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Belgische Vereniging voor Kindergeneeskunde
Société Belge de Pédiatrie

DYNAMICS IN PEDIATRICS

THE EGG, Brussels
21 & 22.03.2019

Gunnar Buyse, MD PhD
Congress President,
UZ Leuven

Anne Malfroot, MD PhD
President BVK/SBP

Belgische Vereniging voor Kindergeneeskunde
Société Belge de Pédiatrie

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Dear colleagues and friends,

It is a privilege and pleasure to welcome you at “DYNAMICS IN PEDIATRICS’, the 47th Annual Congress of the Belgian Society of Pediatrics (BVK-SBP).

As pediatricians and pediatric subspecialists, we all share the passion for bringing the very best pediatric medicine and health care to children and adolescents. Pediatric medicine is fascinating, dynamic and constantly evolving, as is our beautiful profession.

The Scientific Committee has put together a cutting edge scientific program for this congress, with renowned national and international speakers, addressing topics relevant for the clinical practice of both general and specialized pediatricians. The program includes four plenary sessions: ‘Windows of Opportunity’ and ‘Epigenetics & Environment’ on Thursday, ‘Technology & Pediatrics: the Future’ and ‘Growing Up in a Complex World’ on Friday. Four thematic multidisciplinary subspecialty sessions address topics relevant for the practice of a pediatrician: (i) ‘Children and (competitive) sport’, (ii) ‘The role of the pediatrician in the follow-up of the child requiring complex chronic care’, (iii) ‘Dealing with new therapies in daily pediatric practice’; and (iv) ‘Pediatric acute care’. New in this year’s edition of the Annual Congress is a plenary session hosted by pediatric trainees. This session is focusing on technology & pediatrics, on how technology and applications could impact the future of medicine and pediatric clinical practice.

Out of the large number of high-quality abstracts submitted, thirteen were selected for oral presentation (sessions ‘Oral Abstracts I & II’ on Thursday and Friday), and 36 were selected for a short communication (3 parallel ‘Poster Presentation’ sessions on Thursday). The remaining abstracts (posters) are on display in the Poster area (Riverside), please do visit the poster area and interact with the authors.

Please do join us on Thursday evening (- 6.30 pm, immediately after Plenary Session 2 -) for the BVB-SBP Celebration. This is truly ‘a can’t miss moment’, with drinks & snacks, entertainment by JeJe Magic and Tomorrowland DJ Christophe from The Knightriders, and a fun attraction by young VVK. And last but not least: amongst the persons present at the celebration, we will hand out two otherwise impossible-to-get DUO VIP DAY TICKETS for Tomorrowland, the largest dance music festival worldwide.
I wish to express my sincere gratitude to the organizing committee, to the scientific committee, to the board of the BVK-SBP, to ACT-wise, to all sponsors, to our international and national faculty, to all abstract presenters, to the session chairs, to all of you attending this 47th annual meeting of our society.

Sincerely yours,
Gunnar Buyse, MD PhD, UZ Leuven
Congress President BVK-SBP 2019

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BVK/SBP Congress 2019
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A la vie

CARDIOLOGY

Oral presentations

O1. **Univentricular heart malformation: outcome after (prenatal) diagnosis**
L Duchi, S Lesage, K Vandekerckhove, K Van Herck, D De Wolf, J Panzer, H De Wilde, K François, T Bove, K De Groote. UZ Gent

Posters with short oral presentations

OP1. **Effects of chronic ventricular volume deprivation and acute reloading studied in an animal (ovine) model**
B Cools, F Rega, P Claus, L Fresiello, J Duchenne, T Verbelen, M Gewillig. UZ Leuven, KU Leuven

OP2. **Percutaneous lymphangiosclerosis as treatment for protein loosing enteropathy and plastic bronchitis in patients with failing Fontan circulation**
M Gewillig, E Storme, B Eyskens, B Cools, R Heying, D Boshoff, J Hubrechts, G Maleux. UZ Leuven

Posters

P37. **Non-invasive assessment of the liver in pediatric Fontan patients: time for thorough follow-up**
K Vandekerckhove, R De Bruyne, F Hendricx, H Van Overschelde, C Vande Walle, K De Groote, J Panzer, D De Wolf, S Van Biervliet, T Bové, K François. UZ Gent

P38. **Univentricular heart malformation: outcome after surgery and importance of isomerism**
L Duchi, K Van Herck, K Vandekerckhove, D De Wolf, J Panzer, H De Wilde, K François, T Bove, K De Groote. UZ Gent

P39. **Percutaneous closure of a window-like patent arterial duct with an ASD occluder**
K Vandekerckhove, J Panzer, D De Wolf, UZ Gent

P40. **Long term outcome after tricuspid valvectomy in a neonate with candida endocarditis: a case report**
E SI Vanhie, W A Helbing. UZ Gent, Erasmus university Medical Centre - Sophia Childrens' hospital Rotterdam

P41. **Interventional closure of perimembranous ventricular septal defects: Experience with two commonly used occluding devices**
Q Jordens, R Heying, B Cools, B Eyskens, J Hubrechts, M Roggen, M Gewillig. UZ leuven

P42. **Cardiac fibroma in children: single center experience**
AS Alderweireldt, W Decaluwe, K Vandekerckhove, J Panzer, K De Groote, T Bové. UZ Gent, AZ Sint-Jan Brugge

P43. **Percutaneous obliteration of the Right Ventricle in patients witch pulmonary atresia, intact ventricular septum as management strategy to avoid corona**
B Cools, B Eyskens , R Heying, D Boshoff, B Suys, M Gewillig. UZ Leuven

P44. **Complex malignant exercise induced ventricular tachyarrythmia in a girl with Turner Syndrome**
J Van Huffel, AS Crochelet, AS Parent, L Van Casteren, MC Seghaye. Hôpital Universitaire Liège, Université Liège

P45. **Evolution to a native aortic isthmus stenosis in a boy**
MC Seghaye. CHU Liège

Publieksprijs België **86,52€**

Bexsero: het eerste vaccin tegen meningokokken van serogroep B.

Het enige geïndiceerd vanaf 2 maanden.[1,2]

BEXSERO

Vaccin tegen meningokokken van groep B (rDNA, component, geadsorbeerd)

VERKORTE SAMENVATTING VAN DE PRODUCTKENMERKEN Gelieve de Samenvatting van de Produktkenmerken te raadplegen voor de volledige informatie over het gebruik van dit geneesmiddel. ▼ Dit geneesmiddel is onderworpen aan aanvullende monitoring. Daardoor kan snel nieuwe veiligheidsinformatie worden vastgesteld. Beroepsbeoefenaren in de gezondheidszorg wordt verzocht alle vermoedelijke bijwerkingen te melden. Zie rubriek "Bijwerkingen" voor het rapporteren van bijwerkingen. NAAM VAN HET GENEESMIDDEL Bexsero suspensie voor injectie in voorgevulde spuit. Meningokokken groep B vaccin (rDNA, component, geadsorbeerd) EU/1/12/812/001. Farmacotherapeutische categorie: meningokokkenvaccins, ATCcode: J07AH09 KWALITATIEVE EN KWANTITATIEVE SAMENSTELLING Een dosis (0,5 ml) bevat: Recombinant ***Neisseria meningitidis*** groep B NHBAfusieeiwit [1, 2, 3] : 50 microgram Recombinant ***Neisseria meningitidis*** groep B NadAeiwit [1, 2, 3] : 50 microgram Recombinant ***Neisseria meningitidis*** groep B fHbpfusieeiwit [1, 2, 3] : 50 microgram Buitenmembraanvesikels (BMV) van ***Neisseria meningitidis*** groep Bstam NZ98/254, gemeten als hoeveelheid totaal eiwit dat PorA P1.4 bevat [2] : 25 microgram [1] Geproduceerd in ***E. coli***cellen door recombinantDNAtechnologie [2] Geadsorbeerd aan aluminiumhydroxide (0,5 mg Al^{3+}) [3] NHBA (Neisseria heparinebindend antigeen), NadA (Neisseriaadhesine A), fHbp (factor Hbindend eiwit) THERAPEUTISCHE INDICATIES Bexsero is geïndiceerd voor de actieve immunisatie van personen van 2 maanden en ouder tegen invasieve meningokokkenziekte veroorzaakt door ***Neisseria meningitidis*** groep B. Bij het vaccineren moet rekening worden gehouden met het effect van invasieve ziekte bij verschillende leeftijdsgroepen, evenals met de variabiliteit van de epidemiologie van antigenen voor groep Bstammen in verschillende geografische gebieden. Zie rubriek 5.1 van de volledige SPK voor informatie over bescherming tegen specifieke groep Bstammen. Dit vaccin dient te worden gebruikt in overeenstemming met officiële aanbevelingen. DOSERING EN WIJZE VAN TOEDIENING Dosering

Tabel 1. Samenvatting van de dosering

Leeftijd bij eerste dosis	Primaire immunisatie	Intervallen tussen primaire doses	Booster
Zuigelingen van 2 tot en met 5 maanden [a]	Drie doses, elk van 0,5 ml	Niet minder dan 1 maand	Ja, één dosis tussen 12 en 15 maanden oud met een interval van ten minste 6 maanden tussen de primaire serie en de boosterdosis [b, c]
Zuigelingen van 3 tot en met 5 maanden	Twee doses, elk van 0,5 ml	Niet minder dan 2 maanden	
Zuigelingen van 6 tot en met 11 maanden	Twee doses, elk van 0,5 ml	Niet minder dan 2 maanden	Ja, één dosis in het tweede levensjaar met een interval van minimaal 2 maanden tussen de primaire serie en de boosterdosis [c]
Kinderen van 12 tot en met 23 maanden	Twee doses, elk van 0,5 ml	Niet minder dan 2 maanden	Ja, één dosis met een interval van 12 tot en met 23 maanden tussen de primaire serie en de boosterdosis [c]
Kinderen van 2 tot en met 10 jaar	Twee doses, elk van 0,5 ml	Niet minder dan 1 maand	Noodzaak niet vastgesteld [d]
Adolescenten (11 jaar of ouder)en volwassenen*	Twee doses, elk van 0,5 ml	Niet minder dan 1 maand	Noodzaak niet vastgesteld [d]

[a] De eerste dosis moet niet worden gegeven op de leeftijd jonger dan 2 maanden. De veiligheid en werkzaamheid van Bexsero bij zuigelingen jonger dan 8 weken zijn nog niet vastgesteld. Er zijn geen gegevens beschikbaar. [b] In geval van uitstel mag de booster niet later dan op een leeftijd van 24 maanden worden gegeven. [c] Zie rubriek 5.1 van de volledige SPK. De noodzaak voor een boosterdosis na dit vaccinatieschema is niet vastgesteld [d] Zie rubriek 5.1 van de volledige SPK. * Gegevens over volwassenen ouder dan 50 jaar ontbreken. Wijze van toediening Het vaccin wordt toegediend via een diepe intramusculaire injectie, bij voorkeur in het anterolaterale gedeelte van de dij bij zuigelingen, of in de streek van de deltaspier van de bovenarm bij oudere personen. Als meer dan één vaccin tegelijk wordt toegediend, moeten afzonderlijke injectieplaatsen worden gebruikt. Het vaccin mag niet intraveneus, subcutaan of intradermaal worden toegediend, en mag niet worden gemengd met andere vaccins in dezelfde spuit. Voor instructies over het hanteren van het vaccin voorafgaand aan toediening, zie rubriek 6.6 van de volledige SPK. CONTRAINDICATIES Overgevoeligheid voor de werkzame stof(fen) of voor een van de in rubriek 6.1 van de volledige SPK vermelde hulpstof(fen). BIJZONDERE WAARSCHUWINGEN EN VOORZORGEN BIJ GEBRUIK Zoals dat voor alle vaccins geldt, dient ook toediening van Bexsero te worden uitgesteld bij personen die lijden aan een acute, ernstige, met koorts gepaard gaande ziekte. De aanwezigheid van een lichte infectie, zoals verkoudheid, mag echter niet leiden tot uitstel van vaccinatie. Niet intravasculair injecteren. Zoals dat voor alle injecteerbare vaccins geldt, dienen passende medische behandeling en toezicht altijd direct beschikbaar te zijn voor het geval zich na toediening van het vaccin een anafylactische reactie voordoet. Reacties die verband houden met angst, waaronder vasovagale reacties (syncope), hyperventilatie of stressgerelateerde reacties, kunnen in relatie met vaccinatie voorkomen als psychogene reactie op de naaldinjectie (zie rubriek "Bijwerkingen"). Het is belangrijk dat er passende procedures zijn om letsel als gevolg van flauwvallen te voorkomen. Dit vaccin mag niet worden toegediend aan personen met trombocytopenie of een bloedstollingsstoornis die een contraindicatie voor intramusculaire injectie vormt, tenzij het mogelijke voordeel duidelijk opweegt tegen het risico van toediening. Zoals dat voor alle vaccins geldt, beschermt vaccinatie met Bexsero mogelijk niet alle gevaccineerden. Bexsero wordt niet geacht bescherming te bieden tegen alle circulerende meningokokken Bstammen. Zoals dat voor veel vaccins geldt, moet het medisch personeel zich ervan bewust zijn dat een temperatuursstijging kan optreden na vaccinatie van zuigelingen en kinderen (jonger dan 2 jaar). Profylactische toediening van antipyretica gelijktijdig met en meteen na vaccinatie kan de incidentie en intensiteit van koortsreacties na vaccinatie verminderen. Antipyretische medicatie dient te worden gestart volgens de lokale richtlijnen bij zuigelingen en kinderen (jonger dan 2 jaar). Individuen met een immunodeficiëntie, door het gebruik van immunosupressieve therapie, een genetische stoornis, of door een andere oorzaak, kunnen een verlaagde antilichaamrespons hebben bij actieve immunisatie. Immunogeniciteitsgegevens zijn beschikbaar van individuen met complement deficiëntie, asplenie of miltdisfuncties. Er zijn geen gegevens over het gebruik van Bexsero bij personen ouder dan 50 jaar en beperkte gegevens bij patiënten met chronische medische aandoeningen. Wanneer de primaire immunisatieserie aan zeer premature zuigelingen (geboren na ≤ 28 weken zwangerschap) wordt toegediend, moet rekening worden gehouden met een potentieel risico op apneu en de noodzaak van controle van de ademhaling gedurende 4872 uur, vooral bij zuigelingen met een voorgeschiedenis van onvolgroeide longen. Aangezien het voordeel van vaccinatie groot is bij deze groep zuigelingen, moet vaccinatie niet worden onthouden of uitgesteld. De dop van de injectiespuit bevat mogelijk natuurlijk rubber (latex). Hoewel het risico op het ontwikkelen van allergische reacties zeer klein is, moet het medisch personeel de voor en nadelen goed afwegen voordat dit vaccin wordt toegediend aan personen met een bekende voorgeschiedenis van overgevoeligheid voor latex. Kanamycine wordt aan het begin van het productieproces gebruikt en wordt in latere productiestadia verwijderd. Indien aanwezig, bedraagt het kanamycineniveau in het uiteindelijke vaccin minder dan 0,01 microgram per dosis. Veilig gebruik van Bexsero bij personen die gevoelig zijn voor kanamycine is niet vastgesteld. BIJWERKINGEN Overzicht van het veiligheidsprofiel De veiligheid van Bexsero is geëvalueerd in 17 onderzoeken, inclusief 10 gerandomiseerde gecontroleerde klinische studies met 10.565 proefpersonen (vanaf de leeftijd van 2 maanden) die minimaal één dosis Bexsero toegediend kregen. Van de personen die Bexsero toegediend kregen, waren 6.837 zuigelingen en kinderen (jonger dan 2 jaar), 1.051 kinderen (van 2 tot 10 jaar) en 2.677 adolescenten en volwassenen. Van de proefpersonen die de primaire immunisatieserie voor zuigelingen van Bexsero toegediend kregen, kregen 3.285 een boosterdosis in het tweede levensjaar. De meest voorkomende lokale en systemische bijwerkingen bij zuigelingen en kinderen (jonger dan 2 jaar) die in klinische studies zijn waargenomen, waren gevoeligheid en erytheem op de injectieplaats, koorts en prikkelbaarheid. In klinische onderzoeken bij zuigelingen gevaccineerd op de leeftijd van 2, 4 en 6 maanden, is bij 69% tot 79% van de proefpersonen melding gemaakt van koorts (≥ 38**°C**) wanneer Bexsero gelijktijdig werd toegediend met standaardvaccins (die de volgende antigenen bevatten: 7valent pneumokokkenconjugaat, difterie, tetanus, acellulair pertussis, hepatitis B, geïnactiveerde poliomyelitis en ***Haemophilus influenzae*** type b) in vergelijking met 44% tot 59% van de proefpersonen die alleen de standaardvaccins kregen toegediend. Bij zuigelingen die Bexsero en standaardvaccins toegediend kregen, is ook vaker melding gemaakt van het gebruik van antipyretica. Wanneer alleen Bexsero werd toegediend, kwam koorts bij zuigelingen even vaak voor als bij standaardzuigelingenvaccins die tijdens klinische studies werden toegediend. Eventuele koorts volgde in het algemeen een voorspelbaar patroon, waarbij de meeste koortsgevallen de dag na de vaccinatie over waren. De meest voorkomende lokale en systemische bijwerkingen waargenomen bij adolescenten en volwassenen waren pijn op de injectieplaats, malaise en hoofdpijn. Er is geen toename waargenomen in de incidentie of ernst van bijwerkingen bij opeenvolgende doses in de vaccinatiereeks. Tabel met bijwerkingen Bijwerkingen (na primaire immunisatie of boosterdosis) die ten minste als mogelijk gerelateerd aan de vaccinatie kunnen worden beschouwd, zijn naar frequentie ingedeeld. De frequentie is als volgt geclassificeerd: Zeer vaak: (≥1/10) Vaak: (≥1/100, <1/10) Soms: (≥1/1.000, <1/100) Zelden: (≥1/10.000, <1/1.000) Zeer zelden: (<1/10.000) Niet bekend: (kan met de beschikbare gegevens niet worden bepaald) De bijwerkingen worden binnen elke frequentiegroep gerangschikt in aflopende volgorde van ernst. Naast de meldingen uit klinische onderzoeken, zijn ook de wereldwijd ontvangen vrijwillige meldingen over bijwerkingen van Bexsero sinds de introductie op de markt in de volgende lijst opgenomen. Aangezien deze bijwerkingen vrijwillig zijn gemeld door een populatie van onbekende omvang, is het niet altijd mogelijk om een betrouwbare schatting van de frequentie te geven en worden ze daarom hier vermeld met de frequentie Niet bekend. **Zuigelingen en kinderen (tot en met 10 jaar)** Immuunsysteemaandoeningen Niet bekend: allergische reacties (waaronder anafylactische reacties) Voedings en stofwisselingsstoornissen Zeer vaak: eetstoornissen Zenuwstelselaandoeningen Zeer vaak: slaperigheid, ongewoon huilen, hoofdpijn Soms: insulten (inclusief febriele insulten) Niet bekend: hypotoon – hyporesponsieve episode Bloedvataandoeningen Soms: bleekheid (zelden na booster) Zelden: ziekte van Kawasaki Maagdarmstelselaandoeningen Zeer vaak: diarree, braken (soms na booster) Huid en onderhuidaandoeningen Zeer vaak: huiduitslag (kinderen van 12 tot en met 23 maanden) (soms na booster) Vaak: huiduitslag (zuigelingen en kinderen van 2 tot en met 10 jaar) Soms: eczeem Zelden: urticaria Skeletspierstelsel en bindweefselaandoeningen Zeer vaak: artralgie Algemene aandoeningen en toedieningsplaatsstoornissen Zeer vaak: koorts (≥38°C), gevoeligheid op de injectieplaats (inclusief ernstige gevoeligheid op de injectieplaats, gedefinieerd als huilen wanneer geïnjecteerde ledemaat wordt bewogen), erytheem op de injectieplaats, zwelling op de injectieplaats, verharding op de injectieplaats, prikkelbaarheid Soms: koorts (≥40°C) Niet bekend: injectieplaatsreacties (inclusief uitgebreide zwelling van de gevaccineerde ledemaat, blaren op of rondom de injectieplaats en een nodus op de injectieplaats die meer dan een maand kan aanhouden) **Adolescenten (van 11 jaar en ouder) en volwassenen** Immuunsysteemaandoeningen Niet bekend: allergische reacties (waaronder anafylactische reacties) Zenuwstelselaandoeningen Zeer vaak: hoofdpijn Niet bekend: syncope of vasovagale reacties op injectie Maagdarmstelselaandoeningen Zeer vaak: misselijkheid Skeletspierstelsel en bindweefselaandoeningen Zeer vaak: myalgie, artralgie Algemene aandoeningen en toedieningsplaatsstoornissen Zeer vaak: pijn op de injectieplaats (inclusief ernstige pijn op de injectieplaats, gedefinieerd als niet in staat normale dagelijkse activiteiten uit te voeren), zwelling op de injectieplaats, verharding op de injectieplaats, erytheem op de injectieplaats, malaise Niet bekend: koorts, injectieplaatsreacties (inclusief uitgebreide zwelling van de gevaccineerde ledemaat, blaren op of rondom de injectieplaats en een nodus op de injectieplaats die meer dan een maand kan aanhouden) **Melding van vermoedelijke bijwerkingen** Het is belangrijk om na toelating van het geneesmiddel vermoedelijke bijwerkingen te melden. Op deze wijze kan de verhouding tussen voordelen en risico's van het geneesmiddel voortdurend worden gevolgd. Beroepsbeoefenaren in de gezondheidszorg wordt verzocht alle vermoedelijke bijwerkingen te melden via het nationale meldsysteem: **België** Federaal agentschap voor geneesmiddelen en gezondheidsproducten Afdeling Vigilantie EUROSTATION II Victor Hortaplein, 40/40 B-1060 Brussel Website: www.fagg.be e-mail: adversedrugreactions@fagg-afmps.be **Luxemburg** Direction de la Santé – Division de la Pharmacie et des Médicaments Villa Louvigny – Allée Marconi L-2120 Luxembourg Site internet: http://www.ms.public.lu/fr/activites/pharmacie-medicament/index.html HOUDER VAN DE VERGUNNING VOOR HET IN DE HANDEL BRENGEN GSK Vaccines S.r.l, Via Fiorentina 1, 53100 Siena, Italië DATUM VAN DE GOEDKEURING VAN DE TEKST 06/2018(v05) **AFLEVERINGSWIJZE** Op medisch voorschrift.

1. Bexsero SMPC 2. Medini D, Stella M, Wassil J, Vaccine 2015; 33; 2629-2636

BE/BEX/0011/16a(1) – July 2018 - V.U.: GlaxoSmithkline Pharmaceuticals n.v., av Pascal 2-4-6, 1300 Wavre

ENDOCRINOLOGY

Oral presentations

O2. **Morning serum cortisol correlates with fasting glucose in children and adolescents with overweight**
A Martens, D Bünyamin, J Vanbesien, S Verheyden, B Rutteman, K Van De Maele, E Anckaert, I Gies, J De Schepper. UZ Brussel, VUB

Posters with short oral presentations

OP3. **Prediction and prevention of type 1 diabetes (T1D) in neonates: a new trial program in Europe**
O Pollé, P Achenbach, S Aydin, E Bonifacio, R Berner, H Elding Larsson, Å Lernmark, F Haupt, T Hoefs, A Hommel, O Kordonouri, M Lundgren, P Lysy, J Ohli, M Oltarzewski, F Roloff, M D Snape, A Szypowska, J Todd, M Vatish, C Winkler, A Gabriele Ziegler, K Casteels, Leona M and Harry B. Cliniques universitaires Saint-Luc, Kinder- und Jugendkrankenhaus Auf de Bult Hannover, Institute of Diabetes Research München, Center for Regenerative Therapies Dresden, Institute of Mother and Child Warsaw Poland, University Hospital Carl Gustav Carus Dresden, University of Oxford UK, Skåne University Hospital, Technical University Munich, University of Oxford, UZ Leuven, KU Leuven, Lund University, Helmsley Charitable Trust

OP4. **GENEPEDIAB study: multicenter screening of genetic forms of diabetes in cohorts of children and adolescents with type 1 diabetes**
S Welsch, C Daems, H Boughaleb, A Robert, Y Sznajer, J Louis, T Mouraux, N Seret, MC Lebrethon, P Lysy. UCL, ULB, Grand Hôpital de Charleroi, CHU Mont-Godinne, CHC Liège, CHU-Notre-Dame des Bruyères

OP5. **Empagliflozin and GABA improve Î²-cell mass and glucose tolerance in new-onset type 1 diabetes**
C Daems, S Welsch, H Boughaleb, J Vanderroost, A Robert, E Sokal, P Lesy. UCL-IREC-PEDI

Posters

P46. **Feasibility and reproducibility of ultrasound techniques to measure intra-abdominal fat in infants and preschool children**
S Provyn, E Schiettecatte, K Van De Maele, J De Schepper. UZ Gent, UZ Brussel

P47. **Novel PAX8 mutation in two siblings with congenital hypothyroidism due to thyroid dysgenesis**
S Bensliman, E Boros, C Brachet, C Heinrichs. HUDERF

P48. **Thyroid carcinoma presenting with hyperthyroidism and thyroid hot nodules in a female adolescent**
J Van Vlaenderen, K Logghe, E Schiettecatte, H Vermeersch, W Huvenne, J De Schepper. UZ Gent, AZ Delta, UZ Brussel

P49. **Fist description of a *NOTCH2* gene mutation in a newborn with thyroid ectopy**
L Willems, A Van Leynseele, K Keymolen, I Gies, J De Schepper. AZ Portaels, UZ Brussel

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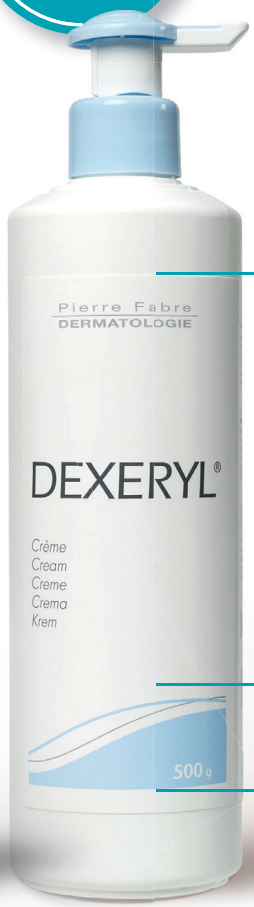

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GASTROENTEROLOGY

Oral presentations

O3. **Long-term outcome of pediatric patients with inflammatory bowel disease receiving immunomodulators**
K Van Hoeve, I Hoffman, M Ferrante, S Vermeire. UZ Leuven

O7. **MyCyFAPP project: Use of a mobile application for self-management of PERT improves gastro-intestinal related quality of life in children with CF**
M Boon, J Calvo-Lerma, I Claes, T Havermans, V Fornes, I Asseiceira, A Bulfamante, M Garriga, E Massip, S Woodcock, C Barreto, C Colombo, P Crespo, E Van der Wiel, H Janssens, J Hulst, S Martinez-Barona, R Nobili, L Pereira, M Ruperto, K De Boeck, C Ribes-Koninckx. UZ Leuven, Instituto de Investigación Sanitaria La Fe Valencia, Associação para a Investigação e Desenvolvimento da Faculdade de Medicina Lisbon, Università degli Studi di Milano, Hospital Universitario Ramón y Cajal Madrid, Erasmus Medical Center, Sophia Children's Hospital Rotterdam

Posters with short oral presentations

OP6. **A prospective observational study of fatigue in children with inflammatory bowel disease compared to a and control group**
L Le Roy, L Braeckeveldt, S Verstraete, E Vande Vijver, S Vande Velde. UZ Gent, UZ Antwerpen

OP7. **How to infuse heterologous human adult liver-derived progenitor cells safely?**
L Coppin, M Najimi, J Bodart, MS Rouchon, P Vandersmissen, S Eeckhoudt, G Dahlqvist, D Castanares, J Heemskerk, S Brouns, C Baaten, S Horman, N Belmonte, E Sokal, X Stephenne. UCL, UCL Saint-Luc, Universiteit Maastricht

OP8. **Effect of an intensive residential rehabilitation program with adjusted nutritional care on body composition in adult patients with cystic fibrosis**
S Buts, S Dereeper, B Würth, H Franckx, A De Guchtenaere, S Van Biervliet. UZ Gent, Zeepreventorium De Haan

OP9. **Implementation of guidelines on diagnosis and treatment of Eosinophilic Esophagitis by Pediatric and Adult GIs in Europe; On our way towards unison?**
G Tourlamain, R Garcia-Puig, C Gutiérrez Junquera, A Papadopoulou, R Elephteria, N Kalach, JH Oudshoorn, C Sokollik, K Karolewska-Bochenek, S Oliva, C Strisciuglio, O Bauraind, MKH Auth, M Thomson, S Otte, O Rok, J Amil Dias, C Tzivinikos, V Urbanos, A Kostovski, N Zevit, S Vande Velde. Ghent University Hospital, Hospital Universitari Mútua Terrassa Barcelona, Hospital Universitario Puerta de Hierro-Majadahonda Madrid, University of Athens, Catholic University of Lille, Gelre Hospital The Netherlands, University of Bern, Medical University of Warsaw, University of Rome, University of Campania Napoli, CHC Liege, University of Liverpool, Sheffield Children's Hospital, Ludwig-Maximilians-University of Munich, University of Ljubljana, Centro Hospitalar S. João Porto, Al Jalila Children's Specialty Hospital Dubai, Vilnius University Children's Hospital, University Children's Hospital Skopje Macedonia, Tel-Aviv University, UZ Gent

OP10. **Pancreatic blunt trauma in children: observations from a monocentric pilot study**
G Sonnino, P Deprez, A Pire, F Zech, R Reding, F Smets, X Stephenne, E Sokal, I Scheers. Université Catholique de Louvain.

Posters

P50. **When hoofbeats are zebras**
L Roels, K Vergaelen, M Van Den Akker, B Hauser. UZ Brussel

P51. **Switching from infliximab originator to a biosimilar is safe in paediatric patients with inflammatory bowel disease**
K Van Hoeve, E Dreesen, I Hoffman, M Ferrante, A Gils, S Vermeire. UZ Leuven. KU Leuven

NIEUW: PAMPERS® AQUA PURE BABYDOEKJES

De zuiverheid van water in het gemak van een doekje

De nieuwe Pampers® Aqua Pure babydoekjes zijn ontworpen om het meest water bevattende doekje te bieden, en daarbij nog steeds de best mogelijke huidbescherming te waarborgen.

Pampers® Aqua Pure babydoekjes bestaan voor 99% uit gezuiverd water, bevatten biologisch katoen en een lotion met unieke pH-buffer functie voor een milde en beschermende reiniging van de gevoelige babyhuid.

- Dermatologisch getest
- Geschikt voor de huid van de pasgeborene
- Bevat biologisch katoen
- 99% gezuiverd water
- 0% alcohol, parabenen, phenoxyethanol, kleurstoffen, parfum

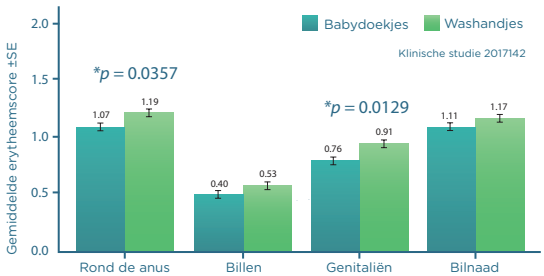
Een nieuwe klinische studie toont aan dat Pampers Aqua Pure-babydoekjes minstens even mild en zacht zijn als een washandje en water

In samenwerking met de ESPD heeft Pampers in een studie bij 130 baby's de invloed van babydoekjes op de luierzone vergeleken met die van een washandje en kraantjeswater.

Dit werd onderzocht in een willekeurig toegewezen, single blind parallel group design studie (dit wil zeggen dat onderzoekers niet weten welke de toegepaste verzorging is). Na een rustfase van één week waarbij enkel washandje en kraantjeswater werd gebruikt, werden de twee verzorgingen vergeleken gedurende een periode van twee weken. De aanwezigheid van erytheem werd daarbij gemeten op vier plaatsen.

Na twee weken gebruik bleken Pampers® Aqua Pure babydoekjes minstens even mild te zijn als washandjes en water. De huid die behandeld werd met babydoekjes, had ook een aanzienlijk lagere pH-waarde dan de huid die verzorgd werd met een washandje en kraantjeswater. Dat zou op lange termijn beter kunnen zijn voor de gezondheid van de huid.

Gemiddelde erytheemscore per meetplaats



Ingrediënten van plantaardige oorsprong die dermatologisch getest werden

- Natriumbenzoaat
- EDTA
- PEG-40 Gehydrogeneerde castorolie
- Citroenzuur
- Natriumcitraat
- Sorbitan Caprylaat

pH-buffer lotion

De lotion bevat een buffer op basis van citroenzuur die het natuurlijke pH-evenwicht van de huid helpt te behouden.[1] Wetenschappelijke studies hebben aangetoond dat de verstoring van het pH-evenwicht door een vuile luier één van de belangrijkste oorzaken van luieruitslag is. De combinatie van urine en stoelgang bevat verteringsenzymen die de huid irriteren. De babydoekjes van Pampers zijn voorzien van een speciaal ontwikkelde lotion die een pH-buffer functie vervult en de pH-waarde van de huid snel herstelt naar het normale niveau van ca. 4,5-6,0.

De Pampers® Aqua Pure babydoekjes bevatten:

Geen alcohol
Geen parfum
Geen parabenen
Geen phenoxyethanol
Geen kleurstoffen
Geen chloorbleekmiddel

[1] Interne gegevens van P&G

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PAMPERS STEUNT DE BELGISCHE VERENIGING VOOR KINDERGENEESKUNDE

European Society for Pediatric Dermatology

Goedgekeurd door ESPD

P52. Case report of a one-year-old girl with repetitive vomiting and loss of consciousness after ingestion of pineapple
C Perceval, E De Wachter, Y Vandenplas. UZ Brussel

P53. Intussuception due to heterotopic pancreas in the intestinal wall: a case report
H Warnier, T Marini, C Richelle, N Bottosso, L Dorthu, F Lardinois, T Carvelli. CHR Verviers

P54. Triple A syndrome revealed by achalasia : case report
K Kotilea, C Charles, T Mahler, H Louis. HUDERF, Centre hospitalier EpiCura, UCL

P55. No two pediatric intestinal polyps are alike
D Vermeulen, S Van Biervliet, M Van Winckel, R De Bruyne, B De Moerloose. UZ Gent

P56. Bilious vomiting: looking for the horse and finding the zebra
E Snoeck, K Vandekerckhove, K De Groote, J Panzer, D De Wolf, S Maesen, S Van Biervliet, M Van Winckel, S Vande Velde, R De Bruyne. UZ Gent

P57. Unusual cause of gastro-esophageal reflux in a 5-month-old girl
M Dirix, O Theeuws, M Lewin, B Massart , P Philippet, O Bauraind. CHC Liège, UCL

P58. two cases of bilious vomiting with duodenal web
C Vandendaele, P Philippet, M Dirix, A Bobarnac, S Colinet. CHC Liège, Université de Liège

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GENERAL PEDIATRICS

Oral presentations

O4. **Physical determinants of weight loss during a residential rehabilitation program for obese adolescents**
G Mets, G Marissens, J Servayge, K Vandekerckhove, B Würth, A De Guchtenaere. UZ Gent, Zeepreventorium De Haan

Posters with short oral presentations

OP28. **Assessment of pedestrian safety around Flemish Schools using Google Street View**
D Coppens, C Faes, J Toelen. UZ Leuven

OP29. **Assessment of cycling helmet use with Google Street View: an international survey**
E Scheurs, J Toelen. UZ Leuven

OP30. **Feasibility and effectiveness of Non-neonatal intubation training for pediatricians in Flanders**
E Janssens, T Schepens, ELIM Duval. UZA

OP31. **Screening for risk factors for developing chronic pain in a tertiary paediatric population**
S Ryckx, A De Jaeger. ZNA K Paola Kinderziekenhuis Antwerpen, UZ Gent

Posters

P59. **Fever & Arthritis: when family history matters**
M Rodesch, C Fricx, F Vermeulen. Hopital Erasme

P60. **The analgesic effect of Virtual Reality in pediatric procedural pain: a systematic review**
J Smeulders, K Vanhonsebrouck, J Toelen. UZ Leuven

P61. **Growth patterns and body composition in former extremely low birth weight (ELBW) infants until adulthood: a systematic review**
C Van de Pol, K Allegaert. KU Leuven, UZ Leuven

P62. **A hernia with a twist: the case of a strangulated ovary hernia**
A Taxhet, V Verdun, MC Seghaye. University Hospital Liège, Regional Hospital Centre Citadelle Liège

P63. **Pulmonary arteriovenous malformations and a neonatal presentation of a Rendu-Osler disease**
C Van Kerkhoven, K El Abd, P Maton, D Brisbois, M Thimmesch. CHC Liège, Université de Liège

P64. **An atypical infantile eruption: let's have a look at Mum!**
O Paduart, F Vermeulen. Hopital Erasme

P65. **A case of spontaneous splenic infarction in a patient with heterotaxy syndrome and polysplenia**
SF Bartelse, FJS van der Velden, S van Gijlswijk. IJsselland Ziekenhuis

P66. **Aquatic activities, an unusual cause of anaphylactic shock**
I De Brouchoven, T Slaouti. Europe Hospitals, Sainte-Elisabeth Clinic

INFECTIOLOGY – PNEUMOLOGY – IMMUNOLOGY

Oral presentations

O5. **Three year follow-up of Streptococcus pneumoniae nasopharyngeal carriage in Belgian children after a PCV13-to-PCV10 vaccine switch**
I Wouters, L Van Heirstraeten, S Desmet, C Lammens, J Verhaegen, H Goossens, P Van Damme, P Beutels, S Malhotra-Kumar, H Theeten. Universiteit Antwerpen, KU Leuven, UZ Leuven

O8. **Retrospective evaluation of the efficiency of azithromycin in the protracted bacterial bronchitis (PBB)-extended**
D Trajman, N Lefèvre. HUDERF, ULB

O9. **Diagnosing enteroviral meningitis via blood transcriptomics: an alternative for lumbar puncture?**
E Bartholomeus, N De Neuter, A Lemay, D Tuerlinckx, D Weynants, K Van Lede, G van Berlaer, D Bulckaert, T Boiy, A Vander Auwera, M Raes, D Van der Linden, H Verhelst, S Van Steijn, T Jonckheer, J Dehoorne, R Joos, H Jansens, A Suls, P Van Damme, K Laukens, G Mortier, P Meysman, B Ogunjimi. UZA, Universiteit Antwerpen, AZ Turnhout, UC, CHU Namur, AZ Nikolaas, UZ Brussel, Gasthuis Zusters Antwerpen, Jessa Ziekenhuis, CHU-ULC Saint-Luc, UZ Gent, Ziekenhuis Netwerk Antwerpen.

Posters with short oral presentations

OP25. **A multidisciplinary approach to diagnose X-linked Hyper IgM syndrome in a boy with acute interstitial pneumonitis**
D Dinneweth, E Linskens, B Denys, J. Philippé, J Willekens, P Schelstraete, T Van Ackere, M De Bruyne, E De Baere, V Bordon, E Dhont, C Bonroy, F Haerynck. UZ Gent, Jan Yperman ziekenhuis Ieper

OP26. **Management of children with tracheal stenosis: a single-center experience**
M Boon, E Ter Haar, F Vermeulen, B Cools, H Decaluwé, F Rega, M Proesmans. UZ Leuven

OP27. **Invasive pneumococcal disease surveillance in Belgium and paediatric pneumococcal conjugate vaccines: have we reached a steady state?**
M Moreira, S Klein, M Khellaf, B Mungall. GSK, Wavre

Posters

P67. **Open your eye to Cat-Scratch Disease...**
L Zambelli, H André, N Ait Salah, J Frère, MC Seghaye. CHR Citadelle

P68. **Non-blanching rash in a toddler- a case report**
E Hoornaert, C Saintes, A de Selys , D Van der Linden, V Selimaj. Clinique Saint Jean Brussels, Clinique Universitaire Saint Luc

P69. **YouTube videos as a source of information about immunology for medical students**
J Van den Eynde, A Crauwels, PG Demaerel, L Van Eycken, R Schrijvers, D Bullens, J Toelen. UZ Leuven

P70. **Side effects of Bexsero®: a systematic review**
AS Lemoine, M Raes, J Toelen. UZ Leuven, Virga Jesse ziekenhuis

P71. **Recurrent laryngeal papillomatosis - From neonatology to future perspectives**
I Rebia. CHU Liege

P72. **Pulmonary function of children after lobectomy**
H Rigolle, De Baets, A Malfroot, M Proesmans. UZ Gent

P73. **Haemophilus influenzae type b cellulitis in a vaccinated and immunocompromised nine-months-old child**
V Hermans, C Schirvel, A Remy, D Michez. Centre Hospitalier EpiCURA

P74. **A linear skin lesion as presentation of long persistent strongyloidiasis**
P Van der Speeten, D Van Brusselen, M Gielen. Sint Vincentiusziekenhuis Antwerpen

P75. **Proteus mirabilis meningitis revealing an intradural dermoid cyst**
MC Nassogne, G Koerts, D Van der Linden, O Chatzis. Cliniques universitaires Saint-Luc, UCLouvain

P76. **Shivering As a Clinical Predictor Of Serious Bacterial Infections in Febrile Children - A Systematic Review**
E Nuyts, J Toelen. UZ Leuven

P77. **About a hereditary pathology of the surfactant: The mutation of the gene encoding surfactant protein C**
K Tazi, A Biver. Centre Hospitalier de Luxembourg, Université Libre de Bruxelles

P78. **'Every sweet has its sour': rare skin lesions in a boy with combined immunodeficiency**
DJ Bogaert, M Hagendorens, M De Bruyne, H Lapeere, A Covents, E De Baere, F De Baets, F Haerynck. UZ Gent

P79. **Brain abscess in a 15 years old teenager: a common clinical presentation?**
J Lorand, E Bodart. CHU UCL Dinant

P80. **YouTube as source for pre-travel health information about malaria**
P D'Hondt, F Peeters, L Pietermans, I Roodhooft, J Toelen. UZ Leuven

P81. **A rare case of invasive Kingella kingae infection**
E Surgun, Y Marchione, S Blumental, A Bondue, C Joris, F Vermeulen. Hôpital Erasme, HUDERF

P82. **Morbidity and mortality of extrapulmonary tuberculosis: two pediatric cases**
S Bottse, P Schelstraete. UZ Gent

P83. **Neonatal granulopenia due to maternal HNA-2 alloimmune autoantibodies: a case report**
FJS van der Velden, SF Bartelse, S van Gijlswijk. IJsselland Hospital

P84. **Food protein-induced enterocolitis syndrome: an infrequent food allergy but not to be missed!**
A Fohn, T carvelli, A Collins. CHR Citadelle Liège, CHR Verviers

P85. **Case-report: A neonate with unilateral lung hypoplasia, complete tracheal rings and hypoplastic left thumb as part of a genetic disease/syndrome?**
J Eelen, A Smits, A Debeer, M Proesmans, E Deloof, K Devriendt. UZ Leuven, Heilig Hart Ziekenhuis Leuven

P86. **Case Report - Perinatal Varicella zoster virus infection in pregnant women: preventive management of the newborn**
A Du Mortier, J Vanclaire, V Selimaj. Clinique Saint-Jean

P87. **Henoch-Schönlein purpura? Open your mind in older ones**
Q Neven, M Malvaux, B Brasseur. Clinique Saint-Pierre, KU Leuven

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NEONATOLOGY – PEDIATRIC INTENSIVE/EMERGENCY CARE

Oral presentations

O6. **Short term survival and survival without severe morbidity in extremely preterm babies: comparison between two birth cohorts in a third level NICU**
A Vicari, C Debauche. UCL Saint Luc, Hôpital Civil Marie Curie Charleroi

O10. **Micro-computed tomography for the longitudinal evaluation of preterm lung injury in a rabbit model**
M Aertgeerts, T Salaets, A Gie, J Vignero, G Van de Velde, J Toelen. KU Leuven

Posters with short oral presentations

OP13. **Proximal tracheoesophageal fistula in esophageal atresia: a diagnostic challenge**
C Rohaert, A Clarysse. AZ Sint-Jan Brugge

OP14. **Both maturational and non-maturational covariates determine neonatal albuminemia on the first day of life**
J Deberdt, L Delemarre, K Allegaert, M Rayyan, G Naulaers, A Smits. KU Leuven, UZ Leuven, Erasmus MC-Sophia Children's Hospital Rotterdam

OP15. **Intermittent CPAP attenuates lung injury in a preterm rabbit BPD model**
A Gie, T Salaets, K Allegaert, J Deprest, J Toelen. UZ Leuven

OP16. **Maternal paracetamol intake and fetal ductus arteriosus constriction or closure: a case series analysis and mechanistic models in support**
K Allegaert, P Mian. KU Leuven, Erasmus MC Sophia Children's Hospital Rotterdam

OP17. **Improved vancomycin exposure in neonates using a population pharmacokinetic model-based vancomycin dosing regimen**
E Desaeger, K Allegaert, A Vandendriessche, V Cossey, A Smits. KU Leuven, UZ Leuven, Erasmus MC-Sophia Children's Hospital Rotterdam

Posters

P88. **Local pulmonary drug delivery in the preterm rabbit: feasibility and efficacy of daily intratracheal injections**
T Salaets, A Gie, O Gheysens, G Vande Velde, K Allegaert, J Deprest, J Toelen. UZ Leuven

P89. **Subgaleaal abces as cause of fever of unknown origin in an 8-day old newborn**
N Van Oost , A Mulder. UZA

P90. **Schizencephaly : how a cerebral lesion can reveal a drug consumption**
C Themelin, P Philippet, P Maton, S Smeets. CHC Rocourt

P91. **Inherited peroxisomal disorders: a case report of neonatal hypotonia and seizures**
E Gkogkou, R Viellevoye, I Broux, V de Halleux, N Hennuy, V Rigo. CHU-CHR Liège

P92. **Aplasia cutis congenita in an infant of an initial twin gestation**
K De Schynkel. AZ Maria Middelares, UZ Brussel

P93. **Case report: should trisomy 18 still be considered as a lethal condition?**
Z Wilderiane, C Coremans, P Philippet, I Loeckx. CHC Liège, Université de Liège

P94. **Methemoglobinemia: a rare side effect of a healthy diet**
P Naessens, T Van Der Heggen, A D'Hooghe, K Sauer. AZ Sint-Jan

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Bexsero : premier vaccin contre le méningocoque de sérogroupe B.

