35 GROWTH IN CHILDREN WITH 45 XO TURNER'S SYNDROME

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Growth records of 64 patients with chromosome constitution 45XO have been analyzed. The extremely short stature of adults with this condition (142.5 cm) appears to be due to the cumulative effects of intrauterine growth failure, a gradual decline in height velocity during childhood and to the absence of a pubertal height spurt. Pubic hair appeared in 62% of the patients but was delayed both in relation to chronological age and to bone age. There was no evidence of a pubertal height spurt even in those children in whom pubic hair appeared.

Treatment with oestrogens in 18 patients caused development of secondary sex characteristics but did not affect final stature. The age of the patient when treatment was given was not of significance in affecting the final height of the patient.

The adult heights of 17 patients were compared with those of their parents; a regression coefficient was found which was identical to that found in 51 normal girls, a roughly constant amount of height being lost through the chromosomal abnormality. This suggests that the genes whose action underlies the variation in adult height amongst the normal population must be located for the most part on the autosomes.

36 EFFECTS ON GROWTH IN LONG TERM TREATMENT WITH HUMAN GROWTH HORMONE (HGH) IN HYPOSOMATROPISM

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Long term treatment with HGH (Crescormone®) in seven children with hyposomatropism according to Hall&Olin Acta Endocr 69:417, 1972 gave the following results:

	N	growth in cm/year mean	range	HGH mg/kg per week mean	range
control	7	3.0	2.0- 4.0	-	
1 year	7	9.7	6.6-15.3	0.26	0.19-0.30
2 "	6	7.4	4.0-10.4	0.27	0.15-0.33
3 "	5	6.0	5.1- 7.6	0.25	0.15-0.38
4 "	5	5.0	2.7- 8.3	0.25	0.16-0.38
5 "	2		5.8- 5.7		0.24 0.15
6 "	2		5.0 4.5		0.29 0.14
7 "	2		5.0 7.0		0.28 0.24
8 "	2		5.0 5.9		0.23 0.27
9 "	2		(7.5) 6.2	(+testosterone)	0.22

The results indicate as sustained and dose dependent response. The long term growth may be compared with an intermittent treatment, Kirkland & al J Clin Endocr Metab 37:204,1972:

HGH given in intermittent-		total growth increment cm		in continuous schedule		
year	N	mean	range	N	mean	range
4	7	20.7	14.6-25.4	5	31.3	19.2-46.0
8	2		39.4 40.2	2		41.3 67.6
9	1		43.5	2		48.8 73.8

Thus continuous treatment with 0.2-0.3mg/kg/w gives a substantial gain in long term growth and is recommended.

37 IRRADIATION OF THE HEAD. IMMEDIATE EFFECT ON GROWTH HORMONE SECRETION IN CHILDHOOD.

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The effect of radiation on the normal hypothalamic - pituitary unit is not known. In the present work an attempt is made to study prospectively the effect of irradiation on the "intact" hypothalamic pituitary axis in children. Nine leukemic children, aged 2½-13 years without clinical or laboratory evidence, of CNS involvement were studied. Plasma growth hormone (PGH) was determined by radioimmunoassay in samples obtained hourly from 8 pm to 7 am before and after a 3-week course of "prophylactic" cranial irradiation (2400 rads). The mean hourly PGH value prior to irradiation (4.1±1.6 ng/ml) was similar to that of 6 endocrinologically normal children (4.09±2.4 ng/ml), a finding strongly suggesting normal pituitary function prior to irradiation. Following irradiation the corresponding PGH value (2.7±1.6 ng/ml) was lower than before ($p < 0.02$). The findings suggest an immediate suppressive effect of irradiation on the apparently normal hypothalamic pituitary axis

