Swiss Society of Neonatology

Schweizerische Gesellschaft für Neonatologie
Société Suisse de Néonatologie
Società Svizzera di Neonatologia

www.neonet.ch

ANNUAL MEETING of the SSN 2019

in collaboration with the

ACADEMY FOR FETAL AND MATERNAL MEDICINE (AFMM)

TUESDAY, JANUARY 22, 2019

KULTUR- UND KONGRESSZENTRUM
AARAU

Abstract book

Discharge teaching quality, readiness for discharge, and post-discharge healthcare utilization in mothers of hospitalized neonates from a Swiss NICU : a correlational descriptive study

Chloé Tenthorey, Laura Rio, Anne-Sylvie Ramelet
Service of neonatal intensive care, Department of Woman Mother and Child- Lausanne University Hospital (CHUV), Lausanne, Switzerland / University Institute of Training and Research in Healthcare (IUFRS), Lausanne, Switzerland

Purpose High quality of discharge teaching should positively affect readiness for discharge and lower negative outcomes such as post-discharge overuse of health services. Here, we describe maternal perceptions of readiness for neonates' discharge, discharge teaching quality and the unplanned use of health services following discharge from a Swiss Neonatal Intensive Care Unit.

Design and methods This cross-sectional correlational descriptive study included mothers of hospitalized neonates about to return home. Readiness for discharge and discharge teaching quality were evaluated with the Readiness for Hospital Discharge Scale parent and the Quality of Discharge Teaching Scale in the 24 hours preceding discharge. Telephone interviews evaluating the unplanned use of health services were conducted 28 days post-discharge.

Results Most of the 71 participants felt adequately taught and ready for discharge but with high heterogeneity of total scores ranging from 54 to 178 for teaching (mean = 129.5 ; SD = 26), and from 116-273 for readiness (mean = 220 ; SD = 29). Unplanned use of health services occurred in 46 % (N = 70) of cases in the month post-discharge. Perceived quality of teaching positively predicted readiness for discharge (R2 = 0.24, P = 0.0004). Post-discharge healthcare utilization was not correlated with readiness or quality of teaching.

Conclusions Unplanned use of healthcare service post-discharge was high even when mothers felt mostly ready and adequately prepared for discharge home.

Implications for practice : Heterogeneity amongst maternal perceptions would suggest that identification of mothers' individual needs before discharge is necessary. Further investigation is needed for exploration of reasons leading to post-discharge healthcare utilization.

Outcome of very preterm infants with grade 3 intraventricular haemorrhage and periventricular haemorrhagic infarction

Philip Thwaites [1], Dirk Bassler [1], Sven Schulzke [2], Juliane Schneider [3], Jean-Claude Fauchère [1], Mark Adams [1], Giancarlo Natalucci [4]
[1] University of Zurich and University Hospital Zurich, Department of Neonatology, Zurich, Switzerland. [2] University of Basel Children's Hospital UKBB, Department of Neonatology, Basel, Switzerland. [3] University Hospital Center- University of Lausanne, Department of Woman-Mother-Child, Lausanne, Switzerland. [4] University of Zurich and University Hospital Zurich, Department of Neonatology- on behalf of the Swiss Neonatal Network and Follow-up Group, Zurich, Switzerland

Aims and objectives In the last decade, studies showed improved prognosis of grade-III intraventricular haemorrhage (IVH-III) and periventricular haemorrhagic infarction (PVHI) when compared to historical cohorts. We aimed to compare the rates of mortality and of neurodevelopmental impairment (NDI) at 2 years between two consecutive national cohorts of very preterm infants affected with one of these two types of brain lesions.

Materials & methods Prospective neonatal and 2-year follow-up data of all infants without major malformations, live-born at a gestational age (GA) <30 weeks in Switzerland in 2002-2007 (period-A) and 2008-2014 (period-B) were analysed. IVH-III and PVHI were diagnosed using cranial ultrasound and NDI at 2 years as : a development index <-2SD, severe cerebral palsy, blindness or deafness. Statistics were adjusted for GA, sex, and birth-weight z-score.

Results Among 5302 live-born infants, 470 developed either IVH-III or PVHI (226 and 244 in period-A and -B, respectively). Mortality rate in period-A (53 %) was slightly higher than in period-B (48 %) but difference did not reach statistical significance (p = 0.305). Of the deceased infants, 89 % and 85 % infants died due to withdrawal of care in period-A and -B, respectively (p = 0.390). We observed an increase in follow up rates between the 2 periods (69 to 79 %) and while this did not reach statistical significance (0.093) the increase was a welcome observation to us. NDI rates of survivors were similar between periods (41 and 42 % respectively, p = 0.861).

Conclusions Despite recent reports of better prognosis in infants with IVH-III or PVHI, we observed no difference in mortality or NDI rate at two years over time in these two consecutive cohorts of very preterm infants. Further research is required to explore the causes of mortality and the end-of-life decision process preceding the withdrawal of care in the studied infants.

Recurrent Pneumatosis in extreme preterm !

Aurélie De Mul, Marie Saint Faust
Neonatal Intensive Care Unit, Hôpitaux Universitaire de Genève

Cow milk protein allergy (CMPA) is the most common food allergy in childhood. Due to non-specific symptomatology in preterm, this entity is difficult to distinguish between necrotizing enterocolitis (NEC). This entity is poorly described in the preterm infants.

