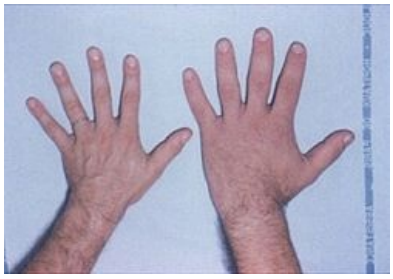
Growth hormone (quantity disorders)

GH excess

Tumors from adenohypophyseal somatotropic cells release large quantities of GH, resulting in:

- in children to **gigantism**,
- in adults to **acromegaly**.

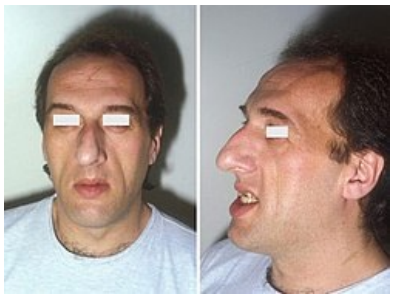


Acromegaly hands

Acromegaly

In patients with acromegaly, hypersecretion of GH is also accompanied by hypersecretion of prolactin (in 20–40 %). Acromegaly can be caused by both intrahypophyseal and extrahypophysal growth hormone-secreting tumors, as well as hypothalamic GRH-secreting tumors (but these are rare). The main findings in acromegaly are manifestations of local action of the tumor (enlargement of the sella turcica, headache, visual disturbances) and further manifestations of GH excess:

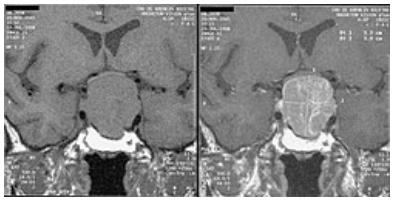
- enlarged legs and arms, protrusion of the lower jaw (prognathism);
- excessive growth of the facial, frontal and basal bones of the facial part of the skull and thus coarse facial expression (*acromegalic facies*);
- body hair increases (hirsutism);
- skeletal changes and a tendency to osteoarthritis;
- lactation even without pregnancy (in 4 %).



Acromegaly prograthism

Gigantism

Pituitary gigantism is characterized by **excessive linear growth**. The growth rate is usually excessive and in addition to a taller figure, other somatic symptoms typical of acromegaly may be present.



Acromegaly pituitary macroadenoma

GH deficiency

The incidence of GH deficiency is estimated at 1 : 10,000 worldwide.

Pituitary disorders

Most patients with growth hormone deficiency appear to lack GRH. Some of them have a sufficient number of adenohypophyseal somatotrophs with significant GH stores. Long-term treatment of GRH in these patients may lead to STH release and improved growth. Patients with pituitary tumors or with congenital pituitary absence (which is rare) do not have somatotrophs. There are also families that lack different parts of the STH gene. These individuals initially respond to the administration of exogenous STH, but many of them soon develop high levels of antibodies, which terminate the favorable course of therapy.

Dwarfism

GH receptor disorder

In another group of nanic patients, plasma GH levels are normal or elevated, but GH receptors are areaactive (or absent) due to a mutation in the receptor gene with loss of its function. The resulting condition is referred to as **growth hormone insensitivity - Laron syndrome**, which is characterized by low plasma concentrations of IGF-1. IGF-1 levels do not rise after administration of exogenous GH, but IGF-1 administration increases growth rate and suppresses GH concentrations. Plasma IGFBP-3 levels are also markedly reduced. This disorder is inherited autosomal recessively.

Defect in IGF-1 production

Similarly, African Pygmies have normal plasma GH concentration, low IFG-1 levels, and normal IGF-2 levels. They do not respond to exogenous GH by improving growth rate and increasing IGF-1 levels because they have a congenital inability to produce IGF-1, which is more important for growth stimulation than IGF-2. Pubertal growth spurt (sudden acceleration of growth) is absent in pygmy children, suggesting that IGF-1 is essential for reaching a normal peak growth rate. IGF-1 treatment is likely to accelerate growth in this population during childhood and puberty, but there are no reports of such treatment.

Clinical manifestations

Congenital deficiency

- normal body length at birth;
- growth rate slows shortly after birth (can be detected by carefully performed measurements during the first year);
- patients are short, obese with an immature facial expression, a high voice, and a delay in bone maturation;
- hypoglycemia and convulsions may be present in neonates or children, and boys may have micropenis.

Intelligence is normal in patients with GH deficiency unless brain development has been impaired by recurrent or severe hypoglycemia. In addition, anatomical defects may occur in the midline: optic hypoplasia with visual disturbances from nystagmus to blindness combined with various hypothalamic disorders (including diabetes insipidus), about half of patients lack septum pellucidum (on CT or MRI), cleft palate affects about 7 % of patients. Milder forms of partial GH deficiency are also described. There are several types of congenital GH deficiency:

1. type IA is inherited autosomal recessively, patients have a disorder in the GH gene, some children have a short birth length;
2. type IB is also inherited autosomal recessively, but there is no gene depletion;
3. type II is inherited autosomal dominantly;
4. patients with type III suffer from X-chromosome-linked GH deficiency.

Gained deficiency

If GH deficiency occurs in late childhood or adolescence, it is an acquired GH deficiency, which can be caused by craniopharyngioma, germinoma, glioma etc. If manifestations of deficiency of other pituitary hormones also occur, it can also be a hypothalamic-pituitary tumor. Also, empty sella syndrome (hypothalamic-pituitary abnormalities) may be associated with GH deficiency, more often in childhood than in adulthood. Irradiation of the head in the hypothalamic-pituitary region in the treatment of head tumors can lead to growth hormone deficiency in 6-24 months as a result of damage to the hypothalamus or pituitary gland. These patients should be closely monitored for growth disorders after irradiation.

Diagnosis of GH concentration

Basal GH levels are **low in healthy children** and in patients with GH deficiency. Therefore, GH deficiency is diagnosed by insufficient increase GH in serum after provocative stimulation.⚠ **Tests may not detect a lack of GH if the GH response to stimulation is normal.**

With the exception of sleep tests, tests should be performed after an overnight fast (because, for example, carbohydrate intake suppresses the GH response, see above). GH secretion is also inhibited in obesity, so fat children may have an apparent lack of GH.

Serum GH levels should rise during sleep at stages 3 and 4 (about 90 minutes after falling asleep) or 10 minutes after intense exercise. After an overnight fast, GH levels should also rise after an infusion of arginine or orally administered levodopa (a dopamine agonist). GH levels also rise in acute hypoglycemia after insulin administration. However, this insulin tolerance test carries the risk of seizures if the glucose level drops excessively. Therefore, the patient must be under the supervision of a physician, must not have a history of hypoglycemic convulsions and must have a normal glucose level at the beginning of the test. Within 20-40 minutes, there is a 50 % drop in blood glucose, which should be followed by an increase in serum GH (and cortisol). Glycemia should be monitored continuously and an intravenous route prepared for infusion of 10-25% glucose solution in the case that the patient loses consciousness and hypoglycemic convulsions (however, glycemia should not exceed the range too high after infusion, otherwise there is a risk of hyperosmolality!).

Links

Related articles

- Growth hormone
- Growth hormone (general)
- Growth hormone (secretion)

Source

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