Background and aims Liquid formulations are easy to swallow but they may have disadvantages such as a bad taste, preservatives or restricted storage conditions. These disadvantages may be overcome by oral solid flexible dosage forms such as powders or mini-tablets. The aim of this study was to investigate the acceptance and preference of four oral dosage forms in children aged 1–4 years in the Netherlands.

Methods Parents administered four different placebo formulations: a 4-mm round, uncoated mini-tablet, powder, suspension and syrup at home to their (healthy) child twice on one day following a randomized cross-over design. They were asked to report the child acceptance by the result of the intake and by a child acceptance score on a 10-cm visual analogue scale (VAS). At the end of the study parents were asked to report the child and parent preference.

Results 183 children were included and 151 children were evaluated. The mini-tablet was fully swallowed in 97% of all cases, the powder 81%, the suspension 86% and the syrup 83%. The mean VAS-scores were: tablet 9.01; powder 8.00; suspension 7.78; syrup 8.04. The mini-tablet was significantly better accepted than the three other dosage forms ($p < 0.05$). The same pattern was observed when only the first intake was considered given a carry-over effect. Children and parents preferred the tablet and syrup over the suspension and the suspension over the powder ($p < 0.05$).

Conclusions All dosage forms were well accepted, but the mini-tablets were the best accepted and preferred dosage form.

218 SOCIOECONOMIC STATUS IN RELATION TO LIPID AND GLUCOSE METABOLISM IN EARLY CHILDHOOD. THE ABCD-STUDY

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Objective The objective of this study was to explore the relations of socioeconomic status to lipid and glucose metabolism as indicators of cardiovascular health in 5–6 year olds.

Methods In 1308 5–6 year old ethnic Dutch children from the ABCD cohort study, lipids (cholesterol, LDL-C, HDL-C, triglycerides), glucose and C-peptide ($n = 974$) were measured after an overnight-fast. Insulin resistance was calculated with HOMA. Using linear regression the association of lipid and glucose metabolism to socioeconomic status as indicated by maternal education and income adequacy was examined.

Results There were no differences in cholesterol, HDL-C, LDL-C, and triglycerides between socioeconomic groups. However, children with low educated mothers had on average a higher glucose ($p = 0.01$), C-peptide ($p = 0.001$), and insulin resistance ($p = 0.001$) compared to children with high educated mothers. These associations could not be explained by birth weight, maternal BMI, breastfeeding duration, and physical activity. Childhood BMI partly explains these associations, but after adjustment for BMI the association between maternal education and markers of the glucose metabolism remained significant (models controlled for age, height, and sex).

Conclusion Socioeconomic status appears to be an independent risk factor for cardiovascular function and seems to emerge in early childhood. In absence of underlying mechanisms these empirical findings are relevant for public health care and further explanatory research.

219 INCIDENCE TESTING OF HUNTER SYNDROME IN A POPULATION AT RISK - FIRST RESULTS OF A BINATIONAL SCREENING PROGRAMME

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Background Hunter syndrome (Mucopolysaccharidosis type II; X-linked inheritance; prevalence rate in Europe approximately 1:77000 male newborns) is a rare, progressive, multisystemic disease, caused by deficiency of the lysosomal enzyme Iduronate-2-sulfatase. Due to the very heterogeneous phenotype Hunter syndrome is often not diagnosed before pre-school age. This is unfortunate, because patients would significantly benefit from the earliest possible start of treatment containing enzyme replacement therapy.

Early screening methods are possible, but due to the rarity of this disease they are too expensive to be performed in all newborns. An at-risk patient population screening provides opportunity for timely identification of the patients. All children with Hunter syndrome have an umbilical hernia and about 60% develop an inguinal hernia in early childhood. This is significantly more than in the general population.

Methods Since February 2012 an at-risk population screening (male sex < 18 years old, presence of an umbilical hernia, surgery for inguinal hernia) is carried out in Germany and Austria. Test centers are over 90 surgical test centers which operate on children routinely. Patients are screened via Iduronate-2-sulfatase enzymatic assay from dried blood spots cards and an additional questionnaire.

Results and conclusion We will present the first results after 6 months study period. In case of positive screening results, a routine screening for the above mentioned at-risk population should be discussed. Simultaneously, an extended study within the European Union can then be planned and organized.

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220 CORD BLOOD CHERIMIN AND OBESTATIN IN LARGE FOR GESTATIONAL AGE INFANTS

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Background and aims Increased neonatal adiposity is associated with childhood obesity and subsequent development of adult diseases. Chemerin was recently introduced as a novel adipocytokine inducing insulin resistance and regulating maternal-fetal metabolic homeostasis during pregnancy. Obestatin is a peptide hormone involved in the control of insulin secretion and adipocyte function. This study aimed to assess circulating concentrations of chemerin and obestatin in fetal samples from large-for-gestational-age-(LGA) and appropriate-for-gestational-age-(AGA) pregnancies and investigate their association with gender, parity and delivery mode.

Methods Cord blood chemerin and obestatin concentrations were prospectively measured by enzyme-linked immunosorbent assay in 40 LGA (9 born from diabetic mothers and 31 born from non-diabetic mothers) and 40 AGA singleton full-term infants.

Results Cord blood chemerin concentrations were significantly higher in LGA compared to AGA neonates, after controlling for confounding factors ($b = 38.91$, $p < 0.001$, SE 9.29). In contrast, no significant differences in obestatin concentrations were observed between groups. Cord blood concentrations of both hormones did not depend on gender, parity or delivery mode.

Conclusions Higher chemerin concentrations in LGA neonates, possibly implying predisposition to insulin resistance, may serve as

an early prognostic marker for later development of the metabolic syndrome. Lack of difference in obestatin concentrations between LGA and AGA groups could possibly suggest that obestatin may not be directly involved in the regulation of fetal adiposity and insulin sensitivity.

