13 CARBOHYDRATE TOLERANCE AND PECULARITIES OF ENDOCRINE SYSTEM FUNCTION IN INFANTS FED WITH TOTAL PARENTERAL NUTRITION /TPN/

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Infants fed with fat-free TPN do receive relatively high glucose and aminoacids intakes. In these patients glucose homeostasis is maintained through deep variations of intermediate metabolism. Various hormones do play a crucial role in this adoptation. A total of 21 infants, 18 days to 8 months old, fed with TPN for an average of 22 days were recently studied. In 12 of these infants plasma insulin, GH and cortisol levels were monitored. Results will be reported discussing separately plasma hormone variations in the first 24 hours, during TPN and after with-drawl from therapy. Soon after starting TPN high insulin and cortisol levels were noticed. Thereafter glucose homeostasis was maintained despite normal insulin levels. GH and cortisol plasma concentrations were normal or below normal. After withdrawl from TPN a drop in insulin levels is usually observed, together with an elevation of cortisol plasma concentrations. When TPN is complicated by infections insulin levels tend to be higher, and glucose tolerance impaired.

14 EARLY FEEDING IN VERY LOW BIRTHWEIGHT INFANTS AND SUBSEQUENT GROWTH

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To groups of very low birthweight infants /birthweight 1500 gms/ were studied; Group I - four infants who required intermittent positive pressure ventilation; and Group II twelve infants who did not reqire IPPV. Attempts were made to feed all infants in both groups on high volumes of milk from the first day of life by nasogastric tube. In Group I feeds were not well tolerated and the mean intake of milk over the first 5 days was: 15,23,36,45,56ml per kilogram. In Group II feeds were tolerated and the mean daily intake over the first 5 days was: 31,51,85,118,149ml per kilogram. The daily intake in these two groups are significantly different. At 2,4,6 and 8 weeks of postnatal age however, the milk intake per kilogram body weight, the rate of weight gain and the levels of plasma albumin in the two groups were not significantly different. Hypoglycaemia was adequately prevented in Group I by intravenous Dextrose. It is argued that such infants may be better off with elective parenteral fluids during the early period of intensive care rather than suffer repeated attemps at naso-gastric feeding.

15 PERIODIC BREATHING IN PRETERM INFANTS: CLINICAL RESPONSE TO INCREASED AMBIENT OXYGEN AND RESULTS OF TRANSCUTANEONSLY OBTAINED ARTERIAL OXYGEN TENSION MEASUREMENTS

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Periodic breathing can be converted to regular breathing by the administration of high inspired oxygen mixtures /F_IO_2/. The amount of F_IO_2 required depends on a/ maturity, b/ postnatal age. The new method of transcutaneous oxygen tension /tc O_2/ measurements provided a simple, non-invasive to test whether the previously found negative correlation between F_IO_2 and maturity is also present with tc O_2. Each infant was tested at laest twice, usually three or four times. Results demonstrate the same general pattern as that previously found with F_IO_2: generally a high tc O_2 /< 200 mm Hg/ is necessary for the conversion of periodic into regular breathing in small preterm infants /< 1800 g/. At 2500 g most infants only need a tc O_2 of 100-150 mm Hg in ordre to convert periodic into regular breathing.

16 5-OXOPROLINURIA /PYROGLUTAMIC ACIDURIA/ AN INBORN ERROR OF GLUTATHIONE METABOLISM

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Two siblings suffering from metabolic acidosis and hemolytic anemia excreted gram quantities of 5-oxoproline in the urine. In erythrocytes, cultured fibroblasts and placenta the levels of glutathione were markedly decreased. The same tissues contained decreased activity of glutathione synthetase, whereas the activities of other enzymes involved in the metabolism of glutathione were normal. Extracts of erythrocytes from the patients and control subjects catalyzed the synthesis of 5-oxoproline from glutamate, provided ATP, Mg ions and cysteine were added. This occurred in two steps: gamma-glutamyl-cysteine synthetase catalyzed the formation of gamma-glutamyl-cysteine, which was converted to 5-oxoproline and cysteine by gamma-glutamyl cyclotransferase. Reduced glutathione specifically inhibited gamma-glutamyl-cysteine synthetase. The following molecular mechanism for 5-oxoprolinuria is proposed: The lack of glutathione synthetase leads to a deficiency of glutathione. The absence of this feedback inhibitor results in increased synthesis of gamma-glutamyl-cysteine which is converted to 5-oxoproline /and cysteine/. The overproduction of 5-oxoproline exceeds the capacity of the 5-oxoprolinase. Therefore 5-oxoproline accumulates in body fluids.

MINERAL METABOLISM

17 MATERNAL VIT D INTAKE AND ITS INFLUENCE ON MATERNAL AND INFANT PLASMA CONCENTRATIONS OF 25-HCC AND ON NEONATAL HYPOCALCAEMIA

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Venous plasma samples were obtained at 24 weeks, 34 weeks and at delivery from 633 women taking a vit D_2 supplement of 400 i.u. /day from the 12th week of pregnancy and from 627 taking a placebo. Initial analyses show that concentrations of total plasma calcium fell significantly from a mean value of 9.46 mg/100 ml at 24 weeks to 9.23 mg/100 ml at term in vit D treated mothers while there was a significant increase from 9.02 mg/100 ml to 9.52 mg/100 ml in untreated women. There were eight times as many hypocalcaemic /< 7.4 mg/100 ml at day 6/ infants born to non-vit D treated mothers. Plasma 25-hydroxycholecalciferol /25-HCC/ concentrations are greater in maternal plasma and umbilical plasma from treated mothers. Maternal plasma 25-HCC concentrations significantly with umbilical 25-HCC values and are significantly greater than them. Maternal vit D deficiency is a causative factor in neonatal hypocalcaemic tetany.

18 25-HYDROXYCHOLECALCIFEROL /25-HCC/ IN NEWBORN INFANTS WITH AND WITHOUT VITAMIN D3.

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The newborn infant is completely dependent on his mother's supply of vitamin D3. In 10 newborn infants after cesarean section we found during late summer time /Sept. through Oct./ 15.1 ng 25-HCC per ml plasma on the 2nd day of life. Compared with 10 newborn infants during winter this level was 4-times higher. During the first 2 weeks of life the summer group did not receive vitamin D3 except 100 I.U. with the daily food. The level of 25-HCC did not change. In the winter group receiving daily 500 I.U. vitamin D3 the level of 25 HCC increased from 3.6 ng/ml to 10.1 ng/ml within 2 weeks. Elevation of calcium after a single high dose of vitamin D3 /5 mg = 200.000 I.U./was higher than after 500 I.U. per day. The level of 25-HCC increased much more after a single high dose of vitamin D3 than after daily 500 I.U. The risk of the so-called "Frühstoss"will be discussed.