A 660-grams male infant was born by cesarean section at 26 weeks of gestation in a context of severe maternal HELLP syndrome and intra-uterine growth restriction. He developed hyaline membrane disease and hemodynamically significant patent ductus arteriosus. Enteral feeding was initiated on day 1 of life with exclusive breast milk (BM). On day 13, a perforation of the right colon was diagnosed and required surgery, resulting in a 2 cm resection and a colostomy. Second surgery was performed after US diagnosis of intra-peritoneal abscesses. Progression toward full enteral feeds with exclusive BM was well tolerated. On day 54, 5 days after introduction of a fortifier and 1 bottle of cow milk formula, he developed abdominal distension and hemorrhagic diarrhea leading to hypovolemic shock. An abdominal x-ray demonstrated a massive intestinal pneumatosis and presence of intraportal venous gas. Blood inflammatory markers were high and eosinophil count increased to 17 %. He was treated as a medical NEC (Bell's stage II) with bowel rest, empirical antibiotherapy and gastric decompression. The x-ray dramatically improved in less than 24 hours. Diet was restricted to a highly amino acid (AA) hydrolyzed formula, according to the hypothesis of CMPA and full enteral feeding was well tolerated. On day 78, 3 days after reintroduction of small amount of BM, he quickly developed the same symptoms, increased count of eosinophilic cells and new radiologic intestinal pneumatosis. The question of maternal daily consumption of CMP was confirmed. The level of serum-specific cow milk IgE was negative. A rapid complete resolution was observed after restarting an exclusive AA formula diet. The evolution and growth were positive, CMP eviction will last 1 year.

CMPA is poorly recognized in the preterm infants. No specific tests are available (non IgE mediated allergy), except a transient peripheral eosinophilia. The dramatic improvement after cow milk eviction diet might be pathognomonic. In breast-fed infants, mothers should start a strict CMPfree diet and attention must be payed to fortifier containing CMP. The presence of late onset or recurrent NEC like illness should raise the suspicion of CMPA.

Characterization of postnatal sodium fluctuation in very preterm neonates

Dr Roland Gerull
University Children's Hospital UKBB, Spitalstrasse 33, 4056 Basel

Aims and objectives Large variations of serum sodium (Na) are associated with increased morbidity and mortality in adults. However, evidence is low in pediatrics and even more so in preterm infants. We aimed to identify factors influencing Na-fluctuations during the first 28 days of life in preterm infants <32 weeks gestational age (GA).

Materials and methods This retrospective study included all preterm neonates born 2007-2014 with GA <32 weeks at the University Children's Hospital Berne, Switzerland. Descriptive modelling included non-linear mixed modelling to fit data and to characterize covariate effects. Secondary parameters, such as predicted Na concentration, were accessed via simulations.

Results A total of 901 preterm neonates with GA of 29.4 [interquartile range 27.4, 30.9] weeks and a total of 20714 sodium measurements were eligible. Na at birth was 133.1 (±1.9) mmol/l and similar across patient groups. Na increase started at postnatal age (PNA) of 1.2 (±0.6) days and a Na peak of 143 (±2.7) mmol/l was observed at 3.2 (±1.5) days. Higher Na peaks were observed in patients with lower GA, after spontaneous delivery and birthweight below 10th percentile. After 10-15 days mean Na concentration was constant around 134 mmol/l. Antenatal steroids, gender and birthweight did not have an influence on sodium at baseline or peak sodium.

Conclusion Na fluctuations were increased in patients with decreasing gestational age, spontaneous delivery or decreased birthweight percentile, while gender, antenatal steroids and birthweight showed no significant effect. Modeling and simulation allowed to characterize individual serum sodium profiles and to identify risk factors in this very vulnerable population.

A Newborn with Bruises

Rahel Kasteler [1], Benedikt Bubl [1], Michael Daskalakis [2], Marc Baumann [3], Jane McDougall [1], Tilman Humpl [1]
[1] Division of Neonatology, Department of Pediatrics, Inselspital, Bern University Hospital, University of Bern, Bern, Switzerland, [2] Transfusionmedicine, Department of Hematology and Central Hematology Laboratory, Inselspital, Bern University Hospital, University of Bern, Bern, Switzerland, [3] Department of Gynecology and Obstetrics, Inselspital, Bern University Hospital, University of Bern, Bern, Switzerland.

Objectives Neonatal Alloimmune Thrombocytopenia (NAIT) is caused by maternal antibodies against alloantigens of fetal platelets.

Case report A term, female infant was born by elective cesarian section to a 36 year old G3 para 1 mother. Within one hour after birth the baby developed rapidly progressive extensive bruising and petechia periorbitally, on the trunk and extremities. The platelet count in the initial blood draw was 1G/l, other coagulation testing was within normal limits. The baby was otherwise well with a normal clinical examination. She was admitted to the neonatology unit for platelet transfusion. Family history for platelet disorders was negative (normal platelet count), thus neonatal alloimmune thrombocytopenia was considered as differential diagnosis. The parents were tested, and maternal antibodies were found against glycoprotein IIa/IIIb of the father, however, the most frequent human platelet-specific antigen (HPA) genotypes could be excluded. This lead to the diagnosis of a NAIT with unknown/rare HPA genotype. The baby required a total of 2 platelet transfusions of non-typed platelet apheresis-concentrates. Cranial and abdominal ultrasound exams did not reveal any hemorrhage.

Discussion Severe NAIT is a rare disease, which requires rapid diagnosis and therapy. An affected child will present with bruises/petechia or other signs of bleeding with isolated thrombocytopenia. NAIT is one of the leading causes of intracranial hemorrhage in full term infants and occurs in 10-20 % of diagnosed children. Therapy with typed platelets is only possible for the most frequent genotype HPA1. Subsequent pregnancies need to be monitored closely for complications and an elective cesarian section is recommended.

Conclusion The diagnosis of NAIT should be considered early in newborn babies, who are otherwise well, but present with severe thrombocytopenia and clinically with bruising and petechia.

