137 SHIGA-LIKE TOXIN (SLT) ASSOCIATED HEMOLYTIC-UREMIC SYNDROME (HUS), A 4-YEARS PROSPECTIVE STUDY.

M Bitzan, M Klent, E Möbius, H Rüssmann, H Karch
Dept. of Pediatrics and Inst. of Medical Microbiology,
University of Hamburg, D-2000 Hamburg 20, FRG.
Patients with classical HUS (n=22; median age 2;2 yrs)
were prospectively studied at our institution since
1986. SLT-association was established by cytotoxicity assays of
fecal filtrates, bacterial isolation or colony-blot-hybridization
with SLT-specific DNA probes. Serial serum samples from 12
pts and appropriate controls were studied for SLT I and II
neutralizing antibodies (NAB) and for hemagglutinating antibodies
(HA) using erythrocytes coated with purified LPS from *E.coli*
O157:H7 and other serovars.

SLT association was demonstrated in 13/22 children (59%);
E.coli isolates (n=6 pts) included serogroups O26, O55, O111 and
O157; fecal SLT alone was present in 7 additional cases. 4 pts
developed rising NAB-titers against SLT I or II; 9 pts (75%)
presented with high LPS-O157-HA at the time of the diagnosis of
HUS that rapidly declined during convalescence.

Thus in Northern Germany an association of the classical HUS
with in-vivo SLT production has been confirmed. Detection of
LPS-O157-AB appears to be a new, valuable serological marker for
this subgroup of HUS. Further studies are needed to elucidate a
possible protective or pathogenic role of SLT- and LPS-specific
AB.

138 NITROGEN AND FAT DEPOSITION IN PRETERM INFANTS FED A FORMULA WITH 5 OR 40% MEDIUM-CHAIN TRIGLYCERIDES(MCT). E.J. Sulkers, H.N. Lafeber, C. Leunisse, J.B. van Goudoever, H.J.Degenhart, P.J.J. Sauer. Sophia Children's Hospital, Rotterdam, The Netherlands.

Formulas specially designed for preterm infants contain up to 50%
MCT. The effects of MCT(C8 and C10) on growth and composition of weight gain
were studied in infants receiving formula (Nutricia, Holland) with 5% or 40% of fat
as MCT, providing 12 g carbohydrates, 3.3 g protein- and 6.8 g fat per kg·day. 72 hr
balance studies were performed at 4 weeks of age. Simultaneously, 6 hr periods of
indirect calorimetry were performed to measure substrate oxidation. Nitrogen and
fat deposition was calculated from intake, losses and oxidation. As mean $\pm$ 1sd:

	40%MCT(n=15)	5%MCT(n=12)		P
Birthweight	1129 $\pm$ 218	1271 $\pm$ 165	grams	NS
Gestational age	31 $\pm$ 1.9	32 $\pm$ 1.8	weeks	NS
Nitrogen stools	59 $\pm$ 23	80 $\pm$ 38	mg/kg·d	NS
Nitrogen urine	85 $\pm$ 23	82 $\pm$ 24	mg/kg·d	NS
Nitrogen deposition	381 $\pm$ 34	372 $\pm$ 45	mg/kg·d	NS
Fat stools	0.87 $\pm$ 0.19	1.5 $\pm$ 0.60	g/kg·d	<0.005
Fat oxidation	1.6 $\pm$ 0.74	2.0 $\pm$ 0.78	g/kg·d	NS
Fat deposition	4.3 $\pm$ 0.71	3.2 $\pm$ 0.91	g/kg·d	<0.01
Weight gain	16.6 $\pm$ 2.4	16.1 $\pm$ 3.2	g/kg·d	NS
%protein of wt gain	15 $\pm$ 2.6	15 $\pm$ 4.4	g/kg·d	NS
%fat of wt gain	26 $\pm$ 4.0	21 $\pm$ 6.1	g/kg·d	<0.005

Conclusions: 1 In stable, growing preterm infants fed 120 kcal/kg·d, addition of
MCT to the feeding results in slightly better fat absorption. 2 However, this does not
promote weight gain or nitrogen retention but only increases fat deposition.