Le seul indiqué dès l'âge de 2 mois.[1,2]

BEXSERO

Vaccin méningococcique groupe B (ADNr, composant, adsorbé)

RÉSUMÉ DES CARACTÉRISTIQUES DU PRODUIT Veuillez vous référer au Résumé des Caractéristiques du Produit pour une information complète concernant l'usage de ce médicament ▼Ce médicament fait l'objet d'une surveillance supplémentaire qui permettra l'identification rapide de nouvelles informations relatives à la sécurité. Les professionnels de la santé déclarent tout effet indésirable suspecté. Voir rubrique « Effets Indésirables » pour les modalités de déclaration des effets indésirables. **DÉNOMINATION DU MÉDICAMENT** Bexsero suspension injectable en seringue prérempliéVaccin méningococcique groupe B (ADNr, composant, adsorbé) EU/1/12/812/001 Classe pharmacothérapeutique : vaccins méningococciques, Code ATC : J07AH09 **COMPOSITION QUALITATIVE ET QUANTITATIVE** Une dose (0,5 ml) contient : Protéine de fusion recombinante NHBA de ***Neisseria meningitidis*** groupe B [1,2,3] : 50 microgrammes Protéine recombinante NadA de ***Neisseria meningitidis*** groupe B [1,2,3] : 50 microgrammes Protéine de fusion recombinante fHbp de ***Neisseria meningitidis*** groupe B [1,2,3] : 50 microgrammes Vésicules de membrane externe (OMV) de ***Neisseria meningitidis groupe*** B, souche NZ98/254 mesurée en tant que proportion de l'ensemble des protéines contenant l'antigène PorA P1.4 [2] : 25 microgrammes[1] produite dans des cellules d'***E. coli*** par la technique de l'ADN recombinant [2] adsorbée sur hydroxyde d'aluminium (0,5 mg Al^{3+}) [3] NHBA (antigène de liaison à l'héparine de Neisseria), NadA (adhésine A de Neisseria), fHbp (protéine de liaison du facteur H) **INDICATIONS THÉRAPEUTIQUES** Bexsero est indiqué pour l'immunisation active des sujets à partir de l'âge de 2 mois contre l'infection invasive méningococcique causée par ***Neisseria meningitidis*** de groupe B. L'impact de l'infection invasive à différentes tranches d'âge ainsi que la variabilité épidémiologique des antigènes des souches du groupe B dans différentes zones géographiques doivent être pris en compte lors de la vaccination. Voir rubrique 5.1 du RCP complet pour plus d'informations sur la protection contre les souches spécifiques au groupe B. Ce vaccin doit être utilisé conformément aux recommandations officielles. **Posologie et mode d'administration** Posologie

Tableau 1. Résumé de la posologie

Age lors de la première dose	Primovaccination	Intervalles entre les doses de primovaccination	Rappel
Nourrissons de 2 à 5 mois	Trois doses de 0,5 ml chacune,	1 mois minimum	Oui, une dose entre l'âge de 12 et 15 mois avec un intervalle d'au moins 6 mois entre la primovaccination et la dose de rappel [b,c]
Nourrissons de 3 à 5 mois	Deux doses de 0,5 ml chacune	2 mois minimum	
Nourrissons de 6 à 11 mois	Deux doses de 0,5 ml chacune	2 mois minimum	Oui, une dose au cours de la deuxième année avec un intervalle d'au moins 2 mois entre la primovaccination et la dose de rappel [c]
Enfants de 12 à 23 mois	Deux doses de 0,5 ml chacune	2 mois minimum	Oui, une dose avec un intervalle de 12 à 23 mois entre la primovaccination et la dose de rappel [c]
Enfants de 2 à 10 ans	Deux doses de 0,5 ml chacune	1 mois minimum	Besoin non établi [d]
Adolescents (à partir de 11 ans)et adultes*	Deux doses de 0,5 ml chacune	1 mois minimum	Besoin non établi [d]

[a] La première dose ne doit pas être administrée avant l'âge de 2 mois. La sécurité et l'efficacité de Bexsero chez les nourrissons de moins de 8 semaines n'ont pas encore été établies. Aucune donnée n'est disponible. [b] En cas de retard, la dose de rappel ne doit pas être administrée au-delà de l'âge de 24 mois. [c] Voir rubrique 5.1 du RCP complet. La nécessité et le moment d'administration d'une dose de rappel n'ont pas encore été déterminés. [d] Voir rubrique 5.1 du RCP complet. * Il n'existe aucune donnée chez les adultes de plus de 50 ans. Mode d'administration Le vaccin est administré par une injection intramusculaire profonde, de préférence dans la face antérolatérale de la cuisse chez le nourrisson ou dans la région du muscle deltoïde du haut du bras chez les sujets plus âgés. Des sites d'injection distincts doivent être utilisés si plusieurs vaccins sont administrés simultanément. Le vaccin ne doit pas être injecté par voie intraveineuse, sous-cutanée ni intradermique et ne doit pas être mélangé avec d'autres vaccins dans la même seringue. Pour les instructions concernant la manipulation du vaccin avant administration, voir la rubrique 6.6 du RCP complet. **CONTRE-INDICATIONS** Hypersensibilité aux substances actives ou à l'un des excipients mentionnés à la rubrique 6.1 du RCP complet. **MISES EN GARDE SPÉCIALES ET PRÉCAUTIONS D'EMPLOI** Comme pour les autres vaccins l'administration de Bexsero doit être reportée chez des sujets souffrant de maladie fébrile sévère aiguë. Toutefois, la présence d'une infection mineure, telle qu'un rhume, ne doit pas entraîner le report de la vaccination. Ne pas injecter par voie intravasculaire. Comme pour tout vaccin injectable, un traitement médical approprié et une surveillance adéquate doivent toujours être disponibles en cas de réaction anaphylactique consécutive à l'administration du vaccin. Des réactions en rapport avec l'anxiété, y compris des réactions vaso-vagales (syncope), de l'hyperventilation ou des réactions en rapport avec le stress peuvent survenir lors de la vaccination comme réaction psychogène à l'injection avec une aiguille (voir rubrique « Effets indésirables »). Il est important que des mesures soient mises en place afin d'éviter toute blessure en cas d'évanouissement. Ce vaccin ne doit pas être administré aux patients ayant une thrombocytopénie ou tout autre trouble de la coagulation qui serait une contre-indication à une injection par voie intramusculaire, à moins que le bénéfice potentiel ne soit clairement supérieur aux risques inhérents à l'administration. Comme tout vaccin, la vaccination avec Bexsero peut ne pas protéger tous les sujets vaccinés. Il n'est pas attendu que Bexsero assure une protection contre la totalité des souches de méningocoque B en circulation. Comme pour de nombreux vaccins, les professionnels de santé doivent savoir qu'une élévation de la température corporelle peut survenir suite à la vaccination des nourrissons et des enfants (de moins de 2 ans). L'administration d'antipyrétiques à titre prophylactique pendant et juste après la vaccination peut réduire l'incidence et la sévérité des réactions fébriles post-vaccinales. Un traitement antipyrétique doit être mis en place conformément aux recommandations locales chez les nourrissons et les enfants (de moins de 2 ans). Les personnes dont la réponse immunitaire est altérée soit par la prise d'un traitement immunosuppresseur, une anomalie génétique ou par d'autres causes, peuvent avoir une réponse en anticorps réduite après vaccination. Des données d'immunogénicité sont disponibles chez les patients présentant un déficit en complément, une asplénie ou une dysfonction splénique. Il n'existe aucune donnée sur l'utilisation de Bexsero chez les sujets de plus de 50 ans et il existe des données limitées chez les patients atteints de maladies chroniques. Le risque potentiel d'apnée et la nécessité d'une surveillance respiratoire pendant 48 à 72 heures doivent soigneusement être pris en compte lors de l'administration des doses de primovaccination chez des grands prématurés (nés à 28 semaines de grossesse ou moins), en particulier chez ceux ayant des antécédents d'immaturité respiratoire. En raison du bénéfice élevé de la vaccination chez ces nourrissons, l'administration ne doit pas être suspendue ou reportée. Le capuchon de la seringue peut contenir du latex de caoutchouc naturel. Bien que le risque de développer des réactions allergiques soit très faible, les professionnels de santé doivent évaluer le rapport bénéfices/risques avant d'administrer ce vaccin à des sujets présentant des antécédents connus d'hypersensibilité au latex. La kanamycine est utilisée au début du procédé de fabrication et est éliminée au cours des étapes ultérieures de la fabrication. Les taux de kanamycine éventuellement détectables dans le vaccin final sont inférieurs à 0,01 microgramme par dose. L'innocuité de Bexsero chez les sujets sensibles à la kanamycine n'a pas été établie. **EFFETS INDÉSIRABLES** Résumé du profil de sécurité La sécurité de Bexsero a été évaluée lors de 17 études, dont 10 essais cliniques randomisés contrôlés portant sur 10565 sujets (âgés de 2 mois minimum) ayant reçu au moins une dose de Bexsero. Parmi les sujets vaccinés par Bexsero, 6837 étaient des nourrissons et des enfants (de moins de 2 ans), 1051 étaient des enfants (entre 2 et 10 ans) et 2677 étaient des adolescents et des adultes. Parmi les nourrissons ayant reçu les doses de primovaccination de Bexsero, 3285 ont reçu une dose de rappel au cours de leur deuxième année de vie.. Chez les nourrissons et les enfants (de moins de 2 ans), les réactions indésirables locales et systémiques les plus fréquemment observées lors des essais cliniques étaient : sensibilité et érythème au site d'injection, fièvre et irritabilité. Dans les études cliniques menées chez les nourrissons vaccinés à 2, 4 et 6 mois, la fièvre (≥ 38 °C) était rapportée chez 69% à 79 % des sujets lorsque Bexsero était co-administré avec des vaccins de routine (contenant les antigènes suivants : pneumococcique heptavalent conjugué, diphtérie, tétanos, coqueluche acellulaire, hépatite B, poliomyélite inactivée et ***Haemophilus influenzae*** de type b), contre 44% à 59 % des sujets recevant les vaccins de routine seuls. Une utilisation plus fréquente d'antipyrétiques était également rapportée chez les nourrissons vaccinés avec Bexsero et des vaccins de routine. Lorsque Bexsero était administré seul, la fréquence de la fièvre était similaire à celle associée aux vaccins de routine administrés aux nourrissons pendant les essais cliniques. Les cas de fièvre suivaient généralement un schéma prévisible, se résolvant généralement le lendemain de la vaccination. Chez les adolescents et les adultes, les réactions indésirables locales et systémiques les plus fréquemment observées étaient : douleur au point d'injection, malaise et céphalée. Aucune augmentation de l'incidence ou de la sévérité des réactions indésirables n'a été constatée avec les doses successives du schéma de vaccination. Liste tabulée des effets indésirables Les effets indésirables (consécutifs à la primovaccination ou à la dose de rappel) considérés comme étant au moins probablement liés à la vaccination ont été classés par fréquence. Les fréquences sont définies comme suit : Très fréquent : (≥ 1/10) Fréquent : (≥ 1/100 à < 1/10) Peu fréquent : (≥ 1/1 000 à < 1/100) Rare : (≥ 1/10 000 à < 1/1 000) Très rare : (< 1/10 000) Fréquence indéterminée : (ne peut être estimée sur la base des données disponibles) Dans chaque groupe de fréquence, les effets indésirables sont présentés par ordre de sévérité décroissante. Outre les événements rapportés lors des essais cliniques, les réactions spontanées rapportées dans le monde pour Bexsero depuis sa commercialisation sont décrites dans la liste ci dessous. Comme ces réactions ont été rapportées volontairement à partir d'une population de taille inconnue, il n'est pas toujours possible d'estimer de façon fiable leur fréquence. Ces réactions sont, en conséquence, listées avec une fréquence indéterminée. **Nourrissons et enfants (jusqu'à l'âge de 10 ans)** Affections du système immunitaire Fréquence indéterminée : réactions allergiques (y compris réactions anaphylactiques) Troubles du métabolisme et de la nutrition Très fréquent : troubles alimentaires Affections du système nerveux Très fréquent : somnolence, pleurs inhabituels, céphalée Peu fréquent : convulsions (y compris convulsions fébriles) Fréquence indéterminée : épisode d'hypotonie-hyporéactivité Affections vasculaires Peu fréquent : pâleur (rare après le rappel) Rare : syndrome de Kawasaki Affections gastro-intestinales Très fréquent : diarrhée, vomissements (peu fréquents après le rappel) Affections de la peau et du tissu sous-cutané Très fréquent : rash (enfants âgés de 12 à 23 mois) (peu fréquent après le rappel) Fréquent : rash (nourrissons et enfants âgés de 2 à 10 ans) Peu fréquent : eczéma Rare : urticaire Affections musculo-squelettiques et systémiques Très fréquent : arthralgies Troubles généraux et anomalies au site d'administration Très fréquent : fièvre (≥ 38 °C), sensibilité au niveau du site d'injection (y compris sensibilité sévère au site d'injection définie par des pleurs lors d'un mouvement du membre ayant reçu l'injection), érythème au site d'injection, gonflement du site d'injection, induration au site d'injection, irritabilité Peu fréquent : fièvre (≥ 40 °C) Fréquence indéterminée : réactions au site d'injection (incluant un gonflement étendu du membre vacciné, vésicules au point d'injection ou autour du site d'injection et nodule au site d'injection pouvant persister pendant plus d'un mois) **Adolescents (à partir de 11 ans) et adultes** Affections du système immunitaire Fréquence indéterminée : réactions allergiques (y compris réactions anaphylactiques) Affections du système nerveux Très fréquent : céphalée Fréquence indéterminée : syncope ou réaction vaso-vagale à l'injection Affections gastro-intestinales Très fréquent : nausées Affections musculo-squelettiques et systémiques Très fréquent : myalgies, arthralgies Troubles généraux et anomalies au site d'administration Très fréquent : douleur au point d'injection (y compris douleur sévère au point d'injection définie par une incapacité à mener à bien des activités quotidiennes normales), gonflement du site d'injection, induration au point d'injection, érythème au site d'injection, malaise Fréquence indéterminée : fièvre, réactions au site d'injection (incluant gonflement étendu du membre vacciné, vésicules au point d'injection ou autour du site d'injection et nodule au site d'injection pouvant persister plus d'un mois) **Déclaration des effets indésirables suspectés** La déclaration des effets indésirables suspectés après autorisation du médicament est importante. Elle permet une surveillance continue du rapport bénéfice/risque du médicament. Les professionnels de santé déclarent tout effet indésirable suspecté via le système national de déclaration : **Belgique** Agence fédérale des médicaments et des produits de santé Division Vigilance EUROSTATION II Place Victor Horta, 40/40 B-1060 Bruxelles Site internet: www.afmps.be e-mail: adversedrugreactions@fagg-afmps.be **Luxembourg** Direction de la Santé – Division de la Pharmacie et des Médicaments Villa Louvigny – Allée Marconi L-2120 Luxembourg Site internet: http://www.ms.public.lu/fr/activites/pharmacie-medicament/index.html **TITULAIRE DE L'AUTORISATION DE MISE SUR LE MARCHÉ** GSK Vaccines S.r.l., Via Fiorentina 1, 53100 Sienne, Italie **DATE D'APPROBATION DU TEXTE** 06/2018(v05) **MODE DE DELIVRANCE** Sur prescription médicale.

1. Bexsero SMPC 2. Medini D, Stella M, Wassil J, Vaccine 2015; 33; 2629-2636

BE/BEX/0011/16a(1) – July 2018 - E.R.: GlaxoSmithKline Pharmaceuticals s.a., av Pascal 2-4-6, 1300 Wavre

P95. **Neonatal thrombopenia with serious complications**
L Goossens, A Keymeulen. UZ Gent

P96. **Fulminant neonatal acute liver failure due to enterovirus: a case of favorable outcome after intravenous immunoglobulin administration**
P Calò, S Blumental, AB Johansson, A Vuckovic. HUDERF

P97. **'How do families cope with premature birth?'**
MR Van Hoestenberghe, K Van Leeuwen. Ziekenhuis Oost-Limburg, KU Leuven

P98. **Postanoxic encephalopathy leading to organ donation after circulatory death in a 12-year-old boy**
P Tshimanga, A Mulder, F Lebrun, I Loeckx, D Ledoux, P Philippet, P Demaret. CHC Clinique de l'Espérance, Universitary Hospital of Liège, CHC - Liège

P99. **Bloody stools - at the maternity ward**
A Smits, A Eerdekens, C Vanhole, L Beckers. UZ Leuven, Imelda

P100. **Bronchocoele in the neonate : conservative or invasive therapy?**
A Clarysse, M Sijmons. AZ Sint Jan Brugge

P101. **Extensive and rapidly developing white matter damage in preterm infant**
S Bogovic, S Van Cauter, MR Van Hoestenberghe. ZOL

P102. **An unusual case of SpO2-PaO2 discrepancy in an intensive care neonate illustrating an important pitfall in routine pulse oxygen monitoring**
H Levi, K De Coen. UZ Gent

NEPHROLOGY

Oral presentations

O11. Stress, anxiety and depression in parents of children with chronic kidney disease and its correlation with protective behavior
E De Bruyne, C Van Herzeele, E Snauwaert, E Holvoet, A Raes, W Van Biesen, L Goubert, E Van Hoecke, S Eloot, J Vande Walle. UZ Gent, Universiteit Antwerpen

Posters with short oral presentations

OP32. Desmopressin in enuresis - predictive factors in therapy response
L Bosschaert, L Dossche, A Raes, J Vande Walle. UZ Gent

OP33. Fecal microbiota transplantation for recurrent clostridium difficile in children with end-stage renal disease and renal transplantation
P D'hondt, N Knops. UZ Leuven

OP34. Promising experience of paediatric acute peritoneal dialysis program using home made fluids in the democratic republic of congo
M Ekulu Pepe, B Nkoy Agath, M Ndiyo Yoli, M Odio Bienvenu, K Betukumesu Dieumerci, K Kazadi Orly, E Levtchenko, B Lepira François. University Hospital Of Kinshasa DR Congo, UZ Leuven, KU Leuven

OP35. Uremic toxin concentrations are related to residual kidney function in the pediatric hemodialysis population
A El Amouri, E Snauwaert, E Holvoet, W Van Biesen, A Raes, G Glorieux, J Vande Walle, S Roels, R Vanholder, V Askiti, K Azukaitis, A Bayasit, N Campolat, M Fischbach, N Godefroid, S Krid, M Litwin, L Obrycki, F Paglialonga, B Ranchin, C Samaille, F Schaefer, CP Schmitt, B Spasojevic, CJ Stefanidis, M Van Dyck, K Van Hoeck, L Collard, R Shroff, S Eloot. Universiteit Gent, A & P Kyriakou Children's Hospital Athens, Vilnius University, Cukurova University Adana Turkey, Istanbul University, Children's Dialysis Center Strasbourg, UCL, Hôpital Necker-Enfants Malades Paris, Children`s Memorial Health Institute Warsaw, Fondazione IRCCS Ca' Granda Ospedale Maggiore Policlinico Milan, Hôpital Femme Mère Enfant Hospices Civils de Lyon, CHU Lille, Center for Pediatrics and Adolescent Medicine Heidelberg, University Children's Hospital Belgrade, UZ Leuven, Universiteit Antwerpen, CHU Liège, Great Ormond Street Hospital for Children NHS Foundation Trust London, UZ Gent

OP36. Chitotriosidase: a novel alternative biomarker for the therapeutic monitoring of nephropathic cystinosis ?
KRP Veys, MA Elmonem, M Van Dyck, MC Janssen, EA Cornelissen, K Hohenfellner, G Prencipe, F Emma, LP van den Heuvel, E Levtchenko. KU Leuven, UZ Leuven, Cairo University, Radboud university medical centre Nijmegen, SudOstBayern Klinikum Traunstein, Bambino Gesù Children's Hospital Rome

Posters

P103. Glomerular developmental delay in the preterm neonatal rabbit
D De Winter, T Salaets, A Gie, K Allegaert, J Deprest, J Toelen. UZ Leuven

P104. Epidemiology of moderate to severe dyskalemia in a paediatric emergency department
O Barbance, D De Bels, P Honoré, K Isamili, D Biarent. HUDERF

P105. The many faces of atypical Hemolytic Uremic Syndrome (aHUS) in Belgium
A Massart, K Claesers, L Weekers. UZA, UZ Leuven, CHU

P106. The effect of Urotherapy in group during youth camp
L Dewulf, K Van den Hende, C Renson, A Raes, J Vande Walle. UZ Gent

P107. Risk factors for recurrent kidney stones in children
C De Ruysscher, L Pien, J Vande Walle, T Tailly, E Van Laecke, A Prytula. UZ Gent

P108. Dental abnormalities and nephrocalcinosis: A case report of a boy with Enamel-Renal syndrome
L Lambrecht, H Van Clooster, M Van Dyck, N Knops. AZ Herentals, UZ Leuven

P109. ADPedKD: A Global Platform on managing childhood Autosomal Dominant Polycystic Kidney Disease
D Mekahli. UZ Leuven, KU Leuven

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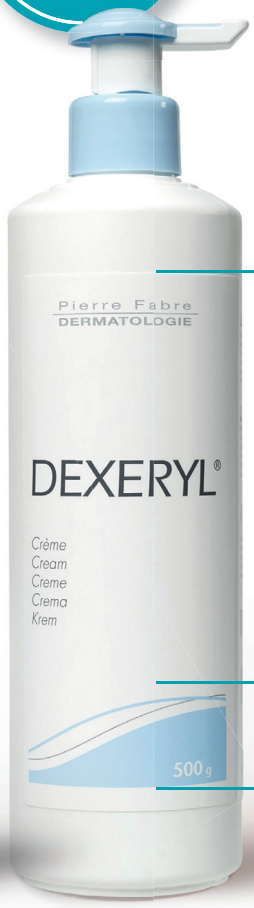

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NEUROLOGY – GENETICS – NEURO-ORTHOPEDICS

Oral presentations

O12. **Three cases of acute flaccid myelitis, a new epidemic?**
A Van Roest, B Ceulemans, S Dekeyser, D Beysen, A Jonckheere, S Kenis, AS Schoonjans. UZ Antwerpen

Posters with short oral presentations

OP18. **Deformational plagiocephaly as a marker for developmental delay: a systematic review**
A Dierx, J Toelen. UZ Leuven

OP19. **Salbutamol - not only in asthma**
C Hardy, A Daron, M Trippaerts, L Servais. CHR Citadelle Liège

OP20. **Scurvy: a forgotten disease. Case report of a 3 year old boy with progressive hypotonia and a very restrictive diet.**
N Willemyns, G Buyse, E Ortibus. UZ Leuven

Posters

P110. **Not just a 'simple' myelomeningocele; A case report of a polymalformative syndrome**
F Derriks, F Vermeulen. Hôpital Erasme

P111. **A case of intracranial bleeding after a vaginal forceps delivery**
AC Gillot, G Delannoy, E Hoornaert, E Nicolaï. Clinique Saint Jean Brussels, Clinique Universitaire Saint-Luc

P112. **Flaccid paralysis of the lower limbs of subacute onset in the context of respiratory infection**
Y Lounis, AS Blecic, O Jadot, N Cajgfinger, M Weerts, A Marschese, JP Misson. CHR Namur, CHU-CHR Citadelle Liège

P113. **Parafalcine subdural empyema - an uncommon complication after tooth extraction: a case report and literature review**
L Delhaise. UZA

P114. **When a heart murmur is hiding a rare genetic disease**
C Fobe, C Baguette, P Philippet, FG Debray. UCL, CHC Liège

P115. **An intracranial haemorrhage in a febrile newborn: About a case**
J Sprumont, R Stevens, M Lewin, H Hoeffelin, L Sprimont, A Gilbert, P Philippet. CHC Liège

P116. **Miller Fisher-syndrome, an atypical presentation of Guillain-Barré**© syndrome
K De Schynkel. UZ Brussel, AZ Maria Middelares

P117. **An infant with a cortical malformation syndrome; A new de novo mutation in the tubulin gene TUBB2A**
K Stouffs, K Keymolen, A Jansen. UZ Brussel

P118. **KBG syndrome, a rare underdiagnosed disorder: about three patients**
L Wulleman, N Revencu, S Ghariani, R El Thary. Cliniques Universitaires Saint Luc

P119. **Live birth neonate with triploidy (69, XXX): a case report**
E Feenstra, J Widdershoven. Jeroen Bosch Ziekenhuis

P120. **Bell's palsy in a 20-day-old newborn with a serology positive for Mycoplasma Pneumoniae**
A Famerie, S Dahalani, A Bocquet, F Houyoux, JP Misson. CHR Namur

NOUVEAU: LINGETTES PAMPERS® AQUA PURE

La pureté de l'eau avec la facilité d'une lingette

Les nouvelles lingettes Pampers® Aqua Pure ont été développées pour offrir une lingette la plus humide possible qui assure à la fois un soin efficace et la meilleure protection de la peau.

Les lingettes Pampers® Aqua Pure contiennent 99% d'eau purifiée, du coton bio et une lotion à effet tampon de pH unique pour un soin en douceur tout en protégeant la peau sensible de bébé

- Testées dermatologiquement
- Conviennent à la peau des nouveau-nés
- organic cotton — A base de coton bio
- 99% d'eau purifiée
- 0% alcool, parabène, phénoxyéthanol, colorant, parfum

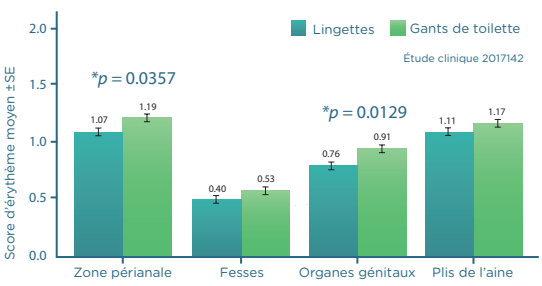
Une nouvelle étude clinique démontre que les lingettes Pampers® Aqua Pure sont au moins aussi douces qu'un gant de toilette imbibé d'eau

En collaboration avec l'ESPD, Pampers a mené une étude chez 130 bébés évaluant l'effet des lingettes pour bébé sur le siège en comparaison avec un gant de toilette imbibé d'eau du robinet.

Cette étude a été réalisée en parallèle en aveugle et à répartition aléatoire (ce qui signifie que les examinateurs ignoraient quels étaient les soins appliqués). Après une phase de repos d'une semaine durant laquelle seul l'usage d'eau du robinet et du gant de toilette était autorisé, les deux types de soins ont été comparés pendant une période de deux semaines en mesurant les scores d'érythème sur 4 sites.

Après deux semaines d'utilisation, il a été démontré que les lingettes Pampers® Aqua Pure sont au moins aussi douces qu'un gant de toilette imbibé d'eau. La peau nettoyée avec des lingettes a également présenté un pH significativement inférieur en comparaison à la peau nettoyée à l'aide d'un gant de toilette imbibé d'eau du robinet, ce qui pourrait procurer des bénéfices à long terme pour la santé de la peau.

Score d'érythème moyen par site



Composants d'origine végétale qui ont été testés dermatologiquement

- Benzoate de sodium
- EDTA
- PEG-40 Huile de ricin hydrogénée
- Acide citrique
- Citrate de sodium
- Caprylate sorbitan

Effet tampon de pH

La lotion contient un système à effet tampon à base d'acide citrique conçu pour préserver l'équilibre naturel du pH de la peau.[1] Des études scientifiques ont démontré que l'une des principales causes de l'érythème fessier est le déséquilibre du pH qui se produit lorsque le lange est souillé. Les langes sales (combinaison urine et selles) contiennent souvent des enzymes digestives qui irritent la peau. Pour contrer cet effet, les lingettes pour bébé Pampers contiennent une lotion spécialement conçue, dotée d'un effet tampon permettant de ramener rapidement le pH de la peau à des valeurs normales comprises entre 4,5 et 6,0.

Les lingettes Pampers® Aqua Pure sont :

sans alcool
sans parfum
sans parabène
sans phénoxyéthanol
sans colorant
sans blanchiment au chlore

[1] Données internes de P&G

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PAMPERS SOUTIENT LA SOCIÉTÉ BELGE DE PÉDIATRIE

European Society for Pediatric Dermatology
Approuvées par ESPD

P121. Kinsbourne syndrome as complication of a Mycoplasma pneumoniae infection
L Adouane, M Hoyoux, J Frère, C Barrea, MC Seghaye. Centre Hospitalier Universitaire de Liège

P122. Cardio-facio-cutaneous syndrome and severe hyperthermia in the child
M Baillat, MA Heng, B Chabrol, C Despineux. UC Louvain

P123. Management of arteriovenous malformations in pediatric population
A Remy, JJ Kengo. Epicura Hornu

P124. Bed for preventing post dural puncture headache, is it efficace?
S Groignet, G de Bilderling, JP Misson. CHR Namur

ONCOLOGY – HEMATOLOGY

Oral presentations

O13. **Long-term auditory effects in patients treated for brain tumors during childhood**
A Rycx, L Bollé, H Keppler, I Van Den Bossche, E Vandecruys, I Dhooge, C Dhooge. Universiteit Gent, UZ Gent

Posters with short oral presentations

OP21. **Retrospective analysis of the incidence and characteristics of pediatric myelodysplastic syndrome and juvenile myelomonocytic leukemia in Belgium**
L De Smaele, M Hofmans, P De Paepe, J Philippé, N Van Roy, V Labarque, B De Moerloose. UZ Gent, AZ Sint Jan Brugge, UZ Leuven

OP22. **Antibacterial prophylaxis with fluoroquinolones in children with acute myeloid leukemia: impact on viridans group streptococci**
T Bauters, L Staels, G Laureys, L Willems, B De Moerloose. UZ Gent

OP23. **Comparison of the incidence of sinusoidal obstruction syndrome in a pediatric stem cell transplantation population receiving oral or intravenous busul**
V Van Keulen, V Bordon, T Bauters, S Van Lancker, A Verstraete, G Laureys. UZ Gent

OP24. **Disease monitoring by liquid biopsies in pediatric patients with solid tumors**
J Messiaen, L Spans, R Sciot, H Segers, A Uyttebroeck, M Debiec-Rychter, I Vanden Bempt, S Jacobs. UZ Leuven, KU Leuven

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O1.

Univentricular heart malformation: outcome after (prenatal) diagnosis

L Duchi, S Lesage, K Vandekerckhove, K Van Herck, D De Wolf, J Panzer, H De Wilde, K François, T Bove, K De Groote. UZ Gent

Objectives

To examine the outcome during staged palliation in patients with a univentricular heart (UVH) and the influence of isomerism.

Methods

Single center retrospective study in liveborn patients with intention to treat, diagnosed with UVH between 2006 and 2018.

Results

A total of 64 patients with intention for surgical palliation were included. 34 had a dominant left and 30 a dominant right ventricle. Five patients had right isomerism.

Five patients did not need first stage surgery, 2 died spontaneously pre-op, and 57 patients underwent first stage surgery (median age: 6 days). Survival rate until second stage surgery was 82% (9 deaths during hospitalization, 1 interstage death).

52 patients, being the 47 survivors after first stage surgery and the 5 patients who did not need said surgery, underwent second stage surgery (median age: 6 months). Survival rate until third stage surgery was 81% (6 deaths during hospitalization, 4 interstage deaths).

Eight patients are still awaiting third stage surgery and 34 patients underwent said surgery (median age: 3 years 9 months). Survival rate after third stage surgery was 100%, with a maximum of 10 person-years follow-up after this surgery.

Overall mortality in patients with intention to treat was 34%. At least 5/20 deaths were non-cardiac related. Mortality in patients with dominant LV (8/34) was lower than in those with dominant RV (14/30) but was not significantly different (p=0.067). However, this might be a potentially clinical relevant difference in outcome after surgery. Patients with isomerism (overall mortality 80%) had a significantly worse prognosis (p=0.044). No correlation was found between type of first stage surgery and mortality up to second stage surgery (p=1.000).

Conclusions

Mortality in the intention to treat group was 34% and is significantly higher in the presence of isomerism. Ventricular morphology was not found to be a predictor for death in the intention to treat group, though it was for overall postnatal mortality (i.e. compassionate care and intention to treat; described in a separate abstract). These results can contribute to a more profound prenatal and postnatal counseling and decision making.

OP1.

Effects of chronic ventricular volume deprivation and acute reloading studied in an animal (ovine) model

B Cools, F Rega, P Claus, L Fresiello, J Duchenne, T Verbelen, M Gewillig. UZ Leuven, KU Leuven

Introduction

Many cardiac conditions cause chronic volume deprivation of the systemic ventricle (e.g. Fontan, mitral stenosis, PAH, large ASD). We created a chronic volume deprived ventricle in an animal model (ovine) to study the effects of chronic volume deprivation and acute reloading.

Methods

In lambs a tight PFTE strip was placed around the inferior and superior caval vein through thoracotomy (n=14). Ten months later the PTFE bands were percutaneously dilated. Cardiac MRI was performed prior and within 48 h after debanding, hemodynamic data and PV loops (CD Leycom) were recorded prior and immediately (30 min) after debanding. Histology was done. Data was compared to age and weight matched healthy controls (n=9).

Results

1/ Survival: 2 animals died after banding (ascites), 2 after debanding (rupture IVC).
2/ Acute hemodynamic effects (PV loop): baseline EDP is elevated 9.0±3.3 mmHg compared to normals 1.0±3.4 mmHg (P <0.05). EDP rises after debanding to 12.4± 4.0 mmHg (P <0.05). The left ventricular CO is 3.3±0.7 pre and 3.0±0.6 L/min post debanding, compared to 3.0±1.2 L/min in normal animals. The EDV is 70.2+-8.7 pre and increases to 81.9+-13.5 ml post debanding (p <0.05), compared to 85±7.2 ml for normal animals. The ESV rises from 33.2±5.4 to 44.5±11.3 ml after debanding (55.6±18.4 ml in normal). Heartrate rises from 88±9 to 101±15 BPM compared to 94+-7 BPM in controls.
3/ Effect after 48 h (MRI): EDV on MRI is 70.5±7.7 ml before and 64.2±10.5 ml 48 h after debanding (P 0.79) ESV is 33.1±5.0 before and 34.1±11.3 ml 48 h later (P 0.14) EDV in healthy controls 76.1±14.1 ml and ESV 41.2±7.7 ml. 62.2±10.5 ml.
4/Histology: mean mass RV 29.2±4.4 g, LV 93.1±16.6 g was not significantly different from healthy controls; mean number of transected myocytes per 0.5 mm was RV 16.2±2.2 and LV 16.4±1.9, no signs of fibrosis.

Conclusion

In a chronic volume deprived ventricle the end diastolic pressure is elevated without marked histologic changes; EDP acutely rises when restoring the preload. Better understanding of this phenomenon may help avoiding/treating decreased ventricular compliance.

OP2.

Percutaneous lymphangiosclerosis as treatment for protein loosing enteropathy and plastic bronchitis in patients with failing Fontan circulation

M Gewillig, E Storme, B Eyskens, B Cools, R Heying, D Boshoff, J Hubrechts, G Maleux. UZ Leuven

Objectives

To determine the feasibility and clinical result of superselective lymphangiosclerosis in Fontan patients with protein loosing enteropathy PLE and plastic bronchitis PB.

Methods

Dilated lymph vessels in periportal (PLE) or paratracheal (PB) position were punctured with a 22G Chiba needle; good intralymphatic position ascertained by water soluble contrast injection with drainage to abnormals 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®) 3-4/1. The effect on symptoms, plasma albumin or expectorations was monitored.

Patients & Results

Four patients with PLE were treated with periportal lymphangiosclerosis; Fontan at 3.4±0.4 years; PLE started 2.3±1.0 (range 1.0-3.3) y after Fontan; time since start PLE 8.2±3.7 (range 3.3-12.2) y; in all patients (1 patient required a 2nd procedure) the lymphangiosclerosis resulted in complete lasting normalisation of albumin levels after withdrawal of all medication (FU 4-8 months). Symptoms of diarrea and abdominal bloating disappeared with significant improvement of quality of life.

One patient (Fontan at 2.9 y; age 16.4y) with PB for 2 years had exacerbation of casts expectorations after a surgical procedure. Inguinal intranodal lymphangiography failed to improve symptoms, but demonstrated peritracheal dilated lymphatics. Direct puncture (left and right parasternal) with paratracheal lymphosclerosis resulted in lasting absence of tracheal casts (FU 3 months).

Conclusions

Periportal/peritracheal lymphangiosclerosis is a promising technique in Fontan patients with PLE/PB. Larger series are needed to determine incidence and reasons of success/failure, with long term results and effects on liver function.

P37.

Non-invasive assessment of the liver in pediatric Fontan patients: time for thorough follow-up

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Introduction

Pediatric data about liver abnormalities after Fontan palliation are scarce. We assessed the prevalence and degree of liver problems through non-invasive investigations suitable for longitudinal follow-up.

Methods

35 Fontan patients (median age 11,8yrs; range 5,2-16,6yrs; 27 boys; median time since Fontan 3,29yrs) were evaluated. A liver ultrasound was performed evaluating nodularity, coarsened echotexture, ascites, liver and spleen size. The diameters of inferior vena cava (IVC), portal vein (PV) in in- and expiration and the IVC collapsibility index (IVCCI) were measured. The pulsatility ratio (PR) of the PV and hepatic vein (HV) and the damping index (DI) were calculated. The resistance index (RI) of the PV, hepatic artery (HA) and superior mesenteric artery (SMA) was examined. Fibroscan (Echosens) was used to perform transient elastography (TE). Blood values of AST, ALT, Î³GT, Alk Phos, bilirubin, total protein, albumin, alpha-foetoprotein, platelet count, cholesterol and Apo-lipoprotein A1 were measured.

Results

Nodularity was found in 2/35 patients and irregularity of the liver surface in 2/35 other patients; hepatomegaly was present in 32% of patients, splenomegaly in 15%. PV mean flow velocity was < 15 cm/sec in 19 (54%) patients, correlating with portal hypertension. 22 patients (63%) showed IVCCI values below 17%, indicative of venous congestion (2). HA RI and SMA RI were inversely correlated with time post Fontan ($p<0,05$; $r2=-0,369$ and $r2=-0,365$ resp.).

Liver stiffness was significantly increased compared to controls, with a median(range) of 12,6 kPa (6,6-25,7) versus 4,6 kPa (2-9,5) ($p<0,001$) from early after Fontan.

AST, ALT, Î³GT and direct bilirubin were abnormally increased in respectively 12 (34%), 5 (14%), 24 (69%) and 7 (20%), platelet count was decreased in 7 (20%).

Conclusion

Non-invasive investigations were not able to confirm or differentiate fibrosis from hepatic congestion. We propose follow-up with serial measurements of lab values (ALT, Î³GT, direct bilirubin, alpha-foetoprotein, platelet count and clotting), US and Doppler parameters (morphology, IVCCI, PV flow velocities, HA RI, SMA RI and PV pulsatility index) and TE. The use of reliable and accurate non-invasive techniques to assess liver fibrosis in children after Fontan remains a major topic for future research

P38.

Univentricular heart malformation: outcome after surgery and importance of isomerism

L Duchi, K Van Herck, K Vandekerckhove, D De Wolf, J Panzer, H De Wilde, K François, T Bove, K De Groote. UZ Gent

Objectives
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Five patients did not need first stage surgery, 2 died spontaneously pre-op, and 57 patients underwent first stage surgery (median age: 6 days). Survival rate until second stage surgery was 82% (9 deaths during hospitalization, 1 interstage death).
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Eight patients are still awaiting third stage surgery and 34 patients underwent said surgery (median age: 3 years 9 months). Survival rate after third stage surgery was 100%, with a maximum of 10 person-years follow-up after this surgery.
Overall mortality in patients with intention to treat was 34%. At least 5/20 deaths were non-cardiac related. Mortality in patients with dominant LV (8/34) was lower than in those with dominant RV (14/30) but was not significantly different (p=0.067). However, this might be a potentially clinical relevant difference in outcome after surgery. Patients with isomerism (overall mortality 80%) had a significantly worse prognosis (p=0.044). No correlation was found between type of first stage surgery and mortality up to second stage surgery (p=1.000).

Conclusions
Mortality in the intention to treat group was 34% and is significantly higher in the presence of isomerism. Ventricular morphology was not found to be a predictor for death in the intention to treat group, though it was for overall postnatal mortality (i.e. compassionate care and intention to treat; described in a separate abstract). These results can contribute to a more profound prenatal and postnatal counseling and decision making.

P39.

Percutaneous closure of a window-like patent arterial duct with an ASD occluder

K Vandekerckhove, J Panzer, D De Wolf, UZ Gent

Background

Patent arterial duct (PDA) is one of the most common congenital cardiovascular malformations. In patients living at high altitude, PDA tend to be larger and increased pulmonary artery (PA) pressure is common, what makes closing the PDA more challenging.
For several years, there is a cooperation between the pediatric cardiologists of the University Hospital of Ghent and a NGO called 'Chaîne de l'espoir' that has pediatric health-projects in different African countries. In some exceptional cases, children are brought to Belgium for the intervention they need.

Methods

We present a case of PDA-closure in a girl from the Democratic Republic of Congo.

Results

A 12 year old girl from Congo was transferred to Belgium to undergo percutaneous closure of a large patent arterial duct. She had clinical signs of heart failure with exertional dyspnea, and a very loud continuous murmur. Echocardiography showed a wide patent duct of 12 mm diameter with a window-like configuration (short ductal segment, large diameter). There was marked left ventricular dilatation (diameter 77mm), with preserved systolic function and only moderate pulmonary hypertension.
During the procedure the window-like configuration was confirmed on angiography and with balloonsizing the appropriate size was determined. Implanting a 12mm ASD-Occlutech, the team succeeded in closing the PDA. During further catheterization normal PA pressure was measured, cardiac function was moderately impaired. The girl was discharged with ACE-inhibition. Echocardiographic follow-up one week later showed a mild, low velocity residual flow through the device, recovery of the left ventricular function (FS 33%) and diminishing of the LVEDd to 66mm. A few weeks later, the girl could safely return to Congo.

Conclusion

In this report we discuss the need for closing a large window-like patent duct, preventing further progression to irreversible pulmonary hypertension, and improving the left ventricular dilatation to preserve cardiac function in a girl of 12 years old from Congo. In this region of the African Great Lakes many of the PDA are notably large without irreversible pulmonary hypertension, a curious finding probably related to living on high altitude, limited access to healthcare and difference in treatment options compared to European countries.
The window-like configuration of the duct makes the normal PDA-devices not suitable for closing, this is why an ASD-device was chosen after careful balloons

P40.

Long term outcome after tricuspid valvectomy in a neonate with candida endocarditis: a case report

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Background

Isolated right sided infective endocarditis is uncommon. First choice treatment is antimicrobial medication. Surgery should be considered in specific conditions. The preferred surgical treatment is valve repair. Because of the long-term risk of developing severe right heart failure, tricuspid valvectomy is only considered in exceptional cases.

Data on tricuspid valvectomy is scarce. Aim of this report is to share a history of over 10 years survival after tricuspid valvectomy in infancy.

Methods

Pubmed research of the literature concerning tricuspid valvectomy in children and adults.

Results

We describe the case of a girl of 5 months old, diagnosed with a thrombus obstructing the tricuspid orifice. Cardiac surgery was performed within 24 hours. The tricuspid valve could not be repaired and valvectomy was performed. She was discharged 5 weeks postoperatively using diuretics and sildenafil 8 mg/kg/d. Ultrasonography at discharge showed a dilated right heart, with normal cardiac output and moderate reversed flow in the hepatic veins.

The following years the girl did surprisingly well. Mild pulmonic regurgitation was noted, without signs of increased pulmonary artery pressure. Sildenafil was gradually reduced and stopped 1 year after surgery. With diuretics as only medication, she had a normal growth pattern and exercise capacity without progressive dilatation of the right heart. Until the age of 11 years old, she stayed clinically stable without cardiac arrhythmias or signs of right heart failure.

Since the age of 11 years, exercise intolerance is present. Cardiopulmonary exercise test showed exercise intolerance and a decrease of peak oxygen uptake over the last 3 years. Cardiac MRI showed non progressive dilatation of the right ventricle and a normal ventricular ejection fraction. Because of suboptimal exercise performance and dilatation of the right heart, the question for surgical intervention arises. Based on criteria including those used in more common situations of right ventricular dilatation, such as pulmonary regurgitation, for now a conservative strategy is implemented, guided by the clinical state of the patient.

Conclusion

This case illustrates survival in good clinical condition for more than 10 years after tricuspid valvectomy performed in infancy. Consensus criteria for timing of re-intervention are lacking. Application of adult tricuspid and pulmonary regurgitation recommendations is helpful but has limitations.

P41.

Interventional closure of perimembranous ventricular septal defects: Experience with two commonly used occluding devices

Q Jordens, R Heying, B Cools, B Eyskens, J Hubrechts, M Roggen, M Gewillig. UZ leuven

Background

Ventricular septal defects (VSDs) are the most common congenital defect observed in new-born children and mostly have a so-called perimembranous localisation. Surgical closure remains the golden standard for closure of these defects, but since 1988 a catheter-based approach gained interest.

Methods

Our study reviewed pre-, per- and post-interventional data of 41 patients < 18 years of age who underwent an interventional percutaneous closure of a perimembranous VSD (pmVSD) by the primarily use of SJM Amplatzer® (22 patients) or PFM Nit Occlud® (19 patients) between January 1, 2003 and July 1, 2017.

Results

The mean age, weight and septal defect diameter are comparable for both groups and only mild pulmonary hypertension was present. Complete occlusion was achieved in 59% of the Amplatzer® group and in 47% of the Nit Occlud® group at day one and respectively 95% and 58% at 6-month follow-up. Haemolysis occurred in 2 patients after placement of Nit Occlud® and complete AV-block occurred in 2 patients after placement of the Amplatzer® device.

Conclusion

In well-selected patients, good results can be expected after catheter-based closure of pmVSD but caution has to be preserved regarding the known complications as complete AV-block and haemolysis.

P42.

Cardiac fibroma in children: single center experience

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Background

Cardiac tumours occur rarely in paediatric patients. Fibromas are the second most common type. They are benign but can affect the adjacent cardiac structures with inflow or outflow tract obstruction, valvular dysfunction, arrhythmias, and sudden death.
Therefore surgical resection is recommended.

Methods

Description of three cases.

Results

Case 1: in 2006, a 8 month-old girl presented with cardiomegaly on X-ray. Additional imaging (MRI) showed a large intraventricular mass, suitable with cardiac fibroma. Surgical excision was performed through left ventricular incision, using blunt dissection. The tumoral cavity remained and during follow-up a double lumen anatomy with localized poor contractility appeared echography. General left ventricular function was adequate.
Case 2: in 2008, a 10 month-old girl presented in exactly the way as case 1. During surgery, the cardiac fibroma was enucleated by blunt dissection, through left ventricular incision. The remaining opening in the decoupled wall of the left ventricle was stitched, using pericard pledgets, fixed with Tissuecol. Follow-up showed a reasonably good restoration of the ventricular cavity and only mild impairment of septal mobility in the region of the resected fibroma.
Case 3: in 2018, A 1 year-old boy presented with an asymptomatic murmur. Ultrasound showed a sharply aligned tumoral mass in the ventricular septum with mild right outflow obstruction. A cardiac fibroma was diagnosed on MRI. During surgery, the residual cavity that remained in the septal wall, after enucleation of the mass, was filled with Tissuecol, the incision stichted. There was an uncomplicated recovery afterwards, with good echographic restoration of both ventricle cavities, and normal cardiac function and mobility of the septal wall.

Conclusion

This report describes a single center experience in surgical excision of cardiac fibromas in children. All patients recovered completely, with good left ventricular function. In case 1 however we see a localized poor contractility of part of the left ventricle due to the remaining tumoral cavity. This opposes a risk for thrombus formation. Therefore we state a surgical restoration of anatomical ventricular cavities superior to solitary resection of the tumor.

P43.

Percutaneous obliteration of the Right Ventricle in patients witch pulmonary atresia, intact ventricular septum as management strategy to avoid corona

B Cools, B Eyskens , R Heying, D Boshoff, B Suys, M Gewillig. UZ Leuven

Background

Patients with pulmonary atresia and intact ventricular septum (PA-IVS) and hypertensive right ventricle RV often present with (multiple) coronary sinusoids to the coronaries. A suprasystemic pressure wave during fetal and early life can damage the coronary system with progressive accelerated hyperplasia of endothelium, resulting in myocardial ischemia and (avoidable) excess mortality.

Method

We aimed to reduce the coronary pressure wave through the sinusoids by abolishing RV stroke volume with percutaneous devices in 4 patients with PA-IVS: median age 26 months (range: 2.5-51). The youngest one had a systemic-to-pulmonary artery shunt, 2 had bidirectional Glenn and 1 Fontan connection.

Procedure

All patients had RV injection and selected coronary angiograms to determine whether significant LV coronary perfusion was RV dependent (=exclusion); singular sinusoidal coronary flow limited to the hypoplastic hypertrofic RV was no contraindication. All 4 patients had dual perfusion with competitive flow from the RV through the sinusoids in both right and left coronary arteries. One presented with coronary sinusoids connected to the circumflex right coronary artery causing large progressive aneurysmal dilatation of 18 mm.
Various types of devices were used: Amplatzer vascular plug II 14mm; vascular occluder 12mm; vascular plug 10 and 16mm; 27 coils (diameter 5-15mm) in the oldest patient.
All patients were monitored for arrythmia or ECG changes during 24 hours in the ICU ward. Echocardiography was performed the day after procedure and clinical follow-up was gathered.

Results

RV gram after cavity obliteration showed no more significant coronary perfusion through sinusoids. There was no mortality nor any major complication; no arrythmia. There were only minor and transient changes in the levels of troponin.
Echocardiography: abolishment RV stroke volume and flow through sinusoids. LV-function was maintained. Long-term follow-up showed no acute incidents. Coronarography at pre-Fontan evaluation showed no progress of coronary abnormalities in 3 patients. The oldest patient showed at 18 years (14 years after procedure) a dystrophic LAD with stenosis, asymptomatic.

Conclusion

In very selected patients, obliteration of the RV cavity by percutaneous devices is safe and abolishes the pressure wave in coronary sinusoids. When performed early, this may halt coronary damage and avoid excess mortality.

P44.

Complex malignant exercise induced ventricular tachyarrythmia in a girl with Turner Syndrome

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Introduction

Patients with Turner Syndrome (TS) carry a higher risk for sudden cardiac death (SCD) than healthy age matched women that is not related to the presence of congenital cardiac disease or to treatment. Higher cardiac arrhythmogenic potential with long QT-interval and QT-wave dispersion has been reported in patients with TS that may be responsible for SCD.