Intestinal atresia – it might be an important clue to cystic fibrosis

Chiara De Angelis [1], Corinne Daester [1], Gabriel Konetzny [1], Peter Klimek [2], Peter Eng [3], Philipp Meyer [1]
[1] Department of Neonatology and Intensive Care Unit, Kantonsspital Aarau, Aarau, (Switzerland), [2] Department of Pediatric Surgery, Kantonsspital Aarau, Aarau, (Switzerland), [3] Department of Pediatric Pulmonology and Allergy, Kantonsspital Aarau, Aarau, (Switzerland)

Introduction Foetal bowel dilatation is often predictive of bowel obstruction requiring surgery and it has been described as marker for many conditions, including jejuno-ileal atresia and cystic fibrosis. Although associated congenital anomalies are less common with jejuno-ileal atresia than duodenal atresia, an increased prevalence of cystic fibrosis in patients with jejuno-ileal atresia (6-13 %) has been described.

Case report We report the case of a term female newborn (38,1 weeks of gestational age, birth weight 2900 g) with a prenatal diagnosis of bowel dilatation and polyhydramnios at 33 weeks of gestation. She was born by vaginal delivery in a secondary hospital. In the first hours of life she presented with abdominal distention, bilious vomiting and an abdominal x-ray highly suspicious for intestinal obstruction. Therefore, she was referred to a tertiary hospital where a type IIIb ileal atresia was intraoperative diagnosed and an end to end anastomosis with skin bridge ileostomy was performed on the 3rd day of life.

The neonatal screening performed on the 1st day of life showed an immunoreactive trypsinogen (IRT) of 146 ng/ml, within normal limits according to age. In the second screening performed on the 9th day of life elevated levels of IRT were detected and at the age of 3 weeks a sweat test confirmed the diagnosis of cystic fibrosis.

The postoperative period was characterized by feeding problems, malabsorption and insufficient growth, which ameliorated after the beginning and adjustment of the pancreatic enzyme replacement therapy, multivitamin supplementation and stoma closure.

The genetic analysis revealed two heterozygous mutations in CFTR genes : F508del and 1717-1G>A.

Conclusion Prenatal diagnosis and neonatal screening are crucial for early diagnosis and treatment of newborns with intestinal obstruction and cystic fibrosis.

The diagnosis of foetal bowel dilatation with polyhydramnios demands the involvement of a multidisciplinary team (perinatal care, neonatology and neonatal surgery) to provide best treatment at birth.

Starting pancreatic enzyme replacement therapy after pathological neonatal screening, before confirmation of the diagnosis of cystic fibrosis could be considered depending on the severity of symptoms.

Increasing awareness of less common clinical presentations of cystic fibrosis such as jejuno-ileal atresia avoids missing or delaying the diagnosis in presence of potentially misleading false negative results of neonatal screening.

Bronchopulmonary dysplasia (BPD): development of a predictive scoring system for premature infants The BPD-Risk-Score

Ikbel El Faleh [1, 2], MD, Mark Adams [3], PhD, Mohamed Faouzi [4], PhD, Roland Gerull [5], MD, Jamel Chnayna [1], MD, Eric Giannoni [1], MD, Matthias Roth-Kleiner [1], MD, for the Swiss Neonatal Network

[1] Clinic of Neonatology, University Hospital and University of Lausanne, Switzerland, and [2] Department of Pediatrics, Hôpital Neuchâtelois, Neuchâtel, Switzerland, [3] Department of Neonatology, University Hospital Zurich, [4] Center of Clinical Epidemiology, Institute of Social and Preventive Medicine, University of Lausanne, Switzerland, [5] Department of Neonatology, University Hospital UKBB, Basel, Switzerland

Objective To develop and validate a predictive risk score for bronchopulmonary dysplasia (BPD), according to two clinically used definitions:

1. New-BPD: need for supplementary oxygen (≥ 12 hours per day) during ≥ 28 cumulated days between birth and 36 weeks of gestation (WG)
2. Old-BPD: need for supplementary oxygen at 36 WG

Method Logistic regression was performed in a national cohort (Minimal Neonatal Data Set (MNDS) of the Swiss Neonatal Network including all infants with birth weight < 1501g and/or between 23 0/7 and 31 6/7 WG in years 2009-2010, n=1488) to identify predictors of BPD. The BPD-Risk-score was built as the sum of these factors, weighted according their ß-coefficients. Internal validation of the score was performed using the bootstrap method (repeated 1000 times). The discriminative properties of the score was analysed by calculating the Area under the ROC (Receiver Operating Characteristics) Curves (AUC). Calibration was assessed with the use of the Hosmer-Lemeshow goodness-of-fit test.

This calculated BPD Risk Score was then applied to another population with the same inclusion and exclusion criteria (MNDS of the years 2014 and 2015, n=2006) in order to obtain an external validation.

Results Incidence of New-BPD varied from 21.5 % to 11.2 % in the national derivation cohort (MNDS 2009-2010) and from 25.1 % to 10.5 % in the validation cohort (MNDS 2014-2015), according to first or second definition respectivily.

Gestational age, birth weight's corresponding z-score, antenatal corticosteroid treatment, surfactant administration, proven infection, patent ductus arteriosus requiring medical or surgical treatment and sum of days of mechanical ventilation were identified as independent predictors of BPD. The AUC of the score in the derivation cohort was 0.90 and 0.89 for 1st and 2nd definition respectively. In the validation cohort AUCs were 0.91 and 0.87. The score was well calibrated in derivation and validation cohorts for both definitions with the Hosmer-Lemeshow test being not significant.

