associated with decreased HRQOL include: comorbidity, older age at transplantation, medication side effects, and parental conflict. Two specific problems in pediatric organ transplantation are adherence to medication and transition from pediatric to adult transplant care. Early disease onset, poor nutritional status, growth deficits, and longer duration of illness prior to transplant have been identified as factors contributing to an adverse cognitive development of these children.

Studies are heterogeneous regarding operationalization of HRQOL, study design, length of follow-up, and age of the children. There are only few prospective multi-center studies, which should be encouraged in future research including specific internationally accepted validated instruments.

Against the background of a new era of immunosuppressive therapy (steroid minimization, individualized therapy), a better long-term outcome in these children could be expected.

161 CHILDREN AS DONORS: A NATIONAL PEDIATRIC INTENSIVE CARE STUDY TO ASSESS PROCUREMENT OF ORGANS AND TISSUES

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Objectives Shortage of size-matched organs and of tissues is the key factor limiting transplantation in children. Empirical data on the procurement process in children is sparse. This study aimed to gain insight into the recognition of potential pediatric donors in the Netherlands and the procurement process.

Methods A national retrospective cohort study in the Dutch pediatric intensive care units. The records of 683 deceased children were analyzed by two independent donation experts and procurement process data were compared with the national protocol.

Results From 2003 thru 2006, 74 (11%) of the deceased children were found to have been suitable for organ donation and 132 (19%) for tissue donation. Sixty-two (84%) potential organ donors had been correctly identified; parental consent had been obtained and donation effectuated in 26/62 children (42%). Sixty-three potential tissue donors (53%) had been correctly identified; parental consent had been obtained and donation effectuated in 17/63 children (27%).

Conclusion Recognition of pediatric organ donors by medical professionals is acceptable; recognition of tissue donors may be improved. Efforts to address the shortage of organs and tissues for transplantation in children should focus on the gap between recognition of donors and parental consent. We suggest such studies should not only assess the process itself, i.e. the competencies of the professional staff (micro-level) but also the influence of legislation, societal views on donation by children, and the potential relevance of children's views on donation (macro-level).

162 CONGENITAL CHLORIDE DIARRHEA: A SINGLE CENTRE EXPERIENCE WITH 43 CHILDREN

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Background Congenital chloride diarrhea(CCD), A rare deadly autosomal recessive disorder of chronic diarrhea in infancy.

Methods Patients diagnosed with CCD between1986–2009 were studied. The demographic data, clinical findings and biochemical findings were collected and statistically analyzed.

Results Forty-three patients (28M/15F) had CCD. Fifteen patients (35%) were diagnosed after one year of age (late referral or misdiagnosis as Bartter syndrome). Premature delivery in 24 cases (55.8%). Polyhydramnios in 26 pregnancies. All patients were distributed among 19 families with 33 children being the outcome of consanguineous marriages. Intractable diarrhea was the presenting symptom in 40patients (93%), Biochemical data revealed: Serum potassium (1.3–4.1, mean 2.4Mmol/l), s. chloride (39–95, mean76.2Mmol/l), s.bicarbonate (22–54) meam-37.6 Mmol/). Fecal chloride (134±21.6, mean±SD)(range 90–205). The fecal chloride over fecal sodium plus potassium ratio was 0.6 (1.1±0.3, mean ± SD) (N.=0.2). Associated disorders were: chronic renal failure 7 (16%), congenital anomalies 8 (19%), mental retardation4 (9.3%) seizures 8 (19%), and brain atrophy 4 (9%). Complications were seen mostly among patients with late referral or poor compliance. At diagnosis, 35 (81.4%) cases were below –2SD for weight for-age, 31 (72%) for weight-for-height, and 31 (72%) for height-for-age. Children under five years of age showed improvement in weight for height as compared with older children.

Conclusions CCD is a treatable cause of intractable diarrhea in infancy.

Awareness, early diagnosis and proper management are essential in preventing irreversible and long-term organ damage and a better outcome compared to those with late referrals.

CCD is to be considered in infants with severe persistent diarrhea where a high rate of consanguineous marriage prevails.

163 LIVING DONOR LIVER TRANSPLANTATION FOR ALAGILLE SYNDROME: RECIPIENT CHARACTERISTICS AND OUTCOME IN A SINGLE CENTER

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Background and aims Alagille syndrome (AGS) is rare in Taiwan. The aim of this study was to review our institutional experience with liver transplantation (LT) for AGS.

