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Evaluation of long term growth response to hGH treatment.

The growth velocity (GV, cm/y) induced by hGH treatment was evaluated in 85 hGH deficient patients classified into 4 groups. Gr. A: Isolated hGH deficiency (IGHD), males, n=22; age at start of therapy (age): 0.8-19.6 y (m=9.6y). Gr. B: IGHD, females, n=19; age 0.4-16.9 y (m=8.6 y). Gr. C: Multiple pituitary hormone deficiencies (MPHD), males, n=30; age 1.3-21.4 y (m=11.7 y). Gr. D: MPHD, females, n=14; age 2.5-18.3 y (m=10.3 y). Follow-up ranged from 1-12 y with actual treatment amounting to 40-60% of the time. Treatment was given in courses of 3-24 months, 2-6 Ux3/wk with intervals of 2-14 mos. The regression equation of GV (cm/y) as a function of age and age² was calculated using program BMDP2R (Dixon & Brown, 1979). The formula is:

$$GV = K_1 + K_2 \times \text{Age} + K_3 (\text{Age} - 12.5)^2$$

| | K ₁ | K ₂ | K ₃ |
|---------|----------------|----------------|----------------|
| Group A | 8.844 | -0.205 | +0.220 |
| Group B | 14.465 | -0.619 | -0.007 |
| Group C | 10.129 | -0.243 | +0.004 |
| Group D | 9.436 | -0.229 | +0.009 |

This formula enables the evaluation of long term response to hGH comparing expected and observed growth velocity.

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Endocrine studies in anorexia nervosa (AN) concurrent with XO-gonadal dysgenesis (GD).

Comprehensive endocrinological investigations were performed in a 15 yr old girl with XO-GD during AN and after recovery. This is the 10th report and first endocrine study of this combination. Basal parameters before, during and after recovery from AN were as follows:

| Plasma | LH | FSH | TSH | PRL | hGH | T ₄ | T ₃ | Ur. MHPG |
|--------|--------|-------|-------|-------|------|----------------|----------------|----------|
| | mIU/ml | | uU/ml | ng/ml | | ug/dl | ng/ml | mg/24h |
| Before | 15.62 | 21.70 | 3.9 | 2.2 | 4.2 | 9.8 | 1.8 | |
| During | 0.23 | 2.36 | 3.7 | 2.0 | 10.6 | 4.8 | 0.8 | 0.33 |
| After | 12.00 | 40.00 | 9.4 | 3.6 | 2.0 | 8.5 | 1.5 | 1.24 |

LH and FSH after LH-RH showed a prepubertal pattern during the anorectic stage and after recovery rose back to the castration range. TSH and PRL increases after TRH were normal but prolonged during AN and several months after recovery. There was a marked paradoxical rise of hGH after TRH, LH-RH and oral glucose, which decreased after recovery. These findings are in concordance with dynamic endocrine profiles we found in 21 pubertal girls with AN. It seems to be the only condition to abolish the hypergonadotrophic state of GD, by reducing the negative feedback threshold back to its prepubertal level. This supports the hypothesis of a reversible hypothalamic dysfunction in AN.

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24-hour Blood Glucose Monitoring in Diabetes Mellitus
Twenty diabetic children aged 3-14 years have had 24-hour plasma and urine glucose estimations.

Two-hourly blood Dextrostix levels were read on a 'Hypocount' meter (duplicates checked in laboratory (r:0.92)). Concurrently glucose excretion was estimated in two 12-hour urine collections. Dextrostix values demonstrated the following patterns which resulted in improved diabetic control:

| Unrecognised nocturnal 'Somogyi' effect | Nature of altered control |
|---|--|
| Apparent nocturnal hypoglycaemia | Smother control with longer acting insulin |
| 'Honeymoon' diabetes | Nocturnal epilepsy diagnosed |
| Poor compliance | Minimal insulin given (high renal threshold) |
| | Correction of type and timing of insulin |
| Poor insulin absorption due to lipotrophy | Injection sites altered |

24-hour Dextrostix monitoring revealed disturbances of blood glucose dynamics which were not always apparent on simple urine testing and which facilitated improved control.

