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CALORIC SUPPLEMENTATION INDUCES GROWTH ACCELERATION WITHOUT Sm-C CHANGES IN A LARON TYPE DWARF.

Circulating Sm-C reflects protein synthesis and the ability to stimulate skeletal growth in close relationship with caloric intake. The occurrence of spontaneous chronic underfeeding in a boy with Laron type dwarfism allowed to study the effect of caloric intake on growth in primary Sm-C deficiency. The 4 yrs old boy had basal GH > 40 ng/ml, Sm-C/IGF I < 0.10 U/ml unresponsive to hGH treatment, typical features with hypoglycemia and severe dwarfism. He was studied over 2 periods. Period I (5 mo) control with spontaneously low caloric intake of 60 Kcal/Kg/d and 1.4 g/Kg/d protein. Period II (5 mo) with 130 Kcal/Kg/d and 3.8 g/Kg/d protein given by constant enteral infusion. Results were the following

	Growth cm/yr	Sm-C/IGF I U/ml
Period I	1.6	0.02 - 0.07 (n=12)
Period II	9.4	0.01 - 0.04 (n=11)

Weight gain paralleled growth acceleration and hypoglycemia attacks disappeared. Refeeding with a normal diet resulted in a substantial catch-up growth in spite of unchanged Sm-C values. In conclusion, this study provided the opportunity to demonstrate that growth may be stimulated in the absence of normal levels of circulating Sm-C/IGF I. It suggests that nutrition may act independently of circulating Sm-C and GH and plays a major role in promoting statural growth.

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HIGH INCIDENCE OF PRIMARY EMPTY SELLA SYNDROME (PESS) IN CHILDREN WITH GROWTH HORMONE DEFICIENCY (GHD)

Computer-assisted tomography (CT) with 2mm axial sections and reconstructions was carried out in 31 children with GHD. Group 1 included 18 pts (11m, 7f, age 2-12yrs, height -3.3 ± 0.8 SD) with idiopathic complete isolated GHD (peak GH on provocative stimuli below 4 ng/ml, otherwise normal pituitary function), group 2 3 pts (2m, 1f, age 6-14yrs, height -1.8 to 2.2 SD) with idiopathic partial isolated GHD (peak GH above 4 and below 7 ng/ml), group 3 2 girls (age 2 and 17 yrs, height -2.0 and -3.1 SD) with idiopathic panhypopituitarism, and group 4 8 pts (6m, 2f, age 7-12yrs, height -2.6 ± 0.9 SD) with acquired hypopituitarism: 4 with isolated hypopituitarism (3 post-radiation, 1 craniopharyngioma), 4 with panhypopituitarism (2 craniopharyngioma, 1 glioma, 1 histiocytosis X). Density in the intrasellar area on CT corresponded to that of cerebrospinal fluid in 12 of 18 pts (group 1), 0/3 (group 2), 1/2 (group 3), and 2/8 (group 4, both postradiation). The overall incidence of PESS in the GHD pts studied was thus over 48%, while in children without endocrine dysfunction, it was only 5/213. It is concluded that PESS is more frequent than was assumed until now and that is most frequently associated with GHD.

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MR IMAGING (MRI) OF PITUITARY GLAND AND STALK IN IDIOPATHIC GROWTH HORMONE DEFICIENCY (IGHD).

To search for the presence of morphostructural abnormalities of the hypothalamus - pituitary region in GH children MRI was performed in 28 IGHD pts (21M, 7F, age 10.1 \pm 3.4, range 4.2-18) and in 15 healthy age matched controls (10M, 5F, age 8.3 \pm 2.9, range 4 to 12.4). Isolated GHD had been demonstrated in 19/28 pts, multiple pituitary hormones deficiency (MPHD) in 9/28; none had ADH deficiency. Hypothalamic or pituitary deficiency was equally distributed in the two groups. We evaluated: presence, size, shape of the pituitary stalk; pituitary and sella volume calculated according to Di Chiro. MRI revealed that in 18/28 pts the pituitary stalk was separate from the gland (Group 1); in 14/18 pts the neurohypophysis was dislocated in the hypothalamic region; the pituitary and sella volumes were markedly reduced in all GHD pts compared with controls. No special endocrinological feature characterized the 2 groups. Conclusions: GHD pts frequently present some degree of abnormality of the hypothalamus-pituitary region; this picture is equally distributed between isolated GHD and MPHD pts. The observed dislocation of the neurohypophysis does not induce ADH deficiency.