Methods We performed a retrospective analysis of transplant records of patients diagnosed as AGS and underwent LT between 1987 and 2010. Nine patients underwent living donor LT.

Results Cholestasis and characteristic faces were seen in all patients. Posterior embryotoxon was seen in 4/9 (44.4%), butterfly vertebrae in 3/9 (33.3%), heart defect (pulmonary stenosis in 2) in 3/9 (33.3%), and renal disease in 2/9 (22.2%) patients. Iminodiacetic acid scans showed no excretion of isotope into the bowel after 24 hours in 4/9 (44.4%). A small gallbladder on ultrasonography was noted in 3/9 (33.3%) and suggested a false diagnosis of biliary atresia. All underwent diagnostic laparotomy and liver biopsy. Liver biopsy showed characteristic features of paucity of interlobular bile ducts in all patients. Kasai portoenterostomy was not performed in any patient before being referred for LT. The mean age at time of LT was 4.6 years. The 5-year overall survival rate after living donor LT was 88.9%.

Conclusions Our conclusion is that the clinical features of AGS are informative. Histological confirmation is important in the diagnosis. These findings support the concept that infants with liver diseases warrant early referral to a specialist center.

164 LINEAR GROWTH AFTER PEDIATRIC LIVER TRANSPLANTATION

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To determine growth patterns in a children undergoing liver transplantation, the outcomes of orthotopic liver transplantations performed in 10 children at Hamad General Hospital between October 2005 and October 2009 were reviewed. The mean age at transplantation was 27 +/– 30 months; 80% of the children were females. The

transplants were from living-related donors. At the time of transplantation the mean height z score was - 1.15 +/– 1.7 and BMI z score was 0.44 +/– 1.8. Eighteen months after transplantation, catch-up growth was seen in 40% of children, 30% had normal linear growth without any catch-up and 30% had slow growth rate after transplantation. Children with evidence of catch-up growth (growth velocity z score >0) had more growth retardation at the time of transplantation, and were receiving lower doses of prednisone at 1.5 years after transplantation. Younger infants (below 6 months) were most likely to demonstrate catch-up growth after transplantation. In summary, a large proportion of children have growth retardation at the time of liver transplantation. Serum albumin increased significantly after (39.8+/–5.2 g/L) vs before (34 +/–11g/L)transplantation, and Alanine transferase (ALT) decreased significantly from (130+/–260U/L) to (30+/–15U/L). Poor growth after transplantation occurred more in those receiving higher doses of corticosteroid. This growth retardation is inversely correlated with age. Growth after transplantation is proportional to the degree of growth retardation at transplantation and inversely correlated to age at transplantation.

165 THE METABOLOMIC PROFILE OF UMBILICAL CORD BLOOD IN NEONATAL HYPOXIC ISCHAEMIC ENCEPHALOPATHY

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Background Hypoxic ischaemic encephalopathy (HIE) is associated with the activation of multiple biochemical pathways. The importance of these pathways individually, and that of their interaction, in the disease process is not fully understood. The aim of this study was to describe and quantify the metabolomic profile of umbilical cord blood samples in a carefully defined population of full-term infants with HIE.

Methods Full-term infants with perinatal asphyxia (with and without HIE) and healthy controls had umbilical cord blood drawn and bio-banked at –80°C, within 3 hours of birth. A combined direct injection and LC-MS/MS assay (AbsolutIDQ p180 kit, Biocrates Life Sciences AG, Innsbruck, Austria) was used for the metabolomic analyses of the samples. The degree of encephalopathy among those with asphyxia was defined using both continuous multichannel-EEG in the first 24 hours, and modified Sarnat score.

Results 142 neonates were included in the analysis (HIE=31, asphyxia without encephalopathy=40, controls=71). There was a significant alteration ($p<0.01$) in 29 metabolites from 3 distinct metabolite classes (Amino Acids, Carnitines, and Phosphatidylcholines) between study groups. 13 of these metabolites were significantly altered between HIE and controls. Cross-validated Partial Least Square Discriminant Analysis models were developed to distinguish between the groups. The HIE model differentiated significantly between HIE, and those without HIE (AUC=0.93, $R^2 = 0.36$, $Q^2 = 0.25$).

