were then kept for another 3 days in media containing either glu or gal alone or combined. SfS of cultures continued at glu 26.1 mM served as control (100%). Gal alone at any concentration (1.3-26.3 mM) was unable to replace glu, SfS remained severely depressed (1.5 to 2.9%). At high glu levels above 12.6 mM addition of gal did not influence SfS, thus excluding a toxic effect. At glu 7.5 mM which left cultures glu-deprived below 0.2 mM for at least 1 day, SfS fell to 35.9±10.6% (SD). Under this condition, SfS could be raised to 52.9±13.5% when gal 0.6 mM was added or even to 67.9±15.6% with gal 12.2 mM (p less than 0.05, resp. 0.001). Therefore, gal may not be necessary for SfS in glu-replenished states. Under glu-deprivation, however, gal could become essential for maintaining structural brain metabolism measured as SfS.

72 A. LUCAS*, T.E. ADRIAN*, S.R. BLOOM* and A. AYNSLEY-GREEN. University Dept. of Paediatrics, John Radcliffe Hosp., Oxford, and Hammersmith Hosp., London. GUT HORMONES IN THE NEONATE.

Gut hormones exert important effects on gut growth, secretion and motility and on intermediary metabolism, and may play a key role in the postnatal adaptations to enteral feeding. Using sensitive radioimmunoassays we have measured 10 entero-insular hormones in the plasmas of (a) 203 healthy term or preterm infants at birth, or during the first 24 days of life either before or at 30, 60 or 120min. after feeding and (b) 10 six-day old preterm neonates who had never been fed orally since birth on account of hyaline membrane disease. Each infant contributed, with ethical approval, only one plasma sample, removed at the time of a routine clinical blood test. Basal levels of gastric inhibitory polypeptide (GIP), pancreatic polypeptide (PP), enteroglucagon (EG), neurotensin (NT), motilin, and gastrin all rose steeply during the neonatal period, the latter four hormones reaching levels which exceeded significantly those seen in healthy fasting adults. In contrast plasma concentrations of GIP, EG, NT, motilin and gastrin did not show postnatal elevation in the unfed group of infants. In addition for 8 hormones; secretin, GIP, PP, NT, EG, motilin, gastrin and insulin, we observed a progressive postnatal increase in the endocrine response to a feed. Profound changes occur therefore in gut hormone physiology after birth and these changes may in part be due to enteral feeding itself. This data is of relevance to the design of optimal feeding regimes for high risk neonates.

73 N.C.R. RÄIHÄ, A.L. JÄRVENPÄÄ*, D. RASSIN*, and G. GAULL*, Depts. Pediatrics, and Obstetrics and Gynecology, University of Helsinki, Finland and Dept. Pediatric Research, N. Y. State Inst. Res. Ment. Retdn., Staten Island N. Y., U.S.A. Milk protein quality: Biochemical and growth effects in full-term infants.

Our previous studies have shown that both quantity and quality of ingested milk protein affects the metabolic homeostasis of low birth-weight infants. In this study, 37 well full-term infants were fed ad libitum for 12 weeks with either breast milk (BM) or with one of two formulas (F_1=1.5% prot., whey prots.: caseins, 60:40 or F_2=1.5% prot.; whey prots.: caseins, 18:82). No differences between feeding groups were found in weight gain, linear growth or head circumference during the study period. Significantly, greater negative base excess and higher BUN were found in the formula fed infants as compared to those on BM. Blood cholesterol at 12 weeks was much higher in the infants on BM when compared to those on F_1 and F_2 ($p<0.0001$). Plasma and urine taurine concentrations were higher in the infants on BM throughout the study and at 4, 8 and 12 weeks of age the urine taurine was 25 to 50 times higher in BM infants than in F_1 and F_2 infants. Plasma valine and isoleucine concentrations were significantly increased in the formula fed infants as was the valine/glycine ratio, suggesting the possibility of protein overloading in the formula fed infants.

74 A.L. JÄRVENPÄÄ*, N.C.R. RÄIHÄ, P. KUITUNEUN*, D. RASSIN* and G. GAULL*, Depts. Pediatrics and Obstetrics and Gynecology, University of Helsinki, Finland and Dept. Pediatric Res., N. Y. State Inst. Res. Ment. Retdn., Staten Island, N.Y. U.S.A. Effect of taurine supplementation to the diet on bile acid conjugation in low birth-weight infants (LBWI).

Previous results have shown that plasma and urine taurine concentrations decrease when LBWI are fed with taurine deficient formulas. In this study, 63 well LBWI with gestational ages of 31 to 36 weeks were randomly assigned to either breast milk (BM) or formula (1.5%, prot. 60:40 whey: caseins) without (F1) or with addition of taurine (30 µmol%, F_2). Duodenal fluid bile acids and glycine-taurine ratio (G:T) were determined with high-performance liquid chromatography. The plasma taurine levels and urine taurine excretion fell significantly in the infants fed with the taurine deficient F_1 formula. At the age of 7 days the G:T ratios were 0.64 (BM), 0.44 (F_1) and 0.47 (F_2). In the infants on BM and F_2 the G:T ratio remained on the same level throughout the study and at the age of 36 to 40 days it was 0.58 and 0.69 respectively on the two diets. In the infants fed with formula F_1, the G:T ratio increased significantly after the age of 21 days being 1.32 at a mean age of 39 days ($p<0.001$). Our results indicate that dietary taurine intake affects duodenal bile acid conjugation.