Clinical case

We report on the case of a 14 year old girl with TS treated by growth hormone.
Previous cardiologic examination done 3 years before was normal. She complained about exercise intolerance and thoracic oppression at exercise, but no syncope or palpitations.
The clinical examination was normal apart of the short stature and overweight.
Echocardiography and ECG were normal with QTc duration of 440 msec.
Stress ECG was performed on ergometric bicycle. Under a load of 1,4 Watts/kg, HR rose up to 178/min. Polymorphic ventricular extrasystoles followed by several episodes of bilateral ventricular tachycardia (VT), a short episode of supra-ventricular re-entry tachycardia (SVT) and ventricular fibrillation occurred. Exercise was immediately interrupted. Patient converted spontaneously in sinus rhythm.
A treatment with the bêta-blocker Nadolol was introduced and titrated up to a dosage of 1,6 mg/kg/d. Magnesium supplementation and spironolactone (to optimize potassium levels) were introduced. Under treatment, stress ECG and Holter-ECG were normal except for prolonged QTc interval at rest (460-480 msec.). QTc 4 minutes after stress termination was 435-455 msec. Genetic testing is pending. The option of ICD-implantation is currently discussed.

Discussion

In this patient with TS we documented a complex cardiac arrhythmia at exercise including polymorphic VES, VT, VF and SVT. While QTc at rest measured several times in the past was normal, under treatment it was prolonged but converted to normal 4 minutes after exercise. Prolonged QT interval, even if it can be assumed that it was not the primum movens of the malignant tachyarrhythmia elicited by catecholamine secretion in this case, must be considered as potentially co-responsible.

Conclusion

Our report confirms cardiac arrhythmogenic potential in girls with TS and demonstrates malignant ventricular tachyarrhythmia related to exercise in such a patient. Follow-up of TS patients should therefore include regular exercise ECG.

P45.

Evolution to a native aortic isthmus stenosis in a boy

MC Seghaye. CHU Liège

Aortic isthmus stenosis (ISTA) that is the narrowing of the aorta located at the level of the aortic isthmus accounts for 5-8% of all congenital cardiac diseases. Besides the critical form that is typically diagnosed in the neonate or infant, the native forms affecting children or adults may be missed for years or decades. We report on the case of a 9 year-old-boy, who was followed-up for relative hypoplasia of the aorta and in whom development of ISTA could be documented over time.

Case

The 2,5-year-old boy was explored for the first time for heart murmur.
Personal history was not contributive.
Clinical examination revealed overweight, a 2/6 non-specific systolic murmur. Heart sounds were normal. There was no intensity difference between humeral and femoral pulses. Arterial pressure (PA) could not be measured due to important anxiety.
ECG was normal. Echocardiography showed normal cardiac cavities- and muscular wall dimensions. The aortic valve was slightly hypoplasic (Z score - 2) without flow acceleration. The dimensions of the ascending aorta, of the aortic arch and the aortic isthmus were in the inferior normal range (Z score -1). The flow measured in the descending aorta was accelerated (2,9 m/sec.) without any diastolic component.
Follow up were conducted twice a year.
The remarkable findings were:
- Apparition of echocardiographic signs of ISTA (accelerated systolic flow velocity with diastolic flow component in the descending aorta (3,6 m/sec.), double systolic flow pattern) at the age of 5,5 years.
- Normalization of the dimensions of the aortic valve and arch at the age of 6 years (Z score 0) but increase of the isthmus stenosis (Z score < -3).
- Apparition of a left paravertebral systolic murmur and of a significant systolic pressure difference between upper and lower extremities (10 mm Hg) at the age of 8,5 years without any pulse intensity difference.
- Stable overweight, absence of arterial hypertension.
Cardiac MRI is scheduled to prepare angioplasty and stenting of the aortic isthmus.

Conclusion

This case documents well the natural history of a native ISTA over a time period of more than 5 years in a now 9 year-old-child, in whom the typical clinical findings of ISTA are currently absent but the slight PA difference between arms and legs and the left paravertebral systolic murmur. It points therefore out the importance of follow-up in cases of borderline measures of cardiac structures, especially of the aortic ones in young children.

O2.

Morning serum cortisol correlates with fasting glucose in children and adolescents with overweight

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Background and aim

Increased serum cortisol concentrations have been found in a variable percentage of obese children. The underlying mechanism as well as the consequences of increased serum cortisol concentrations in childhood obesity have not been well studied. We investigated whether obese children and adolescents with hypercortisolemia would be at higher risk for fasting hyperglycemia, hypertriglyceridemia, hyperhomocysteinemia and hyperleptinemia, as observed in patients with Cushing syndrome.

Methods

Metabolic, hormonal and body composition results of 234 (99 male) overweight (BMI SDS > 1.3) children and adolescents, aged between 4 and 18 years and diagnosed with primary obesity, were retrieved from their medical files. Morning fasting serum cortisol (Cortisol II Cobas), insulin, leptin, glucose, cholesterol, HDL cholesterol, triglycerides and homocysteine were measured by standard automated methods. HOMA-IR and LDL cholesterol were calculated. Body fat was measured by BIA and blood pressure by oscillometry. Correlations were analyzed by Spearman Rank correlation.

Results

Median age (range) age was 9.4 (4-17.4) years, BMI z-score 2.4 (1.4-5.7) and body fat percentage 36 (24-70 %). In total 49 (20 male and 29 female) patients had an elevated morning serum cortisol (> 180 µg/L), whereas 12 patients had a decreased (< 62 µg/L) value. Median serum cortisol was not significantly different between males (130 (47-299) µg/L) and females (129 (36-323) µg/L). No significant correlations between serum cortisol and age, BMI z-score, waist z-score, diastolic and systolic blood pressure z -score were present. Serum cortisol correlated significantly with fasting glucose (r = 0.193 ; $p < 0.005$), triglycerides (r= 0. 143 ; $p < 0.05$), homocysteine (r= 0.145 ; $p < 0.05$) and leptin (r = 0.145; $p < 0.05$), but not with HDL cholesterol, fasting insulin and HOMA-IR. Adjusting for insulin and leptin did not change the correlation between serum cortisol and fasting glucose.

Conclusion

In overweight children and adolescents elevated morning serum cortisol levels were found in 20 % and were associated with higher fasting glucose concentrations, irrespective of underlying insulin resistance. The long-term cardiometabolic consequence of hypercortisolemia in childhood obesity needs further study.

OP3.

Prediction and prevention of type 1 diabetes (T1D) in neonates: a new trial program in Europe

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Background

Type 1 diabetes (T1D) is a common chronic disease in childhood and is increasing in incidence, making primary prevention a major public-health goal. Early identification of neonates and infants who are at risk of T1D is essential and can now be achieved before beta-cell autoantibody markers arise by using genetic markers. In this context, the Global Platform for the Prevention of Autoimmune Diabetes GPPAD has established a screening program (GPPAD-02) to identify high genetic risk newborns and ask them to participate in a primary prevention study, the Primary Oral Insulin Trial (POInT). Afterward, participants will be followed and monitored for early diagnosis of T1D.

Methods

Capillary blood samples are obtained in newborns and infants either at delivery (cord blood), or together with the regular newborn screening, or at a pediatric baby-visit before the age of 3 months. Using a genetic score based on 47 T1D susceptibility SNPs and the first degree-family history for T1D, infants with a high genetic risk for multiple beta-cell autoantibodies (>10%) by the age of 6 years are identified, a sub-group comprising 1% of the general population. Families of high-risk infants receive counselling and are offered participation in POInT, a randomized, double-blinded, multicenter phase IIb primary prevention trial. The primary endpoint of the study is to determine whether daily administration of oral insulin from age 4-7 months until age of 3 years in children with elevated genetic risk for T1D reduces the cumulative incidence of beta-cell autoantibodies and diabetes in childhood. Target is 300,000 screened infants and 1040 enrolled infants in POInT in 3.5 years. To achieve those objectives, the study is carried by 7 main centers located in Germany, Belgium, Poland, Sweden and UK.

Results

Since initiation in October 2017 GPPAD-02, over 51000 infants have been screened with 120 infants randomized in POInT since February 2018. GPPAD-02 has been actively running in Belgium (Universitair Ziekenhuis Leuven) since July 2018 with 520 infants screened and initiation in nine other

OP4.

GENEPEDIAB study: multicenter screening of genetic forms of diabetes in cohorts of children and adolescents with type 1 diabetes

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Background/Aims

Diabetes, which affects 420 million people worldwide with a continuously rising incidence, is defined by a state of chronic hyperglycemia; a criterion referring to a heterogeneous group of diseases with various etiologies and distinct therapeutic options. Besides the two main forms of diabetes (i.e., type 1 (T1D) and type 2 (T2D)), there are rare subtypes of the disease called monogenic diabetes (or formerly MODY) that are hardly diagnosed because of their resemblance to T1D or T2D. Since these monogenic diabetes may appear early in life, a consortium of expert pediatric clinical centers was created under a clinical research initiative (the GENEPEDIAB study) to develop tools for accurate diagnosis of rare diabetes and to propose appropriate care to these children and adolescents wrongly assigned to T1D or T2D cohorts. The GENEPEDIAB study was initiated in the context of a broader collaborative project (DiaType) with the objective to develop personalized diabetes medicine and better patient care.

Methods

For discrimination of patients with monogenic diabetes from those with classical forms of diabetes using the DIAMODIA (DIAgnose MOnogenic DIAbetes) score, patients currently enrolled in the GENEPEDIAB study are being phenotyped and genotyped for T1D risk (HLA typing and 30 SNPs on specific loci). Patients fulfilling sufficient criteria are then genotyped using the routine MODY panel, before being proposed a thorough gene analysis.

Results

As of today, our prospective cohort consists of 817 patients run through our flowchart analysis, with a subgroup of 81 patients identified with atypical diabetes, whereas the retrospective cohort (n=678) was downsized to 40 patients with atypical diabetes. Our current genotyping data revealed a global DIAMODIA score yield of 67%, corresponding to the proportion of patients with atypical diabetes being diagnosed with monogenic diabetes.

Conclusion

Our current work, inside the GENEPEDIAB study and the DiaType project, will enable us to adapt treatment to diabetes etiology and help to provide genetic counseling to patients and their family members. We anticipate that our broad genetic analyses will provide us with important information about the genetic susceptibility of these subgroups of patients with atypical diabetes.

OP5.

Empagliflozin And GABA Improve Î²-Cell Mass And Glucose Tolerance In New-Onset Type 1 Diabetes

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Background/Aims

Presently, the autoimmune character of T1D is challenged, but it is indisputable that inflammation plays a key role in its development. We hypothesized that glucotoxicity could contribute to Î²-cell mass destruction through maintenance of inflammation. Here, the aim is to evaluate empagliflozin (EMPA) potential to protect Î²-cell mass against glucotoxicity, and GABA potential to increase the residual Î²-cell mass after diagnosis of T1D.

Methods

In a streptozotocin-treated mice model of T1D, empagliflozin and/or GABA were delivered during seven days or three weeks by oral gavage or intraperitoneal injection, respectively.

Results

As compared to untreated T1D mice, EMPA-treated T1D mice had a better glucose homeostasis during tolerance tests and decreased FFA levels. EMPA-treated T1D mice had a better islet density, numbers and preservation of islet architecture, compared to T1D mice. T1D mice showed islet with immune infiltration whereas EMPA-treated T1D mice displayed no islet infiltrate. Islets from EMPA-treated mice were also less subjected to ER stress and inflammation, as shown by qPCR analysis. Furthermore, parameters of glucose homeostasis and Î²-cell mass were also improved, as compared to diabetic controls, when T1D mice were treated for 3 weeks with GABA and EMPA. Interestingly, T1D EMPA+GABA mice had higher glucagon levels than T1D mice, without modifications of glucagon area/islet area ratios.

Conclusion

Empagliflozin and GABA, used in monotherapy, have positive effects on Î²-cell mass preservation or proliferation through an indirect effect on islet cell inflammation and ER stress. Further researches are mandatory to evaluate whether empagliflozin and GABA may be a potential therapeutic treatment to protect Î²-cell mass after T1D diagnosis

P46.

Feasibility and reproducibility of ultrasound techniques to measure intra-abdominal fat in infants and preschool children

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Background and aim

Ultrasound is a rapid, non-invasive direct method to measure abdominal fat content. Three ultrasound techniques have been described (Holzhauer et al., Mook-Kanamori et al. and De Lucia Rolfe et al.) and partially validated in children younger than five years. A direct comparison between the three techniques within the same child has not been done. The aim of this study was to compare the three most used ultrasound techniques for the measurement of the abdominal fat in young children, in terms of feasibility, reproducibility and dependence of biometrics.

Methods

34 children, aged between 0.1 to 4.7 years, underwent an ultrasound examination using the Mindray M5 device following the methodology of Holzhauer et al., Mook-Kanamori et al. and De Lucia Rolfe et al. One of the three studied techniques was repeated in 8 or 9 children. The possibility and duration of the image acquisition was tested. The reproducibility was assessed by calculating the correlation coefficient, the absolute difference, the limits of agreement, the coefficient of variation, the intraclass correlation coefficient and the technical error of measurement.

Results

Except for the visceral fat imaging as described by De Lucia Rolfe et al., qualitative images could be obtained by the other techniques, allowing the measurement of the subcutaneous and preperitoneal fat layers. All techniques showed a high correlation between the repeated measures, except for the Mook-Kanamori et al.-technique, in which the probe is kept longitudinally. All values of the subcutaneous measurement, regardless of the technique, indicated strong reliability criteria. The techniques to measure preperitoneal fat described by Holzhauer et al. and Mook-Kanamori et al. (with the probe transversal), had comparable reliability criteria. The subcutaneous fat layer thickness correlated with the subscapular skin fold thickness, whereas the preperitoneal fat correlated with age, bodyweight, body length and the abdominal circumference of the children. The median duration for image acquisition ranged between 23 and 80 seconds.

Conclusion

The three described ultrasound techniques are globally feasible (except for the technique of De Lucia Rolfe et al. for the visceral fat) and reliable methods for the measurement of abdominal subcutaneous and preperitoneal fat layers in children younger than five years. The technique of Holzhauer et al. is recommended for both practical (independant of breathing, speed of execution) and reliability (smallest technical error) reasons.

P47.

Novel PAX8 mutation in two siblings with congenital hypothyroidism due to thyroid dysgenesis

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Background

Approximately two thirds of congenital hypothyroidism (CH) cases result from developmental abnormalities of the thyroid gland (thyroid dysgenesis (TD)), leading to absent, ectopic or normally shaped and located but hypoplasic thyroid. Only in 5% of them, a mutation in a known gene is found. We present on 2 siblings with CH due to TD harbouring the same new PAX8 mutation.

Case report

Patient 1 is a newborn boy from a non-consanguineous family from Morocco (4 healthy siblings) who presented a high level of TSH on the dry blood neonatal screening at day 4 of life(75.7 mU/L). He was born at 38 weeks (BW: 3400g ; BL: 48cm ; HC : 38 cm). On day 6, serum TSH was 139mU/L (N: 0.3 - 4 mU/L) , FT4 : 1.2ng/dL (N: 0.8-2ng/dL) and thyroglobulin : 151µg/L (0-25 µg/L). Thyroid imaging (US and 99T scintigraphy) showed an eutopic non goitrous thyroid gland. Bone age X-ray was antenatal (inferior femoral but no superior tibial ossification point). L-Thyroxin treatment was initiated on day 6 of life. At the age of 3.8 years, his growth and psychomotor development were normal. A L-Thyroxin treatment interruption was then tempted for 3 months, but TSH increased to 283 mU/litre, FT4 at 3.7 pmol/L, thyroglobulin at 34 mcg/L. Imaging confirmed the presence of an in situ relatively hypoplasic gland. Treatment was reinitiated.

Patient 2: Is the young sister of patient 1. She was born at 38 weeks (BW : 3470g, BL : 51cm ; HC : 34cm) presented a high level of TSH on the dry blood spot neonatal screening at day 3 of life(44.7 mU/L).On day 5, serum TSH was 52.1 mU/L , FT4 at 19.3 pmol/L, thyroglobulin 456µg/L . Thyroid gland was eutopic on both ultrasound and scintigraphy. Bone age was antenatal (inferior femoral ossification point present but superior tibial point absent). Both parents had normal thyroid function.

Results

Panel gene analysis identified in both children the same likely pathogenic novel PAX8 mutation c.143 C>A in the heterozygous state. Additionally, a heterozygote variant of unknown significance was found in the DUOX2 gene in both siblings. Genetic analysis is ongoing in parents. Renal ultrasound showed a right renal duplication in patient 1.

Discussion and conclusion

PAX8 is a transcription factor involved in thyroid and renal development. Heterozygous loss-of-function PAX8 mutations have been reported in patients with familial thyroid dysgenesis (mainly thyroid hypoplasia) with an autosomal dominant inheritance. We report a novel PAX 8 mutation

P48.

Thyroid carcinoma presenting with hyperthyroidism and thyroid hot nodules in a female adolescent

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Background

Thyroid scintigraphy is the first line examination for anti-TSHR antibody negative hyperthyroidism. The finding of an isolated nodule or multiple 'hot' nodules is in general comforting, as a benign toxic adenoma or multinodular goitre is the most common diagnosis. We report a thyroid carcinoma in a 12 year old girl presenting a left thyroid nodule, a left and right upper pole tracer uptake at scintigraphy and thyroid hormone overproduction.

Case report

In a 12-years old female, presenting with a painless neck swelling since a month, a large (5 cm) non-tender firm nodule in the left thyroid lobe was detected at physical examination. Slight tachycardia, but no exophthalmos was present. Intermittent sore throat, increasing nervousness and a 2 kg weight loss since several months were reported. Her mother had been operated for a multi-nodular goitre. There was no history of cancer of intestinal polyps in the family. Thyroid function tests showed an elevated FT4 (51 pmol/l) and thyroglobulin (435 µg/l) and a decreased TSH (< 0.005 mU/L). Serum anti-thyroglobulin, anti-thyroidperoxidase and anti-TSH receptor antibodies were undetectable. Doppler ultrasound showed a normal right lobe and a sharp hypervascular solid multilobular mass (longest diameter 5.3 cm) with cystic components in the left lobe, whereas scintigraphy showed a global but heterogeneous hyperfunctioning thyroid gland with excessive uptake at the upper left lobe and upper right lobe. A left tracheal deviation by the left thyroid mass and no cervical lymphnodes or thoracal mass were seen at CT. Fine needle biopsy was refused by the patient. Diagnosis of large toxic adenoma crossing the midline, explaining the tracer uptake in the right upper pole region, was made. Patient underwent a left lobectomy after 2 months of methimazole therapy, normalizing thyroid function already after one month. Histological examination showed a mixed minimal invasive follicular and papillary carcinoma of the left lobe. No evidence of metastasis was found at whole body SPECT I-123 after total thyroidectomy. Results of TSHR gen and GNAS gen analysis are pending.

Conclusion

The presence of a hyperfunctioning thyroid nodule does not rule out thyroid cancer and warrants careful evaluation, even in the absence of cervical lymphnode invasion. The need for fine needle aspiration biopsy in nodules larger than 1 cm to improve diagnosis and first line surgical approach is stressed.

P49.

Fist description of a *NOTCH2* gene mutation in a newborn with thyroid ectopy

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Background and aim

Thyroid dysgenesis is the most frequent etiology of congenital hypothyroidism. In less than 5 % genetic abnormalities have been found in newborns with thyroid dysgenesis. Recently *JAG-1* loss of function variations have been described as a novel predisposing event (de Filippis T, 2016), whereas *JAG1* gene mutations were previously found in 4 % of children with pulmonary valve stenosis/peripheral pulmonary stenosis (Bauer R, 2010). We describe the first case of mutation in the *NOTCH2* gene, coding for a receptor for JAG-1 and together responsible for the Alagille syndrome, in newborn with thyroid ectopy and a congenital pulmonary valve stenosis.

Methods

Next generation sequencing of all coding exons and introns of the *JAG-1* and *NOTCH2* genes were performed.

Case report

The proband was born after an uneventful pregnancy of 39 weeks with a normal birth weight and length. Neonatal screening at day 3 showed an increased TSH value (514 mIU/L). Both parents and a brother are in good health. An increased serum TSH (> 100 mIU/L) together with a severely decreased serum FT4 (0.6 ng/dl) was confirmed at day 10. A systolic ejection murmer at the pulmonary site and a left plagiocephaly was noted at physical examination. Echocardiograhy confirmed a moderate pulmonary valve stenosis. Thyroid scan showed a sublingual tracer uptake. At the age of 2 months , a sudden left convergent strabismus of the left eye developed. A more prominent forehead and pointed chin was noted at follow up examinations. The combination of a congenital thyroid, heart and eye anomaly and facial dysmorphism led to the suspicion of a syndromal disorder. An array based CGH was normal. A *JAG-1* gene and *NOTCH* gene mutation screening was performed by a gene panel NGS, showing a novel heterozygous c.74-2A>G substitution (showing pathogenicity in several model) in the *NOTCH2* gene. Liver tests, X-ray of the spine and slit lamp examination were normal. NOTCH2 gene mutation screening in the parents is pending.

Conclusion

Genetic screening for *JAG1/NOTCH2* gene mutations should be considered in newborns with thyroid dysgenesis in association with a pulmonary valve stenosis, even in the absence of liver disturbances.

O3.

Long-term outcome of pediatric patients with inflammatory bowel disease receiving immunomodulators

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Background

In the era where new powerful biologicals are entering the market, the place of conventional immunomodulators (IMM) in treatment of paediatric inflammatory bowel disease (IBD) is questioned. We studied the long-term outcome of paediatric IBD patients receiving conventional therapy.

Methods

All children with Crohn's disease (CD) or ulcerative colitis (UC) followed at our centre between July 2008 and July 2018 were retrospectively included. Only children receiving conventional therapy including mesalazine, steroids and IMM (thiopurine, methotrexate) at start were studied. Patients requiring rescue therapy (either biologics or surgery) at diagnosis or with a follow-up (FU) <6 months were excluded. The primary outcome was steroid-free clinical remission without need for rescue therapy at 6 and 12 months after diagnosis and at last FU visit. Cox proportional hazard modelling was performed (Hazard risk: HR (95% CI) to determine variables associated with outcomes.

Results

A total of 221 patients (149 CD and 72 UC; 49% male) with a median age at diagnosis of 12 [10-14] years were included. IMM were started in 194 (88%) patients after a median duration of 1 [0-3] month. We excluded 45 (20%) patients due to insufficient FU (n=21), need of biologics (n=22) or surgery at diagnosis (n=2). A total of 176 patients were eligible for the study, with a median FU duration of 5 [2-8] years. Clinical remission rates decreased from 80% at month 6, and 58% at month 12, to 32% at last FU visit. The likelihood of remaining free of rescue therapy was 53% and 72% at 1 year and 27% and 31% at 5 years for CD and UC patients, respectively. For CD patients, higher CRP [HR 1.007 (1.002-1.011), p=0.002], lower albumin [1.045 (1.008-1.080), p=0.016] and growth failure [1.206 (1.011-1.362), p=0.040] at diagnosis were associated with an increased risk of need of rescue therapy. For UC patients, higher PUCAI score at diagnosis [1.037 (1.009-1.065), p=0.008] was determined as a risk factor for rescue therapy.

Conclusion

These real-life data in paediatric IBD show that only 32% of children remain free of biologic or surgery 5-years after diagnosis. Especially children with a high disease burden at diagnosis as witnessed by higher CRP, lower albumin and growth failure for CD and higher PUCAI score for UC were more likely to fail conventional therapy. This type of risk stratification algorithms will help to determine which patients will benefit from accelerated step-up therapy.

O7.

MyCyFAPP project: Use of a mobile application for self-management of PERT improves gastro-intestinal related quality of life in children with CF

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Introduction

Most patients with cystic fibrosis (CF) suffer from pancreatic insufficiency (PI), leading to fat malabsorption, malnutrition, abdominal discomfort and impaired growth. Treatment with pancreatic enzyme replacement therapy (PERT) is effective, but evidence based guidelines for dose adjustment are lacking.

Methods

A mobile APP for self-management of PERT was developed in the context of the HORIZON 2020 project MyCyFAPP. This APP contains an algorithm to calculate individual PERT-doses for optimal fat digestion, based on previous in vitro and in vivo studies on lipid digestion in CF-specific conditions, conducted as part of the project. Based on in vitro experiments a database of theoretical optimal enzyme doses for food products was built, and subsequently validated in vivo.

In addition, the APP includes a symptoms diary and educational material (handbook and games). The APP used by the patient is linked to a professional web tool allowing health care professionals to evaluate patient's data and give feedback.

A 6-month open label prospective multicenter interventional clinical trial was performed to assess the influence of the use of the APP on gastro-intestinal related quality of life (GI QOL) as primary outcome. GI QOL is measured by the modified PedsQL GI (Pediatric Quality of Life score, Gastrointestinal domain Symptoms Module), containing 9 subscales and expressed as a % with higher values indicating better GI QOL. The questionnaire was previously validated for use in children with CF and applied to patients from the age of 5, and to all parents.

Results

A total of 174 patients with CF and PI between 2 and 18 years were recruited in Valencia, Madrid, Rotterdam, Milan, Lisbon and Leuven, 154 patients ended the trial. Scores on the modified PedsQL GI significantly improved from month 0 (M0) (median 84.3, IQR 76.4-90.3) to month 6 (M6) (median 89.4, IQR 80.35-93.5) ($p< 0.0001$). Similar results were obtained in parents, with a significant paired correlation between scores from children and their parents (r 0.64, $p < 0.001$). Scores of most subscales of this modified PedsQL GI improved significantly in both patients and parents. Lower baseline PedsQL GI resulted in a greater improvement at M6 ($p < 0.001$).

Conclusions

MyCyFAPP improved GI QOL during a 6-months interventional trial and may help patients to improve self-management of PERT. The APP may be especially useful for patients with important abdominal symptoms.

OP6.

A prospective observational study of fatigue in children with inflammatory bowel disease compared to a and control group

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Background

Fatigue is a common symptom in patients with inflammatory bowel disease (IBD).
We aimed to assess fatigue and physical capacity amongst IBD children, compared to healthy peers and to evaluate correlation with disease activity scores, fecal calprotectin and quality of life.

Methods

We compared 70 IBD children (7 to 18 years) from Ghent and Antwerp University hospital with 66 age matched children. The patients, controls and one parent filled in the Dutch Multidimensional Fatigue Questionnaire (PedsQL) to assess the degree of general, sleep/rest and cognitive fatigue. Patients and controls performed a six-minute walk test (6MWT). Only patients completed the Impact-III Questionnaire and had fecal calprotectine, PCDAI (pediatric crohn's disease activity index) and PUCAI (pediatric ulcerative colitis activity index) measured.

Results

The proxy-reported fatigue scores were significantly higher than the IBD and healthy self-reported scores (p=0,015). The PedsQL scores of the children with IBD were significantly lower (higher level of fatigue) regarding general fatigue (p<0,000) and sleep/rest fatigue (p=0,015). Cognitive fatigue was not different between IBD children and controls (p=0,121). 6MWT was comparable between IBD group and controls (p=0,071).
Within the IBD group, CD and CU showed no significant difference in PedsQL fatigue and IMPACT scores (P=0,518). The impact score (meaning high impact of the disease on their life quality) was significantly correlated with the self-reported total fatigue score and with general fatigue. No significant correlation was found between the 6MWT and the impact score (p=0,816), or the general fatigue (p=0,751) in the IBD group.
CD patients with a normal level of calprotectine (<250 µg/g) reported a higher PedsQL score (less fatigue) and a higher impact III score (higher quality of life) than children with a high calprotectine level (>1500µg/g) (P=0,048 and P=0,049). This was not seen in children with CU. Elevated (10-35) PUCAI scores are correlated with more fatigue, PCDAI scores not.

Conclusion

Children with IBD report more fatigue than their healthy peers, especially general fatigue. A high level of fatigue is correlated with a low quality of life in both CD and CU. More fatigue in IBD is not a consequence of decreased physical fitness. Calprotectine level is in CD patients linked to fatigue, but not in UC patients. Disease activity scores show conflicting results.

OP7.

How to infuse heterologous human adult liver-derived progenitor cells safely?

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Background and Aims

Mesenchymal stem cell (MSC) infusions are currently evaluated in numerous clinical trials, but therapy-induced thrombi have been described in several patients. Most MSCs in fact express a procoagulant activity (PCA) linked to tissue factor (TF) expression, which is the fuse that ignites the coagulation cascade. The aim of this study was to optimize infusion protocols using Heter-ologous Human Adult Liver-derived Progenitor Cells (HHALPC) without inducing a thrombogenic risk after the infusion.

Methods

First cell dose escalation was studied using in healthy Wistar rat, with or without anticoagulants. Then the crucial role of TF in PCA was confirmed using flow chambers. Finally, we characterized the disseminated intravascular coagulation (DIC) induced by HHALPCs in vitro and investigated how to control the induced thrombotic and haemorrhagic risks in whole blood of healthy and cirrhotic pa-tients.

Results

In vivo we showed that the thrombogenic risk induced by HHALPC infusions is dose dependent. Infusions of high cell doses such as 50 x 106 cells / kg induced DIC 1h after transplantation with a significant decrease in platelets ($p < 0.01$), fibrinogen ($p < 0.001$), and coagulation factors II, V and VIII ($p < 0.01$) compared to control rats infused only with PBS. Infusions of lower cell doses, such as 5x106 cells / kg did not activate the coagulation cascade. Adding anticoagulants during infusions of high cell doses, such as heparin (300 I.U./ 5x106 cells) or a combination of heparin (10 I.U./ 5x106 cells) and bivalirudin could control the thrombogenic risk. Using flowed whole-blood under shear ex vivo, we found that HHAPLC promote fibrin clot formation in a TF-dependent way, inhibited by inactivate factor VII. By tubing loop model HHALPCs activated the coagulation cascade in a less explosive way in decompensated cirrhotic patient's blood, compared to healthy volunteers. HHALPCs only induced a significant decrease in platelets ($p < 0.01$) and fibrinogen ($p < 0.01$), but not in coagulation factors.

Conclusion

Low doses of MSCs (5x106 cells/kg) expressing TF do not induce a thrombogenic risk and could thus be used in future clinical trials treating acute decompensated cirrhotic patients, while monitoring platelet and fibrinogen levels. The thrombogenic risk induced by infusions of higher cell doses can be controlled by adding anticoagulants such as heparin and/or bivalirudin.

OP8.

Effect of an intensive residential rehabilitation program with adjusted nutritional care on body composition in adult patients with cystic fibrosis

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Background

In cystic fibrosis (CF), good nutritional status measured as body mass index (BMI) or as fat free mass (FFM), is strongly associated with pulmonary function and therefore longevity. FFM seems, compared to BMI, to be a superior parameter for malnutrition in CF. The effect of rehabilitation interventions on body composition has not yet been studied. The aim of this pilot study is to assess the impact of a 4-week residential rehabilitation program on body composition in adult CF patients and compare well-nourished and malnourished patients. Primary outcomes were FFM and fat free mass index (FFMI), fat mass (FM) and fat mass index (FMI), as well as forced expiratory volume in 1 second (FEV1).

Method

This study evaluates the impact of a 4-week residential rehabilitation program on body composition measured by dual energy X-ray absorptiometry scan in adult CF patients (2016-2017). European nutritional guidelines were adapted lowering fat intake and focusing on protein intake in general and carbohydrate intake before and after exercise. Two days a week a weighed food intake was noted and the caloric intake per day was calculated.

Results

After informed consent, 17 patients (12/5 male/female, 27.5 years± 6.6) were included. As a result of the program patients gained weight (mean 56 kg ± 10.5 vs 58.2 kg ± 11.3, $p< 0.001$), BMI increased (19.1 kg/m2 ± 2.1 vs 19.9 kg/m2 ± 2.2, $p< 0.001$), and FEV1 improved (49 % (24-76) vs 55 % (22-76), $p< 0.05$). The body composition showed an increase in FFM, fat free mass index (FFMI), fat mass (FM) and fat mass index (FMI). The increase of FFM and FFMI was significantly lower in patients with BMI <18.5 kg/m2.

Conclusion

FFM and FM both significantly increased after a short rehabilitation program consisting of adjusted nutritional care and physical exercise in adult CF patients. There was a greater increase of FM in malnourished patients and of FFM in well-nourished patients. These results seem to be in line with recent studies that suggest more focus on high-quality protein intake and less on high fat intake to increase FFM. We conclude that CF patients could benefit from dietary guidelines adjusted to their individual nutritional status and physical activities. More research is needed to build on these results.

OP9.

Implementation of guidelines on diagnosis and treatment of Eosinophilic Esophagitis by Pediatric and Adult GIs in Europe; On our way towards unison?

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Background

Guidelines for diagnosis and treatment of Eosinophilic Esophagitis (EoE) have changed markedly over the last decade. An international survey by the ESPGHAN EGID Group aimed to analyze current practice and management of Pediatric (PG) and Adult Gastroenterologists (AG).

Methods

Gastroenterologists in 13 European countries were asked to complete a multiple-choice questionnaire gauging physician demographics, EoE diagnosis and management strategies.

Results

Of the 1232 cooperating gastroenterologists, 465 were PG and 697 AG. In contrast to current guidelines on EoE diagnosis, only 41% of gastroenterologists (22% AG vs. 68% PG, $p<0.01$) reported taking biopsies in patients with suggestive symptoms without macroscopic endoscopic abnormalities; 92% (97% PG vs. 88% AG, $p< 0.01$) took biopsies when the symptom was dysphagia. 81% (86% PG vs. 77% AG, $p< 0.01$) sampled multiple esophageal sites when suspecting EoE. High dose PPI administration (68% PG vs. 72% AG), followed by elimination diets (32% and 27% respectively) were the most common first line treatments. In case of failure of initial PPI treatment, the majority opted for topical steroids (56% PG vs. 87% AG, $p<0.01$), however PG utilized food elimination diets as a second line treatment significantly more than AG (44% PG vs. 13% AG, $p< 0.01$). Although proven unreliable, 24% of prescribed food elimination diets were reported to be based on allergy testing (33% PG vs. 16% AG, $p< 0.01$) and up to 83% referred their patients for allergic assessment.

After initiating therapy, the majority monitored therapeutic response endoscopically (86% PG vs. 70% AG, $p< 0.01$). German PG universally reported endoscopic follow-up while Dutch gastroenterologists were least likely to follow this approach.

A greater proportion of PG than AG indicated reading at least one recent international guideline (89% PG vs. 56% AG), but both PG and AG recognize the benefit of national guidelines concerning the diagnosis and treatment of EoE (86% PG vs. 85% AG).

Conclusion

The general practice of pediatric and adult gastroenterologists in Europe differs from international guidelines on diagnosis as well as treatment of EoE. Geographic practice variations are apparent. Although the majority indicated awareness of recent practice standards, strategies to improve the implementation of current guidelines are necessary.

OP10.

Pancreatic blunt trauma in children: observations from a monocentric pilot study

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Background

Pancreatic trauma is the 4th most frequent cause of abdominal trauma in children. Few studies focused on the impact of trauma severity and therapeutic management (surgery, endoscopy or observation) on mid-and long-term outcomes.

The aim was to determine the mid and long-term morbidities related to the grade and the initial management of pancreatic trauma in children.

Methods

The charts of 29 children aged 0-<18y admitted at The Cliniques Universitaires St Luc between 01/2007 and 01/2017 for an abdominal trauma involving the pancreas were retrospectively reviewed. Data about trauma characteristics, clinical symptoms, imaging, therapeutic management and short to long-term complications were recorded. Results: We identified 29 patients aged 2-17(median: 6.9y). Most (18/29,62%) had a mild pancreatic trauma (AAST grade I-II), while 11 had severe (grade III-IV) pancreatic lesions. In 22/29(76%) patients, pancreas trauma was associated to another organ lesion, especially liver injury (17/22,77%). Clinical symptoms are nonspecific to diagnose pancreatic involvement: pain (27/29,93%) and nausea/vomiting (15/29,52%). Imaging by CT-scan suspected ductal involvement in 90% of grade III-IV. Five children were managed by endoscopy (3/5 grade III, 2/4 grade IV), 3 (3/3 grade IV) had surgical management (2 by pancreatico-jejunostomy, 1 for percutaneous drainage) while the other were managed conservatively. Hospital stay was significantly shorter in grade I compared to the severe grades (13vs19.5 days, p=0.025).Mediate complications consisted of pseudocysts; these were exclusively seen in grade III (5/5,100%) and IV (4/5,80%). Endocrine and exocrine pancreatic function was followed (median follow-up: 20.3 months) in 9/11 and 7/11 patients respectively with grade III-IV. Endocrine function was preserved in all of them. Instead, exocrine function was impaired in 3/7 (43%); all complained of intermittent symptoms of abdominal pain and steatorrhea.

Conclusion

This monocentric study showed that compared to grade I-II, the grade III-IV had a higher risk of pseudocysts, required longer hospitalisation and more frequent rehospitalisation for pancreatic reasons and were more likely to evolve to exocrine pancreatic dysfunction. Follow-up of grade III-IV-V is important to detect mid and long-term complications. Extending the study to a multicentric study will enable us to analyse the effect of operative, endoscopy or observation management on the long-term outc

P50.

When hoofbeats are zebras

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Background

Constipation is a common problem of the paediatric population affecting ±30% of children. It usually starts in childhood with a peak prevalence during preschool years and resolves as children get older. In 95% there is no underlying organic cause for the constipation.

Methods

We present a case of a 14-month old boy referred to our centre with the suspicion of an intestinal obstruction. This previously healthy boy was seen 2months before by his physician for new onset complaints of constipation. Despite several dietary changes, followed by enema's and oral laxatives, the defecation difficulties persisted. When he presented at the hospital, it had been 6 days since his last stool. He was painful, less active and had anorexia. Clinically he was dehydrated, pale, somnolent, and irritable with an extended and tender abdomen. Imaging with X ray of the abdomen was suggestive for faecal impaction with dilatation of the colon. He was therefore admitted for rehydration and faecal desimpaction. Unfortunately, he started vomiting and showed a marked increase of his abdominal distention and general clinical deterioration. Rectal enema's were not productive. For those reasons he was transferred to our intensive care unit for treatment and further investigations.
At his arrival we saw a pale, sick boy with a distended tender abdomen with hyperperistalsis. At rectal examination we felt a bulging of the right posterior wall caused by a soft palpable mass compressing the ampulla.

Results

In agreement with the gastroenterologist, radiologist and surgeon, an emergency MRI was performed revealing a large mass at the right side of the rectum with multiple abdominal lymph nodes.
The results of a transrectal biopsy were compatible with a malignant rhabdoid tumor, a rare childhood tumor, most commonly seen in infants and toddlers.
He was transferred to the oncology ward for further staging and treatment. His tumour was aggressive and fast-growing with breakthrough the rectal wall and metastases in the lungs. After 2 months, a palliative treatment was started and he died 2 weeks later.

Conclusion

As physicians we are taught: "If you hear hoofbeats, it's probably a horse, not a zebra". We have to consider the most logical and prevalent diagnosis for a common problem or symptom like constipation which is functional constipation. But we cannot forget that zebras do exist and that in rare occasions constipation can be the first sign of a lethal condition

P51.

Switching from infliximab originator to a biosimilar is safe in paediatric patients with inflammatory bowel disease

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Background

Rising evidence reveals no differences in efficacy and safety between infliximab (IFX) originator and IFX biosimilar CT-P13 in inflammatory bowel diseases (IBD). However, most data are derived from adult patients and data on pharmacokinetics are limited. We evaluated long-term IFX trough levels (TL), immunogenicity and remission rates in children with IBD who switched from IFX originator to biosimilar CT-P13.

Methods

In this single-centre study, all children with Crohn's disease (CD) and ulcerative colitis (UC) receiving maintenance IFX therapy between July 2017 and January 2018 were included. The switch to CT-P13 was imposed by the hospital for all patients regardless of the indication as from January 2018. Demographics, disease activity indices, IFX TL and antibodies to IFX (using Ridascreen IFX Monitoring ELISA) were collected from 6 months before (baseline) till 6 months after switch. Clinical remission was defined as PUCAI/PCDAI <10 and biological remission as CRP ¤5 mg/L and ESR ¤20 mm/h. For paired comparison of data obtained at the different timepoints, a Wilcoxon signed-rank sum test and a McNemar test were used for continuous and dichotomous variables, respectively. All data are presented as median [interquartile range].

Results

A total of 47 children received maintenance therapy with IFX originator at our centre. Forty-two children (26 CD and 16 UC), were eligible for the study as 3 patients were transferred to the adult department and 2 patients stopped IFX just before the switch (due to loss of response or delayed infusion reaction). Included patients had a median duration on IFX originator of 13.5 [6.8-35.5] months prior to switch. No significant changes in IFX TL occurred after switch. The median baseline IFX TL was 5.7 [3.8-9.3] µg/mL vs 6.5 [3.9-8.6] µg/mL at month 6 after switch (p=0.90). The cumulative IFX dose administered over a 6 month period was not significantly different before switch (36.6 [24.0-53.3] mg/kg) compared to after switch (35.8 [26.7-55.6] mg/kg; p=0.21). Antibodies to IFX appeared in 1 patient after switch. The proportion of patients in clinical and/or biological remission did not significantly change after switch. No significant changes were observed in CRP, ESR, albumin or weight and BMI (expressed as z-score) after switch.

Conclusion

Paediatric IBD patients on IFX originator can be successfully switched during maintenance to CT-P13 without affecting efficacy, pharmacokinetics, immunogenicity and safety.

P52.

Case report of a one-year-old girl with repetitive vomiting and loss of consciousness after ingestion of pineapple

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Case report

We report the case of a one-year-old girl presenting at the emergency department with repetitive vomiting, lethargy and loss of consciousness. She was hospitalized. EEG and cardiac ultrasound were normal. A vasovagal syncope after vomiting was suspected. Five weeks later, she presented again with the same presentation of vomiting with temporary loss of consciousness and one bloody stool. Stool sample was negative for bacteria, Giardia lamblia, Cryptosporidium, rotavirus and adenovirus. Brain CT-scan, electrocardiography and abdominal ultrasound were normal. There was a spontaneous resolution of symptoms after administration of IV fluid. She was again hospitalised two months later with excessive vomiting after ingestion of pineapple. Symptoms improved after intra-venous rehydration and ondansetron. At this moment mother realised that vomiting always began after eating pineapple. Allergic testing (IgE, Rast) was negative. An oral provocation test with pineapple revealed the typical clinical signs of a food protein induced enterocolitis syndrome (FPIES): profuse vomiting, paleness and lethargy with recovery after fluid resuscitation.

Discussion

FPIES is a non-IgE-mediated reaction to food, characterized by repetitive vomiting (1 to 4 hours after ingestion), diarrhoea, sometimes accompanied by lethargy and hypotensive shock. The first presentation is often in infancy after introduction of the trigger food, usually rice, cow milk or soy. A recent Australian study determined the annual incidence of FPIES at 15.4 per every 100000 children younger than 2 years.

The pathophysiology of FPIES remains unclear. Intestinal inflammation leading to increased permeability is believed to play a role. The diagnosis is clinical but can be difficult for several reasons: the lack of typical allergic signs like cutaneous and respiratory allergic symptoms. Symptoms often mimic sepsis. An oral food challenge (OFC) is the gold standard for the diagnosis. Resolution of the disease generally occurs in early childhood. Treatment is supportive and fluid resuscitation together with elimination of the trigger food from the diet is the cornerstone.

Conclusion

FPIES is a non-IgE-mediated food allergy. It is not a rarity, but diagnosis is challenging and misdiagnosis is common. Recognition is important since it can potentially present with severe hypotension requiring immediate fluid resuscitation and long term follow-up.

P53.

Intussuception due to heterotopic pancreas in the intestinal wall: a case report

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Clinical Case

A 7 month-old boy has presented bloody stool and loss of appetite for 24h, without fever, crying nor vomiting. Clinical examination is normal. Ultrasonography reveals a large ileo-colic intussuception without alteration of Doppler signal and without obvious lead point. Two successive nonoperative reductions using hydrostatic (contrast) enema are performed, but fails to reduce intussuception completely . The baby is admitted to the operating room for surgical reduction. During the procedure, a lead point is identified, suspected to be a Meckel diverticulum based on macroscopic aspect, but histological examination reveals finally ectopic pancreatic tissue.

Discussion

Intussuception is the most common abdominal emergency in early childhood (6 months to 5 years). The classical clinical triad « pain, palpable sausage-shaped abdominal mass, currant-jelly stool » is seen in less than 15% of patients, and clinical diagnosis remains challenging. Ultrasonography is the best method to detect intussusception, showing the classical « target sign », usually localized in the right lower quadrant for ileo-colic invagination (acouting for 90% of all cases).

Nonoperative reduction (using hydrostatic or pneumatic enema) is the treatment of choice in a stable child with ileo-colic intussuception witout any sign of perforation, with a success rate of 70 to 85 % and a low risk if complication (perforation rate <0.5%). Surgical intervention is indicated if nonoperative reduction fails to reduce completely the intussuception, or in case of perforation, perotinitis or unstable patient.

Intussuception in children are mostly idiopathic, but a lead point (lesion in the intestine wall trapped by peristaltis and dragged into a distal segment) can be found in 25% of cases. The most frequent causes are Meckel diverticulum, polyp, tumor, duplication cyst, hematoma or vascular malformation.

A pancreatic rest (ectopic pancreas/heterotopic pancreas) in the intesinal wall can also cause invagination, as it was the case in our patient. Heterotopic pancreas can be found in the gut submucosa, from distal stomach to jejunum, within a Meckel's diverticulum, or in the gall bladder. Most of cases are incidental findings (during endoscopy or surgery, or occasionally on CT-Scan), but it can also cause abdominal pain, gastrointestinal bleeding, intestinal obstruction or pancreatitis. Rare cases of malignant transformation were described.

P54.

Triple A syndrome revealed by achalasia : case report

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Background

The triple A or Algroove syndrome is a rare autosomal recessive disorder characterised by the triad of alacrima, achalasia and adrenocorticotropic hormone (ACTH) resistant adrenal insufficiency. Neurological abnormalities are frequently associated. It is caused by mutations in the AAAS gene located on chromosome 12q13 which encodes the nuclear pore protein ALADIN (Alacrima-Achalasia- Adrenal Insufficiency and Neurologic disorder). The clinical presentation is variable and the triad may be incomplete at the diagnosis.

Clinical presentation

A four-year-old girl presented dysphagia, vomiting and failure to thrive. She was initially treated by proton pump inhibitors for gastroesophageal reflux disease without improvement. Abdominal sonography, barium swallow study, esophago-gastroendoscopy were performed and finally esophageal manometry confirmed the diagnosis of type 2 esophageal achalasia. Ophtamological testing showed a keratitis due to alacrima which wasn't initially reported by the parents. Albeit the absence of adrenal insufficiency, the clinical diagnosis of triple A syndrome was confirmed by a positive mutation analysis of the ALADIN gene. Dysphagia was successfully treated by Peroral endoscopic myotomy (POEM), a new minimally invasive intervention for achalasia and artificial tears were used for alacrima.

Conclusion

We report a rare case of incomplete triple A syndrome revealed by dysphagia. Paediatricians should keep in mind this diagnosis characterised by a heterogeneity of clinical presentations. Suspicion of triple A syndrome should lead to molecular analysis even if incomplete. We reveal a good short term outcome of Peroral endoscopic myotomy (POEM) for treatment of achalasia.

P55.

No two pediatric intestinal polyps are alike

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Background/Aims

In the pediatric age category, gastrointestinal polyps can manifest in a very different way of which painless red rectal bleeding is the commonest. Most polyps are sporadic, isolated and benign. However, it is important to correctly identify rare inherited polyposis syndromes because of the increased risk of intestinal and extra-intestinal malignancies.

Methods

Between 2016 and 2018, 4 completely different intestinal polyp types were diagnosed by endoscopy. They had different clinical manifestations as well as histopathology. A literature search was made to recapitulate risks for malignancy, probability of underlying genetic disorders and importance of endoscopic or surgical therapeutic consequences such as endoscopic surveillance or preventive colectomy.

Results

The 4 patients included had respectively Peutz-Jeghers syndrome, juvenile polyposis syndrome, familial adenomatous polyposis and Li Fraumeni syndrome. Each syndrome has a different lifetime risk of (extra)-intestinal malignancy and requires a different approach and follow-up. Histopathology and genetic testing play an important role in identifying these syndromes.

Conclusion

Rare inherited polyposis syndromes should be considered and correctly diagnosed in a child who presents with gastrointestinal polyps because of the associated increased cancer risk.

P56.

Bilious vomiting: looking for the horse and finding the zebra

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Background

Bilious vomiting in the pediatric population is a red flag symptom and requires immediate diagnostic work-up and therapeutic approach. The most common gastro-intestinal cause in infants is bowel obstruction due to malrotation with volvulus or intussusception. Here we present a rather unusual cause of bilious vomiting.

