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Plasma glucose-insulin-free fatty acids interrelationships during long term corticosteroid treatment.

The effect of long term corticosteroid treatment on plasma glucose, insulin and free fatty acid levels was studied in 25 children. The fasting blood glucose level and glucose tolerance after oral glucose administration were normal in all cases. The mean plasma insulin concentration on the other hand was significantly higher during the glucose tolerance tests. Free fatty acid levels were moderately lower in the fasting plasma and significantly lower 30 minutes after glucose administration. This shift in the balance between the studied factors may contribute to the fat deposition often seen during corticosteroid treatment. Children- even if treated with corticosteroids for years-are able to maintain a normal glucose tolerance as a result of continualy elevated insulin level. Arginine stimulation also resulted in a trend to higher insulin levels in the corticosteroid treated group but the difference from the normal was much smaller than after glucose administration.

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Beta-D-glucuronidase deficiency mucopolysaccharidosis.

A two year old girl presented with mild facial dysmorphism reminiscent of Hurler disease,moderate mental retardation,and hepatosplenomegaly.Her corneae were clear.Radiologically, there was moderate dysostosis multiplex.Peripheral granulocytes contained abundant,coarse Alder-Reilly granulations.There was an increased urinary excretion of acid mucopolysaccharides.Determination of lysosomal glycosidases showed a markedly decreased activity of ß-D-glucuronidase in cultured fibroblasts,leudocytes,and serum.In the parents,in one grandmother and in one sister,ß-D-glucuronidase activity in serum and leucocytes was decreased to between 30 and 50%. The disorder has been classified as mucopolysaccharidosis,type VII. Our results indicate that a reliable diagnosis of the homozygous and heterozygous deficient state can be made by determining ß-D-glucuronidase in serum.

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An approach to a biochemical definition of low nitrogen and energy transfer across the placental membranes.

Determination of 24 free amino acid levels of plasma by ion-exchange chromatography, accuracy 2-5%. In maternal hypertention and maternal nutritional deprivation there are high foetal levels of alanine, glycine and proline and a resistance to the rapid decline normally seen neonatally. The findings are in agreement with the assumption of decreased gluconeogenesis from amino acids in foetal deprivation. The glycine/valine ratio can discriminate between small size at birth due to low post-menstrual age and low birth weight and crown-heel length in relation to post-menstrual age in maternal hypertention and in low socio-economic groups with maternal nutritional deprivation. The glycine/valine ratio is suggested as a valid parameter in the subclassification of the different anthropometric groups of newborn babies prior to clinical investigation.

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Studies on amino acids (a.a.) in plasma and erythrocytes in patients with hematological disorders.

Erythrocyte and plasma levels of most a.a. are similar. Aspartic acid and glutathione are present in red cells and absent in plasma, while glutamic acid and serine are more concentrated intracellularly. Cystine, methionine and arginine are more concentrated in plasma than in the cells. Plasma and erythrocyte a.a. have been determined in patients with hemolytic anemia and in patients with impaired erythropoiesis. Both in patients and controls, hydroxyproline, proline, cystine, methionine and tryptophan were not detected in the cells. Higher concentrations of valine, isoleucine, leucine, phenylalanine, lysine and arginine were found in plasma than in cells. In the patients, intraerythrocytic glycine, taurine and asparagine were increased, while threonine, tyrosine and histidine were decreased. There was no difference between the 2 groups of anemia. These results suggest that there are other transport mechanisms for the a.a. in the red cell membrane than in the mucosal and kidney tubular cells.