61 RELATION OF INTRAVENTRICULAR HAEMORRHAGE TO HYALINE MEMBRANE DISEASE IN NEWBORN INFANTS

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Using our perinatal data bank, clinical and pathological data on 1o4 sigleton infants who died with germinal layer haemorrhage and intraventricular haemorrhage /IVH/ at Hammersmith were analyzed in comparison with data from babies dying of other conditions, and with the background population. Of 31 cases of IVH without hyaline membrane disease /HMD/ only four infants were born at a gestation of 3o weeks or more, whereas in 73 cases of IVH with HMD 49 were born at over 3o weeks gestation. At all gestations infants who died with HMD + IVH had received more intense alkali therapy than those who died with HMD alone, although there was no difference in acid-base status or PaO_2 measurements. Administration of alkali /THAM and bicarbonate/ in excess of 8 mEq/Kg body weight over any 12 hour period was significantly associated with the development of IVH. In babies of 3o weeks gestation or less the association between IVH and alkali therapy was significant whereas the association with HMD was not. It is suggested that the expansion of blood volume caused by administration of hypertonic solutions for treatment of HMD may be a significant factor in the causation of IVH.

62 FETAL ALCOHOL SYNDROME

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In 1973 Jones et al. were the first to report on the so-called fetal alcohol syndrome. Hitherto 16 cases have been published. Within the last 2 years our group has observed 12 additional cases. All infants were hypotrophic at birth; mean length=39 cm, mean weight=188o, mean gestational lenght=38 weeks. The main symptoms were: microcephaly /11/12/, micrognathia /1o/, high arched palate /6/, epicanthus /7/; clinodactyly /6/, vitium cordis /4/, genital malformations /6/, pathological palmar creases /9/. Postnatally, growth, weight gain, statomotor and mental development were considerably retarded. The alterations of the heart, brain and dermatoglyphs which can be traced back to distinct gestational stages are ample evidence for damaging influences already occurring during the second and third month of pregnancy . Thus, the syndrome is an embryopathy rather than a fetopathy.

63 GLUTARIC ACIDURIA 'TYPE II'.

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A new fetal metabolic disorder is described, mainly characterised by severe acidosis and hypoglycaemia ,a' sweaty-feet' like odour, and massive glutaric aciduria and acidaemia. In addition, urinary excretion of isobutyric and isovaleric acid was increased, as well as that of some dicarboxylic acids. Serum levels of various long-chain and short-chain fatty acids and dicarboxylic acids and plasma levels of valine and lysine were markedly elevated. The degradation of ^{14}C-labelled glutaric acid, the branched-chain amino acids, and two corresponding alphaketoacids in fibroblasts was decreased, while that of pyruvic acid was normal. It is speculated that this metabolic pattern is due to a complex disturbance of different acyl-CoA dehydrogenases, with subsequent inhibition of long-chain fatty acid oxidation and gluconeogenesis.

64 LUNG MECHANICS, LUNG VOLUMES AND DISTENDING PRESSURES IN THE NEONATE.

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The effect of continuous positive airways pressure /CPAP/ on lung mechanics and lung volumes were studied in 12 healthy neonates using a total body plethysmograph. As expected, TGV rose and the resistance fell. Dynamic compliance was calculated from the tidal volume and oesophageal pressure corrected for variations in mouth pressure. The compliance fell in all the infants. One possible explanation was that the CPAP had caused changes in the pulmonary vascular compartment. In a further 12 babies the effect of CPAP was compared to a continuous negative pressure applied to the infant's trunk. Both systems produced similar changes in resistance and compliance. Half the distending pressure was tranferred to the mediastinal structures. Squeezing the chest with a small positive external pressure led to a rise in dynamic compliance. We are unable to explain the fall in compliance produced by the distending pressure. The transfer of half the distending pressure suggests that the compliance of the functioning chest wall is similar to that of the lung over the pressure range studied.

65 IMMUNOREACTIVE INSULINE ACTIVITY AND AMINOACIDS CONCENTRATION IN AMNIOTIC FLUID IN RELATION TO FETAL GROWTH

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AA are performing a study of amniotic fluid parameters for evaluating fetal growth. I.R.I., determined by radioimmunoassay, according with charcoal method, and a few aminoacids /chiefly phenylalanine and tyrosine determined by fluorometry/ are taken into consideration. There is a significant negative correlation between I.R.I. and aminoacids. There is a significant positive correlation between the two aminoacids. The letter increase in fetal distress and in cases of fetal death. Other aspects concerning the relationships between biochemical data of amniotic fluid and fetal growth, the role of insuline for fetal growth and metabolism of phenylalanine and tyrosine in fetal life are discussed.

66 CONGENITAL CHLORIDE DIARRHEA /CCD/, CLINICAL EXPERIENCE WITH 21 CASES

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21 children with CCD have been treated by us since the disease was first diagnosed in Finland in 1961. All were born from a pregnancy complicated by hydramnios /2-6 l/, and 19/21 2 or more weeks before term. Watery diarrhea /about looo ml/m²/day/ was present from the first day of life, with Cl concentration of the fecal fluid exceeding loo meq/l. Absence of meconium was recorded in 14/21. Hyponatremia and hypochloridemia tended to develop, and later hypopotassemia and alkalosis with extreme dehydration. The diarrhea only decreased with dehydration. Normal fluid composition and hydration could be maintained through adequate water and electrolyte replacement. For this a solution of NaCl and KCl was used. The dose and Na/K-ratio were adjusted to maintain normal pH and electrolyte concentration, chloriduria, normal renin and aldosterone activity and normal body content of potassium. Normal development was thus achieved and renal changes could be avoided.