221 EFFECT OF FASTING ON METABOLISM IN TRANSGENIC MICE WITH RESPIRATORY CHAIN COMPLEX III DEFICIENCY

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Background and aim To generate ATP production, mitochondria host crucial metabolic pathways that interact continuously. Therefore, pathological interruptions in one process might disturb entire cell metabolism. To investigate a neonatal mitochondrial disorder (GRACILE syndrome), we developed a mouse model with c.232A>G mutation in *Bcs1l*, resulting in a lethal complex III (CIII) deficiency in homozygotes. Our aim was to analyze how CIII deficiency affects metabolic pathways by pressing the mechanisms with fasting.

Methods Homozygous (*Bcs1l*^{G/G}) and wild type (*Bcs1l*^{A/A}) mice were assessed before and after 4-hour fasting with blood glucose, lactate and ketones, and sacrificed. Liver tissue was obtained for histology (H&E, PAS staining for glycogen and ORO-staining for fat) and ATP measurement.

Results Before fasting, *Bcs1l*^{G/G} had lower glucose (4.3±1.3 vs. 6.6±1.2, p<0.01) and higher ketone (0.6±0.3 vs. 0.3±0.1, p<0.01) levels, but similar lactate values (4.0±2.2 vs. 3.7±1.4 p=0.8). Glycogen depletion and microvesicular steatosis present in *Bcs1l*^{G/G} hepatocytes increased after fasting. After fasting, *Bcs1l*^{A/A} remained euglycemic with increased ketone body production, whereas in *Bcs1l*^{G/G} mice glucose, ketone and lactate were lower. ATP production of *Bcs1l*^{G/G} mice was lower than that of *Bcs1l*^{A/A} (58%±24%).

Conclusion *Bcs1l*^{G/G} mice switched their metabolism to β -oxidation before fasting and failed to build up compensatory metabolic mechanisms to fasting, resulting in low ATP production. These results elucidate mechanisms explaining the deterioration in *Bcs1l*^{G/G} mice. The methods used can be implemented as outcome measures in intervention studies aiming at stimulating mitochondrial biogenesis and metabolism in the mouse model.

222 BRAIN VOLUMETRY IN INFANTS WITH CONGENITAL HEART DISEASE: PRE- AND POST-SURGERY ASSESSMENTS COMPARED TO HEALTHY CONTROLS

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Background and aims MRI studies in neonates with congenital heart disease (CHD) have demonstrated delayed brain maturation and mostly focal brain injury. To better define the distribution of cerebral injuries and regional brain growth in neonates with CHD, we compared volumetric measures from pre- and postoperative MRI of patients to healthy neonates.

Methods Cerebral MRIs of 32 term-born CHD patients, scanned before and after heart surgery (mean age: 6.8 days and 26.8 days, respectively), were manually segmented to measure volumes of total, grey and white matter and of selected brain regions. Results were compared with MRIs of 17 healthy term born neonates (mean age: 23.5 days).

Results Between pre- and postoperative MRI, patients showed significant brain growth, especially in the cortical grey matter (0.25%/day), cerebellum (0.20%/day), and deep gray matter structures (0.10–0.15%/day, all p<0.004). Volume increase of the white matter was 0.05–0.06%/day (left/right; p=0.017/0.003); increase of total brain volume was 0.14%/day (p<0.001).

Compared to healthy controls, the size of all brain structures (except ventricles and right amygdala) was significantly reduced postoperatively. Largest differences were found in deep gray matter structures (13.8–16.8%, p=0.05–< 0.001), cortical grey (12.1%, p=0.01) and white matter (11.8%, p<0.001). Total brain volumes were reduced by 11.3% (p<0.001).

Conclusions In neonates with CHD, significant differences of white and deep grey matter volumes were found postoperatively. Brain growth was high, with notable regional differences. Our results contribute to the knowledge on the timing of cerebral injury in neonates with CHD.

223 CLINICAL UTILITY OF AN AUTOMATED NEONATAL SEIZURE DETECTION ALGORITHM

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Background/Aims EEG is the gold standard for the identification of neonatal seizures as the vast majority of electrographic seizures do not have a clinical correlate. Both under and over diagnosis of seizures is common in the neonatal intensive care unit (NICU). Computer assisted methods of interpreting the EEG have the potential to improve the accuracy of seizure detection. The aim of this study was to determine the clinical utility of our current neonatal seizure detection algorithm (NSDA).

Methods Multi-channel video-EEG recordings of 70 term neonates admitted to the NICU were analysed: 35 babies with seizure (mixed aetiologies) and 35 babies without seizure. The EEGs were annotated by an experienced neurophysiologist. The performance of the NSDA was assessed using time and event based metrics. An additional, clinically relevant, performance metric (based on the number of neonates correctly administered an anti-epileptic drug (AED) as early as possible after electrographic seizure onset) was calculated.

Results The sensitivity and specificity of the NSDA were 83% and 97% respectively when comparing to the experts annotation. The seizure detection rate and false alarm rate were 80% and 0.7/hr respectively. Thirty-four percent of neonates with seizures received an AED within the defined optimal timeframe, while 20% of neonates without seizure received an AED. These results were improved to 71% and 11%, respectively, by supplementing decision making with the output of the NSDA.

Conclusion Current NSDA performance, while not perfect, would greatly improve the efficacy of seizure detection and optimal AED administration in the NICU.

224 AEEG AND NIRS DURING TRANSITION AFTER BIRTH

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Background and aims Easily applicable non-invasive devices to monitor cerebral activity and oxygenation continuously during neonatal transition and resuscitation are lacking. We aimed to identify