Conclusion This BPD-Risk-Score is a simple score to predict respiratory outcome for premature infants using 7 variables at any day of life. The score performed well in an external validation and might be a useful tool for clinical practice and neonatal research to identify early patients with a high risk for BPD.

Evaluation of exposure to vancomycin in neonates with current dosing approaches

Kim Dao [1], Monia Guidi [1, 2], Pascal André [1], Eric Giannoni [3], Aline Fuchs [5], Marc Pfister [5], Thierry Buclin [1], Chantal Csajka [1, 2, 4]

[1] Service of Clinical Pharmacology, Lausanne University Hospital, Lausanne, Switzerland, [2] School of Pharmaceutical sciences, University of Geneva, Geneva, Switzerland, [3] Clinic of Neonatology, Department Mother-Woman-Child, Lausanne University Hospital, Lausanne, Switzerland, [4] Service of Pharmacy, Lausanne University Hospital, Lausanne, Switzerland, [5] Pediatric Pharmacology and Pharmacometrics, University of Basel Children's Hospital, UKBB, Spitalstrasse 33, 4031 Basel, Switzerland.

Aims and objectives There is no single consensus regarding optimal dosing of vancomycin in term or preterm neonates. Current dosing recommendations are based on age, kidney function and/or body weight to define a starting dose. Our objectives were (i) to develop a comprehensive population PK model of vancomycin in the largest cohort of neonates and (ii) to evaluate and compare the performances of current dosing approaches with respect to target attainment, using simulations based on our model.

Materials & methods A total 405 neonates provided 1848 vancomycin concentrations during routine TDM. A one-compartment model with linear elimination incorporating covariates such as age, kidney function and body weight was developed (NONMEM®). The final model was applied to simulate vancomycin exposure for 20 dosing guidelines identified. Proportions of patients within and above target exposure were used as a performance measure. Target attainment meant AUC24/MIC ratio of 400-700 h and minimal drug concentration of 10-20 mg/L on days 1 and 7.

Results Median proportions of neonates within and above target exposure were 44 % (IQR 27–64 %) and 1 % (IQR 0–4 %) on day 1, and 47 % (IQR: 43–54 %) and 16 % (IQR: 7–19 %) on day 7, respectively. Only 2 out of 20 current recommendations (Neonatal Formulary 7 and Neofax® meningitis regimens) ensured target attainment in at least ~60 % of neonates on day 1 and 7.

Conclusions Most current vancomycin dosing regimens failed to achieve target attainment in a majority of neonates. Insufficiently dosed regimens should be avoided, especially in centers with widespread coagulase negative Staphylococci. Adding a loading dose to simple regimens is predicted to increase the proportion of early target attainment. Complex regimens seem to marginally improve exposure.

A case of congenital secretory diarrhea with GUCY2C mutation

T. Restin [1, 4], A. R. Janecke [2], H. Aichner [1, 4], R. Fierling [1, 4], U. Moehrlen [3, 4], D. Bassler [1, 4], T. Müller [2], M. Schmid [1, 4]

[1] Department of Neonatology, University Hospital Zurich, Zurich, Switzerland, [2] Division of Human Genetics, Medical University of Innsbruck, Innsbruck, Austria, [3] Department of Pediatric Surgery, University Childrens' Hospital Zurich, Zurich, Switzerland, [4] University of Zurich, Switzerland

Aims and objectives Congenital sodium diarrhea is a rare cause for severe metabolic acidosis and hyponatremia. This case report focuses on a preterm boy with congenital secretory diarrhea, finally diagnosed with a GUCY2C mutation. The exclusion of possible reasons for severe sodium loss in the newborn helped to find the correct diagnosis.

Materials and methods We report of a preterm boy who was born and treated at the Department of Neonatology of the University hospital in Zurich. The clinical history is presented and put into context with prior publications about this rare congenital defect.

Case report Prenatal ultrasound demonstrated dilated bowel loops and a prominent polyhydramnios suggesting intestinal atresia. The 2-kg-newborn was delivered via cesarian section at 27 6/7 weeks due to pathological umbilical cord doppler ultrasound and pathological cardiotocogram. The newborn showed a poor postnatal adaption (Apgar at 1/5/10min of life: 1/5/5) and was therefore intubated. High respiratory support was applied to ensure adequate oxygenation (maximum peak inspiratory pressure 50, PEEP 10, fiO2 1). Additionally, the baby presented with massive abdominal distension. Circulatory support via fluid application, combined with extensive catecholamine support was needed. Dehydration was suspected despite high fluid substitution of 180ml/kg/day. Metabolic acidosis became worse at day three of life (pH 7.16, Co2 4.8kPa, BE-14mmol/l). Full parenteral nutrition was installed. Gastric fluid levels were widely inconsistent varying between 0 to 50ml every 3 hours and abdominal surgery did not show any intestinal obstruction. On day three of life, sodium levels below 130mmol/l were detected and only sodium substitution higher than 30mmol/kg helped to normalize blood sodium levels. Consequently, congenital sodium diarrhea was suspected. Sequence analysis in genomic leukocyte-derived DNA samples detected a de-novo GUCY2C p.Thr783Ile mutation, confirming the clinical diagnosis.

Conclusions If low sodium levels cannot be explained by increased intravascular water content, sodium loss has to be suspected. These losses may be caused by decreased levels of aldosterone or cortisone, by vomiting, diarrhea or impaired skin barrier (burns). If there is a prenatal history of polyhydramnios and massive bowel dilatation combined with postnatal impressive loss of water and sodium, sodium diarrhea is one – rare – differential diagnosis.

Air leaks and prenatal exposure to magnesium sulfate: coincidence or causality?