Methods

A 2-year-old boy was transferred to the pediatric department because of prolonged fever and bilious vomiting. On admission, he had a bilateral, non-exudative, conjuctivitis and red, fissured lips. Clinical examination revealed a cervical lymphadenopathy and an erythema of the palms and soles with a mild periungal desquamation. Blood tests reported white blood cells 15 850/µL, thrombocytosis 517 000/µL and C-reactive protein 104mg/dL. The main infectious causes were ruled out (EBV, CMV, coxsackie). Abdominal ultrasonography showed circumferrential bowel wall thickening of the horizontal (D3) portion of the duodenum and a jejunal loop. Diagnosis of Kawasaki disease with intestinal involvement was made based on the clinical presentation. Echocardiogram was normal.

Results

Treatment was started with intraveneus immunoglobulins (IVIG) and high-dose aspirin (100mg/kg/day), which was decreased to 5mg/kg/day 24 hours after disappearance of the fever. Abdominal complaints promptly disappeared after initiation of therapy. The boy was discharged and given aspirin for 6 more weeks. An echocardiogram performed 6 weeks after disaese showed no cardiac alternations.

Conclusion

Intestinal involvement in Kawasaki disease is uncommon and may delay the diagnosis and treatment, which is a risk factor for development of cardiac complications. Therefore, pediatricians should consider Kawasaki disease among other diagnoses in children with prolonged fever in association with abdominal symptoms and radiologic findings which can be compatible with pseudo-obstruction.

The most suggested underlying mechanism is a vasculitis and thrombosis of small mucosal arteries involving the intestine, causing vasuclar insufficiency. Management is conservative with high-dose IVIG and aspirin.

P57.

Unusual cause of gastro-esophageal reflux in a 5-month-old girl

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Background

Brief resolved unexplained event (BRUE), previously called apparent life-threatening event (ALTE) is a frequent cause of emergency admission. Gastro-oesophageal reflux disease is one of the possible diagnosis.

Clinical case

We are reporting the case of a 5-month old girl, with an irrelevant past medical history. She presented symptoms of transient loss of consciousness and pallor followed by breathing difficulties. The clinical examination on admission was normal. Since a gastro-oesophageal reflux disease (GORD) was suspected, a 24-hour pH monitoring was thus considered. The chest X-ray showed a deviation of the probe, the trachea and oesophagus on the right, associated with a mediastinal mass. A barium swallow examination confirmed a deviation of the structures, as well as an extrinsic imprint on the oesophagus. The primary hypotheses are oriented towards aortic or bronchial malformations. The performed cardiac ultrasound was normal. A contrast enhanced thoracic CT showed a well-rounded formation of liquid nature. The cyst's proximity to the oesophagus pointed towards a digestive malformation rather than a bronchial cyst. During the surgical exploration by thoracoscopy, the cyst was completely resected from the lower part of the oesophagus and there were no post-operative complications. The anatomopathological analysis confirmed the intramural nature of the duplication cyst. At 8-month follow-up, the infant has normal growth and development and is asymptomatic.

Discussion

Our patient presented a BRUE due to a gastro-oesophageal reflux disease, secondarily to an oesophageal duplication.

Symptoms described in the literature are due to compression of adjacent structures leading to breathing difficulties, disorders of oesophageal motility, retrosternal pain or cardiac arrhythmias.

Duplications of the digestive tract are rare congenital malformations (0.2 % of all children), which 15 to 20% are located in the oesophagus and can be cystic (95% of cases) or tubular. They consist of individualized duplications of the submucosal and muscular layers of the oesophagus. Approximately 80% of oesophageal cysts are diagnosed before the age of 2 years.

Conclusion

Suspicion of gastro-oesophageal reflux disease in BRUE should be followed by a complete assessment to confirm the diagnosis and exclude a secondary cause of GORD. Our case illustrates a rare cause: a cystic oesophageal duplication.

P58.

two cases of bilious vomiting with duodenal web

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Bilious vomiting may be the first sign of intestinal obstruction, so special attention must be paid to it. Indeed, the differential diagnosis is vast and the etiology is not always easy to find.

Patient 1: 6-year-old child with trisomy 21 who presents chronic vomiting. The abdominal ultrasound and X-ray did not demonstrate any specific abnormalities. In the context of persistent vomiting despite a proton pump inhibitor treatment for several months, the esophageal gastroduodenoscopy performed revealed a duodenal obstruction at the second portion of the duodenum and duodenal web has become the most likely hypothesis.
The delayed diagnosis was marked by the insidious symptomatology.
Patient 2: 10 days newborn who presented in the emergency department for postprandial vomiting. The atypical appearance of the radiological examinations and the absence of associated malformation made the diagnosis complex.
The laparoscopic assesment showed a duodenal web
The incidence of duodenal occlusions is 1/5000 to 1/10 000 births with no predominance of sex. The obstruction is most often at the 2nd part of the duodenum.
The differential diagnosis of duodenal occlusions can be divided into two broad categories: extrinsic causes (annular pancreas, common mesentery or malrotation volvulus, Ladd flange, pre duodenal portal vein,..) and intrinsic causes (atresia, stenosis, complete or incomplete duodenal web).
The abdominal radiography and ultrasonography are the exams of choice to exclude the urgent cases of occlusion. The development is mainly done using a baryte abdominal x-ray to highlight a "double bubble" image.
The duodenal web is a rare intrinsic obstruction of the duodenum. There is usually the persistence of a small aperture at the center that differentiating this from a duodenal atresia. In contrast to complete duodenal atresia that is diagnosed early in life, a duodenal web can be diagnosed later depending on the size of the orifice.
In 50% of patients, duodenal atresia is associated with another congenital anomaly, especially Down syndrome.
The treatment is usual surgical (laparoscopic or open repair). For selected cases the endoscopic way may be a possibility.

O4.

Physical determinants of weight loss during a residential rehabilitation program for obese adolescents

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Background and objectives

Comprehensive obesity management programs that emphasize on appropriate nutrition, exercise and behavioral modification generally yield positive results, but with a large variability in outcome. This retrospective cohort study aims to evaluate a possible correlation between physical determinants at baseline and weight loss at completion of a 1 year multidisciplinary residential adolescent rehabilitation program.

Methods

Patient records for 27 boys and 37 girls were selected from available data of adolescents aged 14-18 years that participated in and finished the 1-year weight loss program. Body mass index, body composition and cardiorespiratory fitness (measured as VO2peak in ml/min per kg lean body mass) at baseline and at completion of the 1-year long program were used to investigate a possible correlation with age-, sex- and height-corrected weight loss' calculated as change in BMI standard deviation score (BMIz).

Results

We report a significant mean decrease in BMIz of 1,20±0,5 SD (p<0,001) for boys and 0,87±0,4 SD for girls (p<0,001) after completion of the program, with a similar positive effect on body fat percentage (boys -52,8%; girls -31,5%). Both baseline BMIz (boys R=0,459 ; girls R= 0,432) and baseline VO2peak (boys R=-0,418; girls R=-0,579) are significantly correlated with the change in BMIz. When patients are divided into two groups according to baseline BMIz above or below 3SD, the change in BMIz is smaller for those with BMIz > 3SD (boys: 0,29SD with p=0,116 ; girls: 0,39SD with p=0,001). Those patients starting the program with a BMI >3SD are unlikely (odds ratio 33 and 31,7 for boys and girls respectively) to lose sufficient weight to reach 95th percentile (1,65SD) within a year. When patients are divided into two groups according to physical fitness at baseline, the change in BMIz is significantly smaller for those with below average fitness (boys: 0,42SD with p=0,021 ; girls: 0,36SD with p<0,001).

Conclusion

Less obese and fitter adolescents lose more weight than their heavier and more deconditioned peers during a one-year residential obesity treatment program, advocating early intervention in treating adolescent obesity.

OP28.

Assessment of pedestrian safety around Flemish Schools using Google Street View

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Background/Aims

The improvement of pedestrian safety in the school environment necessitates an accurate catalogue of traffic features in all schools or in a relevant sample. Yet performing this type of survey around schools all over the country is a labor intensive, time consuming and costly affair. In this study we investigated the feasibility of a virtual survey of pedestrian safety in a large sample of Flemish schools.

Methods

We selected 419 (15% of total) basic or secondary schools in Flanders at random in the five provinces. A systematic data collection was performed with assessment of all relevant traffic features (speed zone, presence of pedestrian crossing, modifications to sidewalks). Subsequently all schools were invited to perform a direct visual assessment of the school environment to compare with the virtual survey.

Results

GSV enables a clear identification of pedestrian safety. 71% of school have a painted pedestrian crossing, only 18% have a traffic platform and only 5,9% have traffic lights. 23% of schools have no specific security to cross the street at the school gate. 54% of schools are in an environment with a fixed 30km/h speed limit, 39% have variable 30km/h limit, 7% could not be clearly assessed. There was a good correlation between the virtual and direct observations. The school authorities rated their school environment as very safe (14%), safe (64%) or not safe (22%).

Conclusion

GSV permits researchers to obtain cheap but reliable virtual surveys of demographic characteristics such as traffic situations around schools. These surveys can guide future improvements in pedestrian safety by identifying and prioritizing unsafe school environments.

OP29.

Assessment of cycling helmet use with Google Street View: an international survey

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Background/Aims

The use of a cycling helmet helps to prevent cranial trauma from high impact force during traffic collisions. Differences in legislation lead to differences in cycling helmet use, which government assess using costly surveys. Google Street View (GSV) is a software that enables the virtual assessment of several demographic characteristics that are present in inhabited areas. In this study we investigated the feasibility of a virtual survey to quantify helmet use in several countries.

Methods

We selected three cities in countries with either no legislation (I), obligatory helmet use for children (II) or obligatory helmet use for everyone (III). We subsequently quantified helmet use with GSV for adults and children and compared this to direct surveys and published online surveys.

Results

GSV enables a clear identification of helmet use, age and gender in cyclist in cities on different continents. The interrater variability was very low with a percentage positive agreement of 99,15% with a Ä¸=0.98. Helmet use was highest in the group III (87%), lower in group II (44%) and the lowest in group I (40%). There was no difference in helmet use based on gender, because of the low number of quantified children no statistical analysis in this age group was possible. The quantification with GSV was as accurate as direct observations or recently published online surveys.

Conclusion

GSV permits researchers to obtain cheap but reliable virtual surveys of demographic characteristics such as cycling helmet use. Future research should focus on the use of artificial intelligent pattern recognition software for automated surveys and analysis.

OP30.

Feasibility and effectiveness of Non-neonatal intubation training for pediatricians in Flanders

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Background

The leading cause of preventable death in pediatrics is failure to manage the airway. Children are vulnerable to respiratory failure; their anatomy and physiology makes safeguarding the airway a challenge. Since it is a trend to concentrate specialized care, non-neonatal intubation (NNI) will mostly be performed by anesthetists or paediatric intensivists. Interns spend fewer hours in-hospital, and children in need of intubation decrease due to increasing vaccination rates and non-invasive ventilation support. NNI should be acquired by pediatricians according to the European Academy of Pediatrics (EAP). We argue whether this is still a realistic or desirable goal.

Methods

We developed a survey including items about the importance of NNI in pediatric training and the competency and self-confidence of the participant, and also collected demographic data including information on pediatric life support (PLS) courses. The survey was sent to all Flemish pediatricians (in training), neonatologist and pediatric intensivists were excluded. Primary outcome was the perceived importance to achieve competence in NNI, preparedness and self-confidence rated on a Likert scale. Associations between demographic characteristics and perceived importance or competence were assessed.

Results

The survey was sent to 806 participants. At the moment 233 answers (30% residents) were analyzed. The majority (92%) followed a PLS course. Forty % (strongly) disagreed on whether NNI should be a skill a pediatrician should obtain. A significant difference (p=0,007) was present when comparing pediatricians (51%) and residents (30%). When asked to score self-confidence, a mean of 3.4 (+/- 2.7) was scored on a 0-10 scale. Almost 70% (strongly) disagreed when asked about feeling confident in NNI skills. Only 16% didn't feel confident in managing a child's airway. Of all participants, 75% did not intubate a child in the last 5 years.

Conclusion

Our questionnaire showed low self-confidence in and very low exposure to NNI. Although only preliminary, it seems nowadays many residents will not be able to develop competence in NNI, neither will a pediatrician be able to maintain this skill. Refining of the list of skills required by the EAP could lead to a shift from being able to intubate, to being able to manage the airway of a child. If intubation is still perceived necessary, the best way to teach it should be examined: relying on clinical exposure is clearly not enough anymore.

OP31.

Screening for risk factors for developing chronic pain in a tertiary pediatric population

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Background/Aims

Chronic pain is a common pathology in children and adolescents. This pain has a huge impact on daily functioning of the patient and his family and has also important implications on long term. After reviewing the literature the most important conclusion is that there is little known about the prevalence of chronic pain in children and about the possible risk factors for developing this chronic pain. With this study we tried to know more about it. We explored also if two standardized questionnaires were useful to investigate this population.

Methods

By standardized questionnaires we collected data about the epidemiological situations of the children, the severity (aGCPS – adapted Graded Chronic Pain Scale) and the impact (PPST – Pediatric Pain Screening Tool) of the pain and we screened our population for possible risk factors for developing chronic pain.

Results

493 questionnaires were registered, 53% of these were completed both by the parents and the children. More than half of the children (54%) had experienced pain the past 6 months. No differences between the reports of the parents and the reports of the children were identified. The higher the pain severity was, the higher the impact score was. Fore the group of children who reported pain, we identified significant more 'other family members with pain' and more 'previous traumatic experiences or stressful events'. Girls and younger children reported more pain.

Conclusion

The prevalence of chronic pain in our tertiary pediatric population is about 20%, this is similar to what we found in literature. Both the aGCPS (severity) and the PPST (impact) seem useful in this tertiary population. The most important and most significant risk factor for the development and the persistence of chronic pain is the presence of other family members with chronic pain. Also this is something we found in literature. We conclude chronic pain is an important pathology in children with a huge impact on daily life and the general development of these children. Because the prognosis for these children is very will if there are treated well, we need multidisciplinaire centres. Further research into chronic pain in children is needed.

P59.

Fever & Arthritis: when family history matters

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We present a case of a 4-years-old girl, born Lebanon but living in Belgium since 2016 with her family. She was referred by a general practitioner for a high fever and right knee pain with difficulties to walk but no local inflammatory sign. Laboratory results revealed normal hemogram except slight anemia but a C-reactive protein level of 190mg/L. The ultrasound of both knees was normal, but the ultrasound of the hips revealed a small (6mm; not punctionable) bilateral fluid collection. An MRI of the hips confirmed right hip arthritis. At that point, rheumatoid factor, human leucocyte antigen B27 (HLA-B27) and anti-cyclic citrullinated peptide (anti-CCP) antibodies were all tested negative. After a thorough familial and personal anamnesis, despite language barrier causing important delay, the parents reported that their daughter has fever almost twice a month recurrently and has already suffered from 2 arthritis episodes in Lebanon previously, always in the hips or the knee. Consanguinity was reported in her parents and a history of Familial Mediterranean Fever (FMF) was detected: Paternal grandfather, maternal uncle and aunt were all treated by colchicine. Gene paneling was then performed and mutations in the MEFV gene were found. She has then received a colchicine treatment and does not have any joint swelling or fever anymore since then. A familial genetic counseling was proposed and follow-up was organized. Familial Mediterranean Fever (FMF) is an autosomal recessive hereditary disease caused by pathogenic mutations in the MEditerranean FeVer (MEFV) gene, especially affecting people living around the Mediterranean Sea. Those mutations increase chemotactic activity in serosal tissues causing massive influx of granulocytes during attacks. FMF is characterized by recurrent fever, abdominal pain, pleuritis, synovitis and erysipelas-like erythema. Typically, temperature flares above 38°C for 12 to 72 hours every month. Major long-term complication is secondary amyloidosis, which can lead to terminal renal failure. The attacks and complications can be avoided by lifelong administration of colchicine. FMF is sometimes a tricky diagnosis since family history is sometimes difficult to collect as well as reliable anamnesis regarding the timing of fever or pain. The differential diagnosis of recurrent fever is also very broad including all periodic fever syndromes, systemic vasculitis and rheumatic diseases, infections such as relapsing fever and malignancies.

P60.

The analgesic effect of Virtual Reality in pediatric procedural pain: a systematic review

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Background/Aims

Procedural pain is an important source of fear and distress for children. Distraction is a widely used non-pharmacological approach to manage pediatric pain. A new distraction method is Virtual Reality (VR) technology; it combines multiple senses to provide a feeling of presence into a virtual world. Most reviews so far assessed the effect of VR distraction in adults. This systematic review of randomized controlled trials aims to evaluate the analgesic effect of VR distraction in procedural pain in children.

Methods

A systematic search was conducted using MEDLINE (through PubMed), Embase, CENTRAL and Web of Science from the earliest date until October 2018. By determining a set of inclusion and exclusion criteria, 17 trials were retrieved and qualitatively analyzed using the Cochrane risk of bias tool. Selected studies were grouped by type of procedure.

Results

Children distracted by VR during painful procedures had overall less pain when compared to standard of care. The analgesic effect is better using active VR distraction than passive VR distraction. Mainly for minor procedures (wound care and venipuncture), VR technology seems to be an appropriate technique to redirect children's attention away from the painful stimulus. This was not the case for port catheter access (with EMLA) and lumbar puncture (compared to EMLA topically and sedation).

Conclusion

This study provides support for further implementation of VR technology in daily medical practice on the children's ward.

P61.

Growth patterns and body composition in former extremely low birth weight (ELBW) infants until adulthood: a systematic review

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Background/aims

Preterm infants are lighter and shorter, with smaller head circumferences than normal weight term born peers at birth. Infants born (very) preterm also have a different body composition. Compromised growth can lead to adverse health outcomes. However, reviews regarding growth and body composition in preterm infants throughout childhood and adolescence are rare and mostly do not differentiate between birth weight or age. The purpose of this systematic review is to assemble growth data of extremely low birth weight (ELBW) children.

Methods

PubMed, Cochrane Library, Embase and Web of Science were searched for studies regarding growth and body composition of former ELBW infants until adulthood. We compared their height, weight, head circumference (HC), body mass index (BMI), fat mass (FM), lean mass (LM), fat distribution and body water (BW) with matched normal birth weight (NBW) controls and the World Health Organization (WHO) growth charts.

Results

We included 16 articles and one abstract. Studies consistently reported that ELBW neonates remain shorter and lighter, with smaller head circumferences than NBW children, at each corrected age (CA). It is suggested that ELBW children and adults have a higher percentage total body fat (%TBF), although different results about body composition have been described. ELBW infants seem to have a growth pattern with a height, weight and head circumference around the 25th percentile of the WHO growth charts. Few studies found growth parameters comparable with the 50th percentile. At the age of 8 years and afterwards, their BMI seems to reach the 50th percentile with some values approximating the 75th percentile.

Conclusion

ELBW infants and adolescents are more likely to have lower growth parameters at every stage of their development. Although they exhibit a period catch-up growth, their growth remains retarded later in life. There were only five studies discussing body composition reporting divergent results. Further research and longitudinal studies are needed to investigate body composition and the correlated risk on cardiovascular diseases or metabolic syndrome.

P62.

A hernia with a twist: the case of a strangulated ovary hernia

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Case

A one-month-old girl is brought to the emergency room for a sudden swelling located in the left inguinal region. The mother described the infant as uncomfortable but in no severe pain. The clinical examination confirms the presence of a non-mobile tumefaction above the left labia major, with a slight induration to the touch, without any signs of a local inflammation. The rest of the examination is normal other than an uncomfortable infant that calms easily in the mother's arms.

An inguinal hernia was suspected and confirmed by an ultrasound with color Doppler. The imaging showed a bilateral ovary hernia via the Nück canal. The right hernia was partial, however, the left ovary hernia is total with signs of strangulation.

The infant was operated on within 3 hours. The left ovary showing signs of adequate revascularisation was left in place.

The infant spent 24hours in the intensive care unit for apnoea following anaesthesia.

Discussion

Inguinal hernia is a frequent pathology in paediatric medicine occurring in 1 to 5% in all newborn and up to 11% in premature infants. The incidence is 4 times higher in boy with the right side being more often affected than the left side in both genders.

Although an inguinal hernia is less frequent in females, an incarcerated or strangulated hernia is more likely to contain reproductive organs (ovary, fallopian tube).

The reduction of an inguinal hernia needs be done gently. In any case in which the gentle reduction cannot be done, an ultrasound is necessary to prove the absence of reproductive organs in the hernia. Color Doppler is useful to determine the vascularisation of the herniated tissue or showing signs of ischemia. A prompt consultation with a surgeon is advised for any non-reducible, symptomatic hernia in a female infant.

Surgery is the only effective treatment and should be undertaken more or less rapidly based on the clinical and imaging signs. Any sign of complication grants an expeditious management to preserve the affected tissue.

Conclusion

Inguinal hernia is more commonly found in male patients. It is to be noted that reproductive organs are more often found in the herniated tissue in females.

Any hernia should call for a gentle reduction. If reduction is not obtained, imaging by ultrasound, with colour Doppler shows any signs of complication, including tissue ischemia.

Treatment is surgery that can be needed urgently if there are signs of complications or postponed if manual reduction can be done.

P63.

Pulmonary arteriovenous malformations and a neonatal presentation of a Rendu-Osler disease

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Introduction

Pulmonary arteriovenous malformations (PAVMs) are vascular structures, that directly connect a pulmonary artery to a pulmonary vein, bypassing the capillary bed. These malformations are uncommon during childhood and are often associated with Rendu-Osler disease (ROD) or hereditary hemorrhagic telangiectasia (HHT).

Case report

Our patient was born prematurely at 36 weeks. The immediate neonatal adaptation was good, but a respiratory distress appeared at 10 minutes of life. A X-ray of the chest showed a right para-cardiac mass. An injected chest CT scan shows an arteriovenous malformation in the middle lobe. Oxygen therapy was stopped after 3 days. A mutation of the Endoglin gene confirmed the diagnosis of ROD. At the age of one year, the PAVM afferent artery was embolized with coils under angiographic guidance. Today our patient is one year and 9 months old and is totally asymptomatic.

Discussion

ROD is an autosomal dominant genetic disorder, characterized by spontaneous and recurrent epistaxis, cutaneomucous telangiectasia, pulmonary, cerebral and hepatic arteriovenous malformations. The first symptoms rarely occur before the age of 10. Except for a family screening, this disease is poorly diagnosed during pediatric age. Rendu-Osler type 1 disease is associated with the mutation of the endoglin gene; Rendu-Osler type 2 disease is associated with the mutation of the ALK-1 gene. PAVMs sometimes occur from birth. They are often associated with ROD. PAVMs are very often asymptomatic, but their complications can sometimes be fatal. The rupture of their wall can cause hemoptysis or hemothorax. The absence of a capillary filter can be a source of paradoxical embolism, responsible for strokes or cerebral abscesses. Finally, these PAVMs constitute a right / left shunt, responsible for chronic hypoxemia and cardiac failure. Thoracic CT and contrast echocardiography are the gold standard for diagnosis of PAVM. PAVM treatment is endovascular embolization, when the afferent vessel is larger than 3 mm of diameter, or symptomatic.

Conclusion

PAVM is a rare diagnosis at birth. It should prompt the detection of ROD. An adequate treatment and a regular follow-up can improve the prognosis and quality of life of these patients.

Keywords

Pulmonary arteriovenous malformations ‘Rendu-Osler disease’ hereditary hemorrhagic telangiectasia.

P64.

An atypical infantile eruption: let's have a look at Mum!

O Paduart, F Vermeulen. Hopital Erasme

A 6 weeks-old boy was brought to our department by his Mum with an annular rash exhibited only on the head that started 3 weeks ago with no other medical problems.
The pregnancy was normal and the child was born full term by a Congolese 17 years-old mother, herself without previous medical history. His growth and vitals were normal. His clinical exam was normal with the exception of the erythematous annular lesions all over the head including scalp and face, where they are observed mainly in the periorbital area. The main differential diagnosis of that rash was seborrheic dermatitis, fungal skin infection, neonatal lupus herpes simplex infection and erythema multiforme.
The bloodtest revealed a normal hemogram, no inflammatory syndrome and a moderate cytolysis (AST 163UI/l, ALT 107UI/l, gamma-GT 277 UI/l). Serologic tests were negative for HIV, CMV, mumps, rubella and parvovirus. Antinuclear antibodies (ANA) were positive, with 1/1280 and with anti-SSA/Ro and anti-SSB/La antibodies strongly positive. For the mothers' serum, ANA 1/1280 was measured, however not typable at that point. The sedimentation rate was 120mm/hour, although clinically asymptomatic.
Neonatal lupus is a pathology that can be defined by the presence of anti-SSA/Ro, anti-SSB/La or anti RNP antibodies, in the mother or in the child, with symptoms of typical rash, heart block or hepatic or hematologic manifestations. These symptoms are often the results of a passive transfer of autoantibodies from the mother to the foetus, resulting in foetal or neonatal disease.
We present here a case of neonatal disease diagnosed at 6 weeks of age, usually the mean-age at which a rash can be recognised, with hepatic manifestations but no cardiac or hematologic problems. The child was treated for his rash with topic corticosteroids. Cardiac monitoring was conducted, and it revealed normal results. The cytolysis was followed closely and slowly came back to normal. Both cutaneous and hepatic manifestations are known to be reversible and resolve after maximum six to eight months. Unlike the congenital heart blocks that are rare but really well known by gynaecologists and paediatricians, the diagnosis of neonatal lupus on a healthy child with a rash might be challenging. Multidisciplinary approach is important in neonatal lupus to detect different manifestations, avoid unnecessary biopsy or topic treatment and often the diagnosis of the mother can be done too and she can then be treated quickly.

P65.

A case of spontaneous splenic infarction in a patient with heterotaxy syndrome and polysplenia

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Background

Splenic infarction is a rare cause of abdominal pain in children and symptoms are often very similar to those of appendicitis. Causes include hematologic disease or splenic torsion. For patients with visceral heterotaxy without major congenital heart defects, abdominal pain caused by splenic torsion and/or infarction may be the reason for first presentation. We present a rare case of a boy with spontaneous splenic infarcts associated with heterotaxy syndrome and polysplenia.

Case

A 10-year old boy presented to the emergency department with continuous right-sided abdominal pain, nausea and vomiting since the last four days. His medical history was unremarkable, except for four (self-resolving) episodes of severe abdominal pain in the past year. There was no fever and serum inflammatory markers were normal. An ultrasound was made to exclude appendicitis and revealed visceral situs inversus and a large, ill-defined mass in the right-sided abdomen. To further evaluate, a computed tomography (CT) angiogram was performed and showed right-sided polysplenia with signs of infarction in the two largest out of a total of five spleens along with a midline liver, several enlarged mesenteric lymph nodes and ascites. To detect associated defects, an echocardiogram was made which was suspect for left isomerism and showed an interrupted inferior vena cava with azygos continuation.
In this case, infarction was most likely caused by splenic torsion as a result of polysplenia. There were no clinical indications for and no family history of hematologic disease or thrombo-embolic disorders. The boy was treated with analgesics and after clinical recovery he was discharged from the hospital. Surgical intervention (splenectomy) will be considered if he continues to have significant abdominal pain.

Discussion

The abnormal arrangement of the abdominal organs in heterotaxy syndrome is known to be a risk factor for splenic torsion. Preservation of the spleen is the main goal and therefore, intervention is guided by clinical setting. However, additional investigations are necessary to evaluate heterotaxy associated congenital abnormalties. (i.e. heart defects and anomalies of the gastro-intestinal tract)

Conclusion

Splenic torsion and/or infarction should be considered in patients with heterotaxy syndrome presenting with abdominal pain. Symptoms may mimic appendicitis. Surgical intervention is not preferable unless severe clinical symptoms are present.

P66.

Aquatic activities, an unusual cause of anaphylactic shock

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Introduction

Cold-induced anaphylaxis is a feared complication of cold-induced urticaria, a life-threatening condition occurring after exposure of the skin to a cold stimulus.

Such rare form of physical urticaria presents disturbing symptoms, ranging from pruritis, urticaria to angioedema and ultimately anaphylaxis with haemodynamic collapse.

Case description

During the summer, a 13-year old boy is brought by his mother to the paediatric emergency room with generalised urticaria and dizziness after going to the pool. The boy suddenly lost consciousness at the entrance of the emergency room. The diagnosis of anaphylactic shock was quickly confirmed by the presence of generalised rash, urticaria with hypotension and loss of consciousness. We stabilised the patient after two rounds of epinephrine and volumising injections.

The boy presented the same sequence of events at the swimming pool in the last two years but he never had lost consciousness before.

The etiological assessment pointed out a chronic hepatitis B with insufficient follow-up and high viremia. The ice-cube test performed three weeks after the shock was negative.

Following to the thorough anamnesis and the current clinic, it appeared that the cause leading to the anaphylactic shock was most likely an atypical acquired cold-induced urticaria.

Discussion

Given the rareness of cold-induced urticaria (incidence of 0.05%) and the potential fatal issue of cold-induced anaphylaxis in aquatic activities, it is a case report worth mentioning.

Etiologies' studies have shown that cold-induced urticaria is generally acquired but can also be inherited.

Acquired cold-induced urticaria is most frequently idiopathic but secondary causes, such as autoimmune diseases or viral infections (hepatitis B in our case), are also known to trigger urticarial or anaphylactic cold-induced reactions.

A positive ice-cube test can confirm the diagnosis of acquired cold-induced urticaria. Conversely, a negative test should not infirm this hypothesis but reveals the atypical form of acquired cold-induced urticaria, leading to a higher risk of anaphylactic shock.

The absolute priority is to avoid this complication, especially in water. Avoiding suspected triggers, prophylactic antihistamines and an emergency kit with epinephrine autoinjectors represent the main approaches of prevention.

In front of a clinic of anaphylactic shock without obvious cause, the anamnesis should investigate all the elements in favour of a cold-induced anaphylaxis.

O5.

Three year follow-up of Streptococcus pneumoniae nasopharyngeal carriage in Belgian children after a PCV13-to-PCV10 vaccine switch

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Background

The predominantly '2+1' Belgian infant pneumococcal conjugate vaccine (PCV) programme changed from PCV13 to PCV10 in 2015-2016. A nationwide nasopharyngeal carriage study in children (6-30 months) attending day-care centres (DCC) or with acute otitis media (AOM) was initiated in January 2016. S. pneumoniae (Sp) carriage was evaluated over 3 collection periods.

Methods

Single nasopharyngeal swabs were taken yearly between January 2016 and May 2018 (3 periods of 4-5 months for DCC and 6-7 months for AOM). The collected samples were transported in 1ml STGG-medium. Sp was detected by culture and PCR. Sp-strains were serotyped by Quellung-reaction. Antimicrobial susceptibility against penicillin, erythromycin, tetracycline, cotrimoxazole and levofloxacin was tested. Demographic characteristics and vaccination status were collected. Children were categorised as vaccinated with exclusively PCV13, exclusively PCV10 or a mixed schedule. Chiò or Fisher's Exact Test were used to determine significance at a level of 0.05.

Results

Over the 3 periods, 2364 and 332 samples were collected in DCCs and at paediatric consultations for AOM respectively. In DCC-children, the proportion that was age-appropriately vaccinated with exclusively PCV10 increased from 0.0% in the first to 80.4% in the third collection period. In AOM-children, this proportion rose from 2.6% to 82.0%. At the same time, the PCV13-vaccinated proportion decreased from 73.4% to 2.4% (DCC) and from 74.4% to 1.2% (AOM). Over the three collection periods, the PCR-based Sp-carriage prevalence changed from 80.0% to 75.7% and to 86.5% ($p<0.001$) in the DCC-children. Among AOM-children, the Sp-carriage prevalence did not differ between collection periods (82.1%, 77.7%, 79.1%; $p=0.842$). The culture-based carriage of PCV13 serotypes increased from 5.4% to 9.4% ($p=0.019$) in the DCC-children. Among AOM-children, the increase was not significant; from 7.4% to 9.2%. The frequency of antimicrobial non-susceptibility against at least one of the five tested antibiotics did not change over the study period (DCC: 40%-43%, AOM: 48%-50%).

Conclusion

Among children in DCCs, carriage of Streptococcus pneumoniae increased significantly over the study period, while it remained stable among children with AOM. Furthermore, pneumococcal PCV13 serotype carriage increased as the proportion of children vaccinated with exclusively PCV10 increased, without impact on the overall carriage of antibiotic-non-susceptible strains.

O8.

Retrospective evaluation of the efficiency of azithromycin in the protracted bacterial bronchitis (PBB)-extended

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Introduction

Protracted bacterial bronchitis (PBB) is characterized by an isolated chronic wet or productive cough without signs of another cause, and which usually responds to 2 weeks of an appropriate oral antibiotic covering the comon bacteria of the respiratory tract such as the Haemophilus influenzae or the Streptococcus pneumoniae. Management of the patients who failed to respond the initial treatment and presented a PBB-extended or a recurrent PBB, potentially associated to the development of bronchiectasis, remains controversial.
The purpose of our study was to evaluate the efficiency of a long duration treatment with low doses of azithromycin in the PBB-extended.

Methods

A register was established that included all patients aged between 0 and 16 years who underwent a bronchoscopy at the Queen Fabiola Children's University Hospital between January 2012 and December 2017 for chronic cough and who were treated by low dose of azithromycin for a minimum of 3 months for a PBB extended.

Results

The response rate in our cohort was higher than 80 % after a median treatment duration of 6 months. Less than 30% of the patients experienced a recurrence of PBB within 6 months after stopping the treatment and no side effects were recorded. Multiple hospitalizations for respiratory infections appeared to be the only factor associated with treatment failure.

Conclusions

Our study suggests that azithromycin may be an effective and better tolerated alternative to prolonged classic antibiotic therapy in the treatment of the PBB-extended and, the necessity to confirm our results with a randomized protocol.

O9.

Diagnosing enteroviral meningitis via blood transcriptomics : an alternative for lumbar puncture?

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Introduction

Meningitis can be caused by several viruses and bacteria. Identifying the causative pathogen as quickly as possible is crucial to initiate the most optimal therapy, as acute bacterial meningitis is associated with a significant burden of disease. Bacterial meningitis requires antibiotics, as opposed to enteroviral meningitis, which only requires supportive therapy. Clinical presentation is usually not sufficient to differentiate between viral and bacterial meningitis, thereby necessitating cerebrospinal fluid (CSF) analysis by PCR and/or time-consuming bacterial cultures. However, collecting CSF in children is not always feasible and is a rather invasive procedure.

Methods

In 12 Belgian hospitals, we obtained acute blood samples from children with signs of meningitis (aged between 3 months and 16 years). After pathogen confirmation on CSF, the patient was asked to give a convalescent sample after recovery. Patients with paediatric rheumatic conditions were recruited as controls. 3-mRNA sequencing was performed to determine differentially expressed genes (DEGs) to create a host transcriptomic profile.

Results

We recruited 49 patients with enteroviral meningitis, 2 patients with other causes of viral meningitis, 7 patients with bacterial meningitis and 14 patients with a paediatric rheumatic condition. Enteroviral meningitis cases displayed the largest upregulated fold change enrichment in type I interferon production, response and signaling pathways. Patients with bacterial meningitis showed a significant upregulation of genes related to macrophage and neutrophil activation. We found several significantly DEGs between enteroviral and bacterial meningitis. Random forest classification showed that we are able to differentiate enteroviral from bacterial meningitis adequately with an AUC of 0.975 (using leave-one-out cross-validation). Moreover, using a similar classification strategy, we showed that enteroviral meningitis could be adequately differentiated not only from convalescence samples and bacterial meningitis, but also from non-infectious inflammatory rheumatic conditions (AUC = 0.928).

Conclusion

Enteroviral meningitis has an innate immunity signature in blood with type 1 interferon as key player. Our classifier, based on host transcriptomic profiles of different meningitis cases, is able to adequately ‘diagnose’ enteroviral meningitis. Our results could offer ‘after adaptation for PCR’ a valid alternative for lumbar puncture in specif

OP25.

A multidisciplinary approach to diagnose X-linked Hyper IgM syndrome in a boy with acute interstitial pneumonitis

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Background

A 4-month old boy of healthy non consanguineous parents was admitted at our hospital with acute severe respiratory failure requiring mechanical ventilation. Medical history showed only gastro-esophageal reflux and normal staturoponderal evolution. He has an older healthy sister. CT imaging showed diffuse interstitial lung disease. Differential diagnosis included primary immune deficiency (PID) and congenital interstitial lung disease.

Methods Viral testing, bronchoalveolar lavage (BAL) and first line immunological evaluation were performed. Flow cytometric analysis was done according to the EuroFlow PID screening algorithm. CD40 Ligand (CD40L) expression was evaluated on stimulated T-cells and genetic testing was performed.

Results

Viral testing was negative, BAL detected pneumocystis jiroveci. Immunological evaluation showed normal total white blood count (9160/µL), decreased serum IgG (0.5g/L), absent IgA, mildly elevated IgM (0.43g/L) and a T- and NK-cell lymphopenia (CD4+ 1230/µL, CD8+ 295/µL, NK 73/µL) with normal B-cells (859/µL). The results suggested an underlying PID including severe combined immunodeficiency (SCID) or an immunoglobulin class-switch recombination deficiency (CSR-D) such as hyper-IgM syndrome (HIGM). SCID was excluded based on a normal T-cell maturation and T-cell Receptor Excision Circles. Weak B-lymphocyte maturation according to age, absent switched memory B-cells and preservation of non-switched B-cells is compatible with a CSR-D. Finally, the diagnosis of X-linked HIGM CD40L deficiency was made based on the absence of CD40L upregulation and the identification of a hemizygous CD40L gene missense variant.

HIGM is a rare X-linked PID characterized with recurrent infections, sometimes opportunistic infections, decreased IgG, IgA and normal or increased IgM.

Pneumocystis interstitial pneumonitis was treated with high dose intravenous (IV) cotrimoxazol and IV corticosteroids. IV immunoglobulins were started. Maintenance therapy includes immunoglobulin substitution therapy and measures to prevention of opportunistic infections: low-dose oral cotrimoxazol as pneumocystis prophylaxis and hygienic guidelines to prevent cryptosporidium. Stem cell transplantation is the only curative treatment.

Conclusion

This case shows the importance of stepwise multidisciplinary approach in the work-up to diagnose the underlying disease. Early diagnosis of PID is important to guide patient-specific treatment and to improve patient outcomes.

OP26.

Management of children with tracheal stenosis: a single-center experience

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Background

Congenital tracheal stenosis is a rare anomaly, caused by the absence of the pars membranacea and the presence of complete cartilage rings, ranging from one single tracheal ring to the full length of the trachea and main stem bronchi. Combination with other congenital anomalies (mainly vascular anomalies) is not rare. The surgical management of children with these anomalies is complex.

Methods

This is a single center study of the management of children with congenital tracheal stenosis.

Results

Since 2011, 7 children were diagnosed with congenital tracheal stenosis. Six patients had associated anomalies: left pulmonary artery sling (n=3), VATER association (anal atresia and hip dysplasia) (n=1), severe gastro-oesophageal reflux and feeding problems treated with Nissen fundoplication (n=1), syndromic appearance with facial dysmorphy and mild mental retardation (n=1), minor orthopedic anomalies (n=1), aplasia of the left lung (n=1).

Symptoms at diagnosis were inspiratory stridor at rest and/or with infections/exercise in 6/7 and episodes of desaturation in 1 child. The diagnosis of congenital tracheal stenosis was made by flexible bronchoscopy at a median age of 17.9 months (range 11 days - 12.4 years). OCT (optical coherence tomography) confirmed the diagnosis in 3 cases. Surgery was performed in 5/7 cases at a median age of 4.3 years (range 7 months - 13.5 years): short segment resection with primary anastomosis in 1/7 patients, slide tracheostomy in 4/7 patients, combined with correction of the vascular anomaly in 3/4. One patient was treated conservatively because of good clinical tolerance at the age of 12 years. The patient with left lung agenesis is currently only 6 weeks old and without need for respiratory support.

Post-operative respiratory status was good in 4/5 patients, with only mild inspiratory stridor at time of infections. Three patients needed balloon dilatation after slide tracheoplasty, ranging from once to 8 times. The last patient is still hospitalized because of residual respiratory problems 3 months after surgery. One patient experienced cardiac ischemia during surgery with residual myocardial dysfunction.

Conclusion

Tracheal stenosis is a severe congenital anomaly with a major impact on respiratory function. Long segment stenosis in particular is surgically challenging.

OP27.

Invasive pneumococcal disease surveillance in Belgium and paediatric pneumococcal conjugate vaccines: have we reached a steady state?

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Background

Surveillance of invasive pneumococcal disease (IPD) in Belgium is based on sentinel laboratories sending isolates to the National Reference Centre (NRC) and Paediatric Surveillance (PediSurv) analysis by the Public Health Institute (Sciensano). The 7-valent pneumococcal conjugate vaccine (PCV7) was included in the Belgian national infant immunisation programme (NIP) in 2007 and replaced with PCV13 in 2011. PCV10 (PHiD-CV) replaced PCV13 in 7/2015 (Flanders) and 5/2016 (Wallonia). We reviewed IPD surveillance data in children in Belgium to assess PCV impact.

Methods

Data were collected from Belgian NRC and Sciensano reports.

Results

Before PCV7 introduction, Belgium had one of the highest IPD incidences in <2-year-olds in Europe (156/100000 in 2002-2003). The IPD incidence dropped by 35% after PCV7 introduction (2007-2010 vs 2002-2003) with a further 42% decrease post-PCV13 (2015 vs 2007-2010). Between 2012 and 2015, incidences remained relatively constant in <2-year-olds (42-59/100000), yet still higher than in other European countries. No incidence data are available for 2016-2018.

When examining unadjusted incidences or numbers of cases based on IPD isolates, a temporary rise in IPD in <2-year-olds was seen after PCV7 introduction (153 cases in 2007; 197 in 2009). Fluctuations in the number of IPD cases were also noted in the PCV13/PCV10 period (79-120 cases between 2012-2017). However, confidence intervals for the number of IPD cases in <2-year-olds in this period mostly overlap, suggesting differences were not significant. Fluctuations may also reflect changes in IPD reporting.

Despite effectiveness of PCV13/PCV10 against 19A IPD in children, this serotype still causes IPD in <2-year-olds, with more cases in 2017 vs 2016 (17 vs 2). Without longer follow-up or information on which PCV and how many doses the affected children received, the cause of this rise is unknown.

Conclusions

After 8 years of PCV13/PCV10 use in the Belgian NIP the overall IPD burden remains high in <2-year-olds. Possible causes of this high IPD incidence merit further investigation. The maximal PCV effect may have been reached in children, as reported in other countries. As seen in Belgium, fluctuations in 19A IPD have also been noted elsewhere, including PCV13-using countries. Continued monitoring of IPD is needed and analysis of generated data should be done with reliable methods to assess the real-world PCV impact.

Funding

GlaxoSmithKline Biologicals SA

P67.

Open your eye to Cat-Scratch Disease...

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Case Report

A 4-year-old girl presents to the emergency department with a swelling of her cheek and conjunctivitis of the homolateral eye, both progressing for 5 days. On admission, she is febrile but previously, no fever was measured at home. She has no other symptoms. The current history reveals that she has kittens at home.

On physical examination, she shows erythema of the conjunctiva and an indurated pre-auricular mass, slightly warm and erythematous.

Ultra-sound shows numerous lymphadenopathies without signs of abscess, and a parotidis. Blood analysis indicate mild inflammatory syndrome, serology tests are pending.

A diagnosis of probable bacterial parotidis is made and oral antibiotherapy (amoxycillin-clavulanic acid) is initiated.

Forty-eight hours later, a granulomatous conjunctivitis has appeared while the pre-auricular swelling is stable. The diagnosis of an oculoglandular syndrome related to Bartonella henselae is made. Indeed, the serology comes back positive. Oral clarithromycin is started. The evolution is favorable with the rapid vanishing of the ocular granuloma and the regression of the lymphadenopathy.

Discussion

Parinaud oculoglandular syndrome is characterized by a granulomatous conjunctivitis, accompanied by lymphadenopathy of the pre-auricular, cervical or submandibular ipsilateral region, and, a febrile state. While B.hensalae is its most common cause, this clinical feature occurs only in 2 to 8 % of patients with cat-scratch disease. Inoculation of bacteria happens during a cat bite, scratch or lick near the eye. Serology test confirms the diagnosis. Antibiotics shorten the duration of the illness. The prognosis is excellent.

Conclusion

Parinaud oculoglandular syndrom is an atypical and rare form of cat-scratch disease. The association of follicular or granulomatous conjunctivitis and lymphadenopathy in the drainage area should bring the diagnosis to mind.

P68.

Non-blanching rash in a toddler- a case report

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Introduction

There are many causes of purpura in children, ranging from disruptions in vascular integrity to abnormalities in primary or secondary haemostasis. Purpura may be a complete innocent finding but it can also be a life-threatening disease. Therefore, it is crucial to recognise it and to have an appropriate management of this condition

Methods

We report the case of a two-year-old boy brought by his parents for a non-blanching rash that appeared some hours ago. The day before, general physician diagnosed a viral rhinopharyngitis and the patient was sent home with pain killers. Initially as the boy looked well, and there was only one episode of fever (38°C), diagnosis of Acute Haemorrhagic Oedema of Infancy was made. The patient had a full blood count and was sent home with a phone follow up. The biology showed an important inflammatory syndrome (311 mg/L of CRP) and a high total leukocyte count. Blood culture was negative. At readmission, the toddler had tachycardia, and normal blood pressure. At clinical exam he was pale, conscious, but irritable. He had a pharyngitis and multiple expanding necrotic lesions (on the nasal septum, the cheek and right ear lobule, the left auricular finger and the upper and lower limbs). In this context, intravenous ceftriaxone was administrated. Evolution was marked by necrotic bullae that developed on the purpuric lesions. The swabs of the lesions grew for Streptococcus pyogenes in numerous sites. Unfortunately, we couldn't make further analysis in order to find out whether it was a super antigen producer GAS serotype. As the toddler didn't fill the criteria of a Toxic shock syndrome, the final diagnosis was a GAS associated purpura fulminans. Additional exams were normal (heart echographia and fundus oculi). Once we identified the GAS, ceftriaxone was relayed by intravenous penicillin and clindamycin for a total of 7 days.

Conclusion

Group A streptococcus (GAS) is a common infective agent in children. The spectrum of GAS ranges from superficial to invasive disease. Our case showed that invasive GAS disease can appear with initial low fever and considerably non-ill appearance. However, invasive infection is a life-threatening disease that needs an appropriate management. There are four factors to consider for appropriate management of invasive GAS: aggressive supportive care, early surgical debridement of necrotic tissue, correct use of antibiotics (penicillin and clindamycin) and intravenous immunoglobins

P69.

YouTube videos as a source of information about immunology for medical students

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Background/Aims

The use of the internet as a source of information has grown exponentially in the last decade. YouTube is currently the second most visited website and a major online educational resource for medical students. The aim of this study was to evaluate the quality, accuracy and attractiveness of the information acquired from YouTube videos about two central concepts in immunology.

Methods

YouTube videos posted prior to August 27, 2018 were searched using selected keywords related to either antigen presentation or immunoglobulin gene rearrangement. Video characteristics were recorded and the Video Power Index was calculated. Videos were assessed using five scoring systems: Understandability and Attractiveness, reliability, Content and Comprehensiveness, Global Quality Score and a subjective score. Videos were categorized by educational usefulness and by source.

Results

A total of 82 videos about antigen presentation and 70 about immunoglobulin gene rearrangement were analyzed. Videos had a mean Understandability and Attractiveness score of 6,57/8 and 5,84/8, Content and Comprehensiveness score of 9,84/20 and 5,84/20, reliability score of 1,65/4 and 1,53/4, Global Quality Scale of 3,38/5 and 2,76/5, and subjective score of 2,00/3 and 2,00/3, respectively. The organized channels group tended to have the highest Video Power Index and Global Quality Scale.

Conclusion

YouTube can provide medical students with some useful information about immunology, although content-wise it cannot substitute textbooks and academic courses. Students and teachers should be aware of the educational quality of available videos if they intend to use them in the context of blended learning.

P70.

Side effects of Bexsero®: a systematic review

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Background/Aims

Neiserria meningitis is a bacterium that cause an invasive meningococcal disease (IMD), which can result in severe disability or death within hours. The bacterium had 12 serogroups of which A, B, C, W, X and Y are responsible for most of the infections. Vaccines are available for the serogroups A, C, W and Y, leaving serogroup B as an important cause of IMD worldwide. Serogroup B infection has a case fatality rate estimated at 5,4% for all ages. However, Bexsero® (4CMenB) is been developed, it is a multicomponent meningococcal serogroup B recombinant protein-based vaccine. At present, it is licensed in 39 countries worldwide. Unfortunately, Bexsero is associated with an increased occurrence of adverse effects (AEs).

Methods

This systematic review includes 19 studies reporting the adverse effects or safety of Bexsero in children aged 0-18 years. The intervention consisted of the administration 4CMenB, the control condition was either placebo or other immunizations.