38 ASSESSMENT OF HYPOTHALAMO-ANTERIOR PITUITARY SECRETING CAPACITY IN CHILDREN ON THE BASIS OF A SINGLE TEST.

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A combination of insulin hypoglycemia with administration of the recently synthesised releasing hormones TRH and LH-RH allows almost complete evaluation of the anterior pituitary function in one single test. The combined test was performed in over 60 children with suspected hypothalamo-pituitary disorders. Insulin (0,1 U/kg) was injected intravenously followed by TRH (200 ug) followed by LH-RH (100 ug). Growth hormone, ACTH, LH, FSH, TSH and cortisol were measured by radioimmunoassay and blood sugar by an enzymatic technique. The assays were shown to be specific and there were no cross reactions between the different hormones assayed. The normal response to insulin hypoglycemia is not altered by simultaneous injection of the releasing hormones. Similarly the TSH-response to TRH alone does not differ from that observed during a combined test. The combined test has proved to be useful for the differential diagnosis of short stature in children and various disorders of suspected pituitary hypothalamic origin. The diagnosis of an isolated or combined hormone deficiency can thus be made on the basis of a single test.

39 IS THERE A CORRELATION BETWEEN GH AND LH SECRETION DURING THE NIGHT?

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Spontaneous GH- and LH-secretion during the night was measured by RIA in 27 children and adolescents with normal (n=9) or disturbed pubertal development (precoc.pub., pit.tum., Turner's S.). Incidence and correlation of episodic bursts of these two hormones during sleep and wakefulness were evaluated. Sleep was recorded by observation, blood was drawn every 3o min. from 7 p.m. to 8 a.m. In the 9 controls a total of 29 episodes each of GH- and LH-secretion was recorded. 5 of these GH- and 3 of the LH-bursts occurred following sleep onset (within 6o min.), 19 GH- and 2o LH-peaks during the entire sleepperiod. 14 out of 29 coincided (increase simultaneously or up to 3o min. apart), only 2 of these following the onset of sleep. In the patients with abnormal puberty (n=18), 55 GH- and 38 LH-episodes were found, 7 GH- and 2 LH-peaks followed sleep onset. The rate of coincidence of 15 (total period of observation) and 1 (sleep onset) respectively is proportionally smaller than for the normal adolescents. In none of the 27 a coincidence of all secretory episodes was recorded. If no correlation between GH and LH existed, about 2o% of the episodes (calculated from the relative time they cover) should coincide by chance, but 4o% did. Nevertheless, these data do not yet seem to justify the assumption of a common regulatory mechanism of GH- and LH-secretion in puberty.

40 PREPUBERTAL TESTICULAR RESPONSE TO CHORIONIC GONADOTROPIN IN CRYPTORCHIDISM :

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A delayed maturation in endocrine testicular function of cryptorchids was suggested by previous data. In the present study plasma Testosterone (T) Androstenedione (A) 17 OH-Progesterone (17 OHP) Progesterone (P) and Dehydroepiandrosterone (DHA) were evaluated by specific radioimmunoassay before and after stimulation with HCG, 1500 U every other day, plasma samples being collected the day after 1 (1500 U), 3 (4500 U) and 9 injections (13500 U) in 24 prepubertal cryptorchids and 12 controls aged 4 to 13 years.

In cryptorchids basal levels for all steroids were similar to those of controls. Post stimulatory increase after 1500 U was not significant, or of poor significance in both groups. After 4500 U the increase for all steroids except DHA was highly significant ($p<0.01$) in both groups. The response of cryptorchids for T and A was significantly lower than in controls ($p<0.01$), while 17 OHP and P showed no significant difference. Results were similar after 13500 U. No difference appeared between unilateral (14 cases) and bilateral (10 cases).

It may be concluded : 1/ that 3 injections of 1500 U seem advisable for comparison of the two groups studied ; 2/ that with this dose T and A response is significantly lower in cryptorchids than in controls. The present data suggest a prepubertal impairment of Leydig cell function in uni and bilateral cryptorchidism, possibly related to quantitative and/or qualitative testicular defect.