PRIMEVIEW

ACROMEGALY

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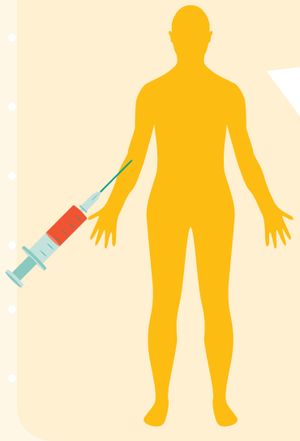
Acromegaly is a hormonal disorder involving increased levels of growth hormone (GH) and, consequently, insulin-like growth factor 1 (IGF1), typically due to a pituitary adenoma. Excess hormone levels lead to progressive somatic disfigurement and various systemic complications.

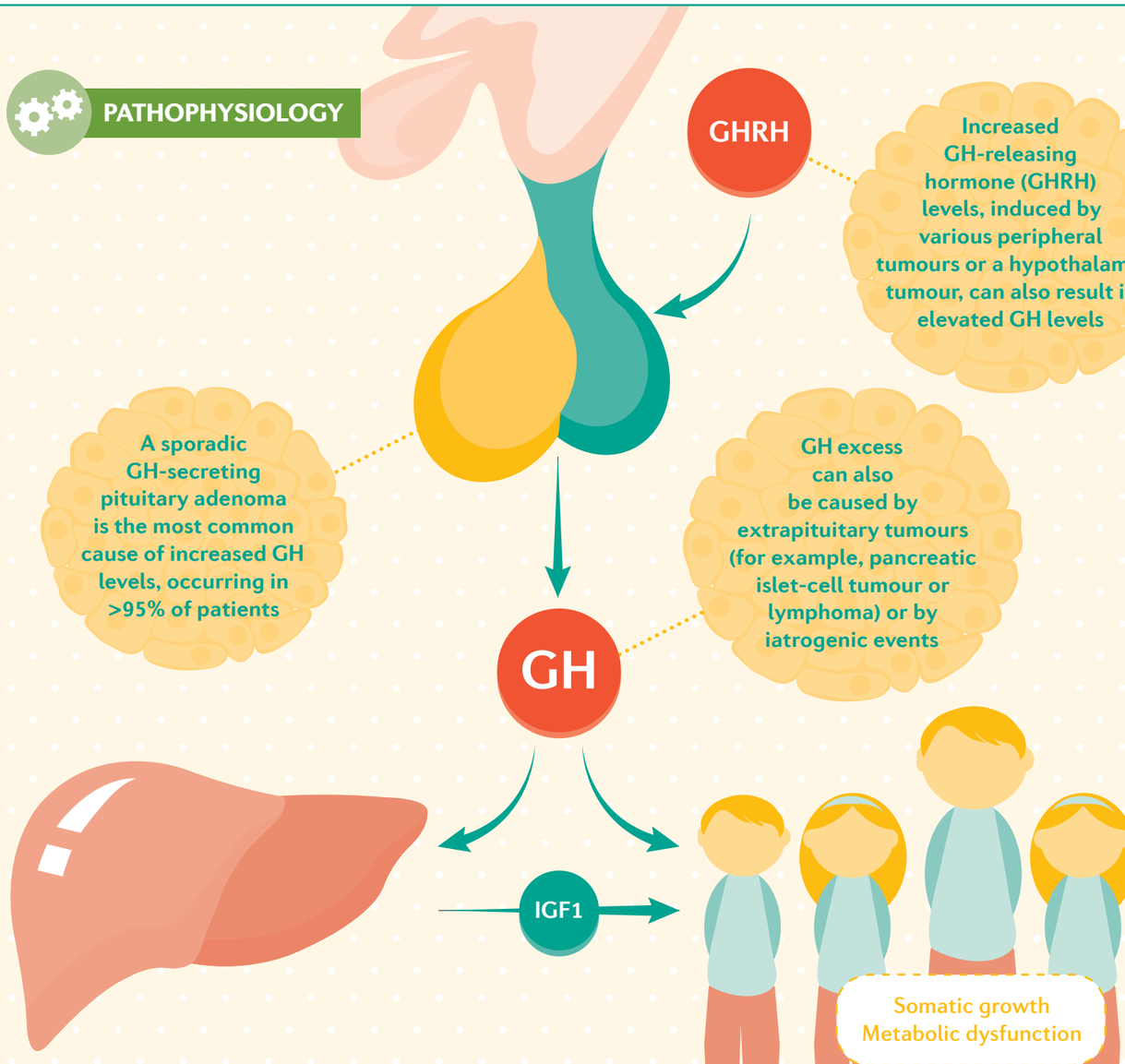
DIAGNOSIS

Progressive overgrowth of the extremities, such as the hands and feet, and changes in facial appearance are the most common outward manifestations of acromegaly and usually lead individuals to seek medical attention. However, various systemic complications and comorbidities (related to excess hormone levels or effects of the tumour mass), such as headaches, athralgia, sleep apnea, glucose intolerance, diabetes and hypertension, are also important for diagnosis. A diagnosis is confirmed biochemically by detection of increased serum levels of IGF1 and high serum GH levels that are not suppressed in an oral glucose tolerance test. MRI is used to detect and assess tumour size and invasiveness. Other pituitary functions (such as thyrotroph, corticotroph and gonadotroph function) are assessed for deficient or excess hormone secretion.



Measurement of serum IGF1 levels, rather than GH levels, is the first-line biochemical diagnostic assay, as only one blood sample (obtained at any time of the day) is required

PATHOPHYSIOLOGY



EPIDEMIOLOGY

Acromegaly is considered a rare disease, with a prevalence of <7 cases per 100,000 individuals reported in the late 20th century. However, some studies in the past 5 years have reported >13 cases per 100,000 individuals.

This increased prevalence might be due to improved diagnosis, greater screening efforts and/or increased patient survival owing to improved treatment, or it might reflect an actual increase in the prevalence of acromegaly.

As acromegaly is usually diagnosed when symptomatic (and later in life (40-50 years)) and the duration of active disease primarily determines the severity of complications, reducing delay to diagnosis is highly desirable.

MANAGEMENT

The main aims of treatment are to reduce GH and IGF1 to normal levels, to control tumour mass and to prevent systemic comorbidities and complications. Treatments include surgery, medical therapy and radiotherapy. Trans-sphenoidal adenomectomy is the first-line treatment for pituitary adenomas. However, in patients for whom surgery fails or is too high-risk, the first-line treatment is medical therapy to normalize hormone levels, including somatostatin receptor ligands (such as octreotide, lanreotide and pasireotide), dopamine agonists (such as cabergoline) and the GH receptor antagonist pegvisomant. Radiotherapy is indicated in patients for whom surgery and medical therapy are unsuccessful or unavailable.

OUTLOOK

Substantial progress in improving diagnosis and in detecting and controlling GH and IGF1 levels have refined acromegaly treatment and improved disease control,which has led to reduced mortality and morbidity and enhanced the quality of life for patients. Various new medical therapies (for example, antisense oligonucleotides that target the GH receptor) or formulations of existing therapies (for example, octreotide oral capsules) that are in development promise to further improve disease control. However, the economic costs and benefits and the effects (positive and negative) on patient quality of life must be considered if lifelong medical therapy is required.

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