Results

Fourteen studies describe the occurrence of AEs within infants, 3 within children and 3 within adolescents. The most frequent local reaction in infants is erythema (median 59%) and pain in children and adolescent (median 92,5% and 40%). The most frequent systemic reaction in infants and children is irritability (median 68% and 40%) and myalgia in adolescents (median 47,5%). There are little to no serious AEs. No deaths occurred.

Conclusion

Bexsero is proven to be effective. It has an acceptable tolerability profile with no major safety concerns. Fully informing doctors and parents is important, leading to well-considered choice.

P71.

Recurrent laryngeal papillomatosis - From neonatology to future perspectives

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Recurrent laryngeal papillomatosis is a scourge for children, in an effort to act upstream and achieve a long term complete resolution, diagnosis and therapeutical management should be as early as possible.

Recurrent laryngeal papillomatosis is a rare disease caused by low-risk papilloma virus (HPV), types 6 and 11. It is characterized by an appearance of condyloma in the air tract which can lead to complete obstruction and severe respiratory failure. The incidence of this disease in the United States is estimated at about 4.3 per 100,000 children. The morbidity for these children is significant while there is low mortality.

To illustrate this disease, we will rely on the clinical history of a patient from his neonatal history until now. Despite the treatments tried and put in place repeatedly, the patient still has lesions that now spread through different anatomical sites.

Several difficulties emerge, first of all, the clinical diagnosis which requires an endoscopic examination and anatomical pathology analysis. These patients often present themselves in a state of severe respiratory failure, which is life-threatening and requires emergency invasive management.

Second, the therapeutic management of patients; this involves first and foremost repeated ORL microsurgery, accompanied by hospitalizations and general anesthesia several times a year. It is thus a heavy load, as much from a psycho-medical point of view for the child himself and his entourage, but also economic in view of invasive acts and specific treatments.

Recurrence here is a major problem for management; We will therefore dissect the long-term therapeutic means for a total eradication, if this is possible, of this recurrent laryngeal papillomatosis. Several daily antiviral treatments and the HPV quadrivalent (type 6, 11, 16 and 18) recombinant vaccine have now been proven in the literature to improve the quality of life of these patients. Today, there are many adjuvant treatments which we will detail the efficiencies described in the literature. The therapeutic levels are set up depending on the severity and extent of this disease.

However, we can not yet talk about total cure given the possible reactivation of the virus and the risk of new papillomatous lesions appearing in later years.

P72.

Pulmonary function of children after lobectomy

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Background

Studies in children evaluating the impact of a lobectomy on pulmonary function later on are rare. The aim of the study is to evaluate lung function parameters in children several years after lobectomy.

Methods

In three Belgian university hospitals, children under the age of 16 year who had a lobectomy between 1996 and 2016 were included. Retrospective data were extracted from patient files. Demographic data, associated anomalies, indications for surgery, surgical procedure, complications and pulmonary function tests were reviewed.

Results

108 patients were included. The most common indication for lobectomy was congenital lung lesions (41%). Seventy seven patients (71%) performed postoperative pulmonary function tests at different time intervals. More than 70% of the population had normal median values for forced vital capacity (FVC) and forced expiratory value (FEV1). Almost two thirds of the study population had mean expiratory flows (MEF) at 25%, 50%, 75% and 25%-75% lower than 80% predicted. The total lung capacity (TLC) was normal in 84% of our study population. Fifty percent of patients had a residual volume (RV) above 120% predicted. There was no mortality due to the resection. 26 patients had a minor complication. The most common complication was a pneumothorax after removing the chest tube. There was no correlation between age at lobectomy and pulmonary function ($p>0.05$).

Conclusions

In conclusion, our results confirm that lobectomy is a safe procedure. After lobectomy the majority of our study population has normal pulmonary function tests for FEV1, FVC and tiffeneau index. We identified lower MEF 25%- 50%- 75% one year after lobectomy. Larger prospective studies are necessary to evaluate pulmonary function.

P73.

Haemophilus influenzae type b cellulitis in a vaccinated and immunocompromised nine-months-old child

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Background

Since introduction of Haemophilus influenzae type b (Hib) conjugate vaccine, invasive Hib infection has successfully decreased. Although, cases of invasive Hib disease in children were still reported and may be associated with vaccine failure.

Methods

We analysed clinical and laboratory data of a nine-months-old child admitted in our center at EpiCURA hospital on November 2018. We reported the case to the infectious disease surveillance (AVIQ) and considered available literature about invasive Hib infection.

Results

We report a case of a previously healthy nine-months-old boy admitted in our emergency unit for fever and abnormal swollen and painful knee. He was irritable, but didn't present any neurological signs. Initial laboratory analysis showed important inflammatory syndrome with hyperleucocytosis. Radiography and ultrasound of his leg were performed and excluded bone fracture and arthritis, but showed signs of diffused cellulitis. Empiric antibiotherapy (ceftriaxone) was started for cellulitis with suspected bacteraemia and was prolonged for 10 days. After 24 hours, haemoculture analyses showed presence of Haemophilus influenzae strain and serotyping of the Haemophilus influenzae strain in blood culture observed a serotype b. The boy was fully vaccinated for Hib and received 3 doses of Hexavalent vaccine. Immunological analyses were performed to detect immunological defect and revealed low immunoglobuline dosis of IgG.

Conclusion

Despite global vaccination coverage programs, this case reports Hib vaccine failure in a fully vaccinated boy and suggests that invasive Hib infection may be associated with immunological defect. Furthermore, Hib cellulitis still appears to be uncommon.

Keywords

Haemophilus influenzae type b, children, invasive infection, immunological defect

P74.

A linear skin lesion as presentation of long persistent strongyloidiasis

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Background

A 7 year old girl, born in Ethiopia and adopted at the age of 7 months is presenting with symptoms of acute stomach ache. She is vomiting weekly for several years. A dentist removed all her milk teeth, because they were severely affected by stomach acid. The patient is a very anxious girl that is quickly tired and does not like to do physical exercise. The mother of our patient tells that she sees a linear skin lesion on the buttocks and belly of her daughter quiet often. It disappears after a few hours spontaneously. She is also known with constipation and asthma which are both treated according to the guidelines. And she follows a cow-milk free and soy free diet because of suspected allergies.

Methods

This clinical presentation suggests a parasitic infection. There are 3 types of migrating larva unter the skin: Larva currens which is seen by a strongyloides stercoralis infection and moves 1-2 cm each hour. Larva cutanea migrans is associated with an ankylostoma infection and moves 1-2 cm each day. Also a Loa Loa infection, which is endemic in Ethiopia is associated with subconjunctivale migration of the larvae.

Results

Biochemical examination showed eosinofilia of 4,9 % with a total total eosinofielia of 700/mm3. Microscopic examination of the faeces for parasites was negative. PCR examination of the faeces for Strongyloides which has a very high sensitivity, was positive in combination with a positive serological test for Strongyloides.

Conclusion

Our patient was diagnosed with Strongyloidiasis. This is an infection caused by Strongyloides Stercoralis. Skin lesions of the migration larvae (larva currens) are pathognomonic for Strongyloidiasis. Gastro-intestinal symptoms like nausea, vomiting, stomach pain and constipation are associated with this parasitic infection.

Pulmonary manifestations include coughing, dyspnea and wheezing. Pulmonair eosinophilia is also seen, because of the migration of the parasites through the lungs. This causes asthma-symptoms that worsen with corticosteroid.

Strongyloidiasis is treated with Ivermectine 0,2 mg/kg/dose in 4 doses (dose administered on two consecutive days and then repeated after two weeks). Ivermectine is a safe medication. No intake of food 2 hours before and after oral administration of Ivermectine because of the possible interactions. Albendazole is no longer used as treatment of stongyloidiasis because of the high resistance profile in Strongyloides stercoralis.

P75.

Proteus mirabilis meningitis revealing an intradural dermoid cyst

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Background

Dermoid cysts are infrequent lesions, usually located in lumbosacral region. They can be suspected in case of cutaneous abnormalities, neurologic deficits or infections like spinal abscess or meningitis. Proteus mirabilis is rarely causing bacterial meningitis but its frequency can rise up to 37% of gram-negative meningitis with a predisposing factor. Furthermore, only 50 pediatric cases of spinal cord abscesses due to a dermal sinus are reported.

Case-report

A 15-month-old child was admitted for irritability, refusal to walk, vomiting and lethargy. He was afebrile. There was no history of traumatism. On clinical examination a stiff neck and positive Kernig sign were reported. A lumbosacral orifice with intermittent discharges was reported from early life but never investigated. Laboratory tests showed a neutrophilic (19060/µL) hyperleukocytosis (22550/µL) with a C-reactive protein within the normal ranges. Blood cultures were negative. Urine culture was positive for Proteus mirabilis sensitive to second and third generation of cephalosporins and to quinolones. Lumbar puncture showed leukorachia (2817/µL), hypoglycorrhachia (8 mg/dL) and hyperproteinorachia (263 mg/dL). Cerebrospinal fluid (CSF) culture was positive for Proteus mirabilis with the same antibiogram. Intravenous Cefotaxim was started at doses of 200 mg/kg/day. Two days later, his neurological status declined with high fever. Magnetic resonance imaging (MRI) of the spine confirmed a dermic sinus at level S2 with collection in front of the cauda equine (CE). There was also peripheral brain enhancement evocating arachnoiditis. Because of the persistence of the fever and MRI results, ciprofloxacine was added on day seven. Thereafter clinical evolution was characterized by progressive disappearance of fever and pain upon five weeks of antibiotics and weaning dose of corticosteroids. One month later a repeated MRI showed an increasing of the collection in front of the CE. Neurosurgery was performed to close the lumbar tract. An intradural dermoid cyst was removed. Post-operative follow-up was uncomplicated.

Conclusions

Occult spinal malformation can be underdiagnosed even in children benefiting from a regular follow-up. In Proteus mirabilis meningitis outside the neonatal period it is crucial to rule-out a lumbosacral malformation. Intradural dermoid cannot be reliably excluded by MRI and by ultrasound. The excision and an exploration of all spinal dermal sinus is the treatment.

P76.

Shivering As a Clinical Predictor Of Serious Bacterial Infections in Febrile Children - A Systematic Review

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Background/Aims

Serious bacterial infections such as bacteraemia are a common cause of fever in children. Estimating the risk of serious bacterial infections remains a challenge for paediatricians. Many studies have examined various clinical characteristics that can be a predictor of an increased risk. Traditionally shivering has been associated with an increased risk for a serious bacterial infection in children. The aim of this study is to evaluate the significance of shivering as a predictor of serious bacterial infections in febrile children.

Methods

A systematic review was conducted using Medline, Embase and CINAHL. Key words 'shivering', 'shaking', 'rigors', 'chills' and 'febrile children' were used. Ages ranged from birth until 24 years. Studies had to examine the association between shivering and a serious bacterial infection.

Results

Eleven studies were included in the analysis. Three different study populations were identified: febrile children without malignancy, febrile paediatric oncology patients with neutropenia or without neutropenia. In febrile children without malignancy no significant association was found in the most recent studies. Shivering was significantly associated with bacteraemia in oncology patients with and without neutropenia.

Conclusion

The presence of shivering in paediatric oncology patients with or without neutropenia does significantly increase the probability for a serious bacterial infection. However, shivering is no clear predictor in febrile children without malignancy. Paediatricians should therefore be alert when a febrile child with malignancy presents itself with shivering. Further research, including large-scale long-term prospective studies are still needed to study the role of shivering as a predictor of serious bacterial infections in febrile children.

P77.

About a hereditary pathology of the surfactant: The mutation of the gene encoding surfactant protein C

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Pulmonary surfactant is essential to prevent alveolar collapse at end-expiration. Its dysfunction can lead to severe neonatal respiratory distress or may remain silent until childhood or adulthood.

Here follows the case of a 5 years old girl admitted to our paediatric intensive care unit for acute respiratory distress. In her medical history, we noted recurrent bronchopulmonary infections. The medical workup highlighted a genetic disorder of surfactant which leads to a surfactant protein C deficiency.

She had 5 days of fever associated with a cough and respiratory progressive respiratory distress. At the admission, she was cyanotic and tachypneic with a bronchial breathing pattern at auscultation and a SpO2 of 73%. A nasopharyngeal wab was positive for Influenza A virus. Chest X-ray revealed diffuse opacities. A severe respiratory failure secondary to a Influenza A bronchopneumonia in the context of hereditary surfactant disease was diagnosed. She developed a septic shock due to Streptococcus pneumoniae and needed 10 days of invasive ventilation. She was treated with ceftriaxone for 3 weeks.

Pulmonary surfactant is composed for 80% of phospholipids and for 15% of proteins including 3% of specific proteins called SP-A, B, C and D. Deficiency of proteins SP-B and SP-C are mainly involved in surfactant disorders. SP-C is encoded by a gene located on chromosome 8 (SFTPC). The mutations of the gene encoding surfactant protein C lead respiratory diseases that may present during the neonatal period, childhood or adulthood.

Several mutations exist and they often appear de novo. The prevalence is uncertain but it is a rare entity. The phenotype associated with the same SFTPC mutation is variable. There exist neonatal forms that are lethal in the first year of life and late forms that only give symptoms in adulthood.

The most common clinical signs in children are cough, tachypnea, hypoxemia and growth retardation.

Broncho-alveolar lavage (to study the profile of surfactant proteins by western-blot), genetic analysis or histology confirm the diagnosis.

There is no clear consensus on therapeutic management. Systemic corticosteroids, hydroxychloroquine and azithromycin are used in the treatment of patients with protein C dysfunction. There are few controlled trials that evaluated the treatment of surfactant disease due to the low prevalence of the disease. Lung transplantation is the only definitive treatment.

P78.

'Every sweet has its sour': rare skin lesions in a boy with combined immunodeficiency

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Background

Primary immunodeficiencies (PIDs) are characterized by an increased susceptibility to infections and immune dysregulation. To date, more than 350 PID disease genes have been reported in literature. For most PIDs there is a poor phenotype-genotype correlation.
Sweet syndrome (acute febrile neutrophilic dermatosis) is a rare skin disease that manifests as painful erythematous nodules or plaques, and is usually accompanied by fever and leukocytosis. About half of cases have a severe underlying disease, mainly malignancy or autoinflammatory conditions. It has been very rarely reported in patients with immune deficiency (chronic granulomatous disease, HIV infection).
We report a boy with combined immunodeficiency (CID) from early childhood who showed an exceptional presentation of Sweet syndrome.

Methods

Immunological lab analyses, skin biopsies and PID gene panel analysis were performed.

Results

A boy, currently 16 years old, suffered from recurrent bacterial respiratory infections and an episode of orbital cellulitis since the age of 3 years. Immunological work-up indicated a CID with low IgG2 levels, reduced antibody response to polysaccharide vaccine, monocytopenia and T, B and NK lymphopenia. At 5 years, he presented with erythematous skin lesions with biopsies confirming Sweet syndrome. Till today, he requires dapsone therapy to control skin eruptions. He also developed bronchiectasis, splenomegaly and general lymphadenopathy. Intestinal lymphoid hyperplasia resulted in ileocaecal invagination at 10 years. Genetic testing revealed a gain-of-function mutation in PIK3CD, encoding the PI3K catalytic subunit p110-delta. Activated PI3K-delta syndrome (APDS) can cause variable humoral and/or T cell defects, and is associated with an increased risk of autoinflammation, lymphoma, growth retardation and developmental delay. Its pathophysiology is incompletely understood.

Conclusion

To our knowledge, this is the first report of an APDS patient with Sweet syndrome. This case demonstrates the broad clinical presentation of PIDs. We emphasize the importance of dermatological manifestations in PID patients to provide additional clues towards the diagnosis. In addition, it is has become clear that inborn errors in the immune system may result in an overall failure of both activating and regulating mechanisms. Consequently, the line between autoinflammatory disorders and PID is beginning to fade and both types of diseases can co-exist in the same patient.

P79.

Brain abscess in a 15 years old teenager: a common clinical presentation?

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Introduction

Brain abscess (BA) is a rare pathology in the paediatric population. The mortality and morbidity decreased during the last 60 years, but BA remains a life-threatening pathology. Despite a typical clinical triad including headaches, fever and focal neurological deficits, the diagnosis is always a challenge.

Aim

We want to highlight by means of a case report and a literature review, the clinical features who must evoke this diagnosis.

Case report

We report the case of a 15 years old teenager admitted in the emergency department for headache, nausea and deterioration of general condition. The headaches have lasted for 1 month. They are pulsatil, localized in the left parietal area and non-improved by classical antalgics. Since 5 days, his symptoms have progressed, the pain wakes him up at night and is associated with photophobia, neck pain extending into the spine and alimentary vomiting without schedule. He didn't shudder and didn't present fever recently at home. In this context, he's followed by an ophthalmologist and an orthodontist and has no evidence of ear pain, other ENT symptoms or trauma.

The clinical examination showed only stiff neck at the end of course and diffuse non-inflammatory infracentimetric lymphadenopathy. His vital signs were regular.

The patient was hospitalized in order to manage the pain and to carry out a complete assessment. We performed a blood sample analysis and a blood culture. During hospitalisation, the clinical examination progressed and showed neurological abnormalities and intracranial hypertension signs. We then performed a CT scan, an MRI and a fundus examination.

Results

The biology showed inflammatory syndrome with a neutrophilic leucocytosis. Imaging tests showed a fluid mass in the right frontal lobe and a right pan sinusitis. The fundus showed bilateral papilledema. Finally, we diagnosed a frontal BA complicating frontal sinusitis even if the patient had no clinical sign of sinusitis. The patient was transferred in a neurosurgical centre.

Conclusion

Even if the BA is a typical complication of sinusitis, the clinical presentation isn't obvious. The clinical symptoms are often non-specific and when faced with prolonged headache in children, the anamnesis remains primordial to not miss elements that can guide the diagnosis. The literature review showed that despite the description of pseudo-typical clinical signs of BA, the clinical presentation is typically atypical.

P80.

YouTube as source for pre-travel health information about malaria

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Background/Aims

As the number of people to endemic malaria countries continues to rise, there is an increasing need for pre-travel health consultation. Due to the convenience of the internet, more and more travelers consult this channel for health information instead of a professional healthcare worker. YouTube uses visual images to convey information. Therefore, it can be seen as an important channel for the exchange of information. Since the uploaded videos don't go through a quality check, the correctness of the information can be doubtful. The aim of this study is to determine whether travelers can find sufficiently clear and medically justified information about malaria prevention when they use YouTube as an information channel.

Methods

The keywords 'Malaria travel', 'Malaria precautions', 'Malaria prevention' and 'Malaria mosquito bite' were used to obtain the videos from YouTube. Each video was scored using a viewer designed checklist and was evaluated on their content, understandability and attractiveness. A comparison in terms of audience parameters with 100 videos of the 'entertainment' and 'travel' YouTube category was done as well.

Results

After exclusion, 113 videos remained for analysis. In comparison with standard videos on YouTube, videos about malaria were significantly shorter, less viewed and less appreciated. The median global content score was 2,3/20 and this showed no correlation with like ratio nor with viewership per day.

Conclusion

The videos about malaria cannot replace a pre-travel consultation with a healthcare professional. At this moment their quality is extremely low. Reliance on these videos could have serious health consequences, considering the continuously growing number of travelers to malaria endemic countries. In the future, the ABCD principle can be very helpful to make a trustworthy video that contains all the important information.

P81.

A rare case of invasive Kingella kingae infection

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Introduction

Kingella kingae (KK) is a gram-negative bacillus. Asymptomatic oropharyngeal carriage is estimated to be more than 70% in the pediatric population. Transmission is from person-to-person. This pathogen mainly affects osteoarticular tissue. This report illustrates a case of invasive KK.

Illustration of the case

An 11-month-old male infant hospitalized for the management of pyrexia and deterioration of the general state. There is a recent history of infection of the upper respiratory tract. Medical history is banal. At admission, the physical examination reveals high heart rate, asthenia, irritability, an erythematous pharynx. Admission biology shows a major inflammatory syndrome and a normal leukocyte count. Microbiological samples demonstrate an altered cerebrospinal fluid cytology without identified microbe and an admission blood culture positive for KK. Large spectrum antibiotic therapy is started and readjusted in view of the bacteriological findings in favor of multi-susceptible KK bacteremia with suspicion of hematogenous meningitis at KK. The afebrile child relapsed fever. The echocardiography objectives an asymptomatic endocarditis with a vegetation adjoining the mitral valve requiring surgery. The clinico-biological evolution is favorable. Later, the study of bactericide shows an ampicillin resistance of KK involved in the recurrence of pyrexia.

Discussion

KK infections are often manifested in a sub-acute clinic. Oropharyngeal viral infections promote hematogenous dissemination of the pathogen, causing the alteration of natural defenses. KK is usually penicillin susceptible; this characteristic is not evident in our patient. In children under 2 years of age, endocarditis with large-scale vegetation without underlying cardiopathy or immunodeficiency is frequently observed. A bone scan and a cardiac ultrasound is recommended because of the tropism of KK. In our patient, bone scintigraphy was not performed in the absence of clinical evidence of osteoarticular involvement.

Conclusion

We describe here a rare case of sub-acute endocarditis associated with KK bacteremia and meningitis in an immunocompetent child without cardiac risk, with favorable outcome. KK can cause multifocal invasive infections. The search for a deep infectious focus and the continuation of the antibiogram study are fundamental in the context of the demonstration of a pathogen rarely isolated and having an atypical evolution of the disease.

P82.

Morbidity and mortality of extrapulmonary tuberculosis: two pediatric cases

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Background

Even though M. tuberculosis (M.tb) is the leading cause of death from a single infectious agent, pediatric TB in Belgium remains relatively rare. Most TB in children involves pulmonary disease or cervical lymphadenitis. We present two extrapulmonary pediatric TB cases.

Methods

Review of Pubmed literature about abdominal and meningeal TB in children.

Results

A 6-year-old girl with a 2-week history of low-grade fever, malaise, night sweats and weight loss was referred to us. Parents were born abroad, but had not travelled recently. Examination showed enlarged cervical and inguinal lymph nodes <2 cm, distended abdomen with massive ascites and hepatomegaly on imaging. Chest X-ray and cardiac ultrasound were normal. Laboratory analysis demonstrated anemia, thrombocytosis, elevated CRP (88 mg/l) and sedimentation rate (51 mm/h), with normal LDH, white blood cell count and bone marrow pathology. Broad spectrum antibiotics were started while awaiting the infectious diseases work-up. Cytological analysis of ascites showed lymphocytic pleocytosis. When the tuberculin skin test (TST) became positive, quadruple therapy was started. M.tb ascites culture returned positive. Afterwards, a visiting grandmother was diagnosed with TB.

A 3-year-old girl, with a 3-week history of fever, anorexia and headache diagnosed as meningitis, was transferred to our PICU with encephalopathy. She was already receiving broad spectrum antibiotics and acyclovir on admission. Blood work and imaging of chest and abdomen were unremarkable, as was the initial CT scan of the brain. CSF analysis revealed 353/µl WBC (mainly lymphocytes) with elevated protein (152 mg/dl) and low glucose (15 mg/dl). After the TST became positive, dexamethasone and quadruple therapy was started (protionamide instead of ethambutol). Xpert ultra M.tb PCR on CSF was positive <6 hours after second lumbar puncture which was subsequently confirmed by culture. Despite seizure treatment, external ventricular drain placement and addition of moxifloxacin and amikacin, intracranial hypertension caused (sub-)cortical and lacunar infarcts. Comfort care was started after extubation. She died one month after admission. So far no source has been identified.

Conclusions

Our cases illustrate the progressive disease course typical for TB. Diagnosing TB in children remains challenging which often affects the outcome. Empirical initiation of TB treatment remains necessary, but new diagnostic tools can be very helpful.

P83.

Neonatal granulopenia due to maternal HNA-2 alloimmune autoantibodies: a case report

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Background

Neonatal alloimmune neutropenia (NAN) has an estimated incidence of 2:1000 livebirths caused by antihuman neutrophil antigen (HNA) antibodies. AntiHNA2 antibodies are found in 3.4% NAN and can cause recurrent infections. Self limiting after 6 months, patients require prophylactic antibiotics or Granulocyte Colony-Stimulating Factor. 20% sepsis rate and 5% mortality rate have been described.

Methods

Patient information was obtained from hospital medical records.

Results

Case: a newborn boy admitted to our ward after vaginal delivery at 36+4 weeks of gestation, Apgar 9/10, birthweight 3260g. At risk for neonatal infection (prematurity and maternal GBS), blood culture was taken, and he received penicillin/ gentamycin intravenously.

Day 2: lab showed CRP <5 mg/L and neutropenia: 3.2x10^9/L leucocytes, 0.1x10^9/L neutrophils, 1.9x10^9/L lymphocytes, 1.1x10^9/L monocytes and 0.1x10^9/L eosinophils. He was diagnosed with neonatal granulocytopenia.

After 36h antibiotics were stopped due to a negative blood culture and CRP. Yet he started prophylactic amoxicillin.

Differential diagnosis: alloimmune neutropenia, drug-induced neutropenia, congenital neutropenia, 1st episode cyclic neutropenia, and postinfectious neutropenia.

Parental blood was taken for alloimmune diagnostics. He didn't have infectious signs and was discharged home.

Day 5: readmission with neutropenic fever of unknown origin, under prophylaxis. Sepsis workup was done, intravenous cefotaxime/acyclovir started. Lab showed CRP 6mg/L and leucocytes 2.6x10^9/L. Next day the viral PCR was negative, acyclovir stopped. Then he developed left ear otorrhea and got ofloxacin eardrops after ENT consultation. Ear swab cultured E.coli.

Blood/liquor/urine cultures were negative. CRP accrued to 80 mg/L on adequate broad-spectrum antibiotics,. Fungal prophylaxis was started.

Day 10: confirmation of maternal anti-HNA2 alloantibodies.

Day 14: CRP 7 mg/L and leucocytes of 10.8x10^9/L with normal differentiation. All medication was stopped. Normal neutrophil count was confirmed by a 2nd blood test. One month later he had ear abscess drainage, due to persistent otorrhea. Recovered well, he was discharged.

Conclusion

NAN is uncommon. Yet, its natural course can be swift with remission after 14 days. Serious complications occur, such as ear abscess. Follow-up is required whilst granulopenic and on prophylaxis and indicated until repeated normal granulocyte count is shown.

P84.

Food protein-induced enterocolitis syndrome: an infrequent food allergy but not to be missed!

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Food protein-induced enterocolitis syndrome (FPIES) is a non-IgE-mediated gastrointestinal food allergy. The pathophysiology is still poorly understood. There is an acute and chronic form of FPIES. The acute form is more common in infants between 4 and 7 months of age and is characterized by the sudden onset of repetitive vomiting within 1 to 4 hours of ingestion of the food, associated with a lethargic state and intense pallor, malodorous and sometimes bloody delayed diarrhea. In case of significant symptomatology there is a risk of dehydration, hypotension or hypovolemic shock. The accurate diagnosis of SEIPA is difficult to realize. It is based on precise food history as well as a detailed description of the symptoms. If the history is not clear, an oral provocation test is sometimes necessary to realize the diagnosis of FPIES. Clinical case report of a 9-month-old patient hospitalized in a pediatric department for recurrent vomiting with a delayed diagnosis.

Result

H. a 9-month-old boy, hospitalized several times for development of relapsing discomforts for 4 months. Repetitive vomiting with discomfort occur then, lethargy, intense pallor and rapid deterioration of the general state with sometimes subsequent bloody diarrhea. Similar episodes recur approximately every 4-5 days.

During hospitalization a complete medical checkup with biology (celiac serology, specific IgE), prick tests, stool analysis (fat search), and abdominal ultrasonography was normal. Following an extensive food history and a detailed description of the history, discomfort seems to occur more regularly after ingestion of fish or kiwifruit. An oral fish challenge confirms the definitive diagnosis of SEIPA. Therefore, a diagnosis of non-IgE-mediated FPIES for fish and kiwifruit was made. The treatment consists of a complete eviction of this food during 2-3 years. This pathology can be spontaneously resolving after 2-3 years. Under these conditions an intra-hospital food reintroduction is recommended.

Conclusion

This clinical case study attest the need for early integration of this diagnosis of FPIES into differential diagnosis in this type of clinical presentation of repetitive vomiting with intense discomfort.

This type of symptomatology should lead the practitioner to realize a detailed food history and specify the clinical history. If clinical history remains unclear, an oral food challenge should be performed. Once the diagnosis is established, an eviction is proposed.

P85.

Case-report: A neonate with unilateral lung hypoplasia, complete tracheal rings and hypoplastic left thumb as part of a genetic disease/syndrome?

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Introduction

Lung agenesis/hypoplasia is a rare congenital abnormality with complete or partial absence of 1 or both lungs. It occurs as an isolated finding or can be part of a syndrome. The Mardini-Nyhan syndrome (MNS) is characterized by a triad of lung agenesis, congenital heart disease (CHD) and ipsilateral thumb anomalies. We present a case of a neonate with hypoplasia of the left lung and hypoplasia of the ipsilateral thumb, of consanguineous parents.

Methods

A girl born at 36 weeks and 1 day, was referred to the neonatal department of the University Hospitals Leuven at the age of 1 week, due to desaturations. Chest auscultation was asymmetric without notable respiratory distress. Chest X-ray showed total opacification of the left hemithorax. A CT-scan revealed hypoplasia of the whole left lung. Bronchoscopy showed severe constriction of the left main stem bronchus and complete tracheal rings with constriction of the lumen towards the carina. Cardiac ultrasound showed an absent left pulmonary artery. Furthermore she had bilateral hydronephrosis, an enlarged spleen, thoracolumbar scoliosis and a left hypoplastic thumb. Due to consanguinity of the parents an underlying genetic cause was suspected. Micro-arrays were normal. We performed a literature search for other cases with comparable phenotypical findings.

Results

Unilateral lung hypoplasia and thumb hypolasia might fit in MNS. This syndrome, originally described in patients from consanguineous families consists of a triad of unilateral, lung agenesis, CDH and ipsilateral thumb anomalies. One other case by Mardini, had unilateral lung agenesis and an atrial septum defect but normal thumbs. Hastings et al described 3 other cases. The sister of one of these patients had complex CHD and bilateral abnormal lung segmentation but normal thumbs. This is the first family in which a recurrence of similar anomalies was observed. One case of MNS with hypoplasia of the upper airway as a new finding was described. The fact of consanguinity present in almost all the cases, suggests an autosomal recessive origin. The cases of Hastings et al, as our case, did have normal chromosomes and no pathogenetic microdeletion or duplication.

Conclusion

Our case partially fit in MNS, a rare disorder in which clinical findings make it possible to suspect the diagnosis but for which genetic etiology is not yet documented. Although there was no CHD in our case we observed additional anomalies that, together with the consanguinity, suggest an autosomal recessive inheritance.

P86.

Case Report - Perinatal Varicella zoster virus infection in pregnant women: preventive management of the newborn

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Abstract

Neonatal varicella remains rare but can lead to severe illness for the mother and her neonate. The incidence of varicella during pregnancy is 0.1 to 0.7 per 1000 pregnancies. In Belgium, this corresponds to one of about 45 cases per year. (1)

The consequences for the fetus and the newborn depend on the time of the maternal disease. Infants with the highest risk of acquiring Varicella zoster virus (VZV) are born to women with acute infection appearing between five days before and two days following delivery.

This type of neonatal varicella acquired transplacentally is generally very severe with a very high mortality rate around 30% due to exposure to the virus without passive protection by maternal antibodies. Additionnaly, the cell-mediated immune response of the neonate is likely insufficient to retard the hematogeneous dissemination of VZV after transplacental spread.

In this particular case, it is necessary to administer preventive treatment as soon as possible after birth. Preventive treatment consists of the administration of specific immunoglobulins (IVIG) intramuscularly within 96 hours. The additional prophylactic treatment with Aciclovir remains controversial. Some articles suggest that Aciclovir should only be given if Varicella is suspected. Huang et al. Suggest that Aciclovir should be administered prophylactically to the neonate 7 days after the beginning of mother's rash, although there is a lack of data showing the effectiveness of this preventive approach. (1) The use of non-specific immunoglobulins waiting to obtain the specific immunoglobulins is also discussed.

We present a case of a newborn whose mother presented a varicella-like rash 3 days before delivery. The child was admitted to the neonatology department and was isolated from the mother until she was no longer contagious. Breastfeeding was also post-posed. The neonate received aspecific immunoglobulins (Multigam) awaiting virus-specific immunoglobulins (Varitect). He also received immediately a treatment with intravenous Aciclovir continued for 7 days. The child did not develop clinical varicella and the serology remained negative.

We present this case to discuss the most appropriate approach in the case of perinatal maternal infection by VZV because transplacental infection of the fetus can lead to a serious neonatal infection with life-threatening conditions.

P87.

Henoch-Schönlein purpura? Open your mind in older ones

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Introduction

*Bartonella henselae (*BH*)*, the major causative agent of cat-scratch disease, may cause a variety of diseases both in immunocompromised and immunocompetent patients. Skin manifestations, which occur in about 5% of infected patients, include urticarial eruption, thrombocytopenic purpura, bacillary angiomatosis and erythema nodosum or multiforme.

We describe a case of simultaneous occurrence of cutaneous vasculitis (CV) and inguinal lymphadenopathy and discuss the etiological role of BH.

Case description

A 14-year-old boy presented a painless unilateral inguinal swelling with fever. Ultrasonography showed two lymphadenopathies. Three days later extensive palpable purpuric lesions appeared on the patient's legs and hands. Despite the lack of abdominal pain, arthralgia and renal involvement, the eruption was first classified as Henoch-Schönlein purpura and treated with rest. Azithromycin treatment was started one week later, with disappearance of the vasculitic lesions in a few days.

Complete blood count and inflammatory markers showed unremarkable results. BH serology test performed at day 18 revealed titers of IgG and IgM to be 1:640 and 1:100, respectively. At follow-up 15 days later, titer of IgG had increased to 1:1280.

Inguinal lymph nodes progressively enlarged and suppurated, accompanied by pain and limp. Complete excision under general anesthesia was performed 4 weeks after the onset of symptoms. Histological examination fitted with the diagnosis of cat-scratch disease, characterized by epithelioid granuloma formation and micro abscesses.

Discussion

Cutaneous vasculitis in children may be secondary to a wide range of disorders including medications, malignancy, systemic diseases or infectious causes.

The role of BH as causal agent of this CV may be supported by four main arguments: (i) the simultaneous onset of the skin and lymph node involvement; (ii) the serological and histological demonstration of acute BH infection; (iii) the prompt resolution after azithromycin treatment and (iv) the absence of other common cause of CV.

Even if BH has a well-known tropism for endothelial cells, this is only, at our knowledge, the third reported case of CV linked to this gram-negative bacilli. We propose that BH could be consider as an infectious agent of secondary CV.

This case also suggests that differential diagnosis of Henoch-Schönlein purpura should be considered in patients with isolated form of CV, especially in children older than 10 years old.

O6.

Short term survival and survival without severe morbidity in extremely preterm babies: comparison between two birth cohorts in a third level NICU

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Aims

Contemporary outcome of survival and survival without major morbidities in a population of extremely preterm (EPT) babies admitted in a third level neonatal intensive care unit (NICU) between 2010 and 2016 (cohort 2), in order to evaluate changes in outcomes since 1999-2003 (cohort 1) and guide possible intervention improvement and parental antenatal counselling.

Methods

We analysed retrospectively data collected on 109 patients born before 27 gestational weeks and admitted in the NICU of Cliniques Universitaires Saint Luc (CUSL), in Brussels, between 2010 and 2016. We compared this population to a previous one of 75 babies born before 27 gestational weeks and admitted in the same unit between 1999 and 2003. The main outcomes were differences in survival and in survival without major morbidities between the two periods.

Results

The two populations were similar in term of gestational age, birth weight and obstetrical complications.

Between the two periods we detected a significant increase in the percentage of women who delivered by cesarean section (from 51% to 78%, p=0,001), of babies intubated (from 69% to 89%, p=0,006) and receiving surfactant therapy (from 20% to 86%, p<0,001) in the delivery room.

The overall in-hospital mortality rate of EPT babies decreased by 4% (32% (cohort 1) vs 28% (cohort 2)) but this is not statistically significant (p=0,66). Neonatal deaths occurred earlier in cohort 1 (66% before 48 hours) compared to cohort 2 (18% before 48 hours). In the first cohort deaths were most frequently attributed to multi-organic failure (42%), in contrast with the second cohort where deaths were most frequently attributed to severe central nervous system (CNS) injury (32%).

Strikingly the overall prevalence of survivors without major morbidities (severe CNS injury, severe bronchopulmonary dysplasia at 36 gestational weeks and/or retinopathy of prematurity requiring laser treatment) decreased by 2% from the 1999-2003 period to the 2010-2016 period (45% vs 43%, p=0,78), but neither this is statistically significant.

Conclusions

Overall, our study showed no consistent advance between the two studied periods, in term of survival and survival without major morbidities in EPT babies. Nevertheless, our results are similar to those of the French and English cohorts. Therefore both figures can be used for parental antenatal counselling.

To add value to this study it would be interesting to do an EPIBEL 2 study 20 years after the first one.

O10.

Micro-computed tomography for the longitudinal evaluation of preterm lung injury in a rabbit model

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Background/Aims

Bronchopulmonary dysplasia (BPD) is a lung disease caused by extreme prematurity. Preterm rabbit pups exposed to hyperoxia can be used to study BPD, as they develop similar morphologic and lung functional features of early BPD in humans. The purpose of this study was to evaluate the utility of in vivo micro-computed tomography (micro-CT) in assessing lung injury in preterm (hyperoxia and normoxia) and term rabbit pups longitudinally. We hypothesized delayed lung growth and thus smaller lung volumes in the hyperoxia exposed groups.

Methods

Rabbit pups are delivered preterm (day 28 of pregnancy, term pregnancy is 31 days) by caesarian section. After delivery they are randomly separated into two groups: one hyperoxia group (FiO2 95%), and one normoxia group (FiO2 21%). A third group of pups, the term group, is delivered full-term (at 31 days) by caesarian section (FiO2 21%). Micro-CT imaging was performed under isoflurane anesthesia on day 0 (D0), D3, D5 and D7 in preterm pups, and on D0, D2 and D4 in term pups (same corrected age) using a low-dose small-animal micro-CT scanner (SkyScan 1278, Bruker micro-CT, Kontich, Belgium). Total lung volume was determined by manual delineation on the micro-CT images and adjusted for birth weight.

Results

Comparison of the normoxia group and the hyperoxia group showed a significantly higher total lung volume in the hyperoxia group at day 7 (p-value = 0,0037), but not at day 0 to day 5. Comparison of the normoxia and the term group showed no significant differences.
Total lung volume also changed significantly over time, from D0 - D7 for preterm pups and D0 - D4 for term pups (p <0,0001).

Discussion

Lung volume increases significantly over time in all groups, indicating growth of the lung. Unexpectedly, lung volumes in hyperoxia exposed animals appeared bigger than that in normoxia littermates, with an increasing difference over time. This finding can be explained by air trapping and was also observed in other disease models (e.g. bleomycine induced lung fibrosis). We conclude micro-CT based lung volumes can be used as a tool for evaluating lung injury in rabbit pups. Future perspectives include the analysis of other micro-CT biomarkers (such as average pixel density or percentage of aerated lung tissue).

OP13.

Proximal tracheoesophageal fistula in esophageal atresia: a diagnostic challenge

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Esophageal atresia (EA) occurs in 1 in 2400-4500 births. There are 5 types of EA based on the Gross classification. Type B is the rarest with an incidence of 1.1%. The proximal tracheoesophageal fistula (PTEF) can easily be missed after birth. With this case, we illustrate the difficulty to diagnose a PTEF and its pitfalls.

A girl with polyhydramnios was born via Caesarean section at 32 weeks gestational age for fetal distress. Birth weight was 1160 gram. There was a prenatal suspicion of EA. At birth, there were episodes of acute desaturation and bradycardia due to oral secretions. Imaging confirmed a long gap EA without distal TEF (DTEF). The infant received nasal CPAP for 5 days before being intubated for respiratory problems. An upper pouch study was performed. Instantly, there was an air bronchogram, which was interpreted as overflow into the larynx. A PTEF could not be confirmed. There were respiratory problems due to copious secretions with desaturation and bradycardia, despite of continuous efficient suction in the proximal esophagus. These spells were also seen when intubated. Surgery at day 16 for construction of a cervicostomy and gastrostomy showed a PTEF while the proximal esophagus was being prepared. The fistula was ligated. Preoperative bronchoscopy was normal.

The exact incidence of type B is underestimated in literature because of the initial underrecognition of PTEF when reported as recurrent fistula after repair of EA. The delay in diagnosis can cause respiratory problems and nerve damage if the presence is not know preoperatively. The main goal is timely recognition. Contrast esophagogram, rigid/flexible tracheobronchoscopy, esophagoscopy and more new methods are described, but there is no clear consensus about a diagnostic preoperative assessment. Contrast esophagogram remains highly debated. It needs a high grade of expertise and can be associated with complications (e.g. aspiration pneumonia). On the contrary, it can give false-negative (e.g. an occluded fistula with mucus) and false-positive results (e.g. aspiration through the larynx). Bronchoscopy and esophagoscopy are other options. Rigid tracheoscopy appears the method of choice in some centres, but other studies conclude the complementarity of tracheobronchoscopy and upper pouch esophagogram in detecting high PTEF. However, PTEF can even be missed during these examinations. Ultrasound, CT, MRI and virtual bronchoscopy are more recent alternatives.

OP14.

Both maturational and non-maturational covariates determine neonatal albuminemia on the first day of life

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Background and aim

Drug protein binding influences drug distribution. Albumin is one of the major plasma proteins which binds drugs. Neonates display a lower drug protein binding compared to children and adults, in part related to quantitative and qualitative (e.g. binding affinity) aspects of albumin. Albuminemia depends on gestational age (GA), and increases during the first days of life. In addition to these maturational factors, we aimed to explore the impact of non-maturational covariates on albuminemia at the first day of life in neonates.

Methods

Albuminemia (g/L) data collected during routine clinical care in neonates, admitted to the neonatal intensive care unit of the University Hospitals Leuven between June 2015 and March 2017, were retrospectively collected. Clinical characteristics at birth [GA, birth weight (BW)] and at the moment of albumin sampling [bilirubinemia (total and direct), C-reactive protein (CRP), mechanical ventilation (yes/no), sepsis (no, suspected, confirmed) were extracted from patient files. To explore the impact of continuous and dichotomous covariates on albuminemia, univariate regression and Mann-Whitney U test (Kruskal-Wallis test in case of 3 parameters) were used. Results were statistical significant if $p<0.05$.

Results

Albuminemia on the first day of life was available in 573 cases [median (IQ range) GA 34 (31-37) weeks, BW 2124 (1465-2990) g, albuminemia 31.6 (28.1-35.2) g/L]. Of these cases, 19% was ventilated, 46.3% had suspected and 5.6% confirmed sepsis. Albuminemia was significantly associated with GA (R^2=40.6%, $p<0.001$), direct bilirubinemia (R^2=4.37%, $p<0.001$) and decreasing CRP ($R^2$1.6%, $p<0.0024$). GA and BW were highly correlated. Median albuminemia was significantly lower in ventilated versus non-ventilated cases ($p<0.0001$) and was significantly higher in cases without sepsis versus suspected or confirmed sepsis ($p=0.001$).

Conclusion

Besides maturational covariates, also disease characteristics (non-maturational covariates) determine neonatal albuminemia on the first day of life. In a large neonatal cohort, the need for mechanical ventilation, sepsis (suspected or confirmed) and increased CRP were significantly associated with lower albuminemia. In a next step, intra-patient variation of albuminemia will be explored. Finally, implementation of albuminemia trends and covariates in physiologically-based pharmacokinetic models might improve knowledge on neonatal drug protein binding and disposition.

OP15.

Intermittent CPAP attenuates lung injury in a preterm rabbit BPD model

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Background/Aims

Prematurely born infants regularly develop respiratory distress requiring treatment with assisted ventilation and oxygen supplementation. Both these live-saving therapies can injure the lung and are associated with the development of bronchopulmonary dysplasia (BPD). BPD disrupts lung development and has pulmonary sequelae that extend into adult life. CPAP is a commonly used method of assisted ventilation in these infants and is one of the few treatment options that have been associated with a decrease in BPD. However there is limited data on the effects of CPAP on the structure and function of premature lungs exposed to hyperoxia.

Methods

Preterm rabbits delivered by caesarean section (day 28 of 31 gestation) were randomized to normoxia, hyperoxia (>90% oxygen) or hyperoxia plus daily intermittent CPAP (5cm H2O for 4 hours daily: days 1-6). On day 7 of life, pressure-volume and forced oscillation pulmonary function tests were performed prior to harvesting the lungs for histological evaluation. Alveolar and vascular morphology, as well as airway smooth muscle content were assessed.

Results

Hyperoxia significantly disrupted lung development. Hyperoxia-reared pups had increased alveolar septal thickness, vascular medial thickness and airway smooth muscle content compared to normoxia-reared pups. Furthermore hyperoxia lead to diminished lung function with decreased inspiratory capacity, poorer lung compliance along with increased tissue damping and tissue elastance. CPAP mitigated hyperoxic lung injury with a reduction in pulmonary artery medial thickness compared to hyperoxic-reared pups. Additionally CPAP rescued lung function changes associated with hyperoxia with a significant improvement in tissue damping and elastance. CPAP treated pups trended to have lower airway smooth muscle content and lower central airway resistance. CPAP had no effect on alveolar morphology or static lung functions.

Conclusion

Intermittent CPAP attenuates structural and functional effects of hyperoxic lung injury in the developing lungs of premature rabbits.

OP16.

Maternal paracetamol intake and fetal ductus arteriosus constriction or closure: a case series analysis and mechanistic models in support

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Introduction

There is increasing literature on the use of paracetamol for postnatal ductus arteriosus closure. Similar to other non-steroidal inflammatory drugs, recent case reports describe an association between maternal paracetamol intake and fetal ductus arteriosus constriction or closure. To put these cases into perspective and explore causality, a structured literature search was conducted.

Methods

The World Health Organization Uppsala Monitoring Center (WHO-UMC) causality tool was applied to the cases retrieved in the structured literature search.

Results

The search resulted in 12 papers with 25 case descriptions, of whom 1 case was classified as unlikely, 9 as possible, 11 as probable and 4 as certain. Consequently, we conclude that a causal relationship between maternal paracetamol intake and fetal ductus arteriosus constriction or closure is likely.

Conclusions

These findings suggest that pharmacovigilance studies on paracetamol safety during pregnancy are warranted to quantify the event and put the current findings into clinical perspective. While analgesia during pregnancy and during the peripartum period is of obvious relevance, alternative analgesics like opioids or other non-steroidal anti-inflammatory drugs also have side effects. Mechanistic models to predict the fetal exposure to paracetamol and the extent of ductus constriction have been developed in the meanwhile.

OP17.

Improved vancomycin exposure in neonates using a population pharmacokinetic model-based vancomycin dosing regimen

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Background and aim

Although vancomycin is used for more than 60 years in neonates to treat late-onset sepsis, the optimal dosing is still unknown. In 2014, we documented that 2 published vancomycin dosing regimens resulted in subtherapeutic exposure (trough level <10 mg/L) in 66.3 and 76.2% of neonates. In November 2017, a new dosing regimen derived from a population pharmacokinetic (PK) analysis and using a loading dose, was implemented in our unit. As part of a prospective validation project, we aimed to investigate if the new dosing regimen results in improved vancomycin exposure (target trough 10-15 mg/L).

Methods

Clinical data and routine vancomycin therapeutic drug monitoring (TDM) in neonates admitted to the NICU of the University Hospitals Leuven and receiving vancomycin for medical reasons, were retrospectively collected from November 2017 to June 2018. The data were pooled with 2 historical vancomycin cohorts [cohort 1 (2011, n=193 observations), dosing based on postmenstrual age (PMA) and creatininemia and cohort 2 (2012, n=101 observations), dosing based on PMA and postnatal age (PNA)]. The new dosing regimen [cohort 3 (2018, n=71 observations)] consists of a loading dose, followed by dosing based on birthweight, PNA and ibuprofen co-treatment [2]. Clinical characteristics and early TDM (24h after start) were compared across the cohorts using the Kruskal-Wallis Test. Results were statistical significant if $p<0.05$.

Results

Clinical characteristic, reported for cohort 1, 2 and 3 respectively, did not differ significantly across the cohorts. Median (IQ range) GA was 32.8 (28.4-37.6), 32.1 (28.5-37.5), 28 (26-38) weeks with p=0.097; PNA 13 (6-26), 12 (7-23), 14 (10-25) days with p=0.15 and creatininemia 0.43 (0.33-0.55), 0.49 (0.33-0.65), 0.45 (0.32-0.57) mg/dL with p=0.15. Median vancomycin trough level was 7.8 (5.1-11.3), 5.8 (4.1-8.7), 13.3 (9.9-17.3) mg/L with p< 0.0001. With the new dosing regimen, 25.4% of trough levels was <10 mg/L, 40.8% was >15 mg/L, and 33.8% was within the target, versus 23.3 and 19.8% on target in cohort 1 and 2 respectively.

