A GUIDE TO

ACROMEGALY

Australian Pituitary Foundation

INCIDENCE

Between 50 and 280 people are diagnosed with acromegaly every year in Australia. This represents around 1 new person diagnosed in every 100,000 - 500,000 population each year[1].

PREVALENCE

Between 715 and 3,500 Australians currently live with acromegaly. This represents around 1 person with acromegaly in every 7,300 – 35,700 Australians[1].

ABOUT ACROMEGALY

Acromegaly is a condition caused by excess production of growth hormone (GH). The most common cause is a benign (non-cancerous) tumour of the pituitary gland. Although rare, acromegaly can also be caused by a tumour in another part of the body that releases growth hormone.

When released, growth hormone stimulates the liver to produce another hormone called insulin-like growth factor (IGF-1). Together, IGF-1 and growth hormone play an important role in controlling bone and muscle mass. The very high GH and IGF-1 levels in acromegaly cause overgrowth of all body tissues, including bone, muscle and skin, and can lead to a number of poor health conditions, like type 2 diabetes and heart disease.

Acromegaly is most often diagnosed in adults aged 40 to 50 years[1].

PRESENTING SIGNS AND SYMPTOMS

The signs and symptoms of acromegaly can develop very slowly. This is why it sometimes takes 5 to 10 years for a diagnosis.

Physical appearance

- Enlargement of the hands, feet and facial features (e.g. nose, jaw, chin, tongue)
- Skin tags
- Excessive sweating and oily skin
- Splaying of the front teeth

General symptoms

- Joint pain
- Fatigue
- Snoring
- Large tumours that press on surrounding tissue may cause headaches and visual problems.

Other conditions or complications often seen with acromegaly

- Carpal tunnel syndrome
- Osteoarthritis
- Sleep apnoea
- Depression
- High blood pressure
- Cardiovascular (heart) disease
- Type 2 diabetes
- Impaired glucose tolerance
- Vertebral (spine) fracture
- Colon polyps (tissue growths in the colon)
- Erectile dysfunction

INVESTIGATIONS

Investigations to confirm a diagnosis of acromegaly:

- **Pituitary function tests** – including measurement of growth hormone (GH) and IGF-1 in the blood. This alone is not enough to diagnose acromegaly, because growth hormone changes across the day, and IGF-1 can be elevated with other conditions.
- **Growth hormone suppression test** – using the oral glucose tolerance tests. An oral glucose tolerance test checks to see whether GH can be suppressed by glucose. This involves measuring GH levels in the blood before and after drinking a sweet drink that contains 75g of glucose. GH levels should fall if the pituitary gland is functioning normally. If GH levels do not fall, acromegaly may be the cause. This test can be performed in a pathology centre or in a hospital clinic as a day-patient.

Investigations to find the underlying cause of acromegaly:

- Magnetic resonance imaging (MRI) – to determine the size and location of the pituitary tumour.
- CT scans may be used instead of MRI if you are claustrophobic or unable to have an MRI (e.g. if you have a pacemaker).

Investigations to check potential side effects of acromegaly[2]:

- Visual field testing – if the tumour is pressing on the optic nerves connecting the eyes to the brain
- Echocardiogram (ultrasound of the heart)
- Sleep study (overnight sleep monitoring)
- Colonoscopy (examination of the colon/bowel)

TREATMENT

The goal of treatment is to reduce growth hormone and IGF-1 levels back to within the normal range. Successful treatment will resolve some of the symptoms of acromegaly and stop further bone growth, however, does not reverse the size of bones.

Treatment will depend on the size and activity of the tumour, and the age and health of the patient. Some people may require a combination of treatments.

1. **Surgery to remove the tumour** is the first line of treatment in most people. Transsphenoidal surgery (surgery through the nose) is performed in most cases. This is best done in a specialist centre by an experienced neurosurgeon. Surgery results in successful remission in 70-80% of cases.

   Sometimes the tumour causing acromegaly is large and invasive, so the surgeon may not be able to remove the entire tumour. In these cases, ongoing medication or radiation may be required to control GH levels.

2. **Medications** are available that act to reduce GH levels or block GH (Table 1). Medications can cause side effects in some people, such as diarrhoea, bloating, nausea, gallstones, fatigue and headaches.

3. **Radiation** can be used to destroy any remaining tumour cells after surgery, or when medication is not effective.

Table 1: Medications available in Australia to treat acromegaly

MEDICATION	HOW IT WORKS	METHOD OF DELIVERY
Somatostatin analogues (lanreotide, octreotide, pasireotide)	These act like the hormone somatostatin, which naturally stops GH release. This reduces GH and IGF-1 levels and can also reduce tumour size.	Monthly injections
Growth hormone receptor antagonist (pegvisomant)	These block the effects of GH in the body. This reduces IGF-1 levels but does not reduce GH or tumour size.	Daily injections
Dopamine agonists (cabergoline, bromocriptine)	These stop the release of GH from the tumour and can reduce levels of IGF-1 and tumour size.	Tablets

ONGOING MANAGEMENT

If acromegaly goes into remission following surgery, yearly check-ups are still needed to monitor GH and IGF-1 levels. This is because GH and IGF-1 levels can rise without any physical symptoms and cause other serious health problems. Keeping these hormones within normal range reduces this risk.

Most patients will have an MRI after their surgery. Repeat MRI scans are only needed if surgery could not remove the entire tumour, or if symptoms return, or if GH and IGF-1 levels rise during follow-up.

Some people may develop hypopituitarism (low pituitary hormone levels) immediately after surgery or several years after radiotherapy. This occurs in around 7% (1 in 14) surgeries3 and can be a permanent or temporary condition. Hormonal replacement therapy may be needed to restore levels of pituitary hormones. More information about hypopituitarism is available from the Australian Pituitary Foundation.

It is very important to have regular monitoring and treatment for health conditions that commonly occur with acromegaly. This includes regular check-ups for depression, headaches, carpal tunnel syndrome, osteoarthritis and pain issues, high blood pressure, heart health, erectile dysfunction, glucose intolerance/diabetes, sleep problems (sleep apnoea), colonoscopