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OGTT in thalassemic major children.

In order to assess early signs of dysfunction of the β-cells in 15 politransfused (mean Hgb=9.1±2.2 g/dl) prepubertal thalassemic major children (Th.),3-14 years old,an OGTT was carried out, after the consent of the parents. It was considered as normal when all glicemic (BS) and insulinemic (IRI) (Biodata) values were within 2 SD in confront to controls. An abnormal BS curve in two (6 and 12 years old) and an abnormally elevated IRI basal value in another two (10 years old) have been observed. The unidirectional Mann-Whitney test was calculated (null hypothesis rejected at P≥ 0.05). No statistically significant differences were found in BS or in IRI values at any time of the test. ΔBS and ΔIRI values and the ΔBS/ΔIRI ratio showed significant differences only at 30 min. BS and IRI peaks were at 60 min. in 9/14 and in 8/15 Th. respectively (3/9 and 3/10 in controls respectively). Our data show a delayed insulin secretion in Th. with abnormal BS or IRI curves in 3/5 Th. older than 10 years and in 1/10 Th. younger than 10 years. The β-cells function seems to be alterated precociously, but after the 10th year of life the impairement is more evident.

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Prolactin secretion disorder in the obese prepubertal and pubertal boy.

A disorder in the prolactin secretion has been demonstrated in the obese adult. No data of this kind exist concerning children.

38 obese boys (13 prepubertal, 25 pubertal) underwent a TRH test with assay of prolactin. Obese prepubertal boys show prolactin levels significantly lower than those of the control group under basal conditions ($p < 0.05$) and after stimulus ($p < 0.01$). In the obese pubertal boys the difference is significantly ($p < 0.05$) only after stimulus.

We can conclude that in the child and adolescent boy obesity induce a hypothalamo-pituitary disorder that also involves prolactin secretion.

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Precocious sexual development in a boy with Addison's disease.

After a short episode of fever and dehydration an 11 year old boy was diagnosed to have idiopathic Addison's disease with an ACTH level of over 4500 pg/ml. Since the age of 6 years he had manifested hyperpigmentation of the skin and buccal mucosa. At admission he showed advanced pubertal signs (P_4 - Tanner) for genitalia and pubic hair, and his bone age was 13 years. The plasma levels of testosterone were 63 ng/dl and the LH and FSH response to LRH stimulation (50 μg/m², i.v.) - 2.2 to 25.0 and 2.6 to 3.5 mIU/ml, respectively - confirmed the diagnosis of central precocious puberty. Basal plasma DHEA (51 ng/dl) and androstenedione (17 ng/dl) levels were very low for age (normal 180 and 40 ng/dl, respectively). It is proposed that the association of precocious puberty in this boy with latent untreated Addison's disease is due to a "drift phenomenon" of LRH and/or gonadotrophins, following the prolonged elevation of ACTH secretion. It is also possible that earlier activation of adrenal androgens by the ACTH contributed to the induction of the precocious pubertal process.

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Characterization of hGH from patients with LTD by a human liver radio-receptor assay (RRA).

A RRA for hGH has been developed using the 100,000 g pellet from human liver tissue homogenate obtained from a donor of kidney transplantation. This assay is sensitive in the physiological range of hGH (1.9 - 30.0 ng/ml). Human prolactin and human placental lactogen did not compete with hGH on its receptor. Dilution curves parallel to the standard curve were obtained by receptor-active hGH found in the plasma of 7 LTD patients, four stimulated hGH healthy children and 2 acromegalics. Examples of results as hGH ng/ml are:

Serum Dilution	Normal Patients	Acromegaly	LTD
1/4	5.6	7.6	4.4
1/8	2.8	4.4	2.8
1/16	1.4	3.0	1.4
1/32	-	2.1	1.0

Our findings using this highly specific RRA for hGH suggest that circulating hGH in LTD is biologically active and that the primary defect lies either in the receptors or in the intracellular mechanism generating somatomedins.

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Plasma glucose and alanine relationship after fast in normal children and children with ketotic hypoglycemia.

Plasma glucose and alanine concentrations were measured after a 24 hour fast in 46 normal children aged 1 to 8 years, and in 15 children aged 1 to 6 years with documented ketotic hypoglycemia (KH). Mean values (± SD) were significantly lower in KH than in controls: 334 ± 149 vs 509 ± 163 mg/dl ($p<0.001$) for glucose and 16.9 ± 4.9 vs 12.6 ± 3.6 μM/dl ($p<0.005$) for alanine. In each group, probit transformation demonstrated a gaussian distribution of both parameters. Additionally plasma alanine values were highly significantly positively correlated with glucose levels in controls ($y = 6.41 + 20.70\ x$, $r = 0.690$, $p<0.001$) and in KH ($y = 6.44 + 18.43\ x$, $r = 0.753$, $p<0.005$), the regression lines in the two groups being not significantly different in slope or position. These data demonstrate further that fasting values of plasma alanine and glucose are significantly lower in KH children than in normals, and are strongly correlated inside the two groups. The lack of any clearcut difference between the two groups demonstrates that KH appears as a borderline state of glycemic regulation and neoglucogenesis during fast in young children.

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HLA - haplotypes investigations in the cases of /AGS/ adreno-genitale syndrome/ children and their parents.

10 affected children /AGS/ and 3 healthy sibs from 11 families were typed for HLA-A and B antigens by the standard NIH lymphocytotoxicity test.

In one family two affected sibs shared both HLA haplotypes. Neither of the healthy sibs had HLA-haplotypes identical to the affected patients. As regards the individual HLA-types, B5/chi² = 1,661/B7/chi²=0,062/, B40/ chi²=1,272/ seem to emerge as more frequent than the average, but with the population frequency based on 222 healthy blood donors' HLA-A-B antigen frequencies there was no significant difference.