Arrizza Chiara, Keller Fabian, McDougall Jane, Humpl Tilman

Department of Paediatrics, Division of Neonatology, University Childrens's Hospital Bern Switzerland

Aims and objectives Pulmonary interstitial emphysema (PIE) and pneumothorax are known complications of mechanical ventilation in preterm infants. However, restrictive use of mechanical ventilation and the implementation of non-invasive respiratory support such as continuous positive airway pressure (CPAP) is also associated with air leaks. Antenatal steroids and surfactant administration are among the most important strategies to reduce the risk of air leaks. We try to identify risk factors in newborn infants with air leaks, which could potentially modify their postnatal management

Materials and methods We retrospectively searched our institutional database for patients born between 2004 and 2018, who developed PIE and/or pneumothorax (PNX). Babies with congenital pulmonary, cardiac, abdominal anomalies or genetic syndromes were excluded.

Results 79 infants were identified with a mean gestational age of 28,2±4,1 (range 24-40 weeks). The rate of completed antenatal steroids was 65,8 %. Spontaneous PNX occurred in 2 infants (2,5 %) before starting any respiratory support, 12 (15,2 %) developed PIE or PNX after introduction of CPAP, 65 (82,3 %) presented with PIE or PNX related to intubation and/or mechanical ventilation. Interestingly, 10/12 (83 %) infants on CPAP were prenatally exposed to magnesium sulfate. Three other infants prenatally exposed to magnesium sulfate needed mechanical ventilation immediately after birth and developed severe PIE in the first hours of life. PIE/PNX in the remaining patients were complications of prolonged mechanical ventilation.

Conclusions We observed a high incidence of air leaks in preterm babies prenatally exposed to magnesium sulfate and requiring CPAP after birth. However, magnesium sulfate still represents the mainstay of therapy for pregnant women with preeclampsia. The neuroprotective effects of magnesium sulfate on the fetus are well-known but little is known about the effects on respiratory outcome. Further studies are needed to assess the possible association between prenatal exposure to magnesium sulfate and air leaks in order to provide a favorable approach to manage respiratory problems in this population.

A surprising cutaneous rash in a well preterm neonate

Cristina Felice-Civitillo [1], Sandra Asner [2], Tarik Azzi [2], Carole Gengler [3], Lydie Beauport [1]

[1] Clinic of Neonatology, Department of Mother Women and Child, CHUV, Lausanne, [2] Clinic of Infectiology, Department of Mother Women and Child, CHUV, Lausanne, [3] Clinic of Pathology, Department of Pathology, CHUV, Lausanne

We report a baby girl born to a 34 year old, gravida 5, para 1, mother. She had two miscarriages, a previous preterm delivery at 29 weeks and a late abortion due to chorioamnionitis. Her pregnancy was threatened by preterm labour at 25,3 weeks, without premature rupture of membranes. She received corticosteroids and antibiotics. No bacteria or yeast were identified in urine or vaginal smear. At 29 weeks, she started labour and a C-section was performed. The baby, weighting 1100 g, required 2 minutes of bag mask ventilation and nasal CPAP with 0.21 FiO2 for mild respiratory distress. Apgar was 1/8/9. Her clinical exam revealed a maculopapular skin rash on her chest. She was started on a 48-hour antibiotic course with amoxicillin and gentamycin for a suspected chorioamnionitis. Laboratory tests were normal with the exception of leucopenia and neutropenia. Candida albicans was documented from placental and newborn's cutaneous swabs. Urine revealed no candida. No lumbar puncture was performed. The suspicion of a congenital cutaneous candidiasis was raised and fluconazole 12 mg/kg IV was started at 36 hours of life. Further evolution was reassuring, even though there was an initial progression of the rash with further desquamations. The leucocyte counts progressively increased to 40 G/l with elevated Beta-D-glucan levels (> 500 pg/ml), raising suspicion of invasive candidiasis. She was continued on fluconazole for a total of 2 weeks. Abdominal and cerebral ultrasounds as well as eye exam were normal. Placental pathology confirmed chorioamnionitis with fetal inflammatory response. Retrospectively, maternal history revealed oral mycosis one week before delivery and lymphopenia since the last pregnancy, raising the suspicion of an underlying immunosuppressive condition.

Congenital cutaneous candidiasis is a rare disorder and can occur regardless of mode of delivery or presence of membrane rupture. Its clinical presentation at birth varies from erythematous to papular or pustular cutaneous rash, but also with peeling, flaking or desquamation. Typical localisations are: back, extensor surfaces of the extremities and skin folds. Chorioamnionitis and funisitis are almost always associated. Preterm and low birth weight babies are particularly at high risk to develop disseminated disease and adverse outcome, especially in case of delayed or inadequate treatment. Prompt systemic antifungal treatment with fluconazole prevents dissemination and reduces mortality.

Unexpected finding in a routine cerebral ultrasound in a preterm infant

Stefan Vorwieger[1,2], B.Gers[3], C. R. Kahlert[4], B.Rogdo[2,5], A.Malzacher[1,2]
[1] Department of Neonatology, Women's Hospital, Kantonsspital, St. Gallen, [2] Perinatal Centre in Eastern Switzerland, St. Gallen, [3] Department of Neurosurgery, Kantonsspital St.Gallen, [4] Department of Infectiology, Children's Hospital in Eastern Switzerland St. Gallen, [5] PICU/NICU, Children's Hospital in Eastern Switzerland St. Gallen

Key words preterm infant, brain abscess, Citrobacter koseri

Citrobacter species are known to cause brain abscesses in preterm infants, a condition associated with severe short- and long-term morbidity and carrying a high risk of mortality.