Discussion

A population PK model-based vancomycin dosing regimen for neonates using a loading dose resulted in improved vancomycin exposure. Prospective validation of PK model-derived dosing regimens in clinical practice is crucial. Although only 25% of trough levels was subtherapeutic, attention and further dosing optimalisation for cases with supratherapeutic exposure is also needed.

P88.

Local pulmonary drug delivery in the preterm rabbit: feasibility and efficacy of daily intratracheal injections

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Background/Aims

Recent clinical trials have successfully used surfactant as a drug carrier for an active compound, in order to minimize off-target exposure. To investigate the translational potential of surfactant-compound mixtures and other local therapeutics, a relevant animal model is required where intratracheal (IT) administration for maximal local deposition is technically possible and well tolerated.

Methods

Preterm rabbit pups (born at 28 days of gestation) were exposed to either hyperoxia or normoxia and randomized to receive daily IT surfactant or saline for 7 days. At day 7, the overall lung function and morphology were assessed. Efficacy in terms of distribution was assessed by microPET-CT on both day 0 and day 7. Lung function as well as parenchymal and vascular structure were altered by hyperoxia, thereby reproducing a phenotype reminiscent of BPD.

Results

Neither IT surfactant nor saline affected the survival or the hyperoxia-induced BPD phenotype of the pups. Using PET-CT, we demonstrate that 82.5% of the injected radio-active tracer goes and remains into the lungs, with a decrease of only 4% after 150 minutes.

Conclusion

The described model and method enable researchers to evaluate IT pharmacological interventions for the treatment of BPD.

P89.

Subgaleaal abces as cause of fever of unknown origin in an 8-day old newborn

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Background

Subgaleal hematoma is a well-known but rare birth trauma in newborns. It is a hemorrhage under the aponeurosis of the scalp and is caused by rupture of the emissary veins. It is often associated with vacuum extraction or forceps delivery. Because the subgaleal space extends across the entire scalp, it can hold up to 260 mL of blood. Therefore it can lead to severe hypovolemia and hyperbilirubinemia. In uncomplicated cases, the blood is resorbed slowly and the hematoma resolves gradually over the course of several weeks. In this case report we present a child born with vacuum-assisted delivery. He had a subgaleal hematoma complicated by infection.

Case report

We report the case of an 8-day old term neonate who presented with fever of unknown origin. Initial physical examination showed fever, irritability and a firm caput succedaneum with otherwise stable vitals. Biochemical examination showed mild elevated CRP and normal WBC. He was admitted to the neonatal ward with administration of cefotaxim and ampicillin. This regimen showed no improvement and the CRP increased. After transfer to the NICU and switch to amoxicillin the infectious parameters declined slowly. However, after discontinuation of antimicrobial therapy, he again developed fever and rising CRP and leucocytes. Diagnostic needle aspiration of the hematoma showed pus and the culture proved an infection with Escherichia Coli. MRI showed a subgaleaal abscess. Antibiotics were switched to cefotaxim and later amoxicillin per os in combination with incision and drainage of the abscess. Rapid recovery followed of both clinical presentation and CRP.

Discussion

Infection of a subgaleal hematoma is known in cases of scalp trauma in adults and children. However, bacterial infection of a neonatal subgaleal hematoma is extremely rare. Diagnosis is relatively ease. Clinical suspicion should arise when local signs of infection or local skin abrasions are present. Diagnosis can be confirmed by needle aspiration. Ultrasound, MRI or CT have little diagnostic value but should be considered to evaluate to the presence of osteomyelitis or intracranial extension. Up to date, a small number of cases have previously been published on this subject. With this case report we want to point out this possible complication. The physician should be aware of the fact that in causes of persistent fever in the newborn period together with a subgaleal hematoma, the cause can be a bacterial infection of the hematoma.

P90.

Schizencephaly : how a cerebral lesion can reveal a drug consumption

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Introduction

Schinzencephaly is a rare congenital cerebral malformation (1/ 65 000 birth) characterized by the presence of abnormal cleft extending from the pial surface of the cerebral hemisphere to the lateral ventricle ; containing heterotopic gray matter. Two types are described: type I or closed-lip where cleft walls are in apposition and type II or open-lip (more frequent) where clefts are separated with CSF communication between ventricle and pericerebral spaces. It tend to occurs in the regions of the rolandic and the sylvian fissures. Multiples causes are described: maternal toxic consumption, environmental issues or familial disorder due to mutation of multiples genes. Clinical feature and prognosis is extremely variable: patients can be asymptomatics or in worse case have spastic quadriplegia. Epilepsy occurs in 50-80% of children and most of them have mental delay.

Case report

A 36-years-old women presents herself in labor at 39 weeks of gestational age.
She had a story of heroin consumption and confessed taking every day Methadone 9mg 1x/d and Lorazepam 2,5mg 1x/d. The newborn had an excellent neonatal adaptation (Apgar score 9/10/10) after vaginal delivery. The birth weight was 2250g (<<3RDP). In the context of small weight for gestational age, the baby got a complete assessment, including a cranial ultrasonography. A type I left schizencephaly has been highlighted. The rest of the work out (cardiac US, abdominal US, PCR CMV) didn't show any abnormalities. The lesion was confirmed by MRI. After explaining to the mother the potential causes of this type of lesion, she revealed having cocaine consumption during pregnancy. According to the neurologist advice, the child was treated with Phenobarbital 4mg/kg/d in prevention of seizure. All the EEG did not showed abnormality. The child remains asymptomatic and has a normal neurological development at the age of 4 months.

Conclusion

Schizencephaly is a rare CNS malformation. We should think about it especially in case of maternal drug consumption. In many cases, neurological manifestations are present such as mental delay or epilepsy. Cocaine consumption during pregnancy may lead to many cerebral malformations. Other negatives consequences may appear: increased preterm labor, mental delay, lower birth weight or behavior disorders. In our case, the schizencephaly has been highlighted by cranial US for IUGR. The child never had symptoms and has normal neurological development at 3 months old.

P91.

Inherited peroxisomal disorders: a case report of neonatal hypotonia and seizures

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Background

Peroxysomes are fundamental for cellular metabolism. More than 15 peroxisomal disorders have been identified, and sorted in 2 main groups: the Zellweger spectrum disorders (ZSDs) where a defect in peroxisome biogenesis leads to loss of multiple peroxisomal functions, and the group of single peroxisomal enzyme deficiencies.

ZSDs are a heterogenous group of autosomal recessive disorders, caused by mutations in PEX genes, and include Zellweger syndrome (ZS), neonatal adrenoleukodystrophy (NALD) and infantile Refsum disease (IRD).

Depending on the genetic defect involved, age of onset and clinical presentation differ. Peroxisomal disorders in neonates should be considered in case of severe neurological abnormalities, craniofacial dysmorphism, liver and kidneys dysfunction and hearing or vision impairment. ZS is the most severe peroxisomal disorder in the neonatal period. Prognosis is poor with death usually in the first months of life.

Methods

We report a girl born at term from consanguineous parents. She presented profound hypotonia, absent reflexes and early neonatal seizures. She had mild dysmorphic features (high arched palate and redundant neck skin folds).

Results

Bilateral fronto-central epileptic seizures without encephalopathy were present on EEG. Association of three antiepileptic drugs only allowed partial seizures control. Brain ultrasound showed sub-ependymal cysts. Cerebral MRI revealed bi-frontal polymicrogyria. Elevated serum very long chain fatty acids (VLCFA) confirmed the diagnosis of peroxisomal disorder. Phytanic and pristanic acids levels were normal. Molecular analysis of the PEX genes and enzymatic activity on fibroblast culture are pending.

Conclusion

Peroxisomal disorders are highly heterogeneous as regard organ involvement, severity of dysfunction, and duration of survival. Sometimes clinical manifestations may be mild or completely atypical. In neonates, peroxisomal disorders should be included in the differential diagnosis of infantile hypotonia, severe neurological dysfunction and dysmorphism. Although treatment is currently only symptomatic, it is important to initiate proper supportive therapy to improve quality of life. The first diagnostic step is biochemical testing, even if some of the peroxisomal disorders may have normal results. Subsequent studies in fibroblasts and genes sequencing are required to differentiate between ZSDs and single enzyme deficiencies. Diagnosis is crucial for genetic counseling.

P92.

Aplasia cutis congenita in an infant of an initial twin gestation

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Background

Aplasia cutis congenita is a rare condition in which there is an absence of all layers of skin tissue at birth, most frequently on the scalp. It can be associated with the intra-uterine death of a co-twin and is most common in monochorionic pregnancies. Numerous causes are postulated, including acute hypovolaemia caused by rapid transfusion from the surviving twin to the demised twin leading to areas of acute ischaemia.

Methods

Case report based on data from the electronic medical records.

Results

A male infant was transferred to the neonatal care unit (N*) with multifocal skin defects on both knees and scalp. He was born at 37 weeks 4 days of gestation by vacuum-assisted vaginal delivery. Birth was induced because of intra-uterine growth retardation (birth weight of 2365g, 3th percentile). The mother had a history of arterial hypertension treated with Methyldopa. Family history was negative for congenital skin diseases and there was no history of infectious disease or trauma in the prenatal period. The pregnancy started as a monochorionic, di-amniotic twin gestation, but deteriorated after the 16th gestational week with the death of a co-twin.

The infant presented with large (4cm diameter), round skin defects on both the knees and a smaller (0.5cm diameter) circular skin defect on the scalp. Lesions appeared as well-demarcated, translucent, ulcerated membranes. Physical examination showed no evidence of other abnormalities except a single umbilical artery. All laboratory tests and abdominal ultrasound were normal. Skin biopsy supported no other theories than aplasia cutis. Conservative treatment with dressings to maintain a moist and anti-bacterial environment was employed, however due to surinfection of the skin, intravenous antibiotics (Amikacin and Amoxicillin) were given during 7 days. Within 1 month, the wounds healed without the need of skin grafting.

Conclusion

In multi-gestational pregnancies with the death of a co-twin, the possibility of aplasia cutis congenita should be considered in the surviving twin with cutaneous defects.

P93.

Case report: should trisomy 18 still be considered as a lethal condition?

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Edwards Syndrome, or trisomy 18, is the most commonly diagnosed autosomal anomaly after trisomy 21 and before trisomy 13. The incidence of live-born children goes from 1/3,000 to 1/10,000, even with pregnancy termination and fetal demise during pregnancy. Children have severe mental deficiency, craniofacial abnormalities and organic anomalies, most of them concerning congenital heart defect. More than half of these patients are still alive after one week and up to 10% after one year.

We report the story of a 5-month newborn diagnosed with trisomy 18 after the sixth months of childbearing. The mother decided to lead the pregnancy to full term without termination although healthcare practitioners delivered the news of this condition being lethal and explained the likelihood of death shortly after birth. She wanted to assist her child in his death and to take advantage of the scarce moments they will be able to live together. This decision led to the parental separation. This patient was not able to undergo a curative cardiac surgery due to diverging opinions in the Belgian medical community. Our patient passed away after being alive for almost 6 months.

The various interventions involving these patients are complex and controversial. Choosing between comfort care and invasive treatment results in tension between perinatal and neonatal communities all around the world. Parental opinion is infrequently understood. With social network testimonies, it has been proved that these children can be happy and tolerably healthy anyways, leading parents to pursue a more aggressive and invasive path of care. Is there a need for a shift in mindset among healthcare practitioners? Is the word «lethal» suitable for this situation?

Keywords: trisomy 18 - lethal - cardiac surgery - congenital heart defect - ethics

P94.

Methemoglobinemia: a rare side effect of a healthy diet

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Background

An 11-month-old girl, without relevant medical history, presented at the emergency department with central cyanosis and an extremely pale skin, noticed by her parents when she woke up from an afternoon nap. She was alert but irritable, there was no shortness of breath. Her oxygen saturation was 80-85% while breathing ambient air and did not respond to high flow oxygen. Chest X-ray was normal. Venous blood gas analysis showed a normal pO2, but revealed methemoglobin (MetHb) levels of 44%. Before sleeping, the girl had lunch with home grown turnips and broad beans.

Methods

A literature-search was done in PubMed with MeSH-terms 'cyanosis', 'methemoglobinemia' and 'child'.

Results

Methemoglobinemia is characterized by increased quantities of hemoglobin containing iron in the oxidized ferric form (Fe3+) instead of the usual ferrous (Fe2+) form. It is an uncommon blood disorder, resulting in a 'functional anemia' with tissue hypoxia. Patients present with a varying degree of cyanosis, proportional to their MetHb level. Symptoms are usually noticed starting from a 15% fraction (normal MetHb fraction is 1%). Levels higher than 70% can be fatal.

The etiology is either congenital or acquired and in most cases due to exposure to oxidizing drugs or toxins, by ingestion or skin contact. In our patient, the presumed causative agent is a high amount of nitrates, a molecule found in well water and some foods, especially in green leafy vegetables and root vegetables. Hereditary methemoglobinemia is rare but should be considered.

The diagnosis is confirmed by direct measurement of MetHb, but can be suspected from a normal pO2 concentration on a blood gas, despite cyanosis and a low oxygen saturation.

If identified, it is important to remove or discontinue the causative agent. Supplemental oxygen should be administered. Treatment with methylene blue is advised if MetHb level > 20%. In critically ill patients, or if MetHb level > 70%, an exchange transfusion or hemodialysis should be considered immediately.

Conclusion

Always suspect methemoglobinemia in children with central cyanosis without respiratory distress or cardiac disorder, not responding to oxygen administration. Low oxygen saturation with normal pO2 on blood gas analysis provides the clue to diagnosis. Usually methemoglobinemia is caused by exposure to certain drugs or toxins that can be found even in vegetables. Methylene blue is the treatment of choice.

P95.

Neonatal thrombopenia with serious complications

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Background

We present a newborn girl with severe thrombopenia, petechiae and symmetric growth retardation on first clinical examination, caused by congenital CMV (cCMV) infection. The mother had already been immunized with CMV prior to the pregnancy. Further examinations revealed a unilateral sensorineural hearing loss (SNHL) of 50 dB and severe MRI abnormalities. So this girl was diagnosed as a severe congenital CMV infection after a non-primary maternal infection. Treatment with oral valganciclovir was started for a total duration of 6 months.

Methods

Literature was obtained by using the PubMed database. The following search terms were used: neonatal thrombo(cyto)penia', 'congenital CMV infection', 'congenital cytomegalovirus', 'reactivation and congenital CMV' and 'non-primary CMV infection'.

Results

Recent evidence suggests that there are more cases of cCMV infections attributed to non-primary CMV than to primary CMV infections. Reactivation of the latent virus or infection with a new strain of the virus can occur during pregnancy in seropositive women, resulting in congenital CMV infection in the newborn. The risk of short and long term sequelae seems to be the same in primary and non-primary infections. Most infants are asymptomatic at birth but sequelae can be severe and may develop many years later. Since there are no vaccines or any therapeutic options prenatally available at this point, we need to educate healthcare providers and (pregnant) women about the possible hygienic measures to reduce the risk of infection. Studies have shown that good education results in an important reduction of the risk of CMV infection during pregnancy.

Conclusion

Congenital CMV infection is the most important congenital infection worldwide with great impact on the child, parents and society. It is the cause of important disability (such as hearing loss, vision loss, cerebral palsy and/or cognitive impairment) in thousands of children each year.

Prenatal therapeutic options are not available at this point so we need to focus on reducing the risk of CMV infection during pregnancy. Educating all pregnant women or women of childbearing age on hygienic measures is of great importance.

As this case shows, not only the seronegative pregnant women should be counseled because seropositive mothers also benefit from adequate prenatal education.

P96.

Fulminant neonatal acute liver failure due to enterovirus: a case of favorable outcome after intravenous immunoglobulin administration

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Background

Neonatal enterovirus infections are usually not apparent or associated with minor symptoms. However, acute liver failure, which combines liver cytolysis and coagulopathy, is a rare but life-threatening manifestation of enterovirus infection.

Clinical case

A 36-week infant born after cesarean section presented at 4 days of life with lethargy, jaundice, and fever. The baby required endotracheal intubation for a comatose state. Electroencephalography showed serious encephalopathy. Laboratory tests revealed severe coagulopathy, moderate hyperammonemia, massive liver cytolysis, anemia, and thrombocytopenia. Five days after infection onset, enterovirus RNA amplified by PCR was found in pharyngeal secretions. Based on extensive and repeated biological tests, other main causes of acute liver failure were excluded, including inborn errors of the metabolism, other viral or bacterial sepsis, fetal alloimmune hepatitis, and hemophagocytic lymphohistiocytosis. Hemodynamic support was transiently needed in the absence of echocardiographic signs of myocarditis. Daily transfusions of plasma and platelets were administered for 7 days. High doses of intravenous immunoglobulins were administered during 3 days. Coagulation tests and platelet count normalized at 22 days of life. Brain magnetic resonance at 3 weeks of life showed stigmas of hemorrhage in the periventricular white substance. Hepatic cytolysis and cholestasis started to improve at 12 and 22 days of life respectively, yet without complete recovery after one month. Liver ultrasonography did not show significant structural anomalies, ascites excepted. At one month of life, the infant was breastfed and exhibited mild axial hypotonia and sleepiness.

Discussion

Enterovirus family is responsible for a wide spectrum of manifestations, from mild symptoms to myocarditis, meningo-encephalitis, and sepsis. Few cases of enterovirus-related fulminant liver failure have been reported in neonates, mainly in the presence of maternal history of respiratory symptoms, fever or diarrhea. Disease severity seems related to serum aminotransferase level, multiorgan failure, and timing of infection. Beside supportive management, high doses of intravenous immunoglobulins could improve survival.

Conclusion

In case of neonatal acute liver failure, enterovirus infection must be ruled out by molecular diagnosis. Early recognition and subsequent intravenous immunoglobulin therapy might improve clinical outcome.

P97.

'How do families cope with premature birth?'

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Background

Premature birth causes a lot of stress and burden to young parents. Fear, guilt, even depression impair families during but also after the NICU-stay. On the other hand a stimulating parent-child interaction plays a key role in brain development, not only in the NICU but also the first years at home. A difficult family situation might hinder this interaction.The study's goal is to see how family life is affected by premature birth and to identify possible ways to support families post-discharge and coach them in positive parent-child interaction.

Methods

This study is part of a single-centre prospective study to evaluate the impact of a long-term intervention on parental stress, parent empowerment and developmental outcome in babies born under the PMA (post-menstrual age) of 32 weeks. The intervention is an age-adapted web-application offered to the parents from NICU-admission until their child's corrected age of 2. We recruited 110 patients: 52 in the pre-intervention group and 58 in the intervention group.

We evaluate the family situation from admission until the corrected age of 1 year old using questionnaires in the first week of NICU-hospitalisation and again during a standardised visit to the neurodevelopmental clinic at the corrected age of 5 months and 1 year old. We question the parents' relationship, their employment status and which persons they turn to for support concerning daily practical matters and childcare. We also ask which professionals they eventually consult for personal support. Finally we ask if the immediate surroundings 'understand' their situation and if the parents experience real support on practical/ emotional field from them.

Results

We present the effect of premature birth on the family situation from week 1 until the corrected age of 1 year old and point out which kind of support these families seek, whether they get it and from whom.

We also evaluate whether the use of the web-application has any effect on the way families try to cope with premature birth and make suggestions on further parent-supporting interventions.

Discussion

There are many interventions that try to support parents during the NICU-stay, but little work is done to evaluate the needs of families after discharge. Interventions to improve the premature's development should include individual assessment of the parents and prolonged parental coaching in order to enhance the parents well-being and facilitate a positive parent-child interaction.

P98.

Postanoxic encephalopathy leading to organ donation after circulatory death in a 12-year-old boy

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Introduction

Postanoxic encephalopathy has a grim prognosis. Patients with severe anoxic brain injuries may be candidates for organ donation (OD). The dead-donor rule states that death must precede OD. That is why OD for patients who are not brain-dead requires an irreversible circulatory arrest. Pediatric OD after circulatory death (DCD) is ethically challenging and is still uncommon in Belgium. Heart DCD is even more unusual. We report the case of a child admitted to our pediatric intensive care unit (PICU) who became a circulatory-dead organ (including heart) donor.

Case report

A 12-year-old boy was admitted to our PICU after hanging. A return of spontaneous circulation had been observed after 15 minutes of resuscitation. Despite usual post-resuscitation care and even though no specific complication occurred after admission, the neurological condition did not improve. Clinical examination and paraclinical tests (monomorphic EEG, unresponsive somatosensory evoked potentials, extensive ischemic lesions at cerebral MRI and high serum neuron-specific enolase) undoubtedly predicted a very bad neurological outcome. The criteria for brain death were not met. At day 9, the decision to withdraw life-sustaining therapies was taken. The option of OD was then offered to the parents. Thanks to a close collaboration with the transplantation center and the adult ICU from Liege university hospital, OD occurred at day 14. Remarkably, besides the kidneys, the liver and the lungs, the heart was recovered and successfully transplanted.

Conclusion

Anoxic cerebral injury is a dramatic event. No test per se is sufficient to assess neurological outcome and a careful multimodal evaluation is mandatory to establish a reliable prognosis. End-of-life management may be challenging when OD occurs, especially for children. The whole process must be centered to the dying child and its family, and OD must not counteract the appropriate end-of-life process. The determination of death must be protocolized so that no doubt arises with respect to the dead-donor rule. Each health-care provider needs to be properly informed of the process in order to alleviate the impact of this potentially stressful and emotionally taxing event. Pediatric DCD is feasible and has the potential to increase organ procurement, including heart transplantation. Moreover, OD may provide solace as parents face the tragic loss of a child. Further work is required to encourage this practice in Belgium and worldwide.

P99.

Bloody stools - at the maternity ward

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We present the case of a one day old infant with bloody stools at the maternity. History and physical examination ware unremarkable. He was made nil per os, a diagnostic work-up was started and he was given broad-spectrum antibiotics. Culture of his stools showed a Campylobacter infection as the cause of his bloody stools, probably with transmission by passing through the birth canal. Other exams were all reassuring. Erythromycine per os was given for five more days after diagnosis and he recovered well.
We discuss the differential diagnosis of bloody stools in infants and describe the work-up that should be done to determine the specific diagnosis. Although most cases are benign, life-threatening pathology should always be excluded.

P100.

Bronchocoele in the neonate : conservative or invasive therapy?

A Clarysse, M Sijmons. AZ Sint Jan Brugge

A pneumatocoele is a gas-filled cyst that develops within the lung parenchyma and is thought to be part of the spectrum of air leaks including pneumothorax, pneumomediastinum and pulmonary interstitial emphysema. Etiology can be infectious, traumatic, congenital or positive pressure ventilation induced. Incidence is 1.8% and can appear at all ages. It is more prevalent in the right lung, mostly in the medial/lower lobe. There is an association with low gestational age and low birth weight. Time to diagnosis is 7 days.

We describe a term male neonate with a postnatal bronchocoele of the right lower lobe. The pregnancy was complicated by a uncontrolled thyroiditis, type Hashimoto. At birth, the baby was pale and bradycardic with opistotonus. Apgar scores were 4, 5 and 6. Normal umbilical blood gasses. There was need for neonatal resuscitation with thorax compressions and intubation with mechanical ventilation. Neonatal total body cooling was commenced for 72 hours for a Thompsonscore of 10. The child was extubated afterwards without further respiratory support. On day 8, the child was in acute respiratory distress. Chest X-ray showed a cystic, air filled mass in the right lung with a lateral pneumothorax. Previous X-rays were normal. The child was reintubated and a CT scan was performed, which showed a big cystic lesion in the right lower lobe with a connection to the main right stem bronchus and compression of the surrounding lung tissue due to mass effect. The pneumothorax and the cyst were drained under X-ray fluoroscopy by placing two pig tail drains. After drainage, a large air leak persisted because of the direct connection with the stem bronchus. Conservative therapy was not possible, so the cyst was surgically removed with a small part of the lower right lobe that was connected with the stem bronchus. The child was extubated and without respiratory support within 24 hours after surgery. Pathology was inconclusive to determine the origin of the cyst, most likely to be a bronchocoele.

Pneumatocoeles and/or bronchocoeles are a rare complication, mostly related to infection or mechanical ventilation. We have to keep it in mind when evaluating a neonate with respiratory distress without a clear explanation. It is important to do a full diagnostic work up when a cystic mass is suspected on medical imaging. Conservative therapy is sufficient in most cases, but rarely there is need for urgent (surgical) intervention or drainage. Complication rate is low.

P101.

Extensive and rapidly developing white matter damage in preterm infant

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Background

White matter damage (WMD) in preterm infants is an unpredictable complication with important implications for future neuro-development. Etiology is multifactorial. Mostly it presents 4 to 6 weeks after the initiating event. Our case shows an early and fulminant course of WMD.

Case presentation

A growth-retarded infant was born at postmenstrual age 30 1/7 weeks. The course was uncomplicated, with normal cerebral ultrasound on day 1 and 14.On day 20 she developed a fulminant necrotizing enterocolitis (NEC) with shock and acidosis. Within 48 h her condition stabilized and cultures of blood, faeces and urine remained negative. Peritoneal fluid obtained during surgery showed minimal growth of Enterococcus Faecalis. Nevertheless there was a strong inflammatory response with C-reactive protein of 249 mg/L. Day 25 she developed marked diastasis of sutures. Imaging showed hyperreflectivity of periventricular white matter with progression to large porencephalic cysts in 4 days' time. A lumbar puncture performed day 48 showed high protein range, no increased cell counts and a negative culture.

Results

Literature shows that development of WMD is multifactorial, with hypoxia, oxidative stress, inflammation and infection as major contributors. The role of infection and inflammation and the exact mechanism are still under investigation. In trials with preterm piglets severe NEC lead to brain barrier disruption and neuroinflammation, also in absence of central infection. Neuroinflammation interferes with oligodendrocyte maturation and myelinisation leading to neuronal loss.On the other hand Gram negative sepsis and encephalitis (e.g. Serratia, Citrobacter) is also a possible entity causing catastrophic WMD. Since lumbar puncture in our patient was only performed later, we cannot exclude encephalitis.Intraventricular debris on MRI imaging suggested ventriculitis. Differentiation from white matter loss due to metabolic disorders and from vanishing white matter disease could be made due to distribution of brain lesions on MRI.

Conclusion

The role of infection and inflammation in development of WMD is still subject to research, the exact mechanism is yet to be revealed. Although there is possibility of an encephalitis in our subject, we suspect that the strong inflammatory response was an important factor in the speed and extent of WMD development.We suggest to consider a lumbar puncture to exclude encephalitis, even in primary NEC without sepsis.

P102.

An unusual case of SpO2-PaO2 discrepancy in an intensive care neonate illustrating an important pitfall in routine pulse oxygen monitoring

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Background

In the last decades, pulse oximetry became an essential part of routine monitoring, ubiquitously present from low to high intensive care units. In most cases, pulse oxygen saturation levels (SpO2) are reliable and reflect the simultaneous partial pressure of oxygen in the blood (PaO2). Nevertheless, specific morbidity, of which methemoglobinemia might be the most well-known, can interfere with this correlation. Clinicians should cautiously interpret SpO2 levels in such cases.

Methods

A case report describing a rare SpO2-PaO2 discrepancy and literature review concerning similar cases.

Results

Progressive hydrops foetalis, caused by Rhesus antagonism and requiring multiple blood transfusions intrauterine, occurred in a premature born (GA 30+2/7w) female infant (G4P4A0). She was hospitalised at the neonatal intensive care unit. Endotracheal intubation, fluid resuscitation and hemodynamic support was necessary. Hemolytic anemia, trombopenia and early-onset hyperbilirubinemia was present. Blood transfusions and intensive phototherapy were given. Progressive cholestasis developed. A severe picture of bronze baby syndrome (BBS), a skin discoloration typically caused by brown photoproducts of accumulated porphyrines (as present in significant cholestasis), followed in the first week of life. During mechanical ventilation, vital parameters were intensively monitored and frequent arterial blood gas analysis took place. At the age of 1 week, prolonged episodes of low SpO2 occurred (82-88%), unresponsive to FiO2 increments. There were no pre- and postductal differences, nor variation with different types of pulse oximeter. Moreover, high PaO2 levels (up to 470 mmHg) were reached with hyperoxygenation, although simultaneous low pulse oxygen saturations remained.

We found only one other case, published 10 years ago, that illustrates a potential but important SpO2-PaO2 inconsistency in bronze baby syndrome. Mechanisms for this interference, e.g. spectral absorbance alterations, have already been proposed, but need further explanation. Our case confirms the possibility of such an underlying interference mechanism.

Conclusions

SpO2 is a widespread means of monitoring in intensive care patients. Pigment dispositions in the skin, such as in bronze baby syndrome, might cause relevant SpO2-PaO2 discrepancies. Our case, only the second in literature, illustrates this important pitfall. Clinicians should cautiously interpret SpO2 levels in such cases.

O11.

Stress, anxiety and depression in parents of children with chronic kidney disease and its correlation with protective behavior

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Background/Aims

The quality of life in children with chronic kidney disease (CKD) is impaired. However, little is known about the impact on the family of having a child with CKD. The aim of this study was to explore the psychological wellbeing in this parent group with special emphasis on parental stress, anxiety, depression, perception of their child's vulnerability, and protective behavior.

Methods

Fourthy-four parents (32 mothers) of children with CKD completed questionnaires regarding their own psychological wellbeing. Parental stress was evaluated by the Parenting Stress Index - Short Form (PSI-SF), symptoms of anxiety and depression were measured using the Hospital Anxiety and Depression Scale (HADS). The Child Vulnerability Scale (CVS) and Parent Overprotection Measure (POM) were used to explore parents' perception of child's vulnerability and protective behavior, respectively. Additionally parents completed a questionnaire regarding their socio-demographic situation. A control group of parents of healthy children (matched for gender, age and socio-economic status) was used.

Results

The socio-demographic questionnaire revealed that 47% of the parents perceives a deterioration of their own health since the CKD diagnosis of their child. In 38% of the families, one parent reduced work activities due to the CKD diagnosis, in 9% both parents reduced work. Averagely parents took 17 days of work leave in the last 6 months because of the CKD treatment. Regarding general parental stress (PSI-SF), parents in the CKD group reported a higher stress level than the controls (59.6 versus 43.5; $p < 0.001$; normal range 43 - 61). Also, this parent group reported higher symptoms of anxiety (7.8 versus 5.1; $p < 0.05$; normal range 0-7) and depression (5.3 versus 3.7; $p< 0.05$; normal range 0-7). Finally, significantly higher scores on child vulnerability perception and protective behaviour could be measured in parents of CKD children with respectively the CVS (8.6 versus 2.5; $p< 0.001$) and POM questionnaire (41.7 versus 36.8; $p< 0.05$).

Conclusion

Parents of CKD patients report more health problems and a significant professional impact. Compared to parents of healthy children, a higher presence of stress, anxiety and depression symptoms can be seen. Also, these parents perceive their child as more vulnerable and use more protective behavior. As the impact of CKD goes beyond the child and affects the entire family, a family-based therapy should be recommended.

OP32.

Desmopressin in enuresis - predictive factors in therapy response

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Background/aims

Desmopressin (dDAVP) is a grade I, level A therapy for monosymptomatic nocturnal enuresis (MNE). Previous research suggests response can be expected in children with nocturnal polyuria (nocturnal diuresis >130% of expected maximum voided volume (MVV) for age) and 'normal' MVV for age (> 65% of expected MVV for age), according to ICCS-definitions. However, this expert-opinion based definition seems applicable to rather a small part of children with MNE. Moreover, up to 60% of children show less than 50% decrease in the number of wet nights despite proven antidiuretic effect. The aim of this study is to identify clinical parameters to predict desmopressin response.

Methods

This study is a retrospective single-centre study, including 76 children who were treated with dDAVP (age 5-18y). 24 patients with MNE and in 52 patients with NMNE. Logistic regressions were performed to verify the predictive power of the different parameters on the response. Furthermore, associations for the nominal parameters were also investigated using Fisher Exact. The parameters studied were sex, age, number of wet nights, average night-time diuresis, nocturnal polyuria, maximum voided volume (MVV) excluding and including morning void, comorbidity and finally the form of administration, duration of therapy and dose.

Results

A higher number of wet nights before dDAVP administration is a significant predictor for a better response in patients with NMNE (OR=1,031; p=0,035). A significant positive association in patients with MNE is found between the use of the lyophilisate as a form of administration and complete response (p=0.024; z=2.8). This was not shown for the tablet form and even negative associated with the nasal spray.

Conclusion

A positive association between the melting tablet and complete response in MNE was shown. This is important since the lyophilisate is proposed as an option in the first line although it has not yet been reimbursed in Belgium. Moreover, in children with NMNE the severity of enuresis, expressed in the number of wet nights, seems a predictive parameter for dDAVP response. The study is limited due to the retrospective design, these findings have to be confirmed in a prospective study.

OP33.

Fecal microbiota transplantation for recurrent clostridium difficile in children with end-stage renal disease and renal transplantation

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Background/aims

Clostridium difficile infection (CDI) is one of the most common causes of healthcare-associated infections. The incidence and severity of CDI in patients with an organ transplant is higher than in the general population. The recommended therapy consists of either metronidazole or vancomycin. If recurrent CDI keeps appearing, a fecal microbiota transplantation (FMT) can be considered. The effect of FMT in children with chronic kidney disease (CKD) with/without immunosuppressive therapy is unknown.

In this study, we describe three children (age range 5-10 years) with CKD and renal replacement therapy that received an allogeneic FMT because of recurrent, life-threatening CDI despite repeated courses of antibiotic therapy.

Case series

A single center, prospective, observation study was performed. Two sisters (one in dialysis and one after renal transplantation) received their FMT in December 2016. The first time the older sister was diagnosed with CDI was in January 2011. After this CDI, she developed 5 recurrences between June 2012 and September 2016 that were all treated with antibiotics (vancomycin or metronidazole) after which a different treatment was chosen, namely FMT. The younger sister developed her first CDI in May 2014. Between July 2014 and October 2016, she had 5 recurrences, all treated with antibiotics and eventually with FMT.

A 5-year old boy after transplantation received his FMT in February 2017. Six months later, a second FMT was performed because of recurrent CDI. In this case, the boy developed 3 recurrences of CDI between June and December 2016 despite courses of antibiotic therapy.

Vancomycin was stopped before the moment of FMT in all. The two children after transplantations were treated with tacrolimus, MMF or azathioprine and corticosteroids.

All three children were discharged one day after the FMT took place. No clinical recurrences of CDI nor adverse events have occurred until now.

Conclusion

In our small series, FMT proved to be a successful treatment to eradicate the recurrent CDI in patients with end-stage renal disease and renal transplantation without serious adverse events.

OP34.

Promising experience of paediatric acute peritoneal dialysis program using home made fluids in the democratic republic of congo

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Background

The Democratic Republic of Congo (DRC) is one of the most challenging environments for health development in sub-Saharan Africa (SSA). Indeed, child mortality in the DRC is one of the highest in Africa. The acute kidney injury (AKI), which is often a fatal complication of the most common diseases of childhood like malaria, diarrhea and sepsis, contributes significantly to the high rate of child mortality because of the conjunction of poverty, the lack of pediatric dialysis programs and the deficiency of qualified health care providers in pediatric kidney diseases.

Methods

In order to reduce preventable deaths by optimizing the treatment of AKI, a team composed of one physician and one nurse from the Pediatric Nephrology Department of the University Hospital of Kinshasa has been recently trained at the University Hospital of Cotonou with the joint support of the KU Leuven through the VLIR Project and the Saving Young Lives (SYL) program, a complementary partnership between 3 international nephrology organizations: International Society of Nephrology (ISN), International Pediatric Nephrology Association (IPNA), International Society for Peritoneal Dialysis (ISPD). The training consisted of learning all aspects of acute peritoneal dialysis (PD) and especially focused on two useful techniques for low income countries namely on bedside PD catheter insertion and the local production of PD fluids.

Results

Upon the return from training, in December 2017, pediatric PD activities have started, remarkably at a very low cost. From January to October 2018, 27 children with AKI mainly due to severe malaria and sepsis were eligible to dialysis. There were 15 boys and 12 girls with the median age of 8 years (4 months - 15 years). The main indications of dialysis were uremic toxicity, prolonged anuria (> 4 days) with fluid overload. Despite the low socioeconomic status of parents, almost all children have been dialyzed and 23 recovered efficient diuresis after an average of 8.0 ±4.1 days of PD treatment. We noted that 1/27 patients developped peritonitis that was effectively treated and 3/27 died of complications of comorbidities.

Conclusions

This promising experience reveals the importance of South-South cooperation with the support of the Northern partners. The final goal of this program is to extend the training to practitioners in other provinces of the country in order to contribute significantly to the 0by25 objective of the ISN.

OP35.

Uremic toxin concentrations are related to residual kidney function in the pediatric hemodialysis population

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Background

Uremic toxins are accepted to play a role in the multisystem disease of children on hemodialysis (HD), but little is known about uremic toxin concentrations in this population.

Aim & Methods

Here, we evaluated concentrations of a broad array of uremic toxins in a large pediatric HD cohort (n=170), and secondly explored the impact of residual kidney function using analysis of covariance and Spearman correlation coefficients (rs).

Results

We found significantly higher concentrations of Î²2-microglobulin (Î²2M), p-cresylglucuronide (pCG), hippuric acid (HA), indole acetic acid (IAA), and indoxyl sulfate (IxS), and overall lower protein-binding capacity in the HD compared to the chronic kidney disease (CKD) stage 4-5 group (n=24). In the HD group, we demonstrated lower concentrations of Î²2M, HA, and 3-carboxy-4-methyl-5-propyl-furanpropionic acid (CMPF), and higher protein-binding capacity of IAA and IxS in the non-anuric compared to the anuric HD patients. Also, residual urine volume was negatively correlated with uremic toxin concentrations of Î²2M, pCG, HA, IAA, IxS, and CMPF (rs -0.2 to -0.5), and overall positively correlated with protein binding (rs 0.2 to 0.3).

Conclusion

Our study shows that residual kidney function plays a major role in the uremic toxin concentrations in the pediatric HD population, and that residual urine volume might determine albumin-binding capacity, and concentrations of a broad array of uremic toxins.

OP36.

Chitotriosidase: a novel alternative biomarker for the therapeutic monitoring of nephropathic cystinosis ?

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Background & Aims

Nephropathic cystinosis is a rare, autosomal recessive lysosomal storage disorder caused by various mutations in *CTNS*, which encodes the lysosomal cystine-proton co-transporter *cystinosin*, leading to lysosomal cystine accumulation and crystal formation throughout the body. It is a multisystem disorder that mainly affects the kidney. The only available disease-modifying treatment is cysteamine and due to its narrow therapeutic window, close therapeutic monitoring is mandatory for which the assessment of the leucocyte cystine level (LCL) is the current gold standard. However, the LCL assay is technically demanding and only available in very few laboratories in developed countries. Since macrophages are among the most commonly affected cells by cystine accumulation and preliminary *in vivo* data have shown significantly elevated levels of chitotriosidase in the cystinosis murine model and in cystinosis patients, we hypothesized that macrophage activation biomarkers could be novel alternative candidates for the therapeutic monitoring of cystinosis.

Methods

A prospective longitudinal multicenter study was conducted, in which 61 nephropathic cystinosis patients on cysteamine therapy were recruited from three European reference centers over the period from October 2015 to January 2018. At baseline, relevant history, clinical and laboratory data regarding the clinical course of cystinosis was collected and during each visit, clinical follow-up data and plasma samples were collected for the assessment of the chitotriosidase enzyme activity.

Results

For each included patient, 2-9 visits were recorded (mean 4.7 ± 2.5). Average follow up duration for recruited patients was 19.3 ± 6.5 months. In a multivariate linear regression analysis of the longitudinal data, chitotriosidase enzyme activity resulted as the only predictor of LCL for all recruited patients. In the cystinosis patients prior to kidney transplantation, chitotriosidase enzyme activity resulted as a significant predictor for kidney function (serum creatinine), in contrast to LCL. Although in adult kidney-transplanted patients a significant correlation could be demonstrated between chitotriosidase and an extra-renal complications score, LCL resulted as a better predictor of extra-renal organ involvement.

Conclusion

Chitotriosidase enzyme activity could serve as a novel alternative biomarker for the therapeutic monitoring of nephropathic cystinosis.

P103.

Glomerular developmental delay in the preterm neonatal rabbit

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Background/Aims

In this study we investigate the effect of preterm birth on postnatal nephrogenesis in preterm neonatal rabbits compared to term. We hypothesize that preterm birth leads to an altered glomerular development in rabbits.

Methods

For this, we analyzed renal morphology, glomerular maturity and functional parameters in different cohorts or rabbit pups: preterm (G28), preterm at day 7 of life (G28+7) and term at day 4 of life (G31+4).

Results

We found no significant differences in kidney volume and weight, and relative kidney volume between the cohorts. Nephrogenic zone width increased significantly over time when comparing G31 + 4 to G28. The renal corpuscle surface area, in the inner cortex and outer cortex, tended to decrease significantly after birth in both preterm and term groups. With regard to glomerular maturity, we found that the kidneys in the preterm cohorts were still in an immature state (presence of vesicles and capillary loop stage). No significant differences at functional level were found.

Conclusion

In conclusion, the results of this study show that the preterm rabbit tends to linger in the immature glomerular stages compared to term but with no functional repercussion in the short term.

P104.

Epidemiology of moderate to severe dyskalemia in a paediatric emergency department

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Introduction

The epidemiology of Potassium abnormalities in the paediatric emergency department (ED) is unknown. The purpose of our study is to identify categories of patients in ED in which dyskalemia occur, their causes and consequences or prognosis.

Methods

Retrospective case control study of all the patients admitted to a single-centre tertiary emergency department over a 30 months period. We compared patients with hypokalaemia (< 3.0 Meq/L), patients with hyperkalaemia (> 6.0 Meq/L) to a normal randomized population with K+ levels between 3.5 and 5 Meq/L, recruited for a 3 to 1 ratio.

Results

Between January 1, 2013 and August 31, 2016, 108209 patients were admitted in our ED. Globally 9342 blood sample with K+ measurements of which 60 hypokalaemia (2.8 ±0.2 Meq/L) and 55 hyperkalaemia (6.4 ± 0.6 Meq/L) were identified. 200 patients with normokalaemia were recruited (4.1 ± 0.3 Meq/L). Main causes were : Inferior respiratory tract infection (23%) and fracture (15%) for hypokalaemia, Inferior respiratory tract (21.8%) and ENT infection (20.0%) for hyperkalaemia. Compared with the controls, patients with hyperkalaemia had an higher creatinine (0.72 ± 1.6 vs 0.40± 0.16, mg/dL, p<0.0001) with lower HCO3- (19.4 ± 3.8 vs 21.8 ± 2.8 mmol/L, , p=0.0001). Patient with hypokalaemia had an higher creatinine (0.66 ± 0.71 vs 0.40± 0.16 mg/dL, p<0.0001). We didn't observe significant differences in pH, PCO2, Base excess and Lactate, nor in de mean duration of hospitalization in general and PICU wards or in the PIM and the PRISM scores.

Dyskalemia is rare in ED patients: 0.64% for hypokalaemia and 0.58% for hyperkalaemia. We observed in all patients a mild degree of renal failure that could be the causes of the K+ disturbances and could be explained by volemic disturbances. The main mechanism could be dehydratation due to digestive losses, polypnea in young patients or poor intake frequently associated with respiratory insufficiency. Feeding or crystalloid infusion easily solve all these ionic anomalies.

Conclusion

Dyskalemia is a rare in ED patients and easily resolved by feeding or perfusion. The plausible mechanism is a transient volemic disturbance typically seen in the pediatric population. Dyskalemia is not predictive of a poor evolution in the paediatric ED population.

P105.

The many faces of atypical Hemolytic Uremic Syndrome (aHUS) in Belgium

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Aims

The Belgian section of the aHUS registry (NCT01522183, sponsored by Alexion, Inc.) constitutes the largest local database available with 78 patients registered as of APR/1/2018. We, herein, report on local disease features.

Methods

All consenting patients with a clinical diagnosis of aHUS and an ADAMTS13 activity >5% (if performed) were eligible. Patients were recruited between 09/2013 and 03/2018 through Belgian academic centers. 'Pediatric' patients were defined as younger than 18 y at enrolment. Results are expressed as % or median and range.

Results

Epidemiology. The registry counts 18% of children (n=14). Median ages at disease onset and diagnosis are 3.1 [0.1, 15] and 3.2 [0.1, 15] y in children and 36 [1.2, 76] and 36 [1.2, 78] in adults. Overall median age at disease onset is 28 y [0.1, 76]. A positive familial history is found in 22%. One third (n=26) of the patients are being treated for end-stage kidney disease (ESKD) (n=3/14 are dialyzed and 1/14 transplanted among children vs. n=9/64 and 13/64 among adults).

Causes of aHUS. A genetic cause of aHUS is identified in 42% (n=33 - CFH in 13 adults + 2 children / 24% among those tested; CD46 in 8+3 / 19%; DGKe in 1+2 /11%; C3 in 4+0 / 9%; CFI in 2+0 / 4%; CFB 1+0 / 3%; THBM in 0+1 / 3% and combination in 3+1 /5%). No mutation is found in 22 patients despite at least 5 genes have been tested. Seven patients (6+1) displayed anti-CFH autoantibodies (12%). Finally, an underlying condition is mentioned in 19% (n=15: malignant hypertension (4), infections (6), systemic lupus erythematosus (1), disseminated intravascular coagulation (1), malignancy (1) or others (2)). CFH variants constitute the most prevalent mutations in ESKD (n=8/26).

Treatment by eculizumab. As of APR/1/2018, 57% of children (n=8/14) and 44% of adults (n=28/64) have already been treated by eculizumab. Five children and 18 adults are still treated. One meningoencephalitis caused by N. meningitidis (serogroup Y) occurred in a transplanted child vaccinated 5 years earlier and taking penicillin as antibiotic prophylaxis. He eventually recovered without sequel.

Conclusion

aHUS is a severe disease concerning both pediatric and adult populations. The wide variety of etiologies found advocates for a large panel of causes to be investigated in each patient with aHUS. Disease genetic background appears quite similar to that of the French cohort but, in contrast with the last one, one Thrombomodulin-related case of aHUS is also reported.

P106.

The effect of Urotherapy in group during youth camp

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Background/aims

Urotherapy is often a successful treatment for daytime urinary incontinence, but the benefit in nocturnal enuresis is unknown. During a voiding youth camp for children with enuresis, standard urotherapy was applied, including standardized fluid intake. The aim of this study was to evaluate the effect on voiding volumes, nocturnal diuresis and achieving continence.

Methods

Data of 161 children (age 8-12 years) who participate in a voiding youth camp were retrospectively evaluated. Voiding volumes and fluid intake were registered during this camp. The two-way ANOVA test was used to investigate if there is a difference between the maximum voided volume (MVV) and/or nocturnal diuresis on the first and last day of camp. Follow-up was provided 6 and 12 months after the camp.

Results

The study sample consists of 49 (30%) girls and 112 (70%) boys. At the entry of the camp, 93 (78%) children had non-monosymptomatic nocturnal enuresis, meaning nighttime-incontinence including lower-urinary tract symptoms at daytime. The children were divided into 2 groups, based on MVV below or above 65% of the expected bladder capacity for age. Children with a small bladder capacity for age demonstrated a mean increase of 51 ml (CI [17,85]) voided volume during the camp, where the group without small bladder capacity for age showed a mean decrease of 14 ml CI [-31,3]. The difference between the two groups was statistically significant (p=0.001). The same test was applied for the difference in nocturnal diuresis. The children with polyuria on day one demonstrated a mean decrease of 148 ml (CI [-224, -71]) nocturnal diuresis during the camp, where the group without polyuria showed a mean increase of 70 ml CI [39,101]. The difference between the two groups was statistically significant (p=0.001). The youth camp also resulted in an increase of dryness. At 6 months and 1-year follow-up respectively 21% and 31% of the children were dry. In addition, respectively 30% and 28% of the children showed a partial response.

Conclusion

In children with small bladder capacity for age, the MVV increases during the youth camp. Since no other therapy as applied, this is most likely due to correction of fluid intake. Also, a decrease of the nocturnal diuresis was observed in children with polyuria. Urotherapy, applied during a youth camp, seems to contribute for achieving dryness if the correct fluid advice is adapted to the underlying cause of enuresis.

P107.

