11 Enzymatic evidence for a medium-chain acyl-CoA dehydrogenase deficiency in muscle of a patient with hypoketotic hypoglycemic dicarboxylic aciduria.
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A female patient came to our hospital at the age of 2 8/12 years with the clinical symptoms (vomiting, diarrhea, hypoglycemia and subcoma) and urinary excretion pattern (C_6-monocarboxylic + C_6-C_{10} dicarboxylic acids and their glycine-conjugates and phenylpropionylglycine) typical for hypoketotic hypoglycemic dicarboxylic aciduria. A muscle biopsy was taken. In the muscle homogenate total carnitine was decreased to 10% of the average controls, but carnitine palmitoyltransferase I and II and palmitoyl-CoA synthetase were normal. In isolated mitochondria the respiratory rates of FAD- and NAD^+-dependent substrates were reduced to 33-47% of controls. Oxidation of ascorbate was 78%. The oxidation of (U-^{14}C)-palmitate in the presence of 0.5 mM carnitine was 50% of normal. It normalized in the presence of 5 mM L-carnitine and CoA. Using hexanoyl-CoA as substrate and PMS + DCIP as electron-acceptors the medium-chain acyl-CoA dehydrogenase activity in freeze-thawed mitochondria was 0.67 nmol/mg protein/min (for 6 controls: 3.32 ± 0.15 nmol/mg protein/min; mean ± SEM). Clinically the patient is in a good condition.

12 Comparison of enterotoxin-inhibitory activity in human milk and bovine milk.
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Human milk contains antibodies as well as non-immunoglobulin components which most likely are of importance for the prevention of diarrhoea in infants. We have previously described the precence in human milk of a ganglioside fraction which inhibited E.coli heat labile enterotoxin in vitro and cholera toxin in vivo, as well as a trypsin-sensitive inhibitory activity of rotavirus.

In this study we have compared the enterotoxin-inhibitory activity in human milk with that of bovine milk and bovine-based formula milk. Ganglioside fractions were obtained by extraction and solvent partition of the milk fat. High performance thin layer chromatography (HPTLC) showed major differences between the ganglioside fractions from human milk and bovine milk.

Toxin inhibitory activity was measured in vitro by ELISA (enzyme-linked immunosorbent assay) and in vivo by rabbit intestinal loop model. In ELISA, enterotoxin was inhibited by human milk, bovine milk and formula milk. In the rabbit intestine, only human milk inhibited the enterotoxin.

13 Low phosphocreatine (PCr)/inorganic phosphate (P_i) ratio in the brain of newborn infants indicates poor outcome. AM DE L COSTELLO*, PL HOPE*, EB CADY*, DT DELPY*, PS TOFTS*, ACM CHU*, PA HAMILTON*, EOR REYNOLDS, DR WILKIE*. University College London School of Medicine, London, WCIE 6JJ.

The PCr/P_i ratio is an index of the energetic status of tissue. To see whether this ratio gave prognostic information, we used phosphorus nuclear magnetic resonance spectroscopy to measure PCr/P_i in the brains of 6 normal infants and on 71 occasions in 30 infants with neonatal neurological abnormalities due, for example, to birth-asphyxia, periventricular haemorrhage and early cerebral infarction. In the normal infants PCr/P_i ranged from 1.10 to 1.71 (mean 1.35). PCr/P_i fell below this range in 24 of the 30 abnormal infants and below 0.8 in 16 of them. 8 of the 16 infants with PCr/P_i ratios below 0.8 died in the neonatal period from predominantly cerebral causes and all 8 survivors were neurodevelopmentally abnormal at a mean age of 7 months. Among the 20 infants whose PCr/P_i ratios were always 0.8 or above, 2 died (one aged 3 weeks with congenital abnormalities including Moebius syndrome and the other, who had Prader-Willi syndrome, as a cot death aged 9 months): 3 infants were neurodevelopmentally abnormal aged 4, 9 and 10 months, and the remaining 15 infants were progressing normally at a mean age of 6 months.

We conclude that PCr/P_i ratios below 0.8 were associated with a very poor prognosis, and may indicate irreversibly deranged cerebral metabolism.

14 Hypercalcemia in infancy. C.HOLMBERG, M.JALONEN* and O.KOSKIMIES. Children's Hospital, University of Helsinki, SF-00290 Helsinki 29, Finland.

