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Dehydroepieandrosterone (DHA), androstenedione (A) and testosterone (T) plasma levels after rapid adrenal suppression and stimulation in prepubertal and pubertal boys.

The supposed role of adrenal secretions in male puberty was investigated by administering dexamethasone (DXM) 1 mg at 9 p.m., then Synacthen (ACTH) 0.25 mg/m2 at 9 a.m., blood beeing collected at 9 a.m. before and after DXM and 1 h. after ACTH. In 11 boys aged 3 to 6 at pubertal stage I (groupe IA) mean DHA levels were (ng/ml) 0.06 before DXM, less than 0.05 after DXM, 0.09 after ACTH. In 21 boys at pubertal stage I aged 7 to 13 (groupe IB) mean DHA levels were respectively 0.76, 0.35, 1.37. In 10 boys at pubertal stage II (groupe II) mean DHA levels were respectively 1.5, 0.82, 2.4. Mean A levels (ng/ml) were respectively 0.11, 0.05, 0.19 in group IA, 0.24, 0.10, 0.28 in group IB, 0.38 0.25, 0.48 in group II. Mean T levels (ng/ml) were respectively 0.05, 0.06, 0.08 in group IA, 0.11, 0.07, 0.14 in group IB, 1.5, 0.70, 0.58 in group II. In the 2 older groups, the variations were highly significant. These data confirm the rise of androgenic capacity of adrenals before puberty and suggest that a large part of circulating T derives both from direct secretion and indirect production by adrenals.

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"Tertiary" adrenocortical tumor and non-rhythmic ACTH excess in untreated congenital adrenal hyperplasia(CAH)

A 60-y-old single woman(46,XX)was admitted Oct. 1975 with evidence of a left adrenal tumor on IVP.The history and finding of urogenital sinus and clitoral hypertrophy suggested CAH.Preoperative steroid excretion: 17-kgs 218,17-ks 91,pregnanetriol 60,DHEA 1.9,11-OH-andro 21,etio 32mg/24h,resp.94,28,14,0.3,5 and 9mg/24h after a standard oral dexamethasone test.Basal CSR 30mg/24h,plasma testosterone 202ng%,plasma ACTH 62 (8h), 54(17h)pg/ml.A large adrenal tumor(410gm)without metastases was removed;normal internal genitalia,including ovarian histology.Steroid excretion fell approx.50%, but remained poorly suppressable.After removal of the right adrenal(58gm,nodular hyperplasia)steroid excretion reached very low values.Multiple plasma ACTH assays during oral cortisol substitution,(Dexter et al. JCE 30,573,1970)revealed little fluctuation (max.280, min.180pg/ml),suggesting pituitary microadenoma.These findings support the need for continued ACTH-suppression in CAH.

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FSH : Evidence for a possible prepubertal regulation of its secretion by the germinal epithelium in cryptorchid boys.

Two groups of prepubertal unilateral cryptorchid boys (UCT) underwent stimulation tests with 1 µg (n=10) or 25 µg (n=8) of LHRH. Two groups of prepubertal bilateral cryptorchid boys (BCT) were similary studied (1 µg, n=6 ; 25 µg, n=8). The number of spermatogonia was determined in both sides in each group after testicular biopsy. UCT groups exhibited FSH and LH responses to LHRH similar to those of prepubertal control groups. Mean numbers of spermatogonia were low on the cryptorchid side (12 ± 6 to 25 ± 12) and normal on the normal side (>50). Both groups of BCT had very low counts of spermatogonia on both sides (means of 7 ± 4 to 14 ± 8). Their FSH responses to LHRH at both dosages were significantly higher than those of the normal prepubertal controls. LH responses were identical. These data suggest that the negative feedback mechanism between FSH secretion and the spermatogonia observed in the adult male is also operative in the prepubertal boy.

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Danazol in the treatment of Precocious Puberty.

Danazol is a synthetic analogue of ethinyl testosterone with a capacity to inhibit gonadotrophin release. Six children (4-10.5 years) have been treated with oral Danazol for up to 17 months. In three females, the precocious puberty is idiopathic, but neurogenic in two other females and one male. Danazol was given in divided doses, 9-17 mg/kg/day, and produced no side effects. In all patients, reduction in breast (testicular) size was noted and further menses were prevented in three patients. Linear growth and bone maturity were not influenced by therapy. Hypothalamo-pituitary maturity was demonstrated prior to treatment by the LH and FSH response to LH/FSH-RH. Reassessment in several children on treatment shows some "blunting" of the peak response to LH/FSH-RH. Improved academic performance was noted in the boy and a useful reduction in sexual drive occurred in the oldest girl. Danazol is effective in controlling some aspects of precocious puberty and further long-term studies are justified.

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Unusual Pseudo-Cushing's Syndrome in a 3-Years Old Boy

In July 75 the patient developed a Quinke edema and a short asthma like period, ceasing after 40 mcg 6-methyl-prednisolone. 3 days later chicken-pox occured. Within few weeks a Cushing like phaenotype was established, including elevated blood pressure. In an outwards children's hospital no further abnormalities were found except two determinations of plasma cortisol: 1.0 and < 1.0 mcg%. At first no special treatment; slight attenuation of Cushing's symptoms. In March 76 a typical Addisonian crisis occured. The boy was set on hydrocortison, 12.5 mcg/day, and lost step for step all pathological signs in his clinical course. Main laboratory data: Cortisol basal (before substitution or at morning before hydrocortisone):<2 mcg%; ACTH basal: lowest limit value; ACTH-test: appropriate response; HGH, TSH, LH, FSH: normal responses; LVP-test: positive response; aldosterone, renin, angiotensin II, transcortin: normal values. Speculation: Viral induced local encephalitis with production of an antibody-like non-steroidal substance with corticoid properties resulting in Cushing's syndrome followed by a hypothalamic lesion resulting in CRF-insufficiency.

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Iodine-deficient goiter in newborns during the course of neonatal screening.

Iodine deficieny in adults causes preferred synthesis of triiodothyronine(T3)an observation not having been reported in newborns with this condition.After starting a local program for screening of congenital hypothyroidism neonatal iodine-deficient goiter with hypothyroidism was observed in a frequency of 1.2 per cent.Mean total thyroxine(T4)in 8 goitrous newborns was 6.2 +1.45 µg/100 ml(+SD)compared to15.5+2.9 µg/100ml in normalweight fullterm newborns at 4 days of life.Mean total T3 in iodine-deficient newborns was 2.59±0.65 ng/ml compared to 1.5 ng/ml in normal newborns of the same age.Serum thyrotropin (TSH) remained elevated during the second half of the first week in iodine-deficient newborns. Percutaneous application of iodine resulted in a quick disappearance of goiter and normalization of T4 and tsh within a few days.T3 returned to normal more slowly within days or weeks.General iodine prophylaxis is needed in Germany.