

MYOCARDIAL METABOLISM IN THE NEWBORN LAMB.

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Data on myocardial metabolism in the newborn are scanty. While "in vitro" studies demonstrated that foetal myocardium of near term monkeys is able to metabolize substrates different from glucose, qualitative data obtained "in vivo" in puppies showed that glucose is the only fuel for the newborn heart. Using a particular preparation-i.e. controlled perfusion of one coronary artery of an isolated newborn lamb heart with oxygenated blood from a donor animal, and total collection of venous drainage-quantitative studies were done on myocardial O_2 -consumption, uptake of glucose and NEFA, and production of lactate and pyruvate under basal and metabolic (glucose load) or hormonal (insulin, hydrocortisone) stimulated conditions. Results showed that newborn heart is able to metabolize NEFA(1), that this fact was enhanced by insulin addition(2) and depressed by glucose load(3), that insulin was a powerful agent to revert ventricular fibrillation sometimes spontaneously occurring in the isolated heart (4).

NEONATAL HYPOCALCEMIA IN INFANTS OF DIABETIC MOTHERS-HORMONAL EFFECTS. L. Bergman, I. Kjellmer and U. Selstam, Pediatric Clinic, Children's Hospital, Göteborg, Sweden.

Infants of diabetic mothers (IDM) are prone to develop neonatal hypocalcemia (NHC) more frequently than control infants. To evaluate the possible role of thyrocalcitonin (TCT) as a cause of NHC a group of 12 IDM was studied and compared with 11 control infants. The IDM had a significantly lower Ca level in serum (both total and ultrafiltrable) than the controls at 24 hrs, despite equal values at birth. Both groups had a significant increase of the TCT level at 24 hrs compared with 0 and 48 hrs. No significant difference between the 2 groups was, however, observed. The Ca level in serum at 24 hrs was significantly correlated to the TCT value in IDM but not in controls. In 2 controls and 3 IDM the concentration of parathyroid hormone in blood was measured at 0, 24 and 48 hrs. All infants had markedly reduced levels, compared to adult standards. One IDM, who developed NHC, increased the concentration of parathyroid hormone 3 times without affecting the serum Ca but with an increased phosphaturia. It is suggested that NHC is related to the high concentrations of TCT which inhibit the mobilizing effect on Ca from the skeleton by parathyroid hormone. The high incidence of NHC in IDM is ascribed to a more rapid turn-over of Ca in the skeleton of these infants--enhancing the effect of TCT.

RENAL GLOMERULAR FILTRATION AND ITS RELATIONSHIP TO HYPOCALCAEMIA OF THE NEWBORN. THE EFFECT OF HYPOCALCAEMIA ON THE DEVELOPMENT OF DENTAL ENAMEL.

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We have shown that in almost all breast fed infants there is a rise in plasma calcium between the first and seventh day, but in artificially fed infants half show a fall in calcium over this period, resulting in 9% actually developing levels of less than 7mgs/100ml. Urinary calcium excretion of 1-2mgs/24hrs. cannot account for the fall in plasma calcium. There is an inverse relationship between plasma phosphate and urinary phosphate, suggesting that high plasma phosphate is due to impaired renal excretion. Renal clearance of phosphate was proportional to creatinine clearance. High plasma phosphate was associated with low plasma calcium. Infants that showed a fall in plasma calcium had significantly lower creatinine clearance than infants that showed a rise. Every child who had had hypocalcaemic convulsions showed very severe abnormalities of the dental enamel. 17% of infants who had had symptomatic hypocalcaemia had similar lesions. The overall incidence of severe enamel abnormalities resulting in loss of the primary dentition by the age of 3-4 years, is between 1.5 to 2% of bottle fed infants.

PRESERVATION OF 2,3-DPG AND ATP IN CITRATED BLOOD

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ATP and 2,3-DPG have been shown to play major roles in the viability of erythrocytes and the preservation of function of hemoglobin A.

In the commonly used ACD preserved blood, concentration of ATP decreases slowly, with a half life of more than 2 weeks but the level of DPG decreases progressively with an half life less than 7 days.