139 CALCIUM(Ca) AND PHOSPHATE(P) METABOLISM IN PRETERM INFANTS ON A FORMULA WITH 5 OR 40% MEDIUM-CHAIN TRIGLYCERIDES(MCT). E.J. Sulkers, H.N. Lafeber, W.P.A. Keyer, J. Lindemans, W.H.L. Hackeng, H.J.Degenhart, P.J.J. Sauer. Sophia Children's Hosp., Rotterdam, The Netherlands.

Formulas specially designed for preterm infants contain up to 50%
MCT because of their almost complete absorption. The effects of MCT(C8 and C10)
on mineral absorption were studied in infants receiving formula (Nutricia, Holland)
with 5%(LCT group) or 40%(MCT group) of fat as MCT, providing 80 mg P-, 152 mg
Ca- and 120 kcal/kg·d, + 600 IU/day vit D. 72 hr balance studies were performed at
4 weeks of age. Values: mean $\pm$ 1sd or median with interquartile range. Plasma levels
(MCT vs LCT) of Ca(2.4 $\pm$ 0.1 vs 2.4 $\pm$ 0.1 mmol/L), P(2.2 $\pm$ 0.3 vs 2.3 $\pm$ 0.2 mmol/L)
and Alkaline Phosphatase (273 $\pm$ 74 vs 295 $\pm$ 43 IU/L) were in the normal range.

	MCT(n=15)	LCT(n=13)		P
Birthweight	1129 $\pm$ 218	1290 $\pm$ 172	grams	NS
Gestational age	31 $\pm$ 1.9	32 $\pm$ 1.7	weeks	NS
Ca % absorption	78 (72-81)	68 (61-71)	percent	<0.005
Ca urine	1.6 (0.65-2.4)	0.92 (0.43-1.3)	mg/kg·d	NS
Ca retention	118 (109-121)	102 (92-107)	mg/kg·d	<0.005
P % absorption	93 (92-95)	93 (92-95)	percent	NS
Purine	3.9 (0.85-7.1)	12 (9.0-15)	mg/kg·d	<0.001
P retention	69 (65-74)	62 (60-64)	mg/kg·d	<0.005
PTH	2.1 (1.1-5.0)	4.7 (4.0-6.7)	pmol/L	<0.01

Conclusions: 1 Both formulas show satisfactory calcium and phosphate retention,
with normal plasma levels. 2 Calcium absorption, but not phosphate absorption was
lower with the LCT formula. 3 The elevation of PTH and of urinary P excretion
suggest that in preterm LCT formula, Ca/P ratio should be slightly higher than 2:1.

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VARIATION OF ERYTHROPOIETIN LEVELS IN CHILDREN WITH CHRONIC RENAL FAILURE: DEFECT OF DIRECT HEMATOLOGIC COMPENSATION IN RENAL ANEMIA

August Khoss, Hans Steger, Andreas Böck, Karola Frenzel, Ferdinand Haschke
Department of Pediatrics, University of Vienna, Austria

Erythropoietin (EPO), the primary humoral regulator of erythropoiesis, an
indicator of direct hematologic compensation in anemia, increases with decreasing hemoglobin
levels. **Material and Method:** To evaluate deficient EPO production or the lack of a regulatory
feedback mechanism between EPO and hemoglobin (Hb), we obtained EPO data with the IBL-
erythropoietin ELISA and hematologic parameters in 144 children (aged 1-19 yrs) with different
anemias. Of these anemic (Hb<12 g/dl) patients 39 were receiving conservative treatment (I), 31
on regular dialysis therapy (II), 34 after transplantation (III); variation of EPO concentration in
children with these various stages of chronic renal failure (CRF) were compared to 40 controls
with nonrenal anemia (IV) and healthy controls (V). **Results (figure):** a relative EPO deficiency
could be detected in I and less after renal transplantation, an absolute deficiency was found on
regular dialysis. An inverse linear correlation between EPO levels and hemoglobin in IV occurred,
however in II the rise of EPO was insufficient compared to IV. In our study we could demonstrate
successfully a feedback mechanism between EPO and oxygen supply (low hemoglobin).

Conclusion: The interrelations
between EPO and Hb in anemic children on
regular dialysis indicates an EPO deficiency
and lack of a regulatory feedback
mechanism due to CRF, possibly an
inhibition of erythropoiesis by uremic
toxins. We emphasize renal anemia could
be correct successfully with small side
effects only by substitution of
recombinant human erythropoietin.