We report an unusual case of a brain abscess caused by Citrobacter koseri in a preterm twin, born at 32 weeks of gestation. The clinical course was uneventful until day 23 of life, when a routine cranial ultrasound revealed two cystic medullary layer lesions situated bihemispheric frontal and parieto-occipital. Previous ultrasound examination was normal. At that time the male infant was asymptomatic, with a completely normal septical work-up, including an unremarkable cerebrospinal fluid (CSF) analysis.

In view of the two cerebral cysts of unknown aetiology and neither clinical nor laboratory signs of an infection, we decided not to start antibiotic treatment directly but prepare a puncture in case the size of the cysts will increase and performed a frequent neuro-imaging follow-up by cranial ultrasound and magnetic resonance imaging. As the cysts showed an increase in size we enforced the diagnosis by ultrasound guided neuro-surgical puncture of one cyst which yielded the diagnosis of a Citrobacter koseri brain abscess by culture.

After diagnostic puncture, intravenous antibiotic treatment was given for 5 ½ weeks, leading to an almost completely regression of the lesions at discharge from our hospital and to the point of disappearance during out-patient follow-up. We conclude that brain abscesses in preterm infants could be present without clinical signs or evident laboratory findings. A pragmatic conservative, observant approach was feasible in this asymptomatic presentation of Citrobacter koseri brain abscesses.

The impact of newborns on their parents' quality of life

Anja Xue[1,2], Pearl de La Marca[2,3], Tanja Karen[1,2], Giancarlo Natalucci[1,2], Barbara Dinten[1,2], Dirk Bassler[1,2], Tanja Restin[1,2]
[1] Department of Neonatology, University Hospital Zurich, Zurich, Switzerland, [2] University of Zurich, Zurich, Switzerland, [3] Department of Psychology, University of Zurich, Zurich, Switzerland

Background, aims and objectives The number of preterm infants is constantly increasing. Preterm infants are at risk for a range of disease conditions such as low birth weight, persistent ductus arteriosus, bronchopulmonary dysplasia, cerebral hemorrhage, retinopathy and necrotizing enterocolitis. This prospective research project assesses the families' quality of life and possible protective factors which contribute to the reduction of anxiety and fear. Differences between parental quality of life of term born versus preterm infants with or without major morbidities will be analyzed during the first three months. The fathers' quality of life and the couple's relationship will be further important topics of research in this project, as previous studies have mainly focused on mothers alone.

Material and methods We apply a questionnaire including standardized questions: SF-12, EPDS (Edinburgh Postnatal Depression Scale), CSI (Couple Satisfaction Index) and TOPSE (Tool to Measure Parenting Self-Efficacy). The SF-12 is a questionnaire used to measure the health related quality of life of adults. The EPDS, CSI and TOPSE will be used to find possible risk and protective factors for families more likely of having a lower quality of life. Additionally, the genoecogram is used for the assessment of parents' social network. We will additionally assess secondary endpoint parameters such as gestational age, birth weight, duration of hospitalization, socioeconomic status, number and age of siblings and complications.

Research plan This is a prospective research project; the recruitment period will take place continuously at the University Hospital of Zurich. Parents of 150 term and 150 preterm infants will be asked to respond to a questionnaire on their quality of life at different points during and after the hospitalization of their child (preterm infants: at birth, at corrected term, after 3 months; term infants: at birth, after 6 weeks, after 3 months).

Hypothesis Our first hypothesis is that the parental quality of life of term infants is higher than those of preterm infants. The second hypothesis is that the parental quality of life of preterm infants with major morbidities is lower than the parental quality of life of preterm infants without major morbidities. We expect that the social support network of parents (number and quality of related friends and family) and their perception of their relationship with the child has a great impact on their quality of life.

Piebaldism: a case report and short review of literature

Ms Selei Hamed
Kantonsspital Aarau, Tellstrasse 25, 5001 Aarau

Introduction Piebaldism is a rare autosomal dominant genodermatosis. Clinical manifestation is present at birth, with typical distribution of depigmented skin areas and a white forelock.

Case A female term infant born by vaginal delivery at 40 4/7 weeks of gestation had a white forelock and areas of depigmented skin on the anterior trunk and extremities, and café-au-lait macules. Further clinical examination was normal. A genetic evaluation revealed suspicion of piebaldism, confirmed by genetic testing which showed a heterozygous mutation of the c-kit gene (KIT:c.2362T>A/p.Cys788Ser). The other family members didn't show signs of piebaldism. The parents have not been genetically tested yet. Today the child is 2 years old with normal development and no other illness noted up to date.

Discussion Incidence of piebaldism is less than 1 : 20 000, no difference in gender distribution. It is due to a mutation in the c-kit proto-oncogene located on chromosome arm 14q12 which encodes KIT, a type 3 transmembrane receptor for mast cell growth factor. This affects the differentiation and migration of melanoblasts from the neural crest during the embryogenesis. At birth, patients present with depigmented skin and hair (forelock) areas due to absence of melanocytes. The white forelock can be the only sign in 80- 90 % of patients. The depigmentation of the skin is typically symmetric, well-circumscribed and irregular, involves often the forehead, the anterior abdomen extending to the chest, the lateral trunk, mid-arms and legs and typically spares the dorsal spine, hands and feets. The white patches are often accompanied by hyperpigmented macules on depigmented as well as unaffected adjacent skin. The disease is typically stable and persists lifelong. Additional hyperpigmented macules may develop. Partial or complete repigmentation can occur spontanesously or after injury. Piebaldism is mostly isolated. Some cases with combination of piebaldism and neurofibromatosis type 1 are known. Differential diagnosis of piebaldism is vitiligo, albinism, Waardenburg syndrome and neurofibromatosis. Treatment is challenging. The use of sunblocking agents are recommended. Approaches mentioned in literature are camouflage, autologous cell suspension transplantation and autologous punch grafting.

