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DEXAMETHASONE INCREASES URINARY EXCRETION OF UREA AND AMINO ACIDS IN PRETERM INFANTS. Timothy M Taylor, Mark G Jones, Ronald A Chalmers & Anthony F Williams. Department of Child Health, St George's Hospital Medical School, London SW17 0RE, England.

We have shown marked increases in the plasma amino acid concentrations of very low birth weight (VLBW) infants treated with dexamethasone (Arch Dis Child 1992; 67: 5-9). Ornithine, citrulline, alanine and glutamine increased most. We now report rates of urinary excretion

Methods: 24-hour urine collections in 6 VLBW infants before and after 48 hours of dexamethasone (0.6 mg kg^{-1} d^{-1}). Amino acids were measured by high performance liquid chromatography (HPLC).

Results: Median values given (n=6 unless indicated).

	Before Steroid	@ 48 hours treatment
Urea	1.5	5.2 mmol 24 h^{-1}
Arginine (n=3)	0.6	12.6 µmol 24 h^{-1}
Ornithine (n=5)	2.1	13.4 µmol 24 h^{-1}
Citrulline	0.5	4.2 µmol 24 h^{-1}
Alanine	15.7	64.1 µmol 24 h^{-1}
Glutamine	16.7	95.0 µmol 24 h^{-1}

Conclusion: Increased excretion of both the end product and intermediate compounds of the urea cycle support the suggestion that changes in plasma amino acid concentrations result from catabolism rather than inhibition of nitrogen disposal.

METABOLISM AND NUTRITION

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PHOTOTHERAPY FOR NEONATAL JAUNDICE: A COMPARATIVE STUDY OF FIBER OPTIC LIGHT AND FLUORESCENT LAMPS. G.P.Donzelli, M.Moroni, M.Paparo, L.Cardellini, C.Vecchi. Department of Pediatrics, NICU. University of Florence, Italy

A new, still to be improved, neonatal phototherapy (PT) device - Biliblanket™ (BB), Ohmeda, USA - has been developed in the form of a fiber optic system, whose radically different method of application, directly to the baby's skin, implies some decrease in the usual PT side-effects (insensible water loss, birth weight decrement) and less physical and psychological stress to the jaundiced neonate. To compare its efficiency to conventional PT, 208 healthy preterm infants presenting non-haemolytic hyperbilirubinemia in the first four days of life - with no statistically difference in gestational age, birth weight, age and bilirubin (BR) levels at onset of PT - were subdivided at random in four study groups receiving respectively fiber optic (BB) PT, Special Blue (SB), White (DA) and Green (G) fluorescent lamp PT. Results (values represent mean ± SEM) are reported in the following tab.

BR (mg/dl)	BB(3.0mW/cm²)	SB(3.2mW/cm²*)	DA(2.3mW/cm²*)	G(2.3mW/cm²*)
after 8 hrs of PT	13.3±0.3	12.24±0.24	12.87±0.26	12.07±0.2
after 24 hrs of PT	12.32±0.31	10.79±0.27	11.62±0.3	10.93±0.23
after 48 hrs of PT	11.14±0.33	11.03±0.29	10.73±0.32	11.06±0.34
after 72 hrs of PT	10.85±0.44	9.72±0.41	10.79±0.48	10.6±0.42
after 96 hrs of PT	10.76±0.5	9.5±0.1	10.48±0.67	10.0±0.15

*Power density at skin surface (Power meter mod. 362, Scientech,USA)

Even though fiber optic PT showed significantly lower efficacy during the 24 hrs of PT compared to all ($p<.001$) but DA fluorescent lamps, after 24 hrs fiber optic light source and fluorescent lamps showed similar efficiency ($p>.5$).

