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NOCTURNAL SECRETION OF GROWTH HORMONE (GH) AND GONADOTROPINS IN 3 PATIENTS WITH CROHN DISEASE (CD) BEFORE THERAPY AND DURING REMISSION

Arrest in somatic and sexual development in patientswith CD is well known. To elucidate the influence of pituitary hormones as GH and the gonadotropins in this process we measured nocturnal secretion of GH, LH, and FSH in 3 patients (1 girl, 12 years old, prepubertal, and 2 boys, 15 and 17 years old) before therapy and during remission. The boys had developed puberty stages 3 and 4 (according to Tanner).

Blood was collected every 20 minutes from 10 p.m. to 6 a.m., sleep was registrated by observation. In all patients GH-secretion during the acute phase of the illness showed no significant nocturnal peaks. Nevertheless baseline GH levels were slightly elevated in atleast one patient. During remission all patients showed restored nocturnal GH-peaks. The gonadotropin levels in the prepubertal girl were undetectable before therapy as well as in remission, no nocturnal pulses were visible. The 2 pubertal boys demonstrated typical pulsatile nocturnal LH secretion. FSH levels were in the pubertal range as well before therapy and during remission. Whether the malnourished situation, the catabolic state or other factors caused the GH-peak depression is yet not clear. In our patients with CD we saw no differences between the spontaneous gonadotropin secretion before therapy and during remission as it is known in other catabolic situationes for instance in anorexia nervosa.

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MAGNETIC RESONANCE IMAGING (MRI) VERSUS CT SCAN IN EMPTY SELLA SYNDROME IN CHILDREN.

Primary empty sella syndrome (PESS) in children is reported in many endocrine disorders, mainly in GH deficiency. In order to better define the anatomical basis of this syndrome, 7 patients (3 girls, 4 boys) presenting with PESS and a complete and isolated GH deficiency (GH peak AITT < 6 ng/ml ; n = 6) or an isolated diabetes insipidus (DI, n = 1) were studied by MRI.

Results :

The patient with isolated DI and a large sella on plain film had a true empty sella filled with CSF. 3 patients with a normal or enlarged sella on plain film had a small pituitary gland, 2 of them with a probable normal posterior lobe. 3 patients with a small sella on plain film, had a normal pituitary gland with an enlarged pituitary stalk. One patient had a normal pituitary gland.

In conclusion :

MRI is superior to CT to demonstrate the anatomy of the pituitary gland. The patient with the isolated DI, with variation in polyuria, shows no identifiable antehypophyseal tissue.

It is our opinion that PESS in children as defined by CT scan, groups heterogenous anatomical anomalies well depicted by MRI.

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AGE-DEPENDENT ALTERATION OF NOCTURNAL SERUM MELATONIN IS A FUNCTION OF INCREASING BODY SIZE IN CHILDREN AND ADOLESCENTS.

Although the biological significance of the pineal hormone melatonin (MLT) in man is still obscure, a certain amount of information on production, secretion patterns and excretion of this hormone has been obtained in recent years. However, data on potential alteration of serum MLT levels during a human lifetime are fragmentary and inconsistent.

We examined day- and nighttime serum MLT in 367 individuals (210 males, 157 females) aged 3 days to 90 years. Blood samples were collected between 7:30 am and 10 am and between 11 pm and 1 am. MLT levels were measured by RIA. Nighttime serum MLT concentrations were low (27.3+5.4 pg/ml; x+SEM) during the first six months of life. They then increased to peak values at age 1 to 3 years (329.5+42.0 pg/ml). From there levels decreased fast until age 15 to 20 years (62.5+9.0 pg/ml). During the following decades MLT declined moderately until old age (29.2+6.1 pg/ml; age 70-90 yrs). This biphasic MLT decline follows two exponential functions with different slopes (from age 1-20 yrs: r=-0.56, $p<0.001$, $y=278.7 \cdot e^{-0.09x}$; from age 20-90: r=-0.44, $p<0.001$, $y=84.8 \cdot e^{-0.017x}$). The decrease of nocturnal serum MLT in children and adolescents (1-20 yrs) correlated with the increase in body weight (r=-0.54; $p<0.001$). At a later age (20-90 yrs) there was no correlation between these two variables. Daytime MLT levels were low without age-related alterations.

This study reveals major alterations in nocturnal serum MLT levels during a human lifetime. The negative correlation between MLT and body weight in childhood and adolescence is evidence that expansion of body size is responsible for the huge MLT decrease during that period.

