53 ENDOCRINOLOGICAL INVESTIGATIONS IN THALASSEMIA MAJOR.

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Hormonal status was assessed in 11 thalassemic children (2 boys and 9 girls) aged 5 to 18 years. All were on a high transfusion regimen and on regular chelation therapy. Investigations were performed in all children prior to transfusion.

The following studies provided normal results: glucose and insulin measurements during oral glucose tolerance test; HGH values during insulin tolerance test; ACTH, T₃, T₄ and TSH basal values. On the other hand, three abnormalities were observed:

1. Cortisol peak values during ITT were low in 4 patients; in these patients, the cortisol response to ACTH was normal.
2. After administration of TRH, mean TSH values were significantly elevated when compared to normal controls.
3. On LHRH test, 5 prepubertal girls out of 6 showed a low response for FSH and 3 out of 6 a low response for LH. One girl with delayed puberty had a low response for both FSH and LH. The 2 girls with normal puberty showed a normal response to LHRH.

It is concluded that gonadotrophin reserve assessment is the best way to detect the endocrinological impairment occurring in thalassemic children. The relationship between these disturbances and the iron overload remains conjectural.

54 ERYTHROCYTE METABOLISM IN ICTERIC TERM AND PREMATURE NEWBORN INFANTS.

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The metabolism of erythrocytes has been explored in premature newborns and in term and premature newborn infants who presented non immune haemolytic anaemia. Results were compared to those of normal term newborn infants.

Activities of erythrocytes hexokinase, 6-Phospho gluconate dehydrogenase, glutathione reductase, level of reduced glutathione and plasmatic level of vitamin E were similar in all groups. Superoxide dismutase activity was the same in icteric term newborns and normal term newborns.

The level of foetal haemoglobin and the activities of glucose-6-phosphate dehydrogenase and pyruvate kinase were significantly increased in premature newborn infants whether icteric or not. Pyrimidine nucleotidase (PN) activity was low in icteric premature infants either when related to reticulocytosis or in absolute value.

Results suggest that decreased activity of PN could be involved in the haemolytic process observed in premature newborn infants.

55 PRE- AND POSTNATAL SERUM ERYTHROPOIETIN (ESF) LEVELS IN MICE AND RATS. RESPONSE TO HYPOXIA.

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Serum ESF levels were investigated in mice and rats pre- and postnatally using a cell culture assay, measuring the formation of erythroid colonies (CFU-E) in methylcellulose after incubating newborn mice liver cells with serum. In fetal rats near term a slight ESF activity was present, with levels comparable to those found within 2 hours after birth. The ESF level then fell to undetectable values during the following 48 h. In mice increasing ESF level was found from the 3d day of life, with the highest concentrations obtained 15 - 20 days after birth. The ESF level thereafter decreased to low levels in adult animals. In fetal rats near term a short-lasting hypoxic period (0.5 atm for 6 h) slightly increased the ESF level, while the ESF level near term was not increased after chronic intrauterine hypoxia (0.5 atm for 8 days). Short-lasting hypoxia caused high ESF levels in newborn mice and rats, and even higher levels in adult animals. After chronic hypoxia the ESF level was not increased in adult animals.

The present results show that hypoxia stimulates ESF production in utero as well as postnatally. The increasing ESF levels postnatally correspond to a period with rapidly increasing myeloid erythropoiesis, decreasing hematocrit level, suckling and rapid growth.