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BILE ACID METABOLISM IN EXTRAHEPATIC BILIARY ATRESIA (EHBA): LITHOCHOLIC ACID IN STORED DRIED BLOOD COLLECTED AT NEONATAL SCREENING. Jan Gustafsson, Gunvor Alvelius, Ingemar Björkhem and Antal Nemeth. Department of Pediatrics, Uppsala University, Uppsala and Departments of Clinical Chemistry and Pediatrics, Huddinge Hospital, Huddinge, Sweden.

Lithocholic acid (LCA) is a hepatotoxic compound. Fetal LCA might play a role in the pathogenesis of neonatal cholestasis/EHBA. Recent results show that fetal liver has capacity for hydroxylations of LCA (Ped. Res. 21:99, 1987).

If the pathogenesis of EHBA involves increased fetal levels of LCA, the condition could be due to impaired fetal liver metabolism of LCA. This should lead to increased levels of LCA at birth.

LCA, cholic acid (CA) and chenodeoxycholic acid (CDCA) were quantitated in stored dried blood from six newborn infants with EHBA and fourteen controls. The blood was collected at neonatal metabolic screening. The bile acids were quantitated by gas chromatography-mass spectrometry using selected ion monitoring.

Results: Mean blood levels (±SD) of LCA were 0.11±0.10 µM in the infants with EHBA and 0.08±0.06 µM in the controls. Mean blood levels (±SD) of CA and CDCA were 15.6±8.7 µM and 7.4±6.1 µM in the EHBA infants and 1.7±1.2 µM and 1.8±1.5 µM in the controls.

Conclusion: The low blood levels of LCA found indicate a normal metabolism of this bile acid in fetuses with EHBA. The increased levels of CA and CDCA in the infants with this disease can be due to cholestasis.

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PROTRACTED DEFICIENCY OF α-TOCOPHEROL (α- TOC) AND ESSENTIAL FATTY ACIDS (EFA) IN ENTEROPATHIC HEMOLYTIC-UREMIC SYNDROME(e+HUS)
O. Amon, C. Hübner, B. Finckh, K. Ludwig, B. Sehringer- Mansour, I. Wernicke, A. Kohlschütter, D. E. Müller- Wiefel

To evaluate lipid peroxidation in the clinical course of e+HUS we investigated plasma of dialysis-dependent children (n= 6;Hb 8.7 +/-1.7 g/dl;platelets 134 +/-117/nL;SCR 4.9+/-0.3 mg/dL;LDH 3402 +/-810U/L) longitudinally from start of dialysis (day 0) up to maximally 14 weeks α - TOC (mg/g total lipids), EFA(weight %) and polyunsaturated fatty acids (weight %; PUFA). (age matched controls = K; N=11)

		K	p≤	Day 0	p≤	Day > 28
EFA:	18:2n-6	28.8 +/- 4.5	0.003	20.7 +/- 2.9	0.01	27.8 +/-3.0
	18:3n-3	0.8 +/- 0.4	0.01	0.4 +/- 0.1	n.s.	0.5 +/-0.1
	20:4n-6	6.0 +/- 1.0	0.05	4.9 +/- 0.6	0.01	8.1 +/-1.8
	PUFA	40.3 +/- 4.6	0.002	28.7 +/- 3.2	0.01	40.9 +/-5.2
	α-TOC	2,5 +/- 0.3	0.01	1.3 +/- 0.2	n.s.	1.9 +/-0.2

(x+/-SD; Mann-Whitney-Test)

In consequence of lipid peroxidation α - TOC, PUFA and thereby EFA were decreased on day 0. They only gradually increased in that way as hemolysis decreased and levelled into the normal range after the disappearance of thrombocytopenia and oliguria. Especially the increase of PUFA up to day > 28 correlated well with the decrease of LDH-activity(219 +/- 52 U/L), demonstrating to hemolysis due to peroxidation of fatty acids of the membrane. Since EFA are essential for cell- and membrane synthesis a protracted course of hemolysis in e+HUS might be the consequence of an EFA deficiency. In this case EFA substitution might be able to shorten the clinical course of e+HUS.
University Children's Hospital Hamburg, F.R.G.

