

# NEURO-ENDOCRINE TUMOR CENTER

AT SAINT JOHN'S HEALTH CENTER

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## **Adenoma sub-type 1: Acromegaly**

Acromegaly is caused by a GH secreting adenoma. The problems associated with acromegaly include the effects of abnormally high GH and IGF-1 levels, and in some instances by the tumor compressing the normal pituitary gland and optic nerves. Untreated acromegaly is a serious condition that can cause dramatic bone and soft tissue changes and serious cardiovascular problems. If the tumor develops before bone growth is completed in adolescence, gigantism is the result. Because of the serious changes resulting from GH excess, treatment is essential.

### ***Symptoms***

The most obvious changes of acromegaly are the external physical changes that typically include enlargement of the hands (increase in ring size) and feet (increased shoe size) as well as frontal bossing (enlargement of the forehead) and prognathism (jaw enlargement). There may also be development of an underbite, spreading teeth, an enlarging tongue, increased snoring and sleep apnea. Carpal tunnel syndrome and excessive sweating are also common. More serious problems can include development of hypertension, diabetes mellitus and an increased risk of colon cancer. With GH-secreting macroadenomas, there may be other problems of visual loss, headaches and problems associated with pituitary gland failure including fatigue, depression, impotence and loss of libido in men and menstrual irregularities and galactorrhea (milk discharge from the breast), in women.

### ***Diagnosis***

Acromegaly is diagnosed by documenting elevated levels of both GH and IGF-1. An oral glucose tolerance test (lack of suppression of GH to oral glucose administration) is often used to confirm excess GH production. Comparing old and recent photographs will often demonstrate dramatic changes in facial appearance. Following hormonal testing that confirms acromegaly, an MRI of the pituitary should be performed to confirm the presence of a pituitary adenoma.

### ***Treatment***

**Transsphenoidal surgery:** Considered the first-line treatment for a GH-secreting tumor. However, cure of acromegaly may not be possible in patients with large or invasive macroadenomas. In such instances, medical therapy and/or radiotherapy may be necessary to control GH levels. In general, the higher the pre-operative

GH level, the lower the chance for cure. Long-term remission of acromegaly after surgery is seen in 80-90% of patients with microadenomas and in 50-60% of patients with macroadenomas.

**Medical therapy:** For patients with persistent GH elevation after surgery, octreotide or pegvisomant treatments, SRS, or both are generally indicated. Octreotide (given three times a day by injection or by one monthly injection) achieves long-acting suppression of GH in about 70% of patients. It causes some degree of tumor shrinkage in 30-50% of patients, and often improves soft tissue swelling, headache, joint pains and sleep apnea. Preoperative use of octreotide may facilitate tumor removal and lessen risks of general anesthesia. Side effects may include loose stools, malabsorption, cholelithiasis (gall stones), and local pain at the injection site. Pegvisomant, a GH receptor antagonist, is also effective in lowering IGF-1 levels although it does cause an elevation in GH levels. Bromocriptine is a "dopamine agonist" which lowers GH secretion in about 15% of acromegalic patients. The major side effect is gastrointestinal upset. Growth hormone lowering and tumor shrinkage are seen in only 10 - 15% of patients with acromegaly.

**Radical surgery (SRS) or Stereotactic Radiotherapy (SRT):** For patients with uncontrolled acromegaly after surgery, SRS (one dose) or SRT (multiple doses), provide precise radiation directly to the tumor, are relatively effective in lowering GH and IGF-1 levels and stopping tumor growth. However, the lowering of GH and IGF-1 levels takes longer with SRT (average 7 years) compared to SRS (average 18 months). Pituitary gland failure often occurs in the years after SRS or SRT. Complications such as visual loss are rare with either SRS or SRT.