Risk factors for recurrent kidney stones in children

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Purpose

Despite a growing incidence of nephrolithiasis in children and its high recurrence rates, there is a paucity of data on the factors contributing to the recurrence of kidney stones. The purpose of this study is to identify risk factors associated with recurrent kidney stones in children followed at a Belgian tertiary centre.

Methods

Medical records of children who were treated and followed for urinary stone disease were retrospectively reviewed. Children with the first episode of nephrolithiasis between 1998 and 2016, followed at the Ghent University Hospital and at least one year follow-up were included. We analysed whether patient characteristics, past medical history, presenting symptoms, the results of laboratory investigations and the applied management strategy were associated with a risk of kidney stone recurrence. The risk of stone recurrence was evaluated by univariate and multivariate analysis.

Results

97 children were eventually included in the analysis. 33 (34%) of them presented with at least one episode of stone recurrence. Body mass index (BMI) above the 85th percentile and asymptomatic stones at the initial presentation were associated with a lower risk of recurrent stones (p=0.020 and p=0.017). In contrast, immobility resulted in a 10 times higher risk (p=0.002, 95% CI 1.968-50.005) and the need for technical treatment was associated with a 3.2 times higher risk (p=0.017, 95% CI 1.297-8.084) of developing recurrent stones.

Conclusion

There was a higher risk of recurrent stones in immobile patients and those who required technical intervention at initial presentation in contrast to patients with a high BMI or asymptomatic presentation, which appears to be a protective factor against the development of recurrent stones. These data can be used to identify children who require an intense follow-up after the 1st episode of nephrolithiasis.

P108.

Dental abnormalities and nephrocalcinosis: A case report of a boy with Enamel-Renal syndrome

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Background/aim

Enamel-Renal syndrome (ERS) is a rare disease, characterized by dental abnormalities in combination with nephrocalcinosis. Since the dental problems are more prominent, patients are initially followed-up by a dentist with a higher risk of delay in the diagnosis/treatment of the associated kidney disease. Alternatively, there is little awareness under MDs concerning the importance of potential dental abnormalities in patients with nephrolithiasis/calcinosis. Our aim is to improve this and also discuss the management.

Methods

We present a 9-year old boy with a consanguineous, North-African, background who was referred to the dept. of Pediatric Nephrology by the dentist because of amelogenesis imperfecta without specific urinary complaints. Two parental sisters also had dental abnormalities, one in combination with kidney stones.

Results

Renal ultrasound demonstrated bilateral medullary nephrocalcinosis. Blood analysis demonstrated an eGFR of 89.9 ml/min/1.73m², without further abnormalities. Urine analysis showed a reduced calcium excretion and a low citric acid/creatinine ratio. Genetic analysis demonstrated a homozygous mutation in the FAM20 gen (c.1369A>T (p.Lys457Ter) in exon 11), confirming the diagnosis of ERS

Conclusion

Nephrocalcinosis in ERS can be asymptomatic and silently progress during childhood, leading to symptomatic stone disease and renal failure at a later age. Early recognition (by dentist and medical doctors) and further treatment, can help in preventing the renal complications of ERS. The exact pathogenesis of the enamel defects and nephrocalcinosis is unknown. FAM20A plays a role in amelogenesis and has been demonstrated to have inhibitory effects on biomineralization. Patients frequently have hypocalciuria and hypocitraturia, the latter of which can contribute to an accelerated growth of calcium oxalate cristals. Therefore, next to increased fluid intake, potassium citrate can be added to the preventive treatment approach.

P109.

ADPedKD: A Global Platform on managing childhood Autosomal Dominant Polycystic Kidney Disease

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Objective

Although literature on Autosomal Dominant Polycystic Kidney Disease (ADPKD) is merging over the last decades, data on paediatric ADPKD are still scarce. Moreover, unified diagnostic, follow-up and treatment approaches regarding modifiable disease factors in order to slow down disease progression are currently lacking for ADPKD children. We therefore aim to build a global multicentre cohort, named 'ADPedKD', from which we will generate data on the incidence and presentation of ADPKD in childhood. Furthermore, we intent to define a paediatric patient risk stratification score, after identification of progression factors.

Methods

Patients diagnosed with ADPKD, based on genetic analysis or a positive familial history and imaging, before the age of 19 years are eligible for inclusion in this observational register study. Ethical committee approval is organized per country as regulations differ internationally. After written informed consent, clinical patient data are pseudonymously introduced in our secured web-based database (www.adpedkd.org), both retrospectively and prospectively, from all participating centres throughout the world. In Belgium, 10 paediatric nephrology departments are involved in the ADPedKD initiative. Our main focus consists of the initial presentation, pre- and perinatal history, genetic analysis, renal function and longitudinal follow-up. Also, we will be able to generate a Belgian consensus based on the current clinical practice, and compare this with those in other European and non-European countries.

Results

We currently have 66 centres registered in ADPedKD, from 27 different countries: Albania, Belgium, Brazil, China, Czech Republic, Egypt, France, Germany, Greece, Iran, Ireland, Italy, Lithuania, Luxembourg, Poland, Portugal, Romania, Russian Federation, Saudi Arabia, Serbia, Singapore, Spain, Switzerland, the Netherlands, Turkey, United Arab Emirates and United Kingdom. This resulted until now in 334 registered paediatric ADPKD patients.

Conclusion

The ADPedKD initiative is the first and the largest international paediatric ADPKD registry that will provide evidence for a paediatric patient risk stratification scoring system from an early disease stage.

O12.

Three cases of acute flaccid myelitis, a new epidemic?

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Background

Acute flaccid myelitis (AFM) is a polio-like neurologic condition and a specific form of acute flaccid paralysis.

Cases

We present 3 cases of AFM in children aged 1 to 3. At initial presentation, differential diagnosis was made with transverse myelitis, Guillain-Barré Syndrome, ADEM, stroke or central infection. Diagnosis of AFM was made based on typical clinical pattern of paresis, hyperintense spinal cord lesions in gray matter and nerve roots at MRI, and pleocytosis in CSV. All of them were negative for extensive additional tests including auto-antibodies. Patients were treated with high doses corticosteroids, plasmapheresis and/or IVIG. During follow-up, a physical and occupational rehabilitation program was started, but until now not or only partial recovery was reached. The etiology of all two of three cases is still open to debate; in one patient an enterovirus D-68 was detected in nasopharyngeal specimen, in another a rhinovirus was isolated and in the third case no pathogen was isolated.

Discussion

In recent years, epidemically hundreds of cases of AFM were identified worldwide, as well in the USA, Europe, Canada as in Japan. The incidence of <1:1.000.000 may be an underestimation because of underregistration and subclinical cases. By the Centers for Disease Control and Prevention (CDC) in the USA, AFM is defined by acute onset of asymmetric multifocal limb weakness and evident spinal cord lesions on MRI. Enterovirus D68 is most frequently associated with AFM. The pattern of tropism for spinal cord motor neurons is clinically, radiographically and electrophysiologically similar to other infectious acute flaccid paralysis syndromes suggesting a common pathophysiology. However, until now the pathophysiology of AFM is incompletely understood, and nowadays no trial data are available to guide therapeutic recommendations. In her expert opinion paper, the CDC discouraged the use of corticosteroids and plasmapheresis in AFM. Therefore, IVIG may be the safest available therapy with potential efficacy if given very early in the course of the disease.

Conclusion

Be aware of a new epidemic or single cases of acute flaccid myelitis. Quickly determining the diagnosis is important because of selecting therapy.

OP18.

Deformational plagiocephaly as a marker for developmental delay: a systematic review

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Background/Aims

Deformational plagiocephaly (DP) and brachycephaly (included in the definition of DP) have become more frequent since the 'back to sleep' campaign was launched in 1992. Many caregivers are concerned that this has had an influence on the child's development as some studies have shown an association between DP and developmental delay. This literature study summarizes the available evidence.

Methods

Three major databases (MEDLINE, EMBASE and CINAHL) were systematically searched in October 2018 for plagiocephaly and developmental delay with relevant search strings. Relevant studies were selected based on specific exclusion and inclusion criteria. We rated each selected study on the basis of its methodological quality using a validated scoring system.

Results

1,276 articles were found following a literature search (after the exclusion of duplicates). 25 final articles were selected on the basis of a number of exclusion and inclusion criteria. The quality of the studies was restricted due to confounding and bias such as selection bias. Most studies had a moderate to high methodological quality. 17 studies showed a positive association between DP and developmental delay (including 4 studies of a high quality). 8 studies found no-association (including one study of high quality).

Conclusion

There appears to be an association between DP and developmental delay in children younger than 2 years old. DP can therefore be seen as a risk factor for developmental delay. Follow up and monitoring is needed in children with DP. Future studies should focus more on older children to investigate whether such delay persists.

OP19.

Salbutamol - not only in asthma

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Case report

A baby born from parents with no particular history, at a gestational age of 38 weeks, presents at birth with generalized hypotonia, ineffective sucking and oxygen desaturations. The child has weak reflexes, ophthalmoplegia but presents with a good contact. The newborn is hospitalized in a neonatal intensive care unit where an infectious and genetic assessment (Prader-Willi, Werdnig-Hoffman, Steinert) turns out to be negative. He is discharged home at day 31. However, at day 33, he presents again with a respiratory distress requiring treatment in intensive care. The severity of his condition is such that a cessation of care is discussed. Considering the possibility of a congenital myasthenic syndrome evoked by the ophthalmoplegia, Salbutamol is tried empirically. In 48 hours, food autonomy is recovered, the child is weaned off invasive ventilation, and a spontaneous mobility appears. In the absence of evidence of mutation in the main genes associated with congenital myasthenia, a muscle biopsy is performed at the age of 9 months and highlights only nonspecific abnormalities. At the age of 15 months, the test of a panel of genes known as associated with muscular diseases reveals a heterozygous composite mutation of RYR (c.1559T>C inherited from the mother, and a mutation c.10627-2A>G inherited from the father). Aged 4 years and still on treatment, the child presents with a normal neurological examination except for an opthalmoplegia.

Discussion

The RYR1 gene codes for the ryanodine 1 receptor, a calcium channel inserted into the sarcoplasmic reticulum membrane, allowing the calcium ions to be released into the cytoplasm. Dominant or recessive mutations give rise to various forms of congenital myopathy with variable phenotypic and pathological spectrum; the recessive forms being frequently more severe. Two studies have reported a potential efficacy of salbutamol in congenital multiminicore myopathy, which can be caused by recessive RYR mutations. However, this treatment is not routinely used in this pathology. Salbutamol has already proved its value in congenital myasthenic syndromes as well as in myotubular myopathies and its efficacy is suspected in spinal muscular atrophy.

Conclusion

We report here a positive and rapid evolution on salbutamol of the symptoms of RYR-related congenital myopathy.

OP20.

Scurvy: a forgotten disease. Case report of a 3 year old boy with progressive hypotonia and a very restrictive diet

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Background/Aims

In our developed countries a severe vitamin C deficiency, also known as Scurvy is a rare diagnosis. The disease can present with a broad spectrum of symptoms and often multiple invasive and unnecessary investigations are done before the correct diagnosis is set. By presenting a case of scurvy in a 3 year old autistic boy, we want to raise awareness for a disease that can be easily diagnosed and threaded, if considered in the differential diagnosis.

Methods

A tree year old boy known with a autism spectrum disorder presented on the emergency ward because of progressive muscle weakness and refusal to walk since tree weeks. In the previous weeks he was seen tree times on the pediatric consultation and the emergency ward of a smaller general hospital. Radiologic imaging of the lower extremities and ultrasound of the hip showed no abnormalities. Computer tomography of the skull was normal. He was seen by a pediatric neurologist and transferred to our centre for further investigation.

He presented with a progressive muscle weakness with disability to walk and rise from a seating position. There was no previous infection or trauma. There was no complaining of pain. Defecation was always difficult. His dietary intake was restricted to chocolate milk, bread with chocolade paste, cookies and a limited intake of vegetable soup. Medical history was negative despite a delayed psychomotor development because of severe autism.

Clinical examination showed a subtle symmetrical diminishing of muscle strenght with an inability to walk and normal tendon reflexes, normal cranial nerves, no altered behavior and the presence of a sacral hematoma and an anal fissure.

Biochemical analysis showed a ferriprive anemia and raised sedimentation. At first a differential diagnosis of several central neurologic inflammatory or auto-immune diseases (Guillain Barré disease, acute disseminated encephalomyelitis …) was made. MRI of the brain and spine showed a subacute subdural hematoma. No signs of infection or inflammation were seen on the lumbar punction.

Because of important constipation with the suspect of a mega rectum, a antegrade colonic lavage was performed. Maintenance treatment with macrogol was started during hospitalization.

The presence of an unexplained neurological condition, subdural hematoma and an anal fissure in a child that could not express himself raised the suspicion of possible child abuse. Therefore a full radiologic examination of the bones was performed, showing multiple atraumatic fractures in a suspected osteopenic bone.

The case of the boy was discussed with a pediatric psychiatrist and passed on to ‘vertrouwenscentrum kindermishandeling’, a trust centre for children with suspected child abuse. An official investigation by the court was initiated.

Despite the fractures, the radiographic examination showed a hypodensity located on the metaphyse of multiple bones. This leaded to a broader differential diagnosis with metabolic diseases, vitamin C deficiency, multifocal osteopmyelitis, cupper deficiency, hypervitaminose A, rickets, leukemia, Menkes syndrome.

Broad blood samples showing low alkaline phosphate, PTH and vitamin D, normalized after administration of vitamin D. There was a severe ferriprive anemia. Iron supplementation was initiated. Peripheral blood smear showed no blastosis. Sedimentation was normal when repeated. High D-dimer count was seen. Coagulation parameters where normale, dispite a lower von willebrand factor antigen. Metabolic screening was initiated for mucopolysacharidose, organisch
acidurie, mucolipidosis, Wilsons disease and osteogenesis imperfecta. Ophthalmologic screening was normal. A bone scan showed no lesions.
Feeding and intake were evaluated by a dietician, showing a limited and insufficient intake of vitamin E, C, D, A, B6 and sink. Motoric stimulation and physiotherapy was done on a daily basis.
Important improvement was seen after start of vitamin supplementation. We administered a dose of 120mg vitamin C, daily.
Result of an indetectable low vitamin C and the rapid clinical respons on the supplementation cofirmed our finale diagnosis of a severe vitamin C deficiency. Follow up on pediatric hematologie, gastro-enterologie and general pediatric was provided.

Results

A diagnosis of scurvy or a severe vitamin C deficiency was made in a boy with a autism spectrum disease and a very restricted diet. He presented with progressive muscle weakness and suspicion of bone pain and disability to walk. There was an important ferriprive anemia based on inadequate iron absorption in the gut. Increased fragility of the blood vessels caused an increased tendency to bleed. There were multiple nosebleeds, gumbleedings and a subdural hematoma. Multiple bone lesions, bone demineralization en atraumatic fractures were seen on radiographic exams.
A rapid recuperation of muscle tone and regain of function was seen, when vitamin C supplementation was started. Full recuperation is expected.

Conclusion

Scruvy is a rare disease that can present with multiple and severe symptoms, but when recognized, is it easy to diagnose and thread. Therefore we advise to have a low threshold to screen for vitamin deficiencies in children with a restrictive diet.

P110.

Not just a 'simple' myelomeningocele; A case report of a polymalformative syndrome

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Background

A 39 year old woman (G8P2) was referred to our hospital at 18 weeks gestational age. During prenatal screening the baby was diagnosed with a myelomeningocele. The parents were consanguine and refused further genetic testing. A MRI at 34 4/7 weeks GA showed a triventricular hydrocephalus with a type 2 Arnold Chiari malformation, a myelomeningocele starting from L1, a hemivertebra at L3, a fusion of apophyses between L2-L3 and L4-L5 and an unilateral clubfoot right. The boy was born at 36 2/7 weeks GA with a birthweight of 3020gr (p50-75), length of 43.5cm (P3-10) and head circumference of 34.5cm (P75-90).

Results

After birth there's a short-term need for respiratory support. The physical exam showed a myelomeningocele covered with a thin translucent membrane and paralysis of both legs.
The parents agreed with further investigations and genetic testing.
Cranial ultrasound confirmed a hydrocephaly, without other anomalies. Cardiac ultrasound was normal. The chest radiography showed an elevation of the right diaphragm and a bifid right collarbone. Abdominal ultrasound revealed the presence of two biliary vesicles.
At day 1 the myelomeningocele was closed. At day 2 the patient deteriorated due to a pleural effusion for which a chest drain was placed. A ventriculoperitoneal shunt was placed at the same day. Further imaging showed an anal atresia with fistula between the descending colon and the bladder for which a colostomy was placed. Cystography showed no passage of contrast. The fistula was closed at day 14. Anoplasty was impossible because of lack of space between the pubic symphysis and coccyx.

Discussion and conclusion

Spina bifida is believed to be caused by a combination of genetic and environmental factors. Several candidate genes and syndromes like Curranino syndrome and Caudal Regression syndrome are associated with a myelomeningocele. However, looking at the phenotype of our patient, it does not match with the other syndromes described before. The array CHG revealed a microdeletion in the CNTNAP2 gene, known for the Pitt-Hopkins-Like syndrome. This genetic defect does not correspond to the clinical phenotype. We are currently waiting for the results of the mendeliome.

P111.

A case of intracranial bleeding after a vaginal forceps delivery

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Introduction

Approximately 2 percent of vaginal deliveries are marked by a fetal injury. The risk increases with: macrosomia (when fetus weight exceeds 4000g), fetus abnormal presentation (especially breech-birth) or the use of obstetrical instrumentation during delivery.

The spectrum of birth injuries is wide, ranging from minor lesions such as (hematomas, lacerations), to more severe and complex lesions such as in depressed skull fractures. Complex lesions may lead to surgery.

Methods

We report the case of a boy born by vaginal breech-delivery in 2018 at Clinique Saint Jean in Brussels.

Results

He was born at 38 weeks and 4 days of gestation with a birth weight of 3,17 Kg. During delivery, his head wedged in the small diameter of the superior strait of the pelvis. A LOVSETT forceps was used for extraction. At birth, the boy presented a left frontoparietal skull depression of 3 cm x 5cm, but his neurological examination was normal. A cerebral CT-scanner was made a few hours later. Computed tomography showed a depressed skull fracture and intracranial lesions: an extradural and intradural hematoma. At less than 24 hours of life, the boy presented seizures due to his intracranial lesions. The boy benefited from neurosurgical intervention on the second day of life.

Conclusion

Depressed skull fractures may be associated with forceps deliveries and present a greater risk of intracranial bleeding. Therefore, Computed Tomography is needed.

With intracranial lesions or skull depression greater than 1 cm, neurosurgical advice should be obtained promptly as these lesions often require surgical intervention. On the other hand, with linear skull fracture and no evidence of intracranial process, conservative treatment and observation is recommended.

P112.

Flaccid paralysis of the lower limbs of subacute onset in the context of respiratory infection

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Introduction

Most respiratory diseases in children are of viral origin and limited, although sometimes very severe and progressing to respiratory distress. Such infection may also be responsible for central nervous system involvement such as encephalitis, myelitis or polyneuritis.

Case report

DE, a 11-month-old girl, was first seen at the emergency room for coughing for 3 weeks and beginning fever. No neurological trouble has been noticed. She was discharged home with treatment for bronchitis and otitis. Two days later, she was admitted again because of progressive loss of spontaneous movements of the lower limbs. Clinical examination revealed an extensive flaccid paralysis of the lower limbs with abolition of reflexes. Because of predominant abdominal ventilation she was transferred to pediatric intensive care unit.
All biological testings including extensive serologies were normal as well as nasopharyngeal aspiration. Chest X-ray showed central bronchopathy. Cerebrospinal fluid examination, including cultures and PCR, was normal. Cerebral and spinal magnetic resonance imaging revealed a hypersignal extending from C3 to T4 compatible with a diagnosis of acute transverse myelitis. Additional stool analysis showed the presence of enterovirus viral RNA. She was then placed on high dose of intravenous (IV) prednisone 30 mg/kg/day followed by a 10-day oral degressive regimen. Soon after the initiation of such treatment she became more alert. She also recovered reflexes as well as some lower limb movement and better sitting position. Six weeks later she continued to recover and starts to stand upright.

Discussion

Enterovirus infection, especially due to D68 (EV68) in children may cause very severe respiratory illnesses associated to bad cough and fever. This polio-like virus is also a neurotropic agent that may cause acute flaccid myelitis (AFM) through inflammation and destruction of the spinal cord anterior horn cells. It is usually preceded by respiratory symptoms in the majority of cases. Several cases of EV68 associated AFM have been already described. Treatment should be prompt and may include high dosage regimen of prednisone, IV immunoglobulin and but less frequent plasma exchange.

Conclusion

This case reminds us that clinical evaluation and follow-up should be rigorous and global even in cases of acute respiratory diseases. Neurological complications requiring special attention and aggressive treatment may also occur subacutely.

P113.

Parafalcine subdural empyema - an uncommon complication after tooth extraction: a case report and literature review

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Background/Aims

Subdural empyema (SE) is an uncommon important intracranial infection. A parafalcine localization is described in less than 10-20% of SE's and has a worse prognosis. Most patients have non-specific symptoms such as a headache, fever and vomiting. This makes diagnosis challenging. In case of preexisting sinusitis, symptoms are even more subtle. We want to raise awareness about this clinical picture as an early treatment is important to prevent life threatening complications.

Methods

The clinical, biochemical and radiographic data of a 15-year-old boy with a parafalcine SE as a complication after tooth extraction are presented.

Results

A few weeks after tooth extraction a 15-year old boy had a headache with altered level of consciousness, a right hemiparesis and fever. He had a leukocytosis and raised C-reactive protein. Magnetic resonance imaging scan (MRI) showed a parafalcine SE on the left side. MRI with gadolinium enhancement is the gold standard for this diagnosis. In this boy the parafalcine SE was caused by a breakthrough sinusitis of odontogenic nature. Other causes include head trauma, otitis media, osteomyelitis of the skull and meningitis. The management of SE necessitates a multidisciplinary approach. In our case intravenous broad-spectrum antibiotics were started and functional endoscopic sinus surgery and trepanation were performed. Adjuvant therapy consisted of preemptive anticonvulsants and corticosteroids. Cultures of the empyema were positive for Streptococcus anginosus. In 67% the cultures are positive for bacteria of the Streptococcus milleri group. Depending on the clinical picture, intravenously antibiotics should be continued for two weeks, followed by six weeks of oral antibiotics. Progressively the hemiparesis regressed and neurological examination at discharge was normal. As was MRI six months later. Nevertheless, neurologic sequelae are common. Morbidity rate is 33% after six months and mortality rate is 5-10%. Factors defining outcome include the neurologic status at admission, time to diagnosis and treatment, a parafalcine localization, patient age, extent of the collection and underlining immunodeficiency.

Conclusion

Parafalcine SE is an uncommon complication after tooth extraction. Symptoms and laboratory changes can be subtle, which makes the diagnosis challenging. Nevertheless, an early multidisciplinary approach is important to prevent complications and determine the outcome.

P114.

When a heart murmur is hiding a rare genetic disease

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Background / aim

Infantile Pompe disease is a genetic disease with an incidence of 1 per 40 000 individuals. It should therefore be considered when hypertrophic cardiomyopathy is detected in newborns or infants. This disease is characterized by a glycogen overload in lysosomes (glycogenosis type II) due to the absence of an enzyme called acid alpha-glucosidase (GAA). This glycogen overload causes progressive muscle weakness with cardiac consequence that would be fatal in the absence of an enzymatic treatment.

Case report

We present the case of a 4 months-old girl, born full term, in which a non-obstructive hypertrophic cardiomyopathy was highlighted by the echocardiography, initially performed to assess a heart murmur. Complementary analyses also revealed a liver injury with increase of TGO / TGP and a myolysis as well as its weight curve showed a posteriori a break from the age of 2 months. A little after she was hospitalized in the intensive care unit for complete left lung atelectasia due to her cardiomegaly and muscular hypotonia.

Enzyme analysis confirmed Pompe disease four weeks after the detection of the cardiopathy (zero activity of de GAA enzyme ; heterozygous status for the c.854C>G, p.Pro285Arg and the c.2331+2T>A, p.? mutation in the GAA gene). Before starting the enzyme replacement therapy (Myozyme), her cross-reactive immunologic material (CRIM) status was assessed which showed a CRIM positive status. To reduce the risk of immune response against ERT, immunomodulation was performed with transient low dose methotrexate before the first three courses of Myozyme, prescribed at the dose of 20 mg/kg every other weeks.

Conclusion

Targeted genetic analysis performed on newborns affected by a cardiomyopathy could prevent the deleterious effects of rare genetic disease such as Pompe disease. In our case, the patient benefitted from a treatment involving a recombinant enzyme which was administered according to the patient's CRIM status. This early treatment gave encouraging first clinical results.

P115.

An intracranial haemorrhage in a febrile newborn: About a case

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Background

When presenting with intracranial haemorrhage the onset of symptoms is typically acute. Symptoms and clinical signs usually reflect the intracranial hypertension and cerebral or meningeal irritation.

Clinical case

A 15 days old newborn was admitted in the emergency department with recent onset fever, gastro-intestinal irritability and vomiting. Clinical examination was reassuring without any neurological or infectious focus. A complete biological and bacteriological screening was realised in the emergency department which showed mild inflammatory parameters. The lumbar puncture was hemorrhagic during the whole tap. Broad spectrum antibiotics, a protein hydrolysate formula and anti-reflux therapy were initiated by hospitalisation. Facing with persisting fever after 5 days of antibiotics and no bacterial of viral pathogen found, another aetiology than the infectious one was suspected.

The hemorrhagic appearance of the lumbar puncture leaded to the realisation of a transfontanellar ultrasound which showed a intraventricular haemorrhage. MRI confirmed a susependymal haemorrhage extending into the cavity of the cavum vergae, into the ventricular system, the cerebral tissue and the pericerebral space. Its aetiology remains unknown and tests are still running. Because of the risk of hydrocephalus, a clinical and radiological follow-up was initially organised but the child was hospitalised again a few weeks later with confirmed hydrocephalus. A ventriculoperitoneal shunt was placed a few days ago.

Discussion

The management of a febrile newborn usually includes the realisation of a lumbar puncture to rule out any central nervous system infection.

Another aetiology than the infectious one should be considered when facing with persisting fever and a hemorrhagic appearance of the CSF because these can be the only signs of meningeal haemorrhage.

In our case radiological imaging showed a susependymal haemorrhage extending into the cavum vergae, a residual embryonic cavity persisting only in 3% of term newborn.

Conclusion

Persisting fever and a hemorrhagic appearance of the CSF in a febrile newborn can sometimes be the only signs to suspect, other than the obvious infectious aetiology, a intracranial haemorrhage.

P116.

Miller Fisher-syndrome, an atypical presentation of Guillain-Barré syndrome

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Background

Miller Fisher-syndrome (MFS) is an atypical variant of the Guillain-Barré syndrome (GBS) in which the triad of the acute onset of ophtalmoplegia, ataxia and areflexia is present. The triad may be present in isolation or may occur along with features of more typical GBS. They are likely to have antibodies directed against GQIb ganglioside detectable in serum and an elevation in cerebrospinal fluid protein concentration.

Methods

Case report based on data from the electronic medical records.

Results

A 3,5 year old girl with a past history of respiratory infections and tympanostomy tubes presented with her parents for evaluation of headache, anorexia and dysarthria since 2 days. 10 days prior to the consultation, she had a respiratory tract infection with fever for which she was treated with Azithromycin during 3 days. She was admitted to hospital for observation of the speech and headache and treatment with intravenous analgesia was started. Headache continued and on day 2 brudzinski's sign was positive. Cranial CT and lumbar puncture were both normal, except for signs of pansinusitis on CT. Further deterioration with ataxia and dysphasia was seen on day 5. A head MRI was performed and was normal, 2nd lumbar puncture showed elevation of proteins. Ganglioside antibodies were negative. On day 6, clinical examination revealed disease progression with fecal incontinence, absent sphincter reflex, absent reflexes in the lower extremities, dysphagia and right facial nerve paralysis. NMR spine on day 7 showed Miller Fisher. Intravenous immunoglobulin therapy was started at 400mg/kg/day during 5 days with improvement of the neurological symptoms. Multidisciplinary revalidation program was initiated.

Conclusion

MFS is a rare subtype of GBS, characterized by the triad of acute onset of ataxia, ophtalmoparesis and areflexia.

P117.

An infant with a cortical malformation syndrome; A new de novo mutation in the tubulin gene TUBB2A

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Background

An infant at the age of 4 months came for a second opinion. She had a failure to thrive and excessive crying from the age of 1 month old. Initially gastroesophageal reflux disease and infantile cramps were considered, but the treatment made no difference. She remained uncomfortable with episodes of hypertonia and opisthotonos. She had already signs of delayed social and motor skills. Physical exam showed some mild dysmorphic features. Cardiac ultrasound was done because of a laryngomalacia, but there was no vascular sling seen, only a patent foramen ovale.

Laboratory research showed persistent elevated aminotransferases, further metabolic research was negative. The brain MRI revealed a simplified cortical pattern, pachygyria, dysgenesis of the corpus callosum and dysplastic basal ganglia. We therefore decided to do a 'malformations of cortical development' gene panel. At the age of 7 months she had epileptic seizures and diagnosed with West syndrome.

Methods

DNA was extracted using standard protocols, after informed consent of the parents. The analysis was performed using gene panel analysis. This analysis was performed at the Center of Medical Genetics, UZ Brussel in collaboration with the BRIGHTcore-facility unit according to standard procedures.

Results

A disordered gyral pattern in combination with other abnormalities of the cerebellum or brainstem together with lissencephaly, pachygyria or polymicrogyria is characteristic for a tubulinopathy. Microtubules play an important role in almost every cell. They are essential during the cortical development. They are highly expressed during the brain formation, suggesting they play a role in neurogenesis, neuronal migration and post migration organization. In our patient we found a variant in the beta tubulin 2A gene (TUBB2A): c.146T>G, p.(Val49Gly) (Refseq: NM_001069.2). So far only 7 case reports have been published regarding variants in TUBB2A. This missense alteration has never been described before. Heterozygous variants in TUBB2A are causing 'Complex cortical dysplasia with brain malformations'. Further analyses of the parental DNA revealed that it concerns a de novo variant in our patient.

Conclusion

An infant with a failure to thrive and a clinical suspicion of a tubulinopathy based on MRI images was confirmed with genetic analysis. A de novo missense variant in the TUBB2A was found. This is associated with a severe global developmental delay and seizures.

P118.

KBG syndrome, a rare underdiagnosed disorder: about three patients

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Background

KBG syndrome is a rare, autosomal dominant disorder, characterized by distinctive craniofacial features, macrodontia of upper central incisors, short stature, behavior problems, developmental delay/intellectual disability with seizures and brain malformations. Recurrent otitis media can also be present.

More than 100 patients have been reported in the literature. The disorder is probably underdiagnosed as the clinical features can sometimes be mild. On the other hand, there is an overlap between KBG syndrome and other disorders, like Cornelia de Lange syndrome.

Causative mutations in the ANKRD11 gene have been identified. Large deletions in the 16q24.3 region, including ANKRD11, have also been reported.

Results

We report 3 patients with KBG syndrome, at different ages.

Patient 1 - 2 year-old girl.

She developed an epileptic encephalopathy with microcephaly, developmental delay, and post natal growth deficiency. She showed chronic constipation. Craniofacial findings include triangular face, hyperthelorism, bushy eyebrows, protruding ears, and anteverted nostrils. Genetic tests showed a de novo nonsense heterozygous mutation in ANKRD11: c.5953C>T; p.(Gln1985*).

Patient 2- 14 year-old boy.

He developed global developmental delay, bilateral strabismus, recurrent otitis media, scoliosis and chronic constipation. Craniofacial findings include triangular face, synophrys, bushy eyebrows, and macrodontia of the upper central incisors. Genetic tests showed a de novo heterozygous deletion of 5 nucleotides in ANKRD11: c.1903_1907del; p.(Lys635fs), creating a premature stop codon.

Patient 3- 51 year-old woman.

She developed mild intellectual disability and epilepsy with microcephaly. The cerebral MRI showed cerebellar vermis hypoplasia. She has bilateral strabismus, scoliosis and recurrent otitis media with hearing loss. Craniofacial findings include triangular face, hyperthelorism, bushy eyebrows, and macrodontia of the upper central incisors. Genetic tests showed a de novo heterozygous nonsense mutation in ANKRD11: c.1372C>T; p.(Arg458*).

Conclusion

KBG syndrome is a rare, but recognizable disorder. Especially, the characteristic dysmorphic features and the macrodontia of the upper central incisors in a child with developmental delay, can point toward the diagnosis. Nevertheless, the syndrome is underdiagnosed above all in young children.

New sequencing approaches are valuable tools for diagnosis in patients with mild or less characteristic phenotype.

P119.

Live birth neonate with triploidy (69, XXX): a case report

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Introduction

Triploidy is a chromosomal abnormality characterized by an extra haploid chromosome set. It is estimated to occur in 1-3% of all conceptions. However, most of these conceptions result in a spontaneously abortion before the end of the first trimester of pregnancy.

Methods

We will describe a case of a live birth infant with triploidy who survived 35 days.

Results

The mother, 30 years old, was gravida 1 and para 0. There was no significant medical history and there was no consanguinity between the parents. The pregnancy was complicated by intra-uterine growth restriction (IUGR). An extensive ultrasound showed beside IUGR no other structural abnormalities and noninvasive prenatal test was negative. A regular control at 36 weeks and 3 days showed an abnormal CTG and an emergency cesarean section was performed. The neonate was female and had a difficult start with the necessity of neonatal resuscitation. The apgar scores were 4 points at 1 minute, 6 points at 5 minutes and 6 points at 10 minutes, she was admitted to our neonatology for respiratory support.
The neonate was extreme small for gestational age with a birth weight of 1445 gram, length 41.5 cm and head circumference of 30.2 cm. Besides, there were dysmorphic features including frontal bossing, hypertelorism, thoracal kyphosis, small sacral dimple with a visable base, malformed feet's and syndactyly of the third and fourth fingers of both hands. Neurological examination showed a general hypotonia and the absence of the sucking reflex. Genetic analysis was performed with SNP array from peripheral blood and showed a triploid karyotype of 69, XXX. An ultrasound of the cerebrum, heart and abdomen did not show any structural abnormalities besides the presumably absence of the gallbladder. The Newborn Screening was twice positive for congenital hypothyroidy and medical therapy was started. After the first days of life the neonate had a cardiorespiratory stable period witch slowly declined. Palliative sedation was started only a few days before she died at the age of 35 days.

Conclusion

In triploidy, many malformations and dysmorphic features have been reported in affected fetuses and neonates. The presence of syndactyly, especially syndactyly of the third and fourth fingers of the hands, is the most distinctive feature. However, there are no obligate malformations or dysmorphic features in triploidy. This case learns that the presence of IUGR in combination with dysmorphic features makes triploi

P120.

Bell's palsy in a 20-day-old newborn with a serology positive for Mycoplasma Pneumoniae

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Introduction

Peripheral facial palsy is common in the pediatric population. The main causes of this condition associated to Bell's sign are viral, traumatic, inflammatory or ischemic even linked to a CNS tumor. In neonates most cases are due to obstetrical traumatisms. Cases secondary to bacterial infections such as Lyme or M.Pneumoniae have been reported in the adult rather less frequent in the pediatric population. We report her the case of a newborn presented to the emergency room because of a facial asymmetry.

She was born after a normal pregnancy. The delivery was uneventful and free of any obstetrical traumatism. The clinical examination was normal. Within 3 weeks old she exhibited a progressive facial asymmetry. Excepted a transient and spontaneously resolutive measles-like at the age of 1 Week, no other complaints have been noticed. The clinical examination showed a lowering of the right labial commissue, an absence of the right naso-genial fold and a right Bell's sign. The general exam was unremarkable.

Biological studies did not show any inflammatory syndrome. The CFS analysis showed 9/µL, 18/µL RBC and a slight hypoglycorrhachia and hyperproteinorachy. The nasopharyngeal aspiration was negative. The empiric IV antibiotic therapy with Cefotaxime and Amoxicillin as well as Acyclovir was first initiated. Complementary results showed blood culture contamination but positive IgM M.Pneumoniae serology. Cefotaxime and Amoxicillin were therefore stopped and switched for Clarithromycin. Other serologies were negative. A symptomatic treatment with artificial tears and a corticosteroid therapy were started. As CSF PCR herpes returned negative, Acyclovir was withdrawn on the 5th day of hospitalization.

The patient was discharged after 5 days, without any clinical improvement. The symptoms were still persistent on day 8 and the M.Pneumoniae serologic control showed a raise in IgM levels with the appearance of IgG. An antibiotic therapy with Clarithromycin was therefore carried on. The symptoms disappeared 5 weeks later.

Conclusion

Facial asymmetry during the neonatal period has many etiologies. It can be associated with syndromic diseases, traumatic and malformative causes. Among the infectious causes, the HSV virus is often listed. Yet, like in the adults, M. Pneumoniae should be part of the investigations even in neonates and in the pediatric population. Treatment shloud be prompt and aggressive and should include steroids.

P121.

Kinsbourne syndrome as complication of a Mycoplasma pneumoniae infection

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Introduction

Kinsbourne syndrome also known as opsoclonus-myoclonus-ataxia syndrome or more commonly dancing-eye and feet syndrome is a rare neurological disorder affecting infants and toddlers previously healthy averagely aged from 6 to 36 months. The pathophysiology is not perfectly understood but seems to involve a dysimmune process; its etiology is either paraneoplastic or parainfectious.

Case report

We describe the case of a five-year old boy without relevant medical history, who presented a left bronchopneumonia complicated by a major pleural effusion. Acute Mycoplasma pneumoniae infection was confirmed by blood serology and by PCR performed on pleural fluid. Significant clinical improvement was observed after starting antibiotic therapy with Clarithromycin and then Moxifloxacin administered for a total of 15 days. From day 10, he abruptly presented rapid, irregular and multidirectional eye movements associated with myoclonias of the extremities of the upper limbs and the face, and orthostatic ataxia. He also demonstrated low frustration tolerance and recurrent nocturnal awakenings that were previously absent. Complementary explorations including abdominal echography, chest x-ray, Neuron-Specific Enolase (NSE) blood and urinary catecholamine tests, and brain MRI excluded a neoplastic etiology such as neuroblastoma. A diagnosis of Kinsbourne syndrome was made. The child quickly showed a spontaneous favorable evolution with a total recovery after 6 weeks.

Conclusion

Few cases of Kinsbourne syndrome secondary to Mycoplasma pneumoniae infection are described in present literature. Here, we present the rare case of a young boy who spontaneously demonstrated a favorable evolution without immunomodulatory treatment.

Key words

Mycoplasma pneumoniae, Kinsbourne syndrome, abnormal movements

P122.

Cardio-facio-cutaneous syndrome and severe hyperthermia in the child

M Baillat, MA Heng, B Chabrol, C Despineux. UC Louvain

Context

Cardio-facio-cutaneous syndrome (CFC) is a rare genetic syndrome, part of the RASopathy group. The principal symptoms are dysmorphism, cardiopathy, cutaneous anomalies, late growth, epilepsy and intellectual deficiency.

Clinical observation

A 23-month-old child with CFC presented during the summer of 2018, very hot, hyperthermia of 42.3°C with septic shock without a specific etiology noted. In the intake biology, there was thrombocytopenia, coagulation abnormalities, increased creatinine and hepatic transaminases, rhabdomyolysis, hyperproteinaemia and hypoalbuminemia as well as ionic abnormalities. Under empirical antibiotic therapy, the child recovered a better general condition but liver damage persisted.

Discussion

This severe hyperthermia with multivisceral dysfunction may be due to "exogenous heat stroke" or, less likely, to a probably viral infection. The cutaneous ichthyosis and cerebral malformation could be factors contributing to the hyperthermia.

Conclusion

CFC children may be more be likely to suffer from a severe hyperthermia at the time of heat waves and/or infection; early treatment could avoid multivisceral failure, especially hepatic failure.

P123.

Management of arteriovenous malformations in pediatric population

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Aims

Arteriovenous malformations (AVMs) is the most common etiology of stroke (47%) in pediatric population, although annual incidence is 1.2-13 cases per 100,000. Literature is very poor compared to the adult population. CT and IRM lead to diagnosis. AVMs carry an annual risk of hemorrhage of approximately 3.2%, a 5-10% mortality rate, and a 50% risk of neurological morbidity.

Methods

Case 1 : A 15-year-old girl presented with acute unilateral headache with left hemianopia, vision disorders, vomiting, but without alteration of consciousness. Non-injected brain CT was performed revealing a large right parietooccipital hematoma. A cerebral arteriogram has confirmed the presence of AVM. She has benefited from an embolization. Currently, the after effects are just reading difficulties. However, the control arteriogram shows that the AVM is still present and requires a second treatment by radiotherapy or surgery.

Case 2 : A 11-yeat-old girls presented with acute onset of headache, vomiting, and alteration of consciousness. CT of the head revealed a massive ventricular hemorrhage without edema. She was intubated. A deep extern drainage has been set up. She proceeded to undergo MRI of the brain and arteriography, which were suggestive of arterioveinous malformations.

Conclusion

Upon initial diagnosis of intracerebral hemorrhage on noncontrast CT, workup and treatment should be initiated without delay. MRI and arteriography should be proceeded in the management. Intensive care and monitoring are indicated. Depending of the location of hemorrhage, if high intracranial pressure is present, an external ventricular drainage or an evacuation of the hematoma must bee proceeded. Surgery depends of the location, size and hematic cavity. Radiotherapy is also possible depending on the stage of bleeding. The embolization is reserved for small lesions. AVMs in children are fraught with a high recurrence rate (as high as 14%), but the critical moment is within 24 hours of bleeding.

P124.

Bed for preventing post dural puncture headache, is it efficace?

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Background

Post-dural puncture headache (PDPH) is one of the most common complication of spinal tap procedures. It is generally defined as pain increasing in the upright position and decreasing in the reclining position after a dural puncture. It's a serious matter because of the consequence on the patient and on family anxiety. That may also influence the length of hospital stay. Our objectif has been to assess throughout a literature review whether or not bed resting is statistically better than immediate mobilization for PDPH prevention.

Methods

Several studies have been reported over the past decades. Main analysis of thes data have been made by Arevalo-Rodriguez et al and published in the 2013 and 2016 Cochrane Database. Their meta-analysis included 24 trials reported up to February 2015 concerning 2996 participants. For the outcome bed rest versus immediate mobilisation, the meta-analysis included 12 studies with 1519 participants. However, of the 12 studies, only one (Ebinger 2004) studied a pediatric population from 2 to 17 years old.

Results

There was no difference between bed rest and immediate mobilization in three of the four separate categories of lumbar puncture: diagnostic lumbar puncture (RR 1.11; 95% CI 0.90 to 1.37; 723 participants; 6 studies), myelography (RR 1.48; 95% CI 0.67 to 3.27; 207 participants; 1 study), and mixed (RR 1.27; 95% CI 0.68 to 2.35; 208 participants; 1 study). There was a small difference between bed rest and immediate mobilization on spinal anesthesia data (RR 1.82; 95% CI 1.19 to 2.78; 381 participants; 4 studies), suggesting an increase in the risk of PDPH with bed rest.

Conclusion

At least in adult population, there is no evidence that suggests that routine bed rest after dural puncture is beneficial for the prevention of PDPH onset. As a result, it seems unnecessary to impose a number of hours lying down rather than evaluate according to the clinical condition of the patient. Unfortunately, there is a lack of pediatrics studies in this domain. However, in pediatrics, it is well recognized that prevention is essential. This includes rigouroussness of indications and explanations to the patient and his family as well as the to be accurate during the procedure. We have the project to design a prospective collaborative study to assess the modalities followed in most pediatric centers and their effictiveness.

** Arevalo-Rodriguez I, Ciapponi A, Roqué i Figuls M, Muñoz L, Bonfill Cosp X. Posture and fluids for preventing post-dural puncture headache. Cochrane Database of Systematic Reviews 2016, Issue 3. Art. No.: CD009199. DOI: 10.1002/14651858.CD009199.pub3.*

O13.

Long-term auditory effects in patients treated for brain tumors during childhood

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Background

Childhood brain tumor survivors are at risk to develop long-term sequelae such as hearing loss (HL). Radiotherapy and platinum agents used in the treatment of childhood brain tumors are known to be ototoxic. The aim of the study was to determine the long-term auditory effects in adult childhood brain tumor survivors. Other risk factors possibly associated to the development of HL and the impact on quality of life were investigated.

Methods

225 survivors treated between 1971 and 2011 in the division of PHO at Ghent University Hospital were invited to take part in the study. Patients (pts) had to be off therapy for at least 5 years. A questionnaire and an extensive audiological evaluation were performed. Four treatment groups were defined: the surgery only group, the group with adjuvant radiotherapy, the chemotherapy group (platinum derivatives) and the group with adjuvant chemo- and radiotherapy. HL was defined as an increase of the hearing threshold of at least 20 dB in at least one frequency and in at least one ear. To define the severity of HL, the CTC and SIOP toxicity scale were used. HL and severity of HL were assessed and compared between the 4 treatment groups.

Results

Response rate was 34,2% and 70 pts were included. 64 pts completed the questionnaire and 57 pts underwent audiological testing. HL was found in 34 pts (59,6%) and was typically sensorineural, progressive and starting at the high frequencies. HL was mostly found in the treatment group that received a combination of chemo- and radiotherapy (71,4%). The majority of pts with severe HL (grade 3-4) were also found in this treatment group. Remarkably, 40% of the pts that only received surgery also had HL. In the patients with HL, mean time between end of therapy and audiological testing was significant longer (p=0.002) than in the patients without HL, even when corrected for age. This suggests the progressive character of ototoxicity. 53 pts reported (very) good hearing but in pts with grade 3-4 HL, questionnaire results were in contradiction with the audiological testing.

Conclusion

Ototoxicity is prevalent in childhood brain tumor survivors. In the group that received a combination of chemo- and radiotherapy the prevalence and severity of HL was the highest. Patients only treated with surgery are also at risk. Ototoxicity may be present early and progress with time. Long-term audiological follow-up is necessary for an adequate detection of this HL and early rehabilitation.

OP21.

Retrospective analysis of the incidence and characteristics of pediatric myelodysplastic syndrome and juvenile myelomonocytic leukemia in Belgium

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Background/Aims

Childhood myelodysplastic syndrome (MDS) and juvenile myelomonocytic leukemia (JMML) are very rare clonal stem cell disorders of early childhood. MDS is characterized by cytopenias and dysplasia and is divided in refractory cytopenia of childhood (RCC) and advanced MDS (MDS with excess blasts (MDS-EB) and MDS-EB in transformation (MDS-EBt)). JMML is a myelodysplastic/myeloproliferative disorder characterized by overproduction of granulocytic and monocytic cells.

The aim of this study is to investigate the incidence, characteristics (clinical and biological) and prognosis of pediatric MDS and JMML in Belgium.

Methods

We retrospectively analyzed data of 49 Belgian patients with MDS and JMML consecutively enrolled in the '2006' study of the European Working Group of MDS in Childhood (EWOG-MDS) between January 2007 and July 2018. Additionally, 7 patients enrolled in the previous '98' EWOG-MDS study were also included. Patient data were prospectively collected in these trials using standardized case report forms and the diagnosis of all children was centrally reviewed in the EWOG-MDS Coordinating Study Centre.

Results

In total, 56 patients were included, of which 41 MDS, 11 JMML and 4 Noonan syndrome. 26 MDS patients presented with RCC, 12 with MDS-EB and 3 with MDS-EBt. 6 patients were diagnosed with secondary MDS (3 patients having underlying BM disease and 3 a history of malignancy). Incidence rates of MDS and JMML in Belgium were 1.5 and 0.4 per million children per year, with a median age of diagnosis of 9.3 years for RCC, 9.5 years for advanced MDS and 2.6 years for JMML. Monosomy 7 was the most common cytogenetic abnormality and could predominantly be found in advanced MDS (33%) and JMML (45%). RCC treatment consisted of immunosuppressive treatment and hematopoietic stem cell transplantation (HSCT), whereas in advanced MDS and JMML only HSCT was a valid treatment option. Event-free survival in RCC (74.5%) was markedly better compared to advanced MDS (58.7%) and JMML (66.7%).

Conclusion

This retrospective analysis demonstrated that pediatric MDS and JMML are very rare disorders with associated morbidity, especially in advanced MDS and JMML. Patient characteristics were heterogeneous, signifying the need for a multidisciplinary diagnostic approach. Systematic collection of patient characteristics and clinical course in an international consortium can lead to a better understanding of these disorders.

OP22.

Antibacterial prophylaxis with fluoroquinolones in children with acute myeloid leukemia: impact on viridans group streptococci

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Background/Objectives

Infections remain an important cause of morbidity and mortality in children with acute myeloid leukemia (AML).