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THE EFFECT OF VALPROIC ACID ON FATTY ACID OXIDATION BEFORE AND AFTER L-CARNITINE SUPPLEMENTATION. Eberhard Schmidt-Sommerfeld, Brian D. Kossak, Duna Penn, James H. Tonsgard, Dale A. Schoeller, Piero Rinaldo, Zhi-Heng Huang and Douglas A. Gage. Departments of Pediatrics, Neurology and Medicine, University of Chicago; Department of Genetics, Yale University, New Haven; Department of Biochemistry, Michigan State University, East Lansing, USA.

Urinary metabolites and $^{13}CO_2$-production from ^{13}C-octanoic acid were studied in 12 patients (3-19 y.o.) with seizure disorders before and during valproic acid (VPA) therapy without and with L-carnitine supplementation. Only one patient had a decreased plasma level of free carnitine (FC) during VPA. Urinary FC concentrations were positively correlated with serum VPA levels ($p<0.01$) indicating a dose-related effect of VPA on the tubular reabsorption of FC. Urinary dicarboxylic acid concentrations relative to plasma β-OH-butyrate and urinary hexanoyl- and butyrylglycine concentrations (stable isotope dilution method) increased during VPA. Changes in cumulative excretion of $^{13}CO_2$ in breath were inconsistent. L-carnitine administration had no significant effect. 11/12 patients excreted octanoylcarnitine (OC) in excess of octenoylcarnitine during VPA. Quantitation of OC by radioisotopic exchange/HPLC showed urinary concentrations of OC comparable to those found in asymptomatic patients with medium chain acyl-CoA dehydrogenase (MCAD) deficiency (x 0.71; R 0.09-14.6 vs. x 0.38; R 0.10-4.21 mmoles/mole creatinine). Gas chromatographic/mass spectrometric analysis revealed that the predominant saturated 8-carbon acylcarnitine in urine was OC with valproylcarnitine being only a minor metabolite. Conclusion: VPA therapy results in an inhibition of β-oxidation partly at the level of MCAD. L-carnitine has no consistent effect.

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CHANGES OF BODY COMPOSITION IN CHILDREN FOLLOWING RENAL TRANSPLANTATION (TX). Franz Schaefer, Babette Hake, Rotraut Schulte, Karl Schärer. Children's Hospital, Heidelberg University, F.R.G.

We have recently demonstrated the validity of bioelectrical impedance (BI) analysis for assessing body composition in healthy and diseased children (*Ped Res 29:350A*). In order to study the effects of renal Tx and corticosteroid treatment on the body composition of children with chronic renal failure, we followed up 15 patients aged 3.2 to 18.5 (mean 11.4) years for the first 12 months after Tx. Measurements of height, weight and BI were taken at 2, 4, 6 and 8 weeks, and 3, 4, 6, 8, 10 and 12 months after Tx. Fat-free mass (FFM) was estimated from BI, according to a formula derived from previous validation studies using ^{40}K plethysmography as reference standard. An age- and sex-independent analysis was achieved by transformation of the raw data to standard deviation scores (SDS) based on regional standards for height, weight and body composition. To allow for the non-Gaussian distribution of weight and fat mass (FM), separate SD values were calculated for subjects above and below the 50th percentile.

Average relative height was subnormal (-1.94 ± 1.8 SDS) and did not change during the first year after Tx. Relative weight for height increased during the first three months from -0.32 ± 1.0 to +0.42 ± 1.0 SDS ($p<0.02$), and changed only slightly thereafter. In contrast, relative FFM for height continued to increase from month 3 (-0.66 ± 0.9 SDS) to month 12 (+0.65 ± 1.2 SDS, $p<0.01$).

Our study demonstrates that during the first 3 months after Tx, when high corticosteroid doses are applied, weight gain is mainly due to an expansion of the FM. Subsequently, the anabolic effect of restored renal function becomes apparent by a marked increase of FFM.