The low pH of ACD is known to cause the decrease in DPG and the maintenance of ATP. On the other hand pH values over 7.2 cause a decrease in ATP while DPG is preserved.

We have investigated the two reciprocal pH dependencies of DPG and ATP and found an optimal pH range of 6.8 to 7.0.

The present study includes a comparison of DPG and ATP maintenance in three different blood preservations: ACD, CPD and a new solution derived from CPD with increased buffer capacity.

The solution enables the maintenance of stored blood pH within the described optimal range and therefore to preserve DPG as well as ATP.

THE EFFECT OF METHEMOGLOBIN ON THE EQUILIBRIUM BETWEEN OXYGEN AND THE HEMOGLOBINS F AND A₁ AT DIFFERENT CONCENTRATIONS OF 2,3-DIPHOSPHOGLYCERATE. H. Versmold & K.P. Riegel*, Univ. Kinderklinik, Munchen, Germany

Brain injury in hereditary NADH-methemoglobin reductase deficiency may result from a highly increased hemoglobin O_2 affinity. Since no data on the interactions of HbF, O_2 , met-Hb, and 2,3-DPG are available, O_2 affinity was determined (gas chromatography) in solutions of HbA₁ and HbF (5 mM in TRIS 0.05 M, pH 7.15; isolated by ion exchange chromatography) at different states of autoxidation. The results (P_{50} Torr at 37°C, pH 7.15; $X \pm$ S.D.)

| | Met Hb % | N no | 2,3-DPG 1mM | 2,3-DPG 5mM |
|------------------|----------|------|----------------|----------------|
| HbA ₁ | 5 | 4 | 12.2 \pm 2.2 | 25.0 \pm 2.1 |
| | 25 | 4 | 11.7 \pm 1.0 | 20.5 \pm 1.3 |
| | 50 | 4 | 9.2 \pm 0.8 | 15.8 \pm 0.7 |
| | 75 | 4 | 7.8 \pm 0.6 | 12.3 \pm 1.1 |
| HbF | 5 | 2 | 14.3 | 19.5 |
| | 65 | 4 | 6.9 \pm 0.6 | 8.8 \pm 1.2 |

show that partial oxidation increases O_2 affinity of HbF more than than of HbA₁. Although 2,3-DPG counteracts this effect, it fails to balance the highly reduced O_2 unloading capacity in severe methemoglobinemia. There is evidence that oxidation of HbA₁ occurs preferentially at the α -chains.

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CONGENITAL ERYTHROPOIETIC PORPHYRIA (CEP) WITH A HITHERTO UNDESCRIBED PORPHYRIN PATTERN. M.F. Seip, L. Eriksen, Depts. Ped. and Physiology, Univ. of Oslo, Norway.

A boy with the typical clinical picture of CEP showed an unusual porphyrin pattern. In addition to the expected very high urinary excretion of uroporphyrin I and high no. of erythroblasts with nuclear fluorescence in bone marrow, he showed large amts. of uroporphyrin III and especially of a 7 carboxylic porphyrin of the isomer III series in the urine. At 2 yrs of age the urine contained 11600 ug porphyrin per day, whereof 4330 ug uroporphyrin, 4330 ug 7-carboxylic porphyrin, 775 ug 6 carboxylic, 860 ug 5 carboxylic, 1270 ug coproporphyrin, and a small amt. of 3 carboxylic porphyrin. Findings of increased amts. of protoporphyrin in plasma and feces are also at variance with the picture seen in classical CEP. These data indicate that we're dealing with a biochemically "new" type of CEP. Meticulous protection against light with wavelengths below 510 nm removed skin lesions and brought about an almost complete compensation of the hemolytic anemia. Under light protection the ratio porphyrinogen/porphyrin in urine was 80/20; under incomplete light shielding 10/90. An important feature is thus the overproduction of porphyrinogen, which on light exposure in the skin is oxidized to porphyrin. Data seem to exclude that impaired activity of isomerase (uroporphyrinogen-III-cosynthetase) is the only or even the most important metabolic defect in this patient.