Hypercalcemic infants were studied to clarify the etiology and mechanisms of this condition. Serum Ca, phosphate, salt, acid-base as well as hormonal (PTH, calcitonin, vitamin-D metabolites, renin and aldosterone) levels and urinary Ca, phosphate, salt and water excretion and transport were measured. 13 infants with hypercalcemia were seen. Age at diagnosis was 3.02±0.5 months (mean±SEM) and the most common symptom (7/13) was growth retardation (weight reduction in SD was -1.3±0.2, length reduction -1.0±0.3) and irritability (4/13). Mean serum Ca concentration was 3.02±0.07 mM/L (normal=2.15-2.70 mM/L) with a low normal protein concentration. Mean phosphate concentration was 2.21±0.09 mM/L (n=11, normal =1.50-2.50 mM/L), PTH (0.26±0.02 µg/L) and calcitonin concentrations were normal. 5 patients had hypo-, 8 hypercalciuria (urinary Ca excretion >4 mg/kg·24h). 2 sisters had a urinary Ca excretion >7 mg/kg·24h and nephrocalcinosis at 2.5 months. Their serum phosphates were low (0.95 and 0.84 mM/L), PTH was normal, U_pV and T_p were low. 2 patients had pseudohypoaldosteronism and 4 idiopathic hypercalciuric hypercalcemia. Their vitamin-D metabolites, calcitonin and PTH concentrations were normal, but PTH was "high for serum Ca level". All 13 children responded to vitamin-D reduction sometimes combined with Ca restriction (pseudohypoaldosteronism was treated with NaCl) and have shown perfect catch-up growth, normal psychomotor development and disappearance of symptoms (the nephrocalcinosis has not progressed in the two sisters).

15 Oxidative Metabolism of 13C Medium chain triglycerid (MCT) in Preterm infants (PT).
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Fat Malabsorption in PT infants improves when part of milk fat is replaced by MCT ; Whether these MCT are stored or oxidized is not well known. 5 PT (BW:1771± 100g; GA= 34± 1WKS, Age at study=24±4d) orally fed a formula containing 50% of fat as MCT were given a known amount of 13C trioctanoin on the second day of a 3 day nutrient balance. Continuous indirect calorimetry was performed during the 24 hours following 13C MCT ingestion, with continuous sampling of expired CO_2 for measurement of 13C/12C ratio. Energy balance, nutrients oxidation and amount of oxidized MCT were then derived. Preliminary results : 1) Nutrient balances are shown in Table (kg/d ; M±SD)

	CHO(g)	FAT(g)	Protein(g)	Energy(Kcal)
absorb.	12.4±5.3	4.9±0.2	2.5±0.3	107± 5
oxid.	11.7±4	1.6±0.6	0.6±1.1	64± 6
stor.	0.7±1.3	3.3±0.8	1.9±0.1	43±10

2) Elimination curves of 13C in expired CO_2 were of similar pattern but of widely different amplitude. Mean MCT oxidation was 21±12% (range 6-43) and represented 31% (range 7-47%) of the total fat oxidation.

Conclusions : 13C CO_2 elimination shows considerable variations between PT infants. No correlation is observed with energy expenditure and with total fat oxidation. The later was even lower than the total amount of MCT given.

16 Nutrient, energy balance and weight gain composition in preterm infants (PT) fed pooled human milk (HM) with or without protein supplementation. PUTET G, FAHMY N, RIGO J, SENTERRE J, SALLE B. INSERM U34 and Neonatal Dept., Lyon France ; Liege Belgique.

Protein content of HM is low and may be inadequate for feeding low-birth-weight infants ; protein supplementation has been proposed to improve its nutritional adequacy. We studied two groups of PT fed isocaloric amounts of either pooled HM (HMgr., n=6; GA=30.5±1.5wks; BW=1378±150g) or cow's based protein supplemented (0.8g/100 ml) HM (HMP gr: n=5; GA=29.6±1.3wks; BW=1378± 156g). A 3 day nutrient balance and an Energy expenditure measurement (indirect calorimetry) was performed on each infant at around 33 weeks post conceptional age (HMgr: 33±1.4wks HMPgr=33.6±0.7wks) along with successive Antropometric measurements.

Results 1) HMPgr has a higher weight gain (16.7±1.7 VS 13.5±1.9; NS) similar length and head circumference gains and lower skinfold increases than the HMgr.

2) Energy and protein balances are shown in table (/kg/d; M±SD). * student t-test $P < 0.05$.

	ENERGY (kcal)		PROTEIN (g)	
	HM	HMP	HM	HMP
E.absorb.	87±11	90±16	2.1±0.4	3.1±0.4
E.oxid.	46± 6	57± 3	0.5±0.1	1.0±0.4
E.stor.	41± 8	33±12	1.6±0.4	2.1±0.1

Conclusion: HMPgr has higher protein retention and lower energy storage than HMgr; weight gain was slighty higher in HMPgr; therefore weight gain composition is different between the groups with higher lean tissue mass and lower fat storage in the HMPgr; this correlates with skinfold measurements.