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HYPERNATRAEMIA-HYPODIPSIA SYNDROME AND HYPOPITUITARISM WITH VARIABLE SIADH AND PARTIAL ADH DEFICIENCY.

A 7 year old girl had pituitary hypothyroidism (T4 37 nmol/l peak TSH response to TRH <0.1 mu/l) and GH deficiency (Peak GH response to insulin hypoglycaemia, 1.7 mu/l). Serum sodium was persistently elevated (155-183 mmol/l). She showed absence of thirst, (fluid intake <500 ml/d), profuse nocturnal sweating and poor temperature control. CT Scan was normal. Maximum urine concentrating ability was 1078 mosm/kg when plasma osmolality was 330 mosm/kg. During a hypotonic saline infusion after oral water loading, plasma osmolality fell from 273 to 258 mosm/kg and free water clearance initially increased from 1.18 ml/100 ml GF to 5.42 ml/100 ml GF before falling to -1.13 ml/100 ml GF. This indicates the development of SIADH. During a hypertonic mannitol infusion plasma osmolality rose from 300 to 307 mosm/kg and free water reabsorption remained subnormal (1.13 ml/100 ml GF) but increased to 3.65 ml/100 ml GF after DDAVP. Increased fluid intake of 2000 ml/m^2 resulted in nocturnal enuresis.
These results indicate profound hypothalamic and pituitary dysfunction, partial ADH deficiency. Temporary SIADH when a hypotonic saline load was administered is previously undescribed in this condition and suggests that forced water loading may be potentially dangerous.

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WILMS' TUMOUR: AUXOLOGICAL AND ENDOCRINOLOGICAL STUDY

In Wilm's tumour, there is frequent spontaneous deletion of chromosome 11 and insulin and growth factor genes are located close to the deletion area. We studied growth and glycemia situation in 42 Wilms tumour cases in complete remission and off therapy for at least 2 years. We also studied adrenal, thyroid and gonad efficiency, as regards adrenalectomy, and irradiation (local, abdominal and abdominal-thoracic). Growth: initial height was normal; non-irradiated subjects' final height resembled chronological, while lower in irradiated subjects (-0.24 SDS±0.95). Differences between standing and sitting height at the latest examination indicate probable spinal column growth deficiency in irradiated subjects. 64.5% responded normally to arginine (like normals). Glycemia situation: in glucose oral load glycemia area is normal, insulin lower ($p < 0.05$), peptide C higher ($p < 0.01$ at 90 mins.). ACTH test: adrenalectomy cases'cortisol response was lower, n.s. compared to others. Thyroid: irradiated prepubertals' FT4 is significantly lower, irradiated pubertals' TSH is lower. Gonads: totally irradiated impubertal males' testosterone is lower; irradiated pubertal females' estradiol is lower. In conclusion an abnormality of chromosome 11 does not seem to be responsible for the alterations observed in our patients; on contrary irradiation and chemioterapy seem to have a real significance

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ISOLATED SALIVARY GLAND INSENSITIVITY TO ALDOSTERONE; A VARIANT OF PSEUDOHYPOALDOSTERONISM

A male infant, the first child of a non-consanguineous parents, presented at age 4 months with salt depletion and dehydration (plasma sodium (Na) 114, potassium 5.1 mmol/l). He quickly responded to I.V. Saline. During hyponatraemia his plasma renin activity (7625 ng $AIC^{-1}hr^{-1}$, NR 472-3130) and plasma aldosterone (PAldo) (3670 pmol/l NR 100-500) levels were appropriately elevated and his renal Na reabsorption was normal (fractional tubular excretion 0.2%, N < 1.0%). A normal Na intake of 3 mmol/kg/day led to a further fall in plasma Na (124 mmol/l), an increase PAldo (16,700 pmol/l) and a fall in urinary Na exception. A search for the source of Na loss revealed sweat Na in the upper normal range (53,58 mmol/l), normal rectal potential difference (60 mV) indicating normal intestinal & Na transport, and a markedly elevated salivary Na (170, 151 mmol/l, NR 4-10). Fludrocortisone 0.1 mg. daily induced no fall in salivary Na. On an intake of Na 6 mmol/kg/day, normal plasma Na levels were maintained and he thrived. His parents' salivary Na levels were < 15 mmol/l. We suggest that salt depletion was caused by salivary Na loss due to selective end-organ insensitivity to circulating aldosterone.