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Serum somatomedin activity (SSA) in children with operated craniopharyngioma and paradoxical growth.

SSA was measured by bioassay in 27 sera from 15 children aged 5 to 15 operated for craniopharyngioma (CP). All had complete GH deficiency, none received hGH. Plasma T4 was normalized by administration of thyroid powder. 14 received hydrocortisone (HC) 8-25 mg/m2/day, 13 did not. Growth velocity (GV) ranged 0 to 7.5 cm/year. SSA ranged 0.18 to 1.5 U/ml. No correlation was found between SSA and : daily dose of HC, insulin peak after arginine, delay from surgery, % of weight excess. A positive correlation was found between SSA and GV ($r = 0.81$, $p < 0.001$). Moreover, in patients receiving no HC therapy, GV correlated negatively to plasma HC ($r = -0.656$, $p < 0.02$).

These data demonstrate 1/ that the generation of SSA after removal of CP is independant from GH secretion and is not related to insulin secretion nor to weight excess or to the therapeutic dose of HC ; 2/ that the paradoxical growth in these patients is closely parallel to SSA and negatively correlated to endogenous plasma HC.

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Radioimmunological determination of somatomedin B in healthy children and patients with HGH deficiency.

In 192 boys and girls, aging from 1 month to 16 years, serum somatomedin B levels were performed by radioimmunoassay of Kabi Inc., slightly modified. In girls the mean value increased from the 1 st. month at the end of the second year of live from 10.5±6.2 to 19.2±9.2 mg/l, in boys at the same time from 10±3.9 to 20.3±6.2 mg/l. The values decreased in girls in the following years up to the age of 14-16 years to 13.7±7.5 mg/l; in the boys to 11.5±4.2mg/l. The beginning of the decrease was in girls two years earlier than in boys. To these values of normal children were compared the values of 8 patients with HGH deficiency. The mean of these cildren was 4.2 mg/l, significantly below the values of the normals. The determination of somatomedin B with the tested radioimmunoassay may be useful for the screening of HGH deficiency in short children.

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Combined virilizing 17-21 hydroxylase deficiency in siblings.

Female pseudohermaphroditism with skeletal abnormalities (cranial and antebrachial) was observed in 3 siblings. Marked hirsutism was observed during pregnancies in the mother. Ambiguous genitalia were present at birth but further virilization did not occur. Saltwasting, hypokalemia or hypertension was not present during long-term observation. Adrenal function studies were performed in 2 of the patients. Basal values of urinary cortisol-metabolites and 17-ketosteroids were normal but the excretion of corticosterone-metabolites and pregnanetriol was increased (ratio cortisol-/corticosterone-metabolites 1:1). After ACTH stimulation additional excretion of pregnanetriol and corticosterone-metabolites was found. Also urinary cortisol-metabolites were increased as were 17-ketosteroids, allthough only moderate. Basal ACTH and 17-OH progesterone values were only slightly increased. These observations suggest the presence of a combined partial 17-21 hydroxylase deficiency with skeletal defects. This clinical entity has not previously been described.

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Corticosteroidreceptors on human lymphocytes and platelets.

About 8ooo receptors are present on human lymphocytes of healthy persons at a dexamethasone concentration of approximately $2x10^{-6}$ M. We studied the behaviour of the corticosteroidreceptors present on lymphocytes and on platelets in dependance on structure and concentration. It was found that the corticosteroidreceptors present on platelets have a higher affinity than those on lymphocytes. This is documented by a higher reactivity and a lower saturation concentration. At 37° the saturation of the receptors occurs on platelets within 15 min. and on lymphocytes after 45 min. at a corticosteroidconcentration of about $2x10^{-6}$ M. The saturation concentration of dexamethasone for platelets is about $5x10^{-7}$ M, for lymphocytes approximately $2x10^{-6}$ M. The affinity of the receptors measured on behalf the saturation concentration depends on the structure of the corticosteroid investigated. We found that 6-α fluorination increases the affinity of a corticosteroid against lymphocyte receptors. The number of receptors however is independant from the corticosteroid used, but varies from person to person. For healthy persons we found about 25 receptors per platelet, and about 6ooo to 8ooo receptors per lymphocyte. At physiologic corticosteroidconcentrations 1 or 2 corticosteroid molecules are bound on 1 platelet and about 5o molecules can be measured on 1 lymphocyte.

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Age at first conscious ejaculation: A milestone in male puberty.

Complete physical examinations and bone age determinations are performed regularly in all children being followed longitudinally in our clinic. Boys approaching puberty are asked at each visit whether there has been a first ejaculation. A survey was made of 158 boys (97 normals (N), 36 with delayed puberty (DP) and 25 with cryptorchidism (Cr)) in whom it was possible to carry out an evaluation within an approximate 3 months of the first conscious ejaculation. Sixty four healthy school boys served as additional controls. The main findings were:

	Age BA - yrs		CA - yrs		Test.vol.
Group	m ± SD	range	m ± SD	range	ml
N	$13^{6}\pm0^{6}$	12^{6} -15^{6}	$14^{3}\pm0^{9}$	12^{6} -16^{6}	10 ± 4
Cr	$13^{6}\pm0^{6}$	12^{6}-15^{6}	$14^{4}\pm0^{7}$	12^{6}-16	11 ± 4
DP	$13^{3}\pm0^{6}$	11^{6}-15^{9}	$16^{1}\pm0^{1}$	14^{9}-19	11 ± 4

It is evident that the first ejaculation occurs within a narrow range of bone age although chronological age shows a much wider variance, as do testicular volume and secondary sexual signs. It is concluded that registration of the first conscious ejaculation may constitute an useful index of maturation in the male.

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Does good diabetic control influence serum lipids or platelet function in diabetic children?

Disturbances in carbohydrate metabolism, lipid pattern and platelet function are potential risk factors in the development of vascular complications in juvenile diabetics. In a prospective study including 29 diabetic children the relationship between these factors has been studied. Carbohydrate control was measured by HbA1, by Clinitest analysis of three urine portions a day, three days a week for two months, and by clinical judgement. Platelet function was studied by Born aggregometry.

Results: The relative number of positive Clinitest results was highly correlated to HbA1 ($r=0.83$). There was a complete accordance between clinical judgement and HbA1. A positive correlation was also found between HbA1 and serum triglycerides ($r=0.61$). The platelets from poorly controlled children often lacked shape change and showed slower aggregation velocity and less pronounced maximal aggregation.

Summary: HbA1 is a very good indicator of carbohydrate control. Serum triglycerides and platelet aggregation are affected by the degree of diabetic control.