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AUTONOMOUS INTERSTITIAL CELL HYPERPLASIA AS A CAUSE OF FAMILIAL PRECOCIOUS PUBERTY.

True precocious puberty (TPP) generally is considered to result from gonadotropin mediated premature gonadarche. However, this report concerns two brothers (46/XY) in whom TPP was caused by autonomous hypersecretion of testosterone (T) from interstitial cell hyperplasia (ICH). Salient clinical features were:

Pat.	CA	HA	BA(yrs)	Testes	Phallus	Pubarche
J.S.	3.3	8.3	10.5	6 cc	11 cm	stage IV
M.S.	2.3	4.3	3.0	6 cc	9 cm	stage II

Basal sex hormone concentrations are listed in the table below:

Pat.	17-KS	T	Free T	DHT	Δ4	DHAS	17-OHP	LH	FSH
J.S.	3.0	595	138	73	52	78	94	<1.5	<1.5
M.S.	1.5	483	96	57	28	7	80	<1.5	<1.5

Turnover studies revealed normal T clearance but excessive T production. Serum androgen concentrations were unresponsive to ACTH stimulation or Dexamethasone suppression. Excessive adrenal androgenesis was further excluded by split adrenal vein catheterization. Basal serum and 24-hour urine LH, FSH, and β-HCG concentrations were undetectable and there was no significant increase of RIA or bioassay LH/FSH in response to GnRH infusion. Testicular biopsy revealed patchy diffuse ICH, with mature spermatogenesis in the 6 y.o. These studies suggest that autonomous ICH may cause TPP in the absence of significant gonadotropin secretion.

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Changes of pituitary and adrenal functions after discontinuation of cyproterone acetate (CPA) therapy in girls with precocious puberty (PP).

In 5 girls with PP who had been treated with CPA (95-133 mg/m^2 x d) for years, the spontaneous secretion (4-hourly) and the pituitary reserves of LH,FSH,TSH,HGH, and ACTH (as measured by cortisol (F) and dehydroepiandrosterone sulfate (DS)) were determined before and 3 months (m) after discontinuation of the drug. All hormones were measured by RIAs previously described. For PRL the Serono Kit was used. - During CPA therapy (tx) the most consistent abnormalities found were elevated PRL levels after stimulation (200/ug TRH and insulin-induced hypoglycaemia (IIH)) and low F concentrations with subnormal responses to IIH. Basal and stimulated (25/ug LH-RH, 200/ug TRH) concentrations of LH,FSH, TSH were variable (low, normal, or normal to elevated). DS concentrations were unmeasurable or low (<25-120/ug/ml). Discontinuation of CPA resulted in a rapid normalization of basal F and PRL. After 3 m off tx the F responses to IIH were normal except in 1 girl (ΔF 59-176 ng/ml). Δ PRL had normalized to 26.2-39.7 ng/ml (during CPA 36.9-95.6 ng/ml). FSH remained unchanged and LH increased in 2 girls. HGH remained subnormal in 1 patient. - We conclude that PRL and F levels are rapidly normalized after discontinuation of CPA, while other parameters are not consistently changed.

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Fertility and hormonal values in adult subjects operated for cryptorchidism during childhood.

An investigation on infertility and hormonal status was carried out in 107 subjects, aged 18-35yrs, who were operated for cryptorchidism during childwood.The group consisted of 50 bilateral and 45 unilateral cryptorchid as well as 1 subject with Klinefelter's syndrome and another with Kallmann syndrome.The sperm count results indicated that 70% of the bilateral and 20% of the unilateral subjects were infertile.There existed a good correlation between the sperm counts and the FSH values before and after LH-RH functional test.The LH values didn't demonstrate any good correlation.The response for LH and FSH after LH-RH in 6 subjects was diminished, although amongst them only 1 bilateral cryptorchid patient was infertile.Plasma testosterone recoreded a range between 69 and 737 ng/100 ml in this group and didn't correlate either to sperm counts or to the gonadotropins.The data, on one hand, show poor fertility prognosis in surgically operated cryptorchid subjects and on the other difficulties in hormonal assestment which could be responsible factors for cryptorchidism.