Conclusion This report describes a newborn with piebaldism and gives a short review of literature. Differentiation of piebald-like syndromes with association to other anomalies is important.

"Blueberry-Muffin-Baby": a misleading clinical presentation of severe Langerhans cell histiocytosis

Rahel Wettstein, Romaine Arlettaz-Mieth, Michael Wolff
Klinik für Neonatologie, Universitätsspital Zürich

Background "Blueberry-Muffin-Baby" describes the presence of dermal erythropoiesis clinically appearing in a variable manner of focal isolated up to generalized papules or hemorrhagic purpuric eruptions. Most commonly associated diseases are connatal infections like TORCH (toxoplasmosis, other, rubella, cytomegalovirus, herpes) or parvovirus, but also malignancy and hematologic disorders.

Langerhans cell histiocytosis (LCH) is a rare disease with a wide spectrum of clinical presentation, from localized to disseminated manifestations. The diagnostic method of choice is biopsy, showing a proliferation of Langerhans cells. The therapy varies depending on the organs involved, but in severe cases chemotherapy is indicated.

Case presentation A male late preterm was born as the first child of healthy parents. During the uneventful pregnancy organ screening was without pathologic findings. Cesarean section performed because of pathologic CTG. The baby presented without spontaneous breathing turning into an Apgar score of 3/4/6. Moreover the whole skin was covered with disseminated red-blueish cutaneous and subcutaneous papules, hemorrhagic crusts and petechia mimicry a Blueberry Muffin phenotype.

The lesions also included the oral mucosa. In addition the boy presented with a distinctive hepatosplenomegaly and developed a pronounced hyperbilirubinemia.

Laboratory findings suggested first a congenital CMV (weak positive PCR in urine) which could be ruled out next to the other important connatal infections. In the peripheral blood count there were no anemia, no signs of leukemic blasts but a severe and lasting thrombocytopenia. The skin biopsy was performed and lead to the post mortem diagnosis of disseminated LCH with severe skin involvement. On the fifth day of life the patient was showing signs of multiple organ failure and died despite maximal intensive care on the sixth day of life.

Conclusion A patient presenting with "Blueberry-Muffin"-phenotype should lead to most common diagnostics to rule out connatal infections. Nevertheless differential diagnosis for further diseases like leukemia, neuroblastoma and LCH should also be considered early. Especially in critically ill patients specialists should be involved early and biopsy of unknown skin lesions should be performed.

Portal vein thrombosis after umbilical vein catheterization – report of cases

Catarina Valpacos, Julie Sommer, Valérie McLin MD, Marie Saint-Faust MD
Pediatric Department, Geneva University Hospitals, Geneva, Switzerland

Umbilical vein catheter (UVC) insertion is a common procedure, daily performed in a neonatal intensive care unit; however, it is not complication-free. Indeed, vein thrombosis, hematomas, abscesses, and parenchymal perforations have been described and are probably underestimated. Both the direct epithelial damage due to the progression of the UVC, and the reduction in the portal flow due to the presence of the catheter in the vascular lumen, put newborns with UVC at high risk of developing portal system thrombi. At the same time, the mere presence of the device increases the risk of sepsis, a pre-thrombotic condition, which interferes with the fragile hemostatic balance of the newborn, particularly in preterm infants. Its incidence is highly variable according to different studies, but it may reach 43 %.

We report 8 cases of left portal vein thrombosis following umbilical vein catheterization. They were all admitted after birth in the unit of neonatology of the Geneva University Hospitals and were followed after discharge. Seven of them were preterm infants (between 23+6 and 34+3 weeks of gestational age), and one was a full-term newborn. All UVC were placed in the first day of life and were removed after an average of 8.13 days (4-18 days). Thrombi were identified after an abdominal doppler ultrasound exam; in half the cases, the procedure was deemed necessary when a thrombocytopenia persisted. Seven patients had a complete recovery with spontaneous vascular repermeabilization after an expectative approach. Three of the 8 infants showed an unfavorable evolution towards a cavernoma, but only 1 patient developed an extra-hepatic occlusion with portal hypertension. Secondary sepsis occurred in 7 cases, all treated by antibiotics; in only 6 infants a microbiological agent was identified in cultures of the UVC tip.

Cases of UVC associated thrombosis require a high level of suspicion at the onset phase, since newborns are frequently asymptomatic or present unspecific clinical signs. Even though spontaneous repermeabilization is the most common outcome, complications as cavernomas, with subsequent portal hypertension, may occur and have a substantial impact on morbidity and mortality. These cases illustrate the need to pay attention to each UVC insertion and show the importance of quality studies in this domain, not only to understand risk factors, but also to stimulate reflection on catheter placement indications and, ultimately, screening guidelines.

Gestational age versus birth weight – evaluating resource consumption und reimbursement in neonatal medicine at a Swiss tertiary care center using routinely collected health data from 2016-2017

Karen Triep[1], Nenad Torbica[1], Luigi Raio[2], Mathias Nelle[3], Olga Endrich[1]
[1] Medical Directorate, Inselspital, University Hospital of Bern, Berne, Switzerland, [2] Department of Obstetrics & Gynecology, University Hospital of Bern, Berne, Switzerland, [3] Neonatology Division, Inselspital, University Hospital of Bern, Berne, Switzerland

Introduction Prematurity is related to a higher risk of health impairment compared to term births. Preterm infants are affected by adverse outcomes and therefore associated to higher costs for health care, especially those small-for-gestational-age. Thus, gestational age (GA) and birth weight (BW) are related to resource consumption. In high income countries data for both variables show high validity. With a preterm birth rate of 4-10 % in these countries the economic impact is high. Different national health care systems using Diagnosis Related Groups (DRG) for reimbursement implemented the variable BW for calculating prices, but only recently the Australian Independent Hospital Pricing Authority has introduced GA. The objective of our study was to evaluate GA as a potent variable to assign costs of inpatient care in comparison to BW. Routinely collected health data from the Inselspital Berne, a Swiss tertiary care center, were used.