In recent years, ciprofloxacin prophylaxis was introduced in our hospital to reduce the risk of infectious complications. Recent reports however, describe breakthroughs of Viridans Group Streptococci (VGS) bacteremia in children with AML receiving fluoroquinolone (FQ) prophylaxis.

The aim of this study is to investigate the impact of FQ-prophylaxis on bloodstream infections in terms of prevalence and type of isolated species in a pediatric AML population who received FQ prophylaxis and those who did not.

Design/methods

Retrospective study in pediatric patients hospitalized in the period 11/2009-4/2017 and treated according to subsequent international AML protocols. Pharmaceutical and laboratory records were analyzed to determine the use of FQs (ciprofloxacin) and to identify positive hemocultures (HCs). Statistical analysis was performed by Chi square tests.

Results

The study included 26 patients with de novo or relapse AML, representing 195 episodes of hospitalization. FQ prophylaxis was administered in 109/195 episodes (55.9%). HCs were positive in 17 episodes (17/195; 8.7%) and in 13 of these episodes (13/17; 76.5%) FQ prophylaxis was used.

When looking at the species isolated from the 17 HC-positive episodes, a mix of VGS/non-VGS (1/4) or a non-VGS species (3/4) was identified in the patients who did not receive prophylaxis. The 13 HC-positive episodes with FQ (13/17; 76.5%) revealed 11/13 (84.6%) VGS or a mix of VGS/non-VGS and 2/13 (15.4%) non-VGS species ($p = 0.022$).

Conclusion

The use of FQ prophylaxis was associated with a significantly higher number of VGS positive HCs. This prompted us to stop FQ prophylaxis in our pediatric patients. A larger retrospective study to validate these findings is currently ongoing.

OP23.

Comparison of the incidence of sinusoidal obstruction syndrome in a pediatric stem cell transplantation population receiving oral or intravenous busul

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Background

Busulfan is an alkylating chemotherapeutic drug used in conditioning regimens for stem cell transplantation (SCT). Sinusoidal obstruction syndrome (SOS), also known as veno-occlusive disease (VOD), is a well-recognized and potentially life-threatening complication that may develop primarily after SCT. Possible factors associated with SOS are young age, liver damage or prior transplantation and hepatotoxic medications amongst others. A high interpatient variability has been described. The introduction and administration of an intravenous (IV) formulation of busulfan was presumed to decrease toxicity and incidence of SOS, because of lower variability in drug exposure and improved control of busulfan levels.
The aim of this study is to investigate if the incidence of SOS is correlated with administration route and/or with high blood levels of busulfan in a pediatric SCT population.

Methods

Monocentric retrospective study (October 2005 till October 2017). Children aged 0 - 17 years who underwent SCT after a (PO or IV) busulfan-based conditioning regimen were enrolled after informed consent. Busulfan was administered at standard dosages (according to body weight) for 4 consecutive days over a total of 16 doses. Levels of busulfan were measured at different time points. The adverse events, admission to the pediatric ICU, incidence of SOS and mortality were recorded. All data were retrieved from medical records and were anonymized by the lead researcher.

Results

Fifty-nine patients were included in the study, 19/59 (32%) received the oral, 40/59 (68%) the IV busulfan formulation. The highest peak blood levels were found in the IV-busulfan group. Thirteen patients (13/59;22%) developed SOS, whereof 5 patients (5/19;26%) in the peroral group, 8/40 (20%) in the IV busulfan treated group. The difference for the development of SOS was not statistically significant between the IV and the PO group (Fisher's exact test p = 0,7).

Conclusions

This study showed no statistically significant difference in the development of SOS after receiving busulfan PO or IV. The development of SOS is due to more cofactors and needs further investigation. Our research team is studying those cofactors and especially the influence of higher peak busulfan levels and higher area under the curve on the development of SOS.

OP24.

Disease monitoring by liquid biopsies in pediatric patients with solid tumors

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Backgound

In recent years, the analysis of tumor-associated copy number variants (CNV) in cell-free tumor DNA (ctDNA) in peripheral blood plasma has gained interest as a noninvasive cancer screening tool. Here, we explored CNV profiling in ctDNA as a potential malignancy biomarker in selected pediatric solid tumor patients.

Methods

We prospectively performed the CNV analysis in ctDNA by genome representation (GR) profiling (median coverage across the whole genome 0.23x) in 25 consecutive patients [with neuroblastomas (NB; n=5), Ewing's sarcomas (ES; n=4), osteosarcomas (OS, n=5), rhabdomyosarcomas (n=5), and others (n=6)]. In patients where an abnormal GR profile was detected, the testing was extended to blood samples collected during routine follow-up in the consecutive 6-24 months from the start of the treatment (surgery/chemotherapy/radiotherapy). Molecular data were linked to the clinicopathologic tumor characteristics and clinical follow-up of the disease.

Results

An abnormal GR profile was detected in 14 patients (five OS, three NB, three ES, one germ cell tumor, one Wilms' tumor and one teratoma). Follow-up blood samples collected during further therapy were available for 12 patients. In nine cases, the aberrant GR profile 'normalized' in successive samples and all were in remission at these time points. In two patients with refractory disease (one NB, one ES), the GR profile remained abnormal during therapy and an increase in the fraction of the genome affected by CNVs was compatible with disease progression. Both patients eventually died of disease. One patient had inconclusive data.

Conclusion

Our results suggest that serial analysis of ctDNA offers a promising approach for the evaluation of treatment response and disease progression in solid tumor pediatric entities. Validation of these findings on a larger group of patients and during longer follow-up period will be required to determine the benefits of this approach in a clinical setting.

P125.

Diencephalic syndrome due to a hypothalamic tumor as a rare cause of failure to thrive in an adolescent

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Background

Diencephalic syndrome is a rare presentation of hypothalamic tumors. It includes failure to thrive with severe weight loss despite normal linear growth, due to increased energy expenditure.

Methods

We present a case report, highlighting diencephalic syndrome as a rare cause of failure to thrive in an adolescent patient.

Results

SH, a 14 years old boy, presented because of dysphagia, decreased intake and weight loss since 4 months. An initial work-up with an abdominal ultrasound, barium esophagram, gastroscopic evaluation and screening blood test did not reveal any abnormalities.
Ten weeks after the first presentation, SH was hospitalized because of ongoing weight loss, vomiting and dysphagia. At this point, a total weight loss of 13 kg was obtained, resulting in a BMI of 15. A blood test showed panhypopituitarism with low levels of free T3, T4, TSH, testosterone, LH, FSH, cortisol, ACTH and IGF-1. The patient did not suffer from headache, there was no visual impairment and a clinical neurological examination was completely normal. An MRI of the brain revealed tumoral lesions in the hypothalamus and the epiphysis, with ependymal seeding. A stereotactic biopsy was performed, confirming the diagnosis of germinoma. SH was treated with craniospinal irradiation and complete remission was obtained.

Conclusion

In children with unexplained failure to thrive, diencephalic syndrome due to a hypothalamic tumor should be considered as a rare cause. Other accompanying alarm symptoms as headache, visual impairment, vomiting or an abnormal clinical neurological examination are not always obvious.

P126.

Choroid plexus carcinoma in Li-Fraumeni syndrome

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Background

Li-Fraumeni syndrome (LFS) is a rare autosomal dominant cancer predisposition syndrome due to a germline mutation in the TP53 tumor suppressor gene. We describe the case of a child diagnosed with a choroid plexus carcinoma (CPC) within a cancer predisposed family.

Case report

A five-years old boy complained of acute headaches, photophobia, vomiting and neck rigidity. Cerebral CT and MRI revealed a large hemorrhagic tumor-like lesion in the left parietal lobe. Staging did not identify spinal metastasis or cerebrospinal fluid invasion. Complete resection was achieved after two surgery. The anatomopathological analysis confirmed the diagnosis of high-grade choroid plexus carcinoma. Adjuvant chemotherapy is indicated.

Family history showed that his mother and his maternal aunt were affected by breast cancer before the of age 40 years old. Both have a germline mutation in the TP53. His cousin has the same mutation. His maternal uncle died of digestive sarcoma at age 16.

Germline TP53 mutation is still researched among the proband. There are family evidences of LFS.

Discussion

Li-Fraumeni syndrome is historically defined in 1969 by clinical criteria. Other Li-Fraumeni-Like syndrome were later defined by Birch et al. (1994), Eeles (1995) and Chompret (2001). They described LFS as a rare autosomal dominant disorder characterized by a familial clustering of tumors before 45 years. Cancer commonly associated with LFS are soft sarcoma or osteosarcoma, breast cancer, adrenocortical cancer, brain tumor and acute leukemia. Most of LFS families have a germline mutation of the TP53 suppressor gene. Penetrance in LFS depends of age, gender and type of cancer. Penetrance is 20% in children, 90% after 70 years of age. Brain tumors (astrocytoma, glioblastoma, choroid plexus carcinoma) are more frequent in early childhood.

Choroid plexus tumors represent 2-4% of all pediatric brain tumors. Choroid plexus carcinoma is a rare highly aggressive malignant tumor designed as grade III according to the World Health Organization. The median age at diagnosis is approximatively 26 to 32 months old. CPC has been strongly associated with LFS. LFS is an aggravating factor of prognosis in CPC.

Conclusion

LFS is associated with a high risk of juvenile cancer or multiple cancer. Various types and location of cancers make screening difficult. Genetic counseling is not easy particularly in children and within families in which many patients are already severely sick.

P127.

Thoraco-Neuroblastoma in a 13 month-old-girl

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We report the case of a 13 months girl who had been presenting an alteration of her general state for one month with asthenia and palor. During the preceding months, she had had several episodes of viral infection. Physical examination showed pale tegument and mild tachycardia. Laboratory examination revealed a normocytosic anemia (7,4g/dl) with low reticulocytosis. The rest of the biology was without any particularity. The concentration of iron, ferritin, vitamin B12, and folic acid was normal. Electrophoresis of hemoglobin was normal as was pyruvate kinase and G6PD. The diagnosis of medullary stinging due to repeated viral infections was considered.

In the following days, she had fever and showed inguinal ganglia and splenomegaly. Laboratory examination confirmed anemia and showed thrombopenia, whithout any inflammatory syndrome. Abdominal ultrasound showed only splenomegaly. On the contrary, chest X-ray demonstrated an enlarged cardiac silhouette. ECG and echocardiography were normal. Bone marrow puncture allowed to exclude leukemia.

In the following course she presented a dysfunction of the left leg. The radiography showed a lytic lesion on the left femoral neck and heterogeneity of the bone structure at the level of the iliac wing. Additional CT imaging show a medullary inflitration of the femoral methaphysis, and the hip. At that time, neuron specific enolase blood concentration was elevated (250 Î¼g/L). A bone marrow biopsy was performed and desmontrated the presence of small blue cells.

The suspicion of a neuroblastoma led to a further examination by MIBGI123 scintigraphy that allowed to identify a left paravertebral mass. It also revealed multiple axial and appendicular bone- and intramedullary attachments. MRI confirmed the left paravertebral mass extending over D4 to D10. Urinary cathecolamines were difficult to interpret because of a small urine sample. Cytogenetics on bone biopsy confirmed neuroblastoma. The patient was directly scheduled for chemotherapy.

50% of newborn malignancies are neuroblastomas, 20% being thoracic, 70% abdominal, 5% cervical and 5% pelvic. 50% have already metastatic involvement at the diagnosis. The diagnosis might be difficult due to the aspecific and confusing clinical signs, as it is demonstrated by this case.

An early diagnosis is crucial. The vital prognosis depends not only of the age, the presence of certain genetic mutations (eg N-MYC) and metastasis, but also on the ability to a complete surgical excision.

P128.

Iron deficiency anemia: the forgotten culprit of thrombocytopenia in female adolescents

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Background

Iron deficiency (ID) is a common, underdiagnosed problem in adolescent females. Iron demand increases due to growth-related consumption and menstrual blood loss. Insufficient intake leads to a decreased capacity of the bone marrow. This can cause anemia but also severe thrombocytopenia.

Case

This case report describes an asymptomatic 15-year-old female with menorrhagia presenting with a bicytopeania (Hb 6.7 g/dL and TBC 92x10E9/L). Both her sister and mother also suffered with ID and menorrhagia. The diagnosis of a severe microcytic, hypochromic anemia with nutritional and menstrual etiologies was made by excluding other diseases such as leukemia, clotting deficiencies and gastro-intestinal pathologies. Oral iron supplementation was started as well as hormonal contraceptives for menstrual regulation. Subsequently a worsening anemia necessitated a red blood cell transfusion. A resolution of the trombopenia followed the normalisation of iron levels.

Conclusion

Multiple etiologies predispose adolescent females at an increased risk of developing ID but can remain subclinical. Symptoms frequently present late with severe sequelae, highlighting the importance of early detection. Further genetic studies are indicated with a positive family history without resolution on adequate oral supplementation. ID is a documented cause of thrombocytopenia by interference of both platelet function and production. This in turn potentiates menorrhagia and further iron depletion. Screening for ID is cheap and accessible by basic laboratory testing. Advice for the prevention of ID in adolescent females is not ready available and should be part of basic health education.

P129.

Monophasic pulmonary blastoma in an adolescent: case report and review of the literature

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Introduction

In children, primary lung neoplasms are uncommon. Pulmonary masses are most frequently caused by benign developmental, reactive lesions or malignant metastasis. Pulmonary blastoma (PB) accounts for 0.25-0.5% of all lung neoplasms. It arises presumably from primitive pulmonary mesenchyme and resemble histologically fetal lung. We report the clinical history of an adolescent with monophasic type of PB.

Case report

A 15-year-old boy presented with chest pain after falling from a trampoline. The chest X-ray performed in the emergency room revealed no parietal injury but the incidental presence of a pulmonary mass. The CT-scan displayed a fleshy and polycyclic tumor (6.7cm x 5.6cm) in the apical segment of the left inferior pulmonary lobe. A first transbronchial biopsy failed to determine the nature of the tumor. A second transthoracic biopsy showed a monophasic PB. The extension workup, including PETscan, confirmed the primary tumor hypermetabolism but no distant metastasis. A surgical inferior lobectomy was performed. Microscopic examination indicated a complete resection, with no pleural invasion, no lymph node infiltration, but a doubt on a possible vascular thrombus. Because of the rarity of this tumor and its poor prognosis, the case was discussed in an international forum of experts. It was decided to administrate adjuvant chemotherapy (3 courses of Etoposide alternating with 3 courses of Cisplatin). Six months after treatment completion, the patient remains in remission and has no therapy related toxicity.

Discussion

PB were initially divided in three subgroups: classic or biphasic PB, well-differentiated fetal adenocarcinoma also called monophasic PB, and pleuropulmonary blastoma. This latter type is currently classified in a separate entity. It's mostly diagnosed before 4 years of age although biphasic and monophasic PB are extremely rare in pediatrics. They are mainly found in the 4th decade of life and associated with smoking. The prognosis of PB is poor but the monophasic type seems to have a better outcome than biphasic variety. No therapeutic guidelines exist. Surgical excision is the treatment of choice and produce better prognosis with adjuvant chemotherapy.

Conclusion

This paper illustrates a very rare case of monophasic PB in children. It also highlights the importance of international expert forum to define the most appropriate therapeutic approach of such rare disease, and so the need to collect more data about these entities.

P130.

Case report: Rectal adenocarcinoma - When age can be deceitful

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Case report

15-year-old Caucasian girl presented with a 4-month history of abdominal pain and 3-month history of diarrhoea. An explorative abdominal laparoscopy revealed signs of pelvic inflammatory disease. Metronidazole was started for a positive Gardnerella culture. However, during the following months her condition deteriorated, fever, weight loss (17 kg) and bloody stools appeared. Initially bacterial gastro-enteritis was suspected and intravenous antibiotics (first amoxicillin clavulanate, later cefotaxime) were started. Laboratory results showed iron-deficiency anaemia and elevated inflammatory parameters (CRP 126mg/L, sedimentation rate 104mm/h). Rectoscopy was performed, showing a large polyp. Under the suspicion of a new onset inflammatory bowel disease (IBD), she was transferred to our centre. Endoscopic biopsy of the 5cm large rectal mass revealed an adenocarcinoma. Carcinoembryonic antigen in the blood was elevated (50.2ug/L). Based on further staging through PET-CT and MRI the tumor was decided to be stage T4N2aM0. Due to the intestinal obstruction sigmoidostomy was necessary. Chemotherapy was started 2 weeks later. Family history was negative for IBD or gastro-intestinal tumours, but there are several relatives with carcinomas of lung, breast and cervix. The results of the genetic testing are expected.

Discussion

In 2016 there were 2407 new diagnoses of rectal cancer in Belgium. The median age at diagnosis is 63 years and rectal cancer is extremely rare at young age (<25 years). Therefore, genetic predisposition for colorectal cancer (especially Lynch syndrome, CMMRD and familial adenomatous polyposis) and cancer predisposition in general (including TP53, BLM and POLD1) need to be investigated. Clinical staging of rectal cancer is done by diagnostic biopsy, physical examination and imaging (CT, MRI, transrectal ultrasound). CEA is not sensitive nor specific enough for screening or diagnostic testing, but has value in the pretreatment staging and follow-up. Surgical resection is the cornerstone of curative therapy, but neoadjuvant chemotherapy followed by chemoradiotherapy can be considered for patients with T4 or N2.

Conclusion

Rectal carcinoma is rare in adolescents, but is important to recognise and genetic predisposition and cancer predisposition need to be explored.

P131.

Klinefelter syndrome and Germ Cell Tumors: review of the literature

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Background

The most common presentation of klinefelter syndrome (KS) is infertility and features of hypogonadism. No consensus currently exists on the risk of malignancy in this syndrome, although several case reports show an incidence of extra gonadal germ cells tumors (eGCT) of 1.5 per 1000 KS patients (OR 50 against healthy population). Malignant germ cell tumors are rare in children, it accounts for 3% of all children cancers. Young patients with germ cell tumors are not usually tested for KS, and therefore this can result in under-diagnosing klinefelter syndrome.

Aims

Literature data suggest a correlation between eGCT and KS. To the best of our knowledge there is still no precise description of the primary locations of germ cell tumors in KS patients. The purpose of this study was to evaluate age groups and primary locations in eGCT in KS patients. With this data we investigated whether it is necessary to perform a cytogenetic analysis for KS in every eGCT patient.

Materials and Methods

This study is based on publications in PubMed/Medline published until 2018 that described 'Klinefelter Syndroom (MeSH) AND/OR Extra Gonal Germ cell tumors (Mesh)'. Publications were included when patients age, location and histology of the germ cell tumor was known. Two dubble blinded reviewers selected the studies.

Results

Until 2018 in total 132 eGCTs in 128 KS patients were published in case-series. Most eGCT were mediastinal or in the central nervous system (respectively 81/132; 63% and 22/132; 17% of all tumors). Mean age at presentation was 17.6 years (StDev +-10,7). Histology based it was most common a teratoma, mixed-type-non-seminomateus GCT and mixed-type-GCT, respectively 32/132; 24%, 26/132; 20% and 14/132; 11% of all tumors.

Conclusion

These data suggest a correlation between primary germ cell tumors and klinefelter syndrome. There appears to be an indication for screening on KS in young patients with an eGCT in the mediastinum.

Keywords

Klinefelter Syndrome, Malignancy, Incidence analysis, extra gonadal germ cell tumor.

P132.

Pedunculated fibrolamellar hepatocellular carcinoma without liver lesion in a 14 years-old girl

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Introduction

Fibrolamellar hepatocarcinoma (FLHCC) is a rare variant of hepatocellular carcinoma (HCC). In contrast to HCC, FLHCC usually occurs in young adults, without underlying liver disease. Final diagnosis is based on morphological characteristics of the tumor, immunohistochemistry analysis and the presence of a DNAJB1-PRKACA fusion gene on FISH analysis. The only curative treatment is complete resection of the lesion. In case of unresectable FLHCC, chemotherapy may be an alternative.

Case report

A 14-years-old girl was admitted to hospital with abdominal pain, bloating and vomiting since a few days. Abdominal ultrasound revealed multiple peritoneal nodules with multi-compartment ascites. Abdominal MRI and 18-FDG-PET CT scan confirmed peritoneal carcinomatosis with bilateral ovarian involvement, without liver lesion. Laboratory investigations showed elevated CA-125 level at 526,8 kU/l and slightly elevated alfa-fetoprotein level at 12,3 µg/l. Histopathologic examination of the tumor revealed large cells with prominent nucleoli and eosinophilic cytoplasm. Immunohistochemistry analysis of the sample also plead for FLHCC. Because of the absence of liver lesion and in order to confirm this hypothesis, PRKACA FISH test was performed, revealing the presence of a DNAJB1-PRKACA fusion gene, conventionally associated with fibrolamellar hepatocarcinoma. These findings allow us to conclude to pedunculated FLHCC.
The girl was treated according to the Paediatric Hepatic International Tumor Trial protocol. Treatment consisted in six cycles of Cisplatin, Doxorubicin and Sorafenib. Abdominal MRI after six cycles showed significant decrease in size and amount of abdominal and peritoneal lesions. On PET CT scan, any active lesions were reported.

Discussion

In our patient, initial presentation of peritoneal carcinomatosis without any liver lesion on abdominal ultrasound and MRI made the diagnosis challenging. Morphological and IHC aspects of the lesion allowed us to suspect FLHCC but the lack of liver lesion had not been previously described. However, it has been reported that HCC may be presenting as a pedunculated tumor from the liver to the abdomen, considered as metastatic lesion. In order to confirm our diagnosis, FISH analysis was performed on the tumor sample, revealing the presence of a DNAJB1-PRKACA fusion gene. It has been recently proven that this fusion gene is associated with FLHCC, conficonfirming our hypothesis of a pedunculated FLHCC.

P133.

Development of a care pathway for the child with a brain tumor: from multidisciplinary to interdisciplinary collaboration

L Willems, E Willems, A Mannaerts, G Laureys. UZ Gent

Background/Aims

The care for children suffering from a brain tumor is complex and involves a lot of different (para-) medical disciplines (e.g. pediatric oncologist, neurosurgeon, radiotherapist, intensive care physicians, rehabilitation physician together with a rehabilitation team of physiotherapist, ergotherapist, psychologist,…). All health care professionals have their own expertise in taking care of a child with a brain tumor.
In 2013, we introduced a nurse-led clinic for children with brain tumors in the pediatric hemato-oncology ward of the university hospital of Ghent. The goal of this clinic was to improve patient care by introducing a nurse consultant as case manager during the pathway of their diagnosis and treatment. Working in this structure made us clear that we needed to align all the concerned disciplines and to insert transparency in the various functions of the team.

Methods

Our first action in the development of our care pathway was to inform each discipline about our purpose and ask for their input. We then critically observed the current workflow of each discipline and spoke and listened to every single partaker of all the different disciplines. Doing this, we enumerated, analyzed and discussed the bottlenecks in the care for children with a brain tumor.

Results

As a result of the above discussions, we decided that a monthly multidisciplinary meeting could improve the communication and co-operation between the health care professionals. We developed a care pathway consisting of a detailed description of all steps in the care of the patient with a brain tumor: registration, diagnosis, treatment and follow up. This care pathway is available throughout the entire hospital, via an easily accessible document managing system in which it is possible to have a quick overview of all tasks for each care professional, in each care-episode.

Conclusion

By developing a care pathway, we accomplished a structure in which the different disciplines involved in the care of children with a brain tumor could co-operate more easily. Furthermore, we found that each different discipline seemed to be of great importance in this process.
Treating a child with a brain tumor according to a care pathway is a large advantage for these patients, as efficient communication between different caretakers has been shown to be of great importance in the care of patients with complex pathologies.

P134.

Survey on satisfaction with the service of a clinical pharmacist on a pediatric hematology and oncology unit

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Background

Pharmacotherapy in pediatric hematology and oncology (PHO) patients is complex. To improve the quality of treatment and care, a team of 4 clinical pharmacists is involved in the PHO unit of the University Hospitals Leuven (UHL) since October 2014. This service can be optimized based on feedback of parents and other healthcare professionals (HCPs).

Methods

An anonymous e-questionnaire on the satisfaction with this service was sent to the parents, nurses and doctors on the PHO unit of UHL. Six items were questioned: overall satisfaction (parents and HCPs), patient-friendly medication list (parents), the counseling conversation (parents), the handling of potentially toxic medication (parents), the added value of the pharmacists' advice (HCP) and the time saving component of the pharmacists' service (HCP).

Results

The response rate was 39% (14/36), 27% (21/77) and 38% (22/58) for the parents, nurses and doctors respectively. Eighty-six percent of parents and 95% of nurses and doctors were satisfied to very satisfied with the overall service of the clinical pharmacist.

For all the parents the counseling is given in an understandable language and 86% receive enough counseling. The patient friendly medication list developed by the pharmacist for an individual patient is clear (100%) and understandable (86%), although some parents advise some minor changes (eg. bigger font, 1 page…). Fourteen percent of the parents don't know how to handle potentially toxic medication (eg. chemo or other immunosuppressants).

The pharmacotherapeutic advice given by the pharmacist to HCPs has added value for 98% of the HCPs, mainly due to the improvement of care for the patient, the obtainment of relevant information, the support that the advice gives to HCPs and the time that HCPs save because they don't have to look up farmacotherapeutic information themselves. Time is also saved because the clinical pharmacists take over the medication counseling of the other HCPs.

Conclusion

Overall, most parents, nurses and doctors are satisfied to very satisfied with the service of the clinical pharmacist on the PHO unit at UHL. However, the pharmacist should focus more on counseling the parents about potentially toxic drugs and on handing over a more patient tailored and patient friendly medication list.

P135.

Paediatric supra-sellar germinoma initially diagnosed as a lymphocytic hypophysitis : a case report

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Introduction

Initial histological/imaging presentation of pituitary tumors can mimic hypophysitis. Because hypophysitis is extremely uncommon in children, the differential diagnosis should always include brain tumors.

Clinical case

We report the case of a 12 years-old girl with a supra-sellar germinoma, initially diagnosed as a lymphocytic hypophysitis. The patient presented growth delay and polyuria/polydypsia. The biological exploration confirmed growth hormone (GH) deficiency based on low level of IGF1 (26 ng/ml) and low GH response after glucagon tolerance test (peak at 0.6 ng/ml). The central diabetes insipidus (DI) was confirmed based on a water restriction test with good response to desmopressin administration. The first brain magnetic resonance imaging (MRI) documented a thickened pituitary stalk (3.5 mm), and the histopathological examination of an in situ biopsy showed a chronic follicular inflammation, compatible with a diagnosis of lymphocytic infudibilo-hypophysitis. Tumoral markers (aFP and b-HCG) were negative in the blood and cerebro-spinal fluid (CSF). The patient was first treated by corticoids, and received substitution by desmopressin than hydrocortisone. A few months later, a rapidly progressing left mydriasis and left oculomotor nerve palsy appeared. A new cerebral MRI showed an increased volume of the patient hypophysis (7.4 mm), with a supra-sellar extension pushing the optic chiasma an left cavernous sinus extension. The histological re-examination documented an important lymphoid infiltrate and immunohistochemistry showed seminoma highly suspected cells, with CD117 positivity. A new CSF examination showed normal values of aFP but increased value of b-HCG (9.7 mUI/mL), proteins (897 mg/L) and nucleated cells (28/mm3). The patient was treated with two cures of polychemotherapy followed by radiosurgery.

Conclusion

Intracranial germinomas are relatively rare malignant tumors (2% of all brain tumors). DI is generally the first symptom but other pituitary deficiencies are often observed. This case illustrates the fact that pituitary stalk thickening caused by a lymphocytic infiltrate may represent the first sign of a host reaction to an occult germinoma. A review of the literature underlines the need for continued monitoring for progressive disease using MRI in patients with apparent lymphocytic hypophysitis, especially in pediatric patients.

P136.

A novel three-way translocation - t(1;7;22) (p13;q21;q13) - in a case of neonatal acute megakaryoblastic leukaemia

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Abstract

Acute megakaryoblastic leukaemia (AMKL, FAB-M7) is a rare disease, occurring mostly in infants and young children. Symptoms at diagnosis consist of bone marrow fibrosis, hepatosplenomegaly and pancytopenia. The chromosomal translocation t(1;22) (p13,q13) resulting in the RBM15-MKL1 (OTT/MAL) fusion gene, is a specific translocation in AMKL in infants.

Here we describe a case of a newborn girl without Down syndrome, with congenital acute myeloid leukemia, type AMKL. At 36 weeks of pregnancy, her mother presented with symptoms indicating fetal distress and labor was therefore induced. At birth, the girl had massive hepatosplenomegaly, and peripheral blood count showed anemia, trombopenia and a leucocytosis with 20% blasts. Immune phenotyping of the bone marrow showed 28% blasts, positive for CD34, CD61, CD42b. Upon these findings, the diagnosis of acute megakaryoblastic leukaemia was made. Karyotyping of these blasts (R-banding) showed a novel chromosomal translocation t(1;7;22) (p13;q21;q13), a three-way variant of t(1;22) (p13;q13). The girl developed multi-organ failure and an intracranial haemorrhage thus initiation of an intense regimen of chemotherapy was renounced.

Up until today, there are only a few cases described exhibiting a variant on this translocation, and to our knowledge, this is the first report of this novel three-way variant in AMKL.

P137.

Bone Marrow Necrosis as presentation of an acute myeloid leukemia

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Background

Bone marrow necrosis (BMN) is a rare histopathology entity defined by necrosis of myeloid tissue and medullary stroma with preserved cortical bone. In more than 90% of cases, it is associated to a malignant disease.

We report the history of a nine-month-old infant with BMN identified in the context of acute myeloid leukemia (AML).

Case report

A nine-month-old boy, was admitted to the pediatric intensive care unit because of thrombocytopenia with purpura, aregenerative anemia, prolonged fever and inflammatory syndrome. A bone marrow puncture and biopsy were performed and showed extensive medullar necrosis. The presence of blasts in peripheral blood suggested the diagnosis of AML FAB-M5. This was sustained by the cytogenetic analysis revealing an inversion and translocation of chromosome 8 with rearrangement of NUP98 gene. Chemotherapy according to the NOPHO-DBH AML 2012 protocol started 72 hours after admission. Despite persistent BMN during the first treatment course a regression of blasts in the peripheral blood was observed and a complete medullar remission was confirmed after 42 days. The patient received all subsequent treatment courses and remains in complete remission two month after end of treatment.

BMN is a rare entity mostly associated with hematological malignancies in particular with acute lymphoblastic leukemia. While BMN has been associated with poor prognosis in adults, in pediatrics this presentation seems not to affect the prognosis of the underlying disease

Conclusion

Our case illustrates the rarity of BMN as well as the challenge difficulties and highlights the challenge in diagnostics. The frequent association with hematological malignancies should trigger an extensive search for such an underlying disease. In this regard cytogenetics and molecular biology appear to be helpful tools.

OP11.

National survey about the current and ideal practice for children and adolescents with chronic pain in Belgian hospitals

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Introduction

Chronic pain in children and adolescents is a significant problem worldwide and the management of pain is definitely a hot topic, even in pediatric medicine. Chronic pain is recognized as a healthcare problem on itself and ample literature describes the advantage of a specialized multidisciplinary approach in order to augment quality of life in these children and their families. Nevertheless Belgian clinicians are facing difficulties to provide this 'state of the art' care to children with chronic pain in the absence of a national management strategy and financing.

Objectives

The objective of this survey is to identify the current approach and treatment of children and adolescents with chronic pain in Belgian hospitals. A second goal is to describe the ideal practice in Belgium.

Method

An online survey was send to 100 pediatric departments in Belgian hospitals, 3 rehabilitation centers for children and 35 specialized multidisciplinary teams (MPC) for the management of chronic pain (for adults). The survey contained 18 questions about the current practice for children with chronic pain in the hospitals and about the type of patients with chronic pain that are most frequently seen.

Results

A total of 50 pediatric centers, 12 MPC and 3 rehabilitation centers fully completed the survey. 3 pediatric and 2 rehabilitation centers have a written standard and multidisciplinary team for the management of chronic pain in children. Of these centers 1 has an outpatient program, 1 is only specialized in chronic abdominal pain and 1 is a small non-specialized center. Top 3 chronic pain problems in children in the Belgian hospitals are chronic abdominal pain, headache and musculoskeletal pain. Most participants answered that a specialized unit with a multidisciplinary team would be the ideal care for these children. Described shortcomings in the current treatment for children with chronic pain are the lack of a national standard, limited education for caregivers and financial support.

Conclusion

This study reveals the limited healthcare facilitations for children with chronic pain in Belgium. It seems there are 2 rehabilitation centers specialized in inpatient care and 1 academic hospital specialized in the treatment for all chronic pain problems in children. This study gives an insight in the problems caregivers experience today and can be used as a tool for Belgian policy makers to start a national standard for the care for children with chronic pain.

OP12.

Executive functioning in inherited intoxication type metabolic diseases: A comparison of phenylketonuria and other intoxication type metabolic disease

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Background

Phenylketonuria (PKU) has been extensively studied. However, the executive profiles of other intoxication type metabolic disorders still need to be described, in order to provide optimal care to patients and families.

Aim

To describe three executive domains: 1) planning and organization, 2) inhibition and 3) cognitive flexibility in patients with an intoxication type metabolic disorder, and to compare these findings with the executive profiles of patients with PKU.

Methodology

In this retrospective, descriptive, quantitative study, test results of 38 patients with a (developmental) age of at least 8 years, recruited from the Centre for Inherited Metabolic Disorders Antwerp database, and diagnosed with an intoxication type metabolic disorder, were analyzed. Only test results of the most recent testing moment on which the three included executive functions were assessed, were used in the descriptive analyses. A clinical neuropsychological assessment was made per metabolic disorder per subtest and statistical comparisons between the results per subtest of patients with PKU and patients with other intoxication type metabolic disorders were performed.

Results

1 patient with 3-methylcrotonglycin aciduria (3-MCG), 1 patient with hyperinsulinemia/hyperammonemia syndrome, 1 patient with maple syrup urine disease, 1 patient with tyrosinemia type I, 7 patients with methylmalonic acidemia, 5 patients with ornithin transcarbamylase deficiency and 22 patients with PKU were included. As a group, the non-PKU patients had a low IQ, but did not show deficits in the studied executive domains. The patient with 3-MCG showed deficits in cognitive flexibility and inhibition. Planning and organization and inhibition were deficient in tyrosinemia type I. Other intoxication type metabolic disorders showed no executive dysfunctions. When taken as a group, PKU patients had a below average IQ and did not show any executive deficits. Statistically, there was no significant difference between both groups.

Conclusion

There is no evidence that patients with intoxication type metabolic disorders should be followed up regarding their executive functioning, or that preventive and therapeutic measures used for patients with PKU can be extrapolated to other intoxication type metabolic disorders. Further research to complete the executive profiles of patients with an intoxication type metabolic disorder, taking into account the quality of treatment and follow-up, is needed.

P138.

Hair Collar sign: the tree that hides the forest

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Introduction

The hair collar sign, defined as a ring of long, thick, coarse hair surrounding a nodule, cyst and/or atrophic hairless area on the scalp is known to often be a sign of dysraphism revealing an underlying neuroectodermal defect, such as heterotopic brain tissue or cephalocele.
We here report on a hair collar sign revealing a sinus pericranii associated with an atretic cephalocele.

Case report

A 5-month-old girl presented with an abnormal tuft of hair on the scalp. Examination of the midline vertex showed a 1-cm soft nonpulsatile flat nodule with surrounding dark terminal hairs forming an irregular hair collar. Other syndromic features were absent.
Cerebral US and MRI gadolinium first diagnosed an anterior accessory sinus pericranii. Posteriorly, a subcutaneous fluid sac was associated with a cerebellar tentorium deviation and a persistence of the primitive falcine sinus, defining an atretic parietal cephalocele.
The imaging and neurosurgical joint discussion concluded a wait and see decision as a first approach.

Discussion

A sinus pericranii (SP) is a rare vascular malformation in which the extracranial veins are connected to the intracranial venous circulation. Although it is generally considered benign, the outcome may not be predicted easily. It can be dominant or accessory and isolated or associated with other malformations, such as an atretic cephalocele. It typically appears as a soft mass with a blue hue on the scalp. The lesion is mostly asymptomatic and treatment conservative.
A cephalocele is defined as a herniation of cranial contents through a defect in the skull. The term atretic cephalocele is used when a rudimentary connection is present such as a fibrous tract to bone or dura.
In this case report, the hair collar sign draws attention to the location of a SP and a meningocele as the subcutaneous part of an atretic cephalocele. A surgical treatment with resection of the atretric cephalocele, ligation of the SP and cutaneous plasty could be suggested by the age of one year.

Conclusion

The hair collar sign is at high risk for cranial dysraphism as a sinus pericranii or a cephalocele. An MRI gadolinium must be performed in these patients not to miss the hidden part of the iceberg. Neurosurgical treatment could only be considered if the sinus pericranii is not dominant and mainly to prevent bleeding or infectious complications.

P139.

Uracal remnant: Another cause of abdominal pain

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Aim

The purpose of this abstract is to describe a not very well known and studied pathology, more frequent than we think and raise the awareness about its potential in the differentials of chronic abdominal pain in children.

Case report

We present the case of an eleven years old girl who consulted for chronic abdominal pain with an exacerbation during the last three days. The pain has been increasing and no additional symptoms were associated (no fever, vomiting, diarrhoea nor urinary). The preteen patient looked well. The physical examination showed a diffuse sensibility mostly in the hypogastric area extending to both flanks. In order to exclude acute abdomen, an abdominal ultrasound was realised showing a hypo echogenic tubular structure laying between the hypochondria and the bladder; an image compatible with urachus structure. A CT scanning confirmed an urachus remnant either a cyst or a sinus.
A surgical excision has been programmed for our patient due to her age and the low possibility of spontaneous resolution.

Definition and epidemiology

The urachus is a connection between the foetal bladder and the allantois. The descent of the bladder toward the pelvis stretches the urachus, eventually leading to obliteration of its lumen. The median umbilical ligament is the resultant fibrous cord which runs from the umbilicus to the dome of the bladder. Occasionally, this obliterative process is incomplete, leading to a persistent urachal remnant. The urachal abnormalities are divided in five groups: urachal cyst (central part of the tract is patent and fills with fluid),patent urachus (communication between the umbilicus and bladder), urachal sinus (umbilical end is open but there is no communication with the bladder), urachal diverticulum (forms a cap on the dome of the bladder) or urachal chorda (the entire tract persists as a cord) .
The incidence of urachal abnormalities is unknown but due to increased awareness in the paediatric community as well as increased detection with improved ultrasonography, this incidence is growing.
A study showed that the median age of patients at diagnosis was 1.8 months, mean age was 1 ± 2.4 years, and male in majority.

Symptomatology and detection methods

According to the literatures mo

P140.

Pediatric Laparascopic Sleeve Operation in ZNA Antwerp: Short-Term Results.

L Hendrickx , K Maes ,E Engels , S Heyman. Koningin Paola Kinderziekenhuis Antwerp, ZNA

Obesity in children and adolescents is a global chronic and progressive epidemic disease, that causes 2.8 million deaths per year. The prevalence of overweight and obese children increases wordwide with an estimation of 60 million obese children in 2020. The definition of morbid obese (BMI 35 kg/m² or BMI 1.2 above the 95 th percentile.
The aim of this poster is to present the short-term results of a laparoscopic sleeve gastrectomy (LSG) in 4 adolescents in our patient population.

Methods
Adolescents who underwent LSG in 2017-2018 were descibed. Charts were investigated retrospectively and short-term weight loss together with comorbidity before and after the LSG were analyzed.

Results:

Pat.	Age (y)	Sex	Before	Surgery	After	Surgery			Result
					1 months		3 months	6 months	
			Weight (kg)	BMI (kg/m²)	Weight	BMI	Weight/ BMI	Weight /BMI	
1	14	M	140	47.9	130	44.5	143/48.9	145/47. 9	+ 5 kg (6 m)
2	13	F	114	48.1	102	43	99.5/41.7		-14.5 kg (3 m)
3	15	F	110	40.4			89.5/32.7	88/32.3	-20.5 kg (6 m)
4	14	F	138.5	50	126	45.7			-12.5 kg(1m)

Comorbidity Before LSG Patient	OSAS	HT	Fatty Liver	Hyperinsulism	Hypogonadism/ Menstrual Disturbances	Mental Problems
1 M	+	+	+	+	+	+++
2 F	+	+	+	+	+	++
3 F	+	+	+	+	+	++
4 F	+	+	+ (bile stone)	+	+	+++

Comorbity After LSG Patient	OSAS	HT	Fatty Liver	Hyperinsulism	Hypogonadism/ Menstrual Disturbances	Mental Problems
1 M	+	++	++	++	+	+++
2 F	-	-	+	+	-	+
3 F	-	-	-	-	-	+
4 F	-	-	+	+	-	+

Conclusion

Even though the follow-up period was short and the number of patients was small, LSG is a and promising surgical method for morbidly obese children with a lowered life expectancy, though to multiple comorbidities. A multidisciplinary approach and lifelong healthy lifestyle together with support of behavior therapy are the key messages for success.

P141.

Lymphocytoma Cutis: Don't Forget Lyme Disease

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Introduction

Borrelial lymphocytoma is a rare and benign cutaneous manifestation of Lyme disease consisting of a purple-red nodule usually located on the earlobe or the nipple. It mainly occurs in the early disseminated stage (secondary stage).

Here we report on two cases of borrelial lymphocytoma in children.

Case reports

First, a seven-year-old girl presented with a 5-month recurrent erythema and swelling of the left earlobe. A transient erythema on the left cheek gradually spread over the entire left side of the face. About 4 months later, the patient developed daily left-parietal headaches and fatigue.

The MRI, ophthalmological and ENT investigations were unremarkable. Two 15-day spaced anti-borrelia titers were positive for IgG and negative for IgM. White blood cells in the cerebrospinal fluid and intrathecal secretion of anti-borrelia IgG were revealed. There was no personal history of tick bite.

The diagnosis of borrelial lymphocytoma and neuroborreliosis was established.

The treatment consisted of intravenous cefotaxime (200mg/kg/day) followed by oral doxycycline (4mg/kg/day) for a total of 28 days. Within 15 days, the earlobe nodule and the headaches had disappeared but the hemifacial erythema persisted although appearing less frequently.

A second eight-year-old boy presented with a one-month purple nodule located on his left breast areola. A 15-days erythema around the nodule extended on the left over the back. There was no personal history of tick bite. The patient confessed attending a summer scout camp a few months earlier. Anti-borrelia titers were positive for IgG and negative for IgM.

A borrelial lymphocytoma was suspected and a treatment with amoxicillin 500mg 4 times a day was started for a total duration of 3 weeks. The patient was re-evaluated 15 days later and a substantial decrease of the breast areola nodule was noted and the surrounding erythema had disappeared.

Conclusion

Borrelial lymphocytoma is a rare and delayed cutaneous manifestation in Lyme disease.

The location on the earlobe or on the breast areola is highly suggestive of Lyme disease. So it is necessary to investigate for a Lyme disease and start an appropriate treatment before considering a cutaneous lymphocytic malignancy. Early recognition may also avoid the evolution to later stages and a neuroborreliosis.

P142.

Screening of anxious and depressive symptoms in youths with high functioning autism

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Background

Previous works have shown that anxiety is the most frequent psychiatric comorbidity in autistic children and adolescents, followed by depression. Prevalence range is estimated from 30% to 50%, which is superior to the general pediatric population. Experts seem to agree on the lack of a large scale prospective study which would compare these findings to a control group. Such a study would allow us to establish more objective results on the prevalence of these comorbidities in high functioning autistic patients. Also, compared to the general pediatric population, practitioners lack specific tools to screen anxiety and depression in the autistic population.

Objective

Our study aims to assess the prevalence of anxious and depressive symptoms in a population of adolescent patients with high functioning autism, using the survey «Echelle Composite de Dépression pour Enfants» (MDI-C).

Patients and methods

Prospective non-interventionnal study, realized between December 2015 and August 2017 in the Infant-Juvenile Psychiatry Unit of Cliniques Universitaires Saint-Luc Brussels. Ten subjects were included in the experiment.

Results

With a maximum score of 80 points, the mean value was 22 ± 8,90, none of the subjects attained the level of 56 points indicating symptoms of depression. Also, with a maximum score of 11 points, the mean value was 4,1 ± 2,19, none of the subjects attained a level indicating symptoms of anxiety. Following this tool, we didn't show any symptomatology in the studied group. These results are not consistent with what is reported in the literature.

Conclusion

The survey used in this study (MDI-C) seems to present limitations to be used in the population with autism because of its cotation system. The number of subjects included in the study could also be a bias. The aspects of this survey, as well as the statistical results and the methods are discussed in this work.

P143.

A huge ovarian mucinous cystadenoma in a 15-year-old girl

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Background

Ovarian mucinous cystadenoma (MCA) rarely occur in children. They may present as large abdominal masses and lead to compression symptoms or torsion, haemorrhage and rupture or be asymptomatic. We report a case of a 15-year-old girl diagnosed with a massive ovarian MCA.

Case report

A 15-year-old postmenarchal girl complained of abdominal distension evolving for at least 2 years, without other associated symptoms (only a little embarrassment). It had not been noticed by her parents, but the girl deliberately hid her belly. Physical examination showed a distended abdomen and a mass extending from the pelvis to the xiphoid process. The abdomen was tense but painless. Abdominal ultrasound revealed a voluminous cystic mass (35x20 cm) and a right hydronephrosis. MRI revealed a voluminous multiloculated cystic mass (27x14.6x33.7cm), likely mesenteric, it pushes the digestive structures without any sign of invasion. A right uretero-hydronephrosis with pyelic dilatation was highlighted. Right ovary was multifollicular, left ovary was not visualized. Imagery concluded to a cystic lymphangioma in first hypothesis. At surgical exploration, the tumor was found to originate from the left ovary (9kg in weight). Frozen section revealed an ovarian MCA and a left salpingo-oophorectomy was performed. Tumor markers CA 19-9, CA 125, hCG and Î±FP were normal.
The histological analysis confirms the diagnosis of MCA. There is no further treatment needed.

Discussion

Ovarian MCA are benign tumors from the epithelial surface of the ovary. The clinical presentation is not specific, so that it might be difficult to diagnose. Patients may be asymptomatic or present abdominal distention in case of large cystadenoma or present complications.
The management consists of performing a laparoscopic cystectomy. Nevertheless, due to the size of the mass, a conservative surgery could not be considered. A cyst rupture during the procedure or an incomplete excision of the cyst is associated with a risk of recurrence.

Conclusion

Ovarian tumors represent less than 2% of all tumors in girls under the age of 16. In this age group 60% of the ovarian tumors are neoplastic (from germ cells) and 20% derive from the surface epithelia of the ovary. Most of mucinous tumors (75%) are benign, (10%) borderline and (15%) carcinomas. The clinical presentation is not specific so the diagnosis is often delayed. The management consists to perform a prompt and fertility-preserving surgery.

P144.

An episode of sudden blindness after a minor head trauma in an 8-year-old girl

C De Windt, C Karimi, M Mounir, F Vermeulen. Hôpital Erasme

An 8-year-old girl was admitted to the emergency department (ED) after a fall on her occiput at school. Stress and hunger preceded the loss of consciousness. When she regained consciousness a xanthopsia (yellow vision) occurred, followed a few minutes later by total blindness associated with a holocranium headache, sleepiness, back pain and pain in her occipital protuberance.

In the ED, her vitals were normal with a heart rate at 90 bpm, a pulsed oxygen saturation at 99 %, a blood pressure at 125/90 mmHg and a blood glucose level of 116 mg/dL. Her physical examination was normal except for total blindness. Her pupils were isocorous, reactive and occulomotricity was maintained.

The differential diagnosis was cortical blindness versus peripheral blindness. In our case the preservation of the photomotor reflex suggests retrochiasmatic involvement, thus a cortical blindness. The brain and cervical spine scanner as well as the electroencephalogram were normal. After excluding neurological emergencies, the ophthalmologist diagnosed a transient post-traumatic cortical blindness.

Transient cortical blindness is a complication of a minor head trauma usually involving the occipital region. Despite an unknown pathophysiology, two hypotheses are described: the first involves a vasospasm in the pre-geniculate region, the second a cortical edema and ischemia in the occipital region. A benign outcome is linked with pediatric age, mild trauma, brief or no loss of consciousness, absence of neurological deficits other than the blindness, onset of blindness within hours of the trauma lasting less than 24 hours. If these criteria are met, a brain scanner does not seem necessary since it always returns negative.

In our case total recovery occurred within 5 hours after the trauma.
This typical case of post-traumatic blindness highlights the benign side of this little-known, frightening and spectacular presentation.

Conclusion
Transient cortical blindness occurring brutally after a minor head trauma is usually benign and does not necessarily require a brain scanner after the advice of an ophthalmologist who confirms the presence of the photomotor reflex and the integrity of the optic nerve.

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