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ACCELERATION OF THE SWITCH FROM FETAL TO ADULT HAEMOGLOBIN SYNTHESIS BY EXTRAUTERINE LIFE IN HUMAN NEWBORN INFANTS. Juchschmid, P., H. Frischknecht, G. Schubiger and G. Duc. Dept. of Paediatrics, University of Zurich and Childrens Hospital Lucerne; Switzerland.

Synthesis of human haemoglobin is directed by α -, β -, δ -, γ - and ϵ -genes. The switch from fetal to adult
erythropoiesis includes a genetically controlled suppression of the
 γ - and activation of the β - and δ -chain synthesis. The two γ -genes
are not equally inactivated. In animals external influences on the
switch could be demonstrated, but no influence of extrauterine (EU)
life on the switch in human newborns could be clearly demonstrated.
The long half life of red cells hampers the detection of subtle
influences on the switch by comparing the concentrations of HbA, HbF
and HbA₂. We studied therefore the de-novo synthesis of α -, β -,
 γ - and ϵ -chains by measuring the incorporation of ³H-Leucine into
reticulocytes and subsequent separation of the globin chains by HPLC.
228 single measurements were obtained from 88 newborns (gestational
age 26.3 to 42.1 weeks) at the age of 0 to 116 days; 16 babies were
studied longitudinally. The results show that the switch $\gamma \rightarrow \beta$ is go-
vernated by the biological age (BA = intrauterine + EU age). A sig-
nificant acceleration of the switch by EU life could be demonstrated:
1. The BA when 16 babies reached 50% (33%) of the full switch was not
constant but correlated with GA: $p < 0.01$ ($p < 0.05$). 2. When 31 babies
were studied at a BA of 40 to 43 weeks, a significant ($p < 0.02$) acce-
leration of the switch was found in those with an EU age of 6 to 7
weeks (47 $\pm$ 21%, n=14) when compared to babies of 1 week or less
of EU age (37 $\pm$ 25%, n=14). No comparable changes of the α -, β -,
 γ -pattern was observed during the observation time of 26 to 43 weeks.
Conclusion: $\gamma \rightarrow \beta$ -switch is accelerated by factors of EU life. Inacti-
vation pattern of α -, β -, γ - is not concerted with the $\gamma \rightarrow \beta$ -switch.

SCREENING FOR ALPHA-1 ANTITRYPSIN DEFICIENCY IN NEWBORN INFANTS IN SPAIN

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M. García-Fuentes, V. Madrigal, J. Alonso, M. J. Lozano y J. L.
Alvarez-Granda.
Facultad de Medicina. Hospital Valdecilla. Santander. Spain.

Alpha-1 antitrypsin (AAT) deficiency is an hereditary metabolic disorder which
predisposes affected individuals to juvenile liver disease and to pulmonary
emphysema towards the third or fourth decade of life. Since prevention and
treatment of this pulmonary complication is at present perfectly feasible, we have
carried out a pilot study of neonatal screening for AAT deficiency in order to
determine the incidence of this deficiency in Spain and to evaluate the pa-
rents' reactions on being told of this metabolic disorder.

Fifteen thousand four hundred spanish newborn infants were screened for AAT
deficiency. The study was carried out using the eluate from dried blood samples
provided by the Cantabria University and the Basque Country Health Service Units
for the detection of metabolic disorders. The screening test was based on a
semiquantitative electroimmunoassay for AAT together with transferrin. For all
cases suspected of AAT deficiency, the Pi phenotype was determined.

With the screening test, 58 suspect cases were detected. Of these, the
phenotype study showed 32 AAT deficient cases (8 Pi Z and 24 Pi SZ)
representing a global incidence of 2.08 $\pm$ 0.72 per thousand. In 4 Pi Z cases there
was clinical and laboratory evidence of liver disease. The parents of the
AAT-deficient children were informed of the potentially harmful effect of tobacco
smoke on the child. Their reaction towards this information was positive.

Bearing in mind the high incidence of AAT-deficiency, and the existing
preventive and therapeutic measures for pulmonary emphysema in AAT-deficient
cases, mass-screening for this deficiency may be advisable. This screening could
be considerably facilitated by recently developed genome analysis techniques.