Materials & methods The study included anonymized inpatient cases of the Inselspital from 2016 – 2017 admitted by delivery, transfer and readmission <28 days of age. Stillborns (n=40), cases with costs for operation room procedures (n=195), early deaths and transfers (until 5 days of age; n=128) and cases with missing values for GA (n=222) were excluded. Ethic consent was not required, the study was regarded as quality surveillance.

The study population (n=5198) was categorized by GA groups (extremely preterm <28, n=42; very preterm 28 0 – 31 6, n=155; moderately preterm 32 0 – 33 6, n=196; late preterm 34 0 – 36 6, n=581; early term/term >37 0 wks, n=4224) and by BW groups (SwissDRG categories <750, n=15; 750-999, n=44; 1000-1249, n=59; 1250-1499, n=78; 1500-1999, n=287; 2000-2500, n=437;

>2500g, n=4285). We conducted linear and multiple regression models with predictors GA, BW, GA+BW, GA*BW for each categorized group and calculated VIF, AIC, BIC, QQ Plots.

Clinical cases & summary results Both BW and GA are significant predictors for case related costs in the linear regression model (BW R2=0.35, p<0001 ; GA R2=0.47, p<0001). By testing GA and BW for multicollinearity we could show, that it does only slightly impair the efficacy of the model (VIF 1.14).

BW categorization by SwissDRG groups : GA as a predictor is significant in the BW-categories (1250-1499g, R2=0.32 ; 1500-1999g, R2=0.36 ; 2000-2500g, R2=0.315 ; all p<0.0001), whereas BW is not (R2=0.06/0.12/0.03). There is a remarkable difference of R2 especially in the group of 2000-2500g between GA and BW as a single predictor (R2=0.32 versus 0.03). GA+BW combined show a better impact than GA alone (<750g, R2=0.62 ; 750-999g, R2=0.10 ; 1000-1249g, R2=0.21 ; 1250-1499g, R2=0.33 ; 1500-1999g, R2=0.38 ; 2000-2500, R2=0.32). The group of cases >2500g shows no significant prediction for GA and/or BW.

GA categorization : GA as a predictor is significant in each GA-category (p<0.0001). However, classified by GA, GA+BW show highest prediction compared to BW or GA, especially in the group of late preterm and very preterm (R2=0.32/0.41). The combination of variables GA+BW is most effective in all GA groups.

Conclusion(s) The hypothesis that GA is a potent predictor of case related costs could be confirmed. Within the different classes an assessment of either GA or BW or a combination, probably even introducing a factor, would be highly suitable to predict costs and refine the DRG systems especially concerning specific groups as the late preterm infants.

Comparing all models, GA categories with the variables GA+BW combined give the best prediction to costs. However, with R2 being below 0.5 in most groups and models, we state that case related costs can be explained by either variable GA/BW only partially.

When the murmur does not come from the heart

Laureline Barrielle, Celine Fischer-Fumeaux, Guillaume Saliou, Anita C. Truttmann
CHUV, Av. Pierre Decker 2, Lausanne

A baby girl was born after an eventful pregnancy by C-section after 38 weeks of gestation in a private clinic. Birth weight was 3100g (P10-25), height 49cm (P25) and head circumference 36cm (P75-90). She showed immediate severe RDS for which she was put under CPAP. The initial Xray was suggestive of a Wet Lung, and a slight cardiomegaly was noted. Due to the respiratory distress, baby girl was transferred to a neonatal level III unit. The evolution was at first suggestive of a Wet Lung, for the CPAP was stopped after 3 days, but she remained tachypneic with a failure to thrive. A cardiac ultrasound performed on DOL 3 showed tricuspid regurgitation, calling to mind a minor Ebstein. High Flow Nasal Canula were started on DOL 7 due to recurrence of respiratory distress.

On DOL 9, she showed signs of decompensation due to a cardiogenic shock. The clinical exam also found a murmur heard on the fontanelle and a macrocephaly. The cardiac ultrasound showed supra-systemic pulmonary hypertension without morphologic abnormalities. The cerebral ultrasound showed then anormal Doppler flow in the left occipital lobe. She had to be then intubated and given aminergic support to treat cardiac high output failure. MRI on DOL 10 showed a malformation of the dural sinuses located next to the left occipital lobe, and explaining the high output failure.

On DOL 12, an embolisation of the malformation was performed after catheterization of the right femoral artery. 80 % of the malformation was successfully embolized. Initial evolution seemed good with extubation on DOL 15, but on DOL 18, she showed again signs of cardiac failure, leading to another intubation. A second embolization was performed on DOL 19 via the left femoral artery. Thereafter she was started on diuretics, as well as anticoagulant to prevent excessive clotting in the malformation. She was extubated on DOL 26, and was put under CPAP.

Evolution was slowly improving and CPAP was stopped on DOL 54. The neurological examination at 2 months was unremarkable with good tone, smiles, target following, the hearing tests were normal as well. MRI at 2 months of age showed important reduction of the dural sinuses malformation. Baby girl was finally discharged from the hospital when she was 2 months and 7 days old.

This case illustrates a very rare form of cerebral vascular malformations, involving the dural sinuses. This malformation carries a very high mortality and morbidity, but when treated rapidly and adequately, can be life saving. We discuss the different types of cerebral vascular malformations and their potential treatment and outcomes.