5 Postheparin plasma lipoprotein and hepatic lipases and serum carnitine in newborn infants during parenteral nutrition. L. ROVAMO*, E.A. NIKKILA*, K.O. RAIVIO. Children's Hospital and IIIrd Department of Medicine, University of Helsinki, Finland

Lipoprotein lipase hydrolyzes triglycerides in plasma and thus regulates the clearance of fat from the circulation. Lipoprotein lipase activity has been estimated in infants during parenteral nutrition by measuring postheparin lipolytic activity (PHLA). PHLA is, however, an inadequate measure of lipoprotein lipase because a substantial part of PHLA results from hepatic lipase. Carnitine is essential for facilitated transport of long-chain fatty acids across the mitochondrial membrane. With specific methods we measured lipoprotein and hepatic lipase activities. Nine newborn infants were operated on because of gastrointestinal anomalies. Parenteral nutrition was built up in three days whereafter infants received 3/g/kg/day of fat (intralipid) at a constant rate. On the average, weight gain started at the age of 5 days and was 16 g/day. The duration of parenteral nutrition was 1-23 weeks. During the first week lipoprotein lipase activity increased from 14 to 35 µmol FFA/ml/h whereas hepatic lipase activity remained at 40 µmol FFA/ml/h during parenteral nutrition. Serum free carnitine decreased from 25 to 11 µmol/l and acylcarnitine from 9 to 2 µmol/l during the first three weeks of parenteral nutrition; urinary excretion of carnitine decreased from 114 to 68 nmol/mg of creatinine. Serum triglycerides, free fatty acids and blood beta-hydroxybutyrate remained, however, practically unchanged during parenteral nutrition. The results suggest that neither lipoprotein lipase activity nor carnitine availability are rate-limiting for the utilization of fat in newborn infants during parenteral nutrition.

6 Lipoprotein and hepatic lipase activities in postheparin plasma of preterm infants. L. ROVAMO*, E.A. NIKKILA*, K.O. RAIVIO. Children's Hospital and IIIrd Department of Medicine, University of Helsinki, Finland

Fat tolerance tests suggest that the disposal of lipids infused is slower in preterm than term infants. This has been attributed to low lipoprotein lipase activity because postheparin lipolytic activity (PHLA) has been found to be low in very-low-birth-weight infants. Hepatic lipase, however, accounts for about 70% of PHLA, which makes PHLA an inadequate measure of lipoprotein lipase. With specific methods we measured lipoprotein lipase and hepatic lipase activities in postheparin plasma of eleven preterm neonates. Blood samples were taken fifteen minutes after a heparin bolus of 100 IU/kg given before an exchange transfusion performed with fresh heparinized blood because of hyperbilirubinemia, blood group incompatibility or sepsis. The infants were 1-4 days old and had birth weights (range 1210-3490 gm) appropriate for gestational age (range 28-36 weeks). Eight infants (group 1) were in good clinical condition while three infants (group 2) suffered from septic shock. In group 1 lipoprotein and hepatic lipase activities were 27 and 64 µmol FFA/ml/h; both are higher than the activities found in term infants (Rovamo et al. 1984 Pediatr. Res. in press). In group 2 lipoprotein and hepatic lipase activities were 1.8 and 11 µmol FFA/ml/h; both are considerably lower than the activities found in Group 1. Our results indicate that lipoprotein lipase is not the reason for slow clearance of fat from the circulation in preterm infants except in septic shock.

7 D- Thyroxine Treatment in Glycogen Storage Disease Type VIa.
W. ENDRES, Y.S. SHIN, M. RIETH*, K. ULLRICH, F. KOLLMANN, J. SCHAUB.
Children's Hospital, Universities of Munich, Muenster, Frankfurt and Kiel, Federal Republic of Germany.

Phosphorylase b kinase (PK) deficiency is considered as a relatively benign glycogen storage disease (GSD VIa). Garibaldi et al. (Helv. Paediat. Acta 33, 435 (1978) reported that the treatment with dextro-thyroxine (DT_4) resulted in normalization of liver size, triglyceride concentration and transaminase activities in serum of four boys with GSD VIa. We treated three patients with GSD VIa over periods of 39, 19 and 18 months respectively with 60-330 µg DT_4 per kg b.w. per day. In two patients liver height (sonographically measured in the right medioclavicular line) decreased by seven and five cm respectively. Glycogen concentration in erythrocytes also diminished accordingly in these patients. Clinical response in one of these two patients was remarkable showing a decrease in transaminase activities, triglyceride levels in serum as well as an increase in growth during the treatment. However, there was no significant activation of PK in erythrocytes by DT_4 in all three patients. This individual difference in responses to DT_4 treatment in GSD VIa may be due to heterogeneity of this disease. It is necessary to study further the possible heterogeneity of this disease in order to apply efficiently and correctly DT_4 in the treatment of GSD VIa.

8 Cytochrome (cyt) c_1 deficiency in liver and muscle mitochondria

H PRZYREMBEL, HR SCHOLTE[X], HFM BUSCH[X], W BLOM[X], WFM ARTS[X] IEM LUYT-HOUWEN[X], JA BERDEN[X]. Dept. Pediatrics, Neurology and Biochemistry I, Erasmus University, Rotterdam, BCP Jansen Institute, University Amsterdam (The Netherlands).