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The importance of radial metaphyseal band width.

The authors compared the relatively radiodense metaphyseal band width at distal end of the radius /Edlin, Whitehouse, Tanner, 1976/ and cortical thickness and diaphysal diameter of the metacarpal bones /Bonhards, 1968/ of the same wrist roentgenograms. On the basis of comparing the viewpoints of sensitivity, specificity, reproducibility and simplicity Edlin's method was found more utilisable which was carried out in the following groups: hypothyroidism/12 cases/: the T₄ serum level showed a close correlation with ratio /eliminating the influence of age and sex/ of the radial band width/correl. coeff.: 0.93/; Graves disease without treatment/10/: 1.25±0.267; dwarfism without treatment/27/: 0.522±0.229; dwarfism before and during anabol steroid treatment/12/: 0.800±0.366; 1.447±0.744 resp.; GH deficiency before and during hGH treatment/12/: 0.701±0.379, 1.788±1.157 resp.; obesity with advanced high and bone age between the ages of 4-10/11/: 1.217±0.261. On the basis of the results gained and the comparison of literature data the authors assume the relation of Somatomedins to radial metaphyseal band width.

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Evaluation of the combined test of hypothalamic-pituitary function.

In 47 children with disturbances in growth and/or puberty a test was performed with simultaneous injection of insulin (0.1 U/kg), TRH (3-5 µg/kg) and LHRH (100 µg). The mean bone age retardation was 39 months and the mean standard deviation score - 2.8 SD. In 11 children a Growth Hormone deficiency (GHD) was demonstrated: GH peak value 3.34 mU/l and Somatomedin 0.63 U/ml (mean) - 5 of these were patients with intracranial tumours. The other 36 children had GH peak value 20.44 mU/l and Somatomedin 1.27 U/ml (mean). ACTH response was subnormal in 15/32. In these 15 patients 13 had GHD or subnormal GH response. In the 15 children in the highest age group (girls > 12 years, boys > 14 years) a normal response to LHRH was found, regardless of pubertal development or coexisting GHD. In the 24 children in the youngest age group was found a definite abnormal response in 4, two of which had GHD. TSH response was low in 6/45, three of which were hypothyroid and had GHD. Defect of 3 pituitary axes were found in 5 patients, 3 of these had intracranial tumour. In conclusion: The combined test of pituitary function seems to be of limited value. The essential information is acquired by the hypoglycemia to establish GHD and ACTH deficiency (these 2 are often coexisting). Stimulation with LHRH is valuable in older children without signs of puberty and may be of prognostic value in young prepubertal children with GHD. Stimulation with TRH does not give any supplemental information.

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Male pseudohermaphroditism - asymmetrical gonadal dysgenesis.

A child with ambiguous genitalia at birth was studied over 17 years. Both a penis with hypospadias and a vagina was shown to be present. Laparotomy at the age of one year showed an uterus - on the right side no gonad, but a hypoplastic fallopian tube could be seen - on the left side a gonad, shown to be infantile testis and a normal fallopian tube. The sex of rearing changed during time: girl/boy/girl. At the age of 14 hormonal study showed a very raised FSH and LH, in spite of the presence of a testis producing testosterone causing progressive virilization. The left testis was removed, it showed Sertoli cells and a few germinative cells in the tubules and Leydig cell hyperplasia. A biopsy from the place of the ovary on the right side showed ovarian stroma, no primordial follicles. This sort of hermaphroditism with a dysgenetic testis and a "streak gonad" is called mixed gonadal dysgenesis or asymmetrical gonadal dysgenesis. Chromosome analysis showed a mosaic of 71% 46 XY and 29% 45 XO (blood) cells. Further analysis on the patient and the parents made a structurally abnormal Y-chromosome, a chimera and mosaicism due to simple non-disjunction improbable. It is concluded, that this case of asymmetrical gonadal dysgenesis must be due to anaphase lag. The course and the sex of rearing is discussed.