella catarrhalis* and *Staphylococcus aureus*) are associated with naso-pharyngeal carriage making diagnosis difficult. Thus, antibiotic treatment is decided on empirical basis, despite variations in antibiotic susceptibility among pathogens and without knowing if the infection is viral or bacterial. This leads to a high level of treatment failure, increases risk of adverse effects and contributes to the global problem of antibiotic resistance. The goal of this project is to develop a rapid 'point of care' (POC) molecular diagnosis test which will identify the bacterial etiologic agent and its antibiotic resistance genes from a blood drop. This test will help discriminating between a carriage state and an acute infection by detecting RNA of virulence genes in patient blood.

Methods

The first part consisted in the identification of the target genes and experimental validation of the primers drawn. We began by performing a bioinformatics analysis of databases (Genbank, EBI) and a literature review to select the best molecular targets. Our research focused on the detection of conserved genes encoding for virulence factors (in order to distinguish pathogens and carriage).

Results

By in silico analysis, we have identified 1-4 target genes for each pathogen. We have designed specific primers/probes and validated them experimentally on genomic DNA and complementary DNA by qPCR. We have also analysed their specificity on clinical and reference strains collection for each pathogen.

Conclusion

Etiologic diagnosis of AOM is difficult leading to empirical antibiotic treatment. We are working on the development of a rapid molecular diagnostic test which could guide clinicians about the causal pathogen and the appropriate treatment for their patients. We have already identified at least one target per pathogen to use in a future POC diagnostic test.

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