BIOGENESIS OF GALACTOSE: EVIDENCE IN GALACTOSEMIC INFANTS. R. Gitzelmann & R. Gaurth Hansen (Zurich, Switzerland & Logan, Utah)

In several galactosemic infants (hereditary deficiency of galactose-1-phosphate uridyltransferase) high levels of red cell galactose-1-phosphate have persisted for months despite rigid exclusion of galactose from diet. In vitro conversion of UDP-galactose to galactose-1-phosphate by erythrocyte lysates had been demonstrated earlier. It now is certain that such conversion is due to the pyrophosphorolysis of UDP-galactose. This reaction is usually thought of as providing a bypass for the transferase reaction in the conversion of galactose to glucose. It is now conceivable that in the young galactosemic under treatment, this bypass reaction is used in the reverse direction i.e. not for the breakdown of galactose-1-phosphate but for its buildup from glucose. Whether this will also take place in tissues other than the red cell i.e. in the central nervous system is unknown but must be considered since self-intoxication of these patients would ensue.

GLUCOSE FORMATION FROM ALANINE IN EARLY NEONATAL PERIOD.
A.Dacou-Voutetakis,M.Xanthou,H.Mameli,D.Anagnostakis and D.Nicolopoulos.

Alanine increases glucagon secretion and is considered a key aminoacid in gluconeogenesis.The hyperglycemic response to alanine load was studied in 50 newborns aged 18 hours to 9 days.During the first 48 hours of life the blood glucose increment following oral administration either of alanine (0.25g/kg) or saline was similar.At the end of the first week however a significant rise in blood glucose concentration was observed following alanine load.The defective glucose formation from aminoacids during the first 48 hours of life may indicate immaturity of the enzymes involved in gluconeogenesis or decreased responsiveness of pancreatic alpha cells to alanine, resulting in diminished glucagon secretion.

THE INTRAVENOUS L-ALANINE TOLERANCE TEST AS A MEANS FOR TESTING GLUCONEOGENESIS.
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The intravenous administration of L-alanine can be used to test gluconeogenesis.It is important that the glycogen stores of the liver are depleted before the test is performed. Only then can an increase of blood glucose be ascribed to gluconeogenesis from L-alanine, glycogenolysis being of minor importance. Therefore, we extended the fasting period previous to the test until the blood glucose concentration was 3 mmol/l approximately. We performed the test on 3 children with ketotic hypoglycemia, 4 with hepatic glycogenosis and 1 with a deficiency of hepatic fructose-1,6-diphosphatase. The former two groups responded with a marked increase of blood glucose, whereas blood lactate hardly varied. The results pointed to unimpaired gluconeogenesis. In the patient with a fructose-1,6-diphosphatase deficiency blood glucose did not increase, the elevated fasting lactate increased further and L-alanine elimination was delayed. These data pointed to impaired gluconeogenesis.

THE TOLERANCE OF NEWBORN INFANTS TO FULL PARENTERAL NUTRITION. B.S.Lindblad,B.Persson,G.Settergren.The Paediatric Dept. of Karolinska Institutet at St. Göran's Children's Hospital, Stockholm, Sweden.

During the last year, full parenteral nutrition has in our intensive care unit been given for shorter periods to 20 children. Out of these, 12 have been newborn. Their tolerance to the program given has been studied during the simultaneous infusion of 18 amino acids, glucose,fat, minerals,electrolytes and vitamins. The detailed composition of the i.v. alimentation program will be presented. The tolerance has been studied by ion-exchange chromatography of plasma amino acids, triglycerides, FFA, ketone bodies, insulin of plasma, puruvate, lactate and glucose of blood, as well as routine water, electrolyte and acid-base balance. Serious intolerance to amino acids, glucose and fat was seen in 3/12 newborn infants, in 2 cases with homocystine appearing in plasma, corrected by vitamin B6 administration.Sepsis complicated the treatment in 3 of the newborn children.The indications and contra-indications will be commented.The need for qualified supervision of parenteral nutrition to newborn infants will be stressed,in view of our limited knowledge of requirements in this age-group,as well as in liver-kidney-heart disease, respiratory insufficiency, trauma, infection and undernutrition, all conditions prevailing among the newborn infants requiring parenteral nutrition.

TOTAL PARENTERAL ALIMENTATION (TPA) IN THE NEWBORN: METABOLIC ALKALOSIS RESULTING FROM THE ADMINISTRATION OF AN EXCESS OF ANIONIC AMINO-ACIDS (a.aa.). G. Maffei-Negrin, R. Agostino, S. Nodari, G. Marzetti, P. Colarizi, and G. Bucci. Institute of Pediatrics, University of Rome Medical School, Rome, Italy.

36 newborns (28 weighing <2500 g) were given TPA because of extreme prematurity, apnoeic spells, RDS, or feeding difficulties. Owing to the composition of the commercial amino-acid mixture used for TPA, the full TPA regimen included the administration of an excess of a.aa. (aspartic+ glutamic ac.= 5.1 mmoles/kg/day) over cationic aa. (lysine+hystidine+arginine= 2.6 mmoles/kg /day), and an excess (of 4.0 mEq/kg/day) of fixed cations (mostly Na^+) over fixed anions. After a few days of TPA, hyperbasemia developed in most patients. These results show that TPA with an excess of a.aa. is able to produce a negative metabolic H^+ balance, and are in keeping with observations of Heird et al (New Engl J Med, 287; 943, 1972), showing that TPA with an excess of cationic aa. results in metabolic acidosis. Other pertinent clinical and laboratory findings will also be presented.

RESPONSES OF O_2-CONSUMPTION,RESPIRATORY QUOTIENT & SUBSTRATE UTILIZATION TO INTRAVENOUS NUTRITION OF PREMATURE INFANTS. J.Mestyan.Pécs,Hungary.

The examinations were perfomed on 7 premature infants on the 1st or 2nd day after birth.Aminosol glucose was infused at an average rate of 0.11 ml/kg/min representing a mean caloric load of 8.45 kcal/kg for an observational period of 4 hours.Urine was collected for determination of the non-protein RQ.O_2-consumption and CO_2-production of the infant kept at 34-36C were continuously recorded by the Kipp diaferometer.

The cumulative heat increment for a 4-hr.period of observation was found to be 0.79 kcal/kg representing 13.4% of the basal calories produced and 9.7% of the calories infused.The preinfusion RQ was low in all premature infants indicating that mainly lipids were metabolizad during the metabolic transition from intrauterine to extrauterine life.Aminosol glucose profoundly changed this metabolic state; glucose became the dominant energy-yielding substrate and the participation of amino acids as sources of energy also increased substancially. This shift in substrate utilization was quite independent of the changes in heat production indicating that the metabolism of the substrates may be accompanied by variable utilization of their energy content.