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Basal,stimulated and circadian hormone levels in Turner's Syndrome

Apart from hypogonadism several endocrine disorders have been reported in patients with Turner's syndrome.Therefore 14 patients with true XO-syndrome aged 13.3-18 years were studied.Every 1-2 hours by day and half-hourly at night blood was obtained for hormone assays. In addition LH-RH- and TRH-tests were performed. Thyroid function was in the normal range except for one patient. Remarkable high sleep-induced HGH values were observed.- Basal LH was 6.9 times and FSH 25.1 times higher than normal.There was a great circadian variation in LH and FSH levels as found in postmenopausal women. In our hypogonadal patients mean LH levels were significantly increased ($2p < 0.01$) during sleep in accordance with LH secretion in normal puberty whereas in normal prepubertal and adult persons no sleep effect on LH concentrations is seen. This pubertal gonadotropin secretion pattern seems to be independent of ovarian sexual steroids. LH-RH resulted in a 312.6% increase in LH levels and in a 163.4% increase in FSH levels,elevationswhich were significantly less ($2p < 0.02$) than normal. There was a highly significant strong correlation between mean 24-hour-gonadotropin concentrations and LH- and FSH-peaks following LH-RH.Thus, pituitary response to exogenous LH-RH was closely related to spontaneous secretion of LH and FSH.-In animals LH secretion is suppressed by melatonin.In our patients,however,high levels of LH and melatonin were found.

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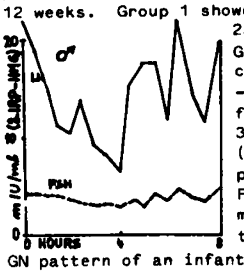
Pulsatile Secretory Pattern of Gonadotropins ir Infants

In adult males and females luteinizing hormone (LH) and to a lesser degree follicle stumulating hormone (FSH) are secretedin a pulsatile fashion: an abrupt rise of the hormone is followed by a slower decline during 1 to 3 hours. Prior to puberty gonadotropin (GN) levels are low and show little fluctuation. Recently high GN levels as well as gonadal activity were demonstrate in infants. The following examination was carried out to elucidate the type of GN secretion in early infancy. LH and FSH were determined every 30 minutes over a period of 8 hours in three diffenrent groups: Group 1: 2 male and female adults, Group 2: 2 male and female prepubertal children, Group 3: 3 male and 3 female infants, age 6-12 weeks. Group 1 showed a clear pulsatile secretion of LH (4,5-23,5 mIU/ml(range)) and FSH(6,0-16,0 mIU/ml) Group 2 demonstrated a rather constant secretion of LH (1,5-2,3 mIU/ml) and FSH (1,6-4,9 mIU/ml). Group 3: In male infants we found a pulsatile secretion of LH (3,6-34,7 mIU/ml) and to a lesser degree of FSH (1,8-4,6 mIU/ml). In female infants the pulsatile secretion was more pronounced in FSH (6,5-22,7 mIU/ml) than in LH (1,5-4,7 mIU/ml). In conclusion, the secretory pattern in early infancy shows a pulsatile type.



GN pattern of an infant

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Primary empty sella syndrome with micropenis and hyperprolactinemia.

Empty sella syndrome is a rare entity in paediatrics. A case is presented in a 13-year old boy who was referred for a micropenis. He was born at term by breech delivery and recovered rapidly from a neonatal subarachnoid hemorrhage. Psychomotor development was normal. Physical examination showed a micropenis with normal prepubertal testes and a delayed growth with height at - 3 standard deviations. There was no sign of puberty and no gynecomastia or galactorrhea. Endocrinological studies indicated growth hormone, TSH, ACTH and gonadotropin deficiencies contrasting with elevated basal plasma levels of prolactin (260 µg/l) which failed to increase with TRH stimulation. Skull roentgenograms, tomography of the sella turcica and computerized tomographic brain scan were normal. There were no abnormalities of the fundi and visual fields. Diagnosis of empty sella was made on pneumoencephalogram showing the absence of a sella mass. Treatment with bromocryptine led to a normalisation of the plasma prolactin levels but failed to modify the other hormonal pituitary dysfunctions. Increased statural growth was obtained under substitutive therapy with human growth hormone, thyroxine and hydrocortisone.