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Epidemiological and pathogenetic aspects of growth hormone deficiency in the Netherlands.

In a nationwide survey 121 boys and 51 girls with growth hormone deficiency were found. There were 14 boys and 6 girls with a tumour or other organic cerebral disease. The case histories of the remaining 107 boys and 45 girls with idiopathic growth hormone deficiency were examined for birthweight, duration of pregnancy, type of delivery and parity of the mother. There were 60 boys and 20 girls with multiple pituitary hormone deficiency (MPHD); and 47 boys and 25 girls with isolated growth hormone deficiency (IGHD). Breech delivery occurred in 55 % of the children with MPHD and in 15 % of the children with IGHD. Fifty-three of the children (34,9 %) were firstborn. The average birthweight and duration of pregnancy was normal; the number of prematurely born children was not increased. Neither birthweight, nor duration of pregnancy or parity appeared to be factors in the aetiology of GH-deficiency. Breech delivery is an important factor, although the incidence of GH-deficiency in breech deliveries is less than 1/1000.

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Developmental analysis of voice deviations in children with congenital hypothyroidism.

A harsh, low pitch voice is evidently one of the hallmarks in congenital hypothyroidism. We have described the significance of voice analysis in a developmental study of three newborn and young infants and seven older children of congenital hypothyroidism. The voice analyses have been made with Cry Analyser (Vuorenkoski, L. et al: Acta Paediat Scand 1973: Suppl 236, 27) and with FO Indicator (Vuorenkoski, V. et al: Arch Dis Child 1978:53, 201). The data from the three infants followed up from the beginning of the treatment show a rapid primary development: the mean length and the mean number of cry sounds are normalized in 1-3 weeks but the development of the pitch and harshness is slow. The clinical follow-up data indicate the possibility that the latency in the onset of normalization may have a prognostic value. The analysis of the voice pitch after the first year of life from seven patients (age 2-13 years) show that the pitch in these children remains below the normal level.

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Familial male pseudohermaphroditism due to C_{17}-C_{20} desmolase deficiency.

Three sibblings were seen for various degrees of abnormal sexual morphology. The first was stade II of Prader, the 2nd stade III, the third only had a bifid scrotum. In the two elder, basal levels (ng/dl) of 17αOH Progesterone (OHP), 17αOH Pregnenolone (Δ^5OHP) and Progesterone (P) were 10-20 fold above normal while those of Testosterone (T), Δ^4-androstenedione (Δ^4), DHA and DHAS were low. None of these hormones including cortisol increased significantly under ACTH suggesting high endogenous ACTH levels. Dexamethasone (Dex) treatment suppressed all hormones. After hCG, T rose slightly (7→8 and 12→85) while OHP and Δ^5OHP reached respectively 1065, 1050 and 662, 405 in each child. hCG + Dex confirmed the low T response (7→63 and 1→50 in the 2 children), abnormal increase of OHP (1→90 and 14→47), Δ^5OHP (22→65 and 15→78). In the 2nd child after Fluoxymesterone P, OHP, Δ^5OHP levels decreased to almost normal values while T, Δ^4, DHA were unchanged. It is concluded that both testis and adrenals exhibit the enzyme defect. That the last newborn sibbling was also affected by the same defect was suggested on abnormally low levels of DHA (210) and T (15) and high levels of P (32156), OHP (1350) and Δ^5OH (2382) at birth.

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Relationship between structure of a corticosteroid and its biologic activity, studied on human lymphocytes.

The biologic activity of a steroid depends widely on its binding properties against plasma proteins and against cell receptors. Because of their use for treatment of leucemia, we are interested to study the influence of the structure of a corticosteroid on its binding properties against lymphocytes and plasma proteins. Specific binding on plasma proteins is diminished by 6-α fluorination and inhibited by 9-α fluorination, unspecific binding on albumin and specific binding on lymphocytes however is increased. Nevertheless no direct correlation could be found between binding on lymphocytes and suppression of lymphocyte transformation by corticosteroids, if measured in lymphocyte cultures stimulated with phythemagglutinine.

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How should we assess thyroxine dosage?

Serum T4, T3 uptake, T3, basal TSH and TSH response to TRH were measured in 19 children aged 1-17 yrs with agoitrous hypothyroidism on thyroxine. Normal values were established using 15 well children aged 2-16 yrs with constitutional short stature.

The pituitary-thyroid axis appeared to function normally in the hypothyroid children; the response to TRH was not influenced by the age of onset of hypothyroidism or duration of therapy. Δ TSH was the best single determinant of abnormal biochemical thyroid status, but the serum T4 and basal TSH used in combination were equally satisfactory providing the child was not suffering from intercurrent infection.

Four children had normal TRH respons (Δ TSH 4-21 mU/L) 5 exaggerated (Δ TSH 50-1125 mU/L) and 10 suppressed responses (Δ TSH -1 to +1 mU/L). There was a poor correlation with the clinical impression of thyroid status, and with growth velocity and bone age.

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Ectopic thyroid as a cause of hypothyroidism in children

In 52 hypothyroid children thyroid scintigraphy (131 I or Tc-99m) permited to detect ectopic thyroid in 25 cases (48,1%), athyrosis in 5 cases (9,6%), and thyroid at the normal site in 22 cases (42,3%). Ectopic thyroid was also found in two children at the time of operation, before the addmitance to our hospital. The group of 27 children with ectopic thyroid consisted of 17 girls and 10 boys from 6 months to 18 years old. Signs of hypothyroidism were evident before the end of the first year in 21 children (77,8%), but only 4 (14,8%) were diagnosed at that time. Serum thyroxine (n=18) ranged from 0,8 to 2,8 µg/dl (mean 1,35, SD±0,51). Values of TSH (n=10) ranged from 25 to 500 µU/ml. IQ (n=17) ranged from 36 to 101. Conclusion: 1/ Ectopic thyroid is a prime etiological factor of hypothyroidism in our group. 2/ Ectopic thyroid becomes insufficient within the first year of life in 77,8% cases.