A 890 g boy was born by caesarean section because of intrauterine distress and growth retardation at 29 weeks of gestation. He developed tyrosyluria, dicarboxylic aciduria, hyperammonemia, and lactic acidosis (12 mM). Blood lactate was not influenced by fasting, glucose, biotine, thiamine, dietic measures. With 1.5 g vitamin C/day lactate is 2 to 5 mM. Clinical features are failure to thrive, muscular hypotonia, spasticity and mental retardation. Open liver and muscle biopsies at age 8 months showed morphological evidence of lipid storage and abnormal mitochondria. In serial studies in isolated mitochondria respiratory rates were decreased with all substrates except ascorbate+TMPD. Residual succinate oxidation was little inhibited by antimycin. Succinate cyt c reductase activity was reduced in liver (258nmol cyt c/min; control 3088±536, n=12) and muscle homogenate (0; control 1634±152, n=8). Cyt b and aa_3 were present in normal amounts in muscle mitochondria. Cyt $c+c_1$ content was decreased (128 pmol/mg protein; control 419±42). The normal capability to oxidise ascorbate+TMPD in combination with a slow reduction of cyt b and $c+c_1$ by succinate+KCN indicates a defective cyt c_1, meaning that all cyt $c+c_1$ measured is probably cyt c. Carnitine was decreased in muscle (1.86 µmol/g wet weight; controls 3.96±0.09) and liver (1.33; control 2.86). The patient is alive at 32 months of age, when a second muscle biopsy was taken.

9 A comparative study of the maturation of NaK-ATPase activity in different nephron segments. Anita Aperia, St. Göran's Children's Hospital, Box 12500, S-112 81 STOCKHOLM, Sweden.

In the kidney the development of deep nephrons preceeds the development of superficial (sf) nephrons and the structural maturation of proximal tubular (PT) segments preceeds the structural maturation of distal tubular segments. NaKATPase is present in all nephron segments. The comparison between the development of NaKATPase activity in PT and thick ascending limbs of Henle (TAL) in deep and sf nephrons can therefore yield information about the relative importance of genetic and environmental factors for enzymatic differentiation. NaKATPase activity was determined in isolated rat tubular segments with Doucet's method (AJP 1979). NaKATPase activity increased in both PT and TAL till the age of 40 days. In PT development was linear while in TAL development was accelerated between 16 and 20 days. The developmental pattern for NaKATPase was the same in cortical and medullary TAL and the same in sf and deep PT.

Serum corticosterone was determined with RIA. It was low till 16 days, then increased rapidly to reach adult values at 20 days. Adrenalectomy inhibited the development of NaKATPase. Treatment with betamethasone (up to 60 µg/100 g) precociously induced NaKATPase in TAL at 16 days of age and in PT at 10 to 20 days of age. PT concentration of cytosolic glucocorticoid receptors determined with isoelectric focusing was significantly higher in 20- than in 40-day-old rats.

Conclusion: The enzymatic differentiation is typical for each cell type. In a given cell type extracellular factors appear to influence the enzymatic differentiation simultaneously and irrespective of embryonic age.

10 Importance of adrenocortical hormones for maturation of colonic NaKATPase activity.
Yigael Finkel, Anita Aperia, St. Göran´s Children´s Hospital, Box 12500, S-112 81 STOCKHOLM, Sweden.

NaKATPase activity is low in the immature rat colon and increases till the 40th postnatal day. Studies from this laboratory have shown that immature renal tubular cells are more sensitive to the inductive effect of adrenocortical hormones on NaKATPase activity than mature cells. The aim of this study has been to examine whether endogenous fluctuations of aldosterone (Aldo) and/or corticosterone (CS) induced by changes in sodium balance can precociously increase NaKATPase activity in the immature colon. Young and adult rats were therefore given a normal (C) or low sodium (E) diet for 4 days and studied at the age of 20 and 40 days, respectively.

		s-aldo pg/ml	S-CS nmol/l	NaKATPase µmol Pi/mg prot H proximal	distal
20 d	C	246 ± 46	144 ± 24.2	6.33 ± 0.5	8.41 ± 0.96
	E	>1200 a	274 ± 58.8	13.65 ± 1.43 a	14.86 ± 1.71 a
40 d	C	283 ± 161	197 ± 81	10.59 ± 0.83 b	7.85 ± 0.7
	E	>1200 a	240 ± 22.1	13.67 ± 2.05 a	10.38 ± 0.44 a

a) $p < 0.05$ compared to c (same age)
b) $p < 0.05$ compared to c (20 days)

The effect of NaKATPase increase in 20 days secondary hyperaldo rats was evaluated using in vivo perfusion of colon. Net Na-absorption increased from control value 190 ± 91 µmol/min/dry g colon to 428.5 ± 97.2 ($p < 0.05$) and net fluid absorption increased from 1.02 ± 0.67 µl/min/dry g colon to 2.64 ± 0.75 ($p < 0.05$). These data indicate that aldosterone increases NaKATPase activity in proximal and distal colon, stimulates net sodium and water transport in large intestine and that the immature large intestine is more sensitive to aldosterone stimulation.