75 T. TIKANOJA* and O. SIMELL* (Intr. by J. Perheentupa). Children's Hospital, University of Helsinki, Helsinki, Finland. Plasma amino acids in healthy newborns after a feed of breast milk or formula.

Postprandial concentrations of plasma amino acids were followed in 20 full-term healthy infants, aged 4.5-8 days, to quantitate differences in response to a physiologic feed of breast milk or formula. The study had been approved by the ethical council of the hospital; the parents of each infant had given their informed consent. After a 4 hr fast, 10 infants received from bottle a normal feed (1/36 of body wt) of pooled breast milk; 10 infants received milk formula. The true protein contents of the milks were 0.8% and 1.5%, respectively. Venous blood samples were drawn just prior and 30, 60, 120 and 210 min after the beginning of the feed. Peak plasma concentrations were reached with both milks in 30-60 min and starting values in 210 min. Valine, leucine, tyrosine, lysine and arginine showed highest molar increased amounting 50-100% above their fasting concentrations. The rises in plasma amino acids were approximately equal after both meals despite differences in the proteins of the milks. Thus, (1), the absorption of amino acids is significantly more efficient from breast milk than from formula, or (2), the postprandial amino acid curves are strictly regulated by the metabolic clearances and poorly reflect absorption in the newborns.

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Orotic aciduria, an indicator of adequacy of treatment in lysinuric protein intolerance (LPI).

The basic defect in LPI is in the transport of the diamino acids in kidney tubuli, jejunum and liver cells. This leads to a deficiency in the liver of the urea cycle intermediates arginine and ornithine, a malfunction of the cycle and an accumulation of ammonia. Like in the other conditions in which hyperammonemia is associated with accumulation of carbamyl phosphate, urinary excretion of orotic acid is excessive. Orotic acid is an intermediate of the synthesis of the pyrimidines from carbamyl phosphate. We measured orotic acid excretion in controls and patients with LPI. The controls in all situations excreted less than 20 µg/kg/hr. The patients excreted normal amounts during fasting (4.6, 2-8, 5 for mean, range, number of subjects). Their excretion was increased in 24-hr urines during a self-chosen low-protein diet (125, 3-366, 7), in 4 to 6 hr urines after a milk load with 0.5 g of protein/kg (488, 251-1747, 3), in 2-hr urines after oral ammonium lactate, 2.5 mmoles/kg (212, 15-1126, 10), and in 6-hr urines after iv alanine, 6.6 mmoles/kg in 90 min (790, 47-1831, 11). If the loads were given with an iv infusion of arginine, ornithine or citrulline, the orotic aciduria did not appear. Given orally, citrulline was the most efficient of the three in preventing orotic aciduria. Orotic aciduria thus is a reliable indicator of the function of the urea cycle in LPI and enables us to monitor the home treatment.

77 M. DURAN, F.A. BEEMER, S.K. WADMAN, J.L. JOHNSON, W.R. WAUD, K.V. RAJAGOPALAN, University Children's Hospital Het Wilhelmina Kinderziekenhuis, Utrecht, The Netherlands and Duke University Medical Center, Dept. Biochemistry, Durham NC, USA.
Combined deficiency of sulfite oxidase and xanthine oxidase as a result of defective synthesis of molybdenum-cofactor.

A girl is presented with congenital abnormalities including asymmetry of the skull and slight mediofacial dysplasia. The eye lenses were dislocated. Severe neurological abnormalities and deep mental retardation were observed. On routine screening a low serum urate (0.01-0.07 mmol/l) was observed. Chromatography of urinary purines revealed xanthinuria. In addition the sulfite test in the urine was repeatedly positive. Sulfite oxidase deficiency was suggested by high excretions of S-sulfocysteine, taurine and thiosulfate and low levels of urinary sulfate. Deficiencies of both xanthine oxidase and sulfite oxidase were demonstrated in a liver biopsy specimen. Because sulfite oxidase and xanthine oxidase are known to require an activated form of molybdenum (Mo-cofactor), investigations of Mo metabolism were carried out. A complete absence of hepatic Mo-cofactor activity was demonstrated and analysis by atomic absorption revealed a severe depletion of hepatic Mo as well. A near normal amount of inactive xanthine oxidase protein was present, but several immunological techniques failed to detect any sulfite oxidase apo-enzyme. It is suggested that a defective synthesis of Mo-cofactor causes the biochemical abnormalities. Treatment with oral supplements of molybdenum did not result in biochemical or clinical improvement.

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5-phosphoribosyl-1-pyrophosphate (PRPP) synthesis in glucose-6-phosphate dehydrogenase (G6PD) deficiency.

The supply of ribose for nucleotide synthesis is considered a major function of the oxidative pentose shunt (PS). We have evaluated this function on the basis of ribose-5-phosphate (R5P) and PRPP concentration and generation in normal and G6PD-deficient fibroblasts. PS in normal cells accounts for 0.8% of glucose utilization. It can be stimulated over 10-fold by methylene blue (MB) and inhibited 85% by 6-aminonicotinamide (AN) without affecting PRPP levels or generation. PS in mutant cells is 30% lower than normal and barely affected by MB or AN. Basal R5P,