PATIENTS	PIT.VOL. mmc	SELLA VOL. mmc	PIT.VOL./SELLA VOL.
GROUP 1	69.7 \pm 39.2	159.6 \pm 85.4	0.43
GROUP 2	175.5 \pm 43.4	271.5 \pm 65.1	0.64
CONTROLS	256.8 \pm 53.7	390.1 \pm 95.5	0.73

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GROWTH HORMONE INSUFFICIENCY ASSOCIATED WITH A SMALL NODULE OF THE PITUITARY STALK-NEUROHYPOPHYSEAL ECTOPY?

Two prepubertal boys without any neurological signs nor diabetes insipidus were studied at age 12 and 15 years (bone age 9 and 12 years) because of declining height velocity. Arginine and insulin failed to induce GH release whereas TRH test results and cortisol response to insulin were normal. Skull radiographies did not show any abnormalities. On CT scan the pituitary stalk was suspected to be abnormal, but only with magnetic resonance imaging (MRI) a small round posterior nodule of the pituitary stalk was clearly demonstrated in both cases. Its qualitative appearance was equal to neurohypophyseal tissue. The lower part of the stalk was reduced to a filament leading to a hypoplastic pituitary gland which seemed to lack the posterior lobe. This suggests that in our patients the neurohypophysis may be located in an ectopic position attached to the pituitary stalk. Biopsy or surgical removal was avoided. The patients responded markedly to hGH therapy. At present, a malformation of the pituitary stalk seems to be a rare cause of GH insufficiency, but it is likely that a more extensive use of MRI for evaluation of apparent idiopathic hypopituitarism may reveal a higher incidence of such anomalies.

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TRANSIENT GROWTH HORMONE DEFICIENCY DURING BROMOCRYPTINE TREATMENT FOR HYPERPROLACTINEMIA

Growth hormone (GH) deficiency has been demonstrated in several patients with prolactinoma. Very often it was attributed to tumor expansion and found to be permanent after surgery. In a 14-year-old boy with galactorrhea, arrested puberty (testes volume 4ml) and poor growth (3cm/year) elevated prolactin (PRL) levels (5800 mU/l) were found. MR imaging showed no abnormalities of the pituitary gland. Nighttime sampling revealed persistent PRL elevation and low GH peaks (max. 12 mU/l). GH response to insulin induced hypoglycemia was also low (max. 10 mU/l) but GH increased normally after GRF injection (38 mU/l). Basal and stimulated LH and FSH values were prepubertal. 3 h. after the administration of 10 mg bromocryptine (B) PRL dropped to normal values and GH increased to 22 mU/l. After six months of treatment with B, height velocity doubled and normalization of GH secretion was observed. Basal LH, FSH and testosterone values remained prepubertal. In conclusion, GH deficiency in hyperprolactinemia can be restored during B treatment. The acute GH response to B may be predictive of the subsequent normalization of GH secretion during long term treatment. In isolated hyperprolactinemia a central dopaminergic defect is probably responsible for this deficient GH secretion.

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PROLACTIN SECRETION IN 149 GH DEFICIENT CHILDREN.

One hundred forty nine children and adolescents with growth hormone (hGH) deficiency were reviewed retrospectively in relation to plasma basal and TRH stimulated prolactin (Prl) levels before and during long-term hGH therapy. The following groups were distinguished: Gr. I - IGHD (n=62, 41M, 21F); Gr. II - MPHD (n=62, 44M, 18F); Gr. III - Intra- or suprasellar tumors (n=25, 17M, 8F). High basal Prl (> 20 ng/dl) were found in 6 IGHD patients (9.7%). During hGH therapy Prl normalized in all six. Nineteen (30.7%) MPHD patients had high basal Prl. Following L-T4 therapy Prl normalized in 14 of them. All MPHD patients with high basal Prl had higher than normal TSH response to TRH. High basal Prl levels were unrelated to stress and E2 therapy. It is assumed that in some hypopituitary patients hGH deficiency is due to hypoactivity of the catecholaminergic tone leading to GH-RH deficiency and high Prl levels. This is reversed by hGH therapy. A low response of Prl was found in 10 IGHD and 10 (16.1%) MPHD patients. A pituitary lesion was proven in these patients by a low response of GH to GH-RH and TSH to TRH. It thus seems that some IGHD patients are actually MPHD being deficient in both hGH and Prl. In the tumor patients, 2 had high Prl preoperatively. Post-op 5 had a low Prl response to TRH indicating complete hypophysectomy and 6 developed high Prl levels indicating hypothalamic or stalk injury. It is concluded that dynamics of hPrl secretion should be investigated in all hypopituitary patients, including IGHD.