Conclusion The description of the metabolomic profile from umbilical cord plasma and the specific metabolite signature associated with HIE, offers insight into the disease mechanism and the possibility of an early screening test.

166 SIMULTANEOUS ^{31}P-MAGNETIC RESONANCE AND NEAR-INFRARED SPECTROSCOPIC INVESTIGATION OF BRAIN TISSUE OXYGENATION, CYTOCHROME-C-OXIDASE AND INTRACELLULAR METABOLITES DURING PERINATAL CEREBRAL HYPOXIA-ISCHAEMIA

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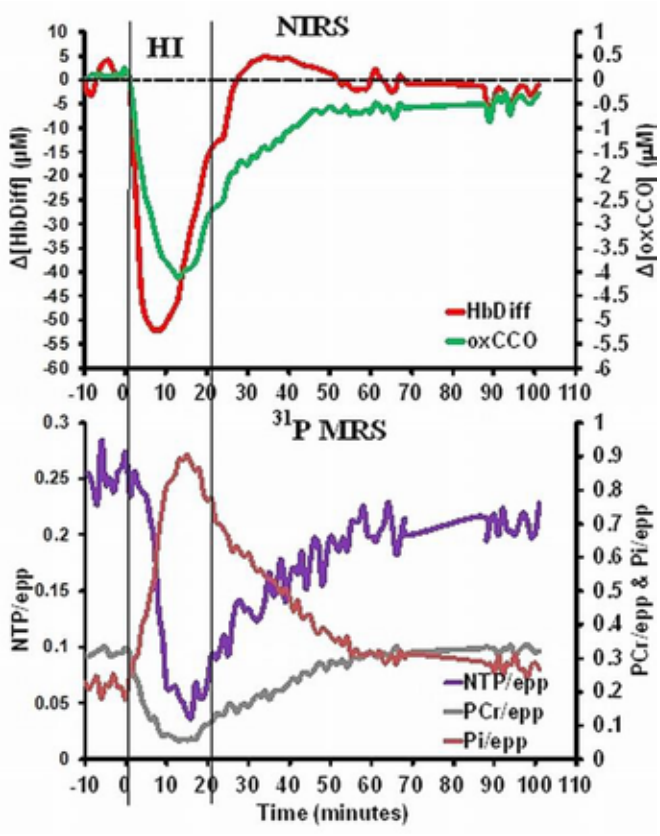
Background Hypoxic-ischaemic neonatal encephalopathy is associated with high mortality and morbidity rates worldwide.

Aims To investigate brain haemodynamic, cytochrome-c-oxidase (CCO) and energy-resource changes during transient hypoxia-ischaemia (HI) and recovery using simultaneous broadband near-infrared spectroscopy (NIRS) and phosphorus (^{31}P) magnetic resonance spectroscopy (MRS).

Methods Nine healthy piglets (aged < 24 hr) were anaesthetised and physiologically monitored. Transient cerebral HI (duration 20 minutes) was induced by reducing the inspired oxygenation and reversibly inflating bilateral carotid artery occluders. Using ^{31}P MRS we measured inorganic phosphate (Pi)/epp, phosphocreatine (PCr)/epp, and nucleotide triphosphate (NTP)/epp where epp=exchangeable phosphate pool=Pi+PCr+3NTP. NIRS measured cerebral concentration changes of oxy-haemoglobin (HbO_2) and deoxy-haemoglobin (HHb), and cytochrome-c-oxidase oxidation state changes (Δ[oxCCO]).

Results Simultaneous ^{31}P-MRS and NIRS results are shown. HI rapidly reduced brain oxygenation as shown by changes in haemoglobin difference (Δ[Hbdiff]=Δ[HbO_2]-Δ[HHb])) closely followed by a fall in Δ[oxCCO]. PCr/epp fell, and Pi/epp rose, quickly while NTP/epp was buffered initially and only declined when Δ[oxCCO] was significantly lowered.



Abstract 166 Figure 1

Discussion During transient HI, CCO becomes reduced due to oxygen depletion; adenosine triphosphate levels are initially preserved by the creatine kinase reaction leading to PCr decline whereas energy utilisation without oxidative phosphorylation leads to increased Pi. Complementary MRS and NIRS enable better understanding of the cerebral metabolic response to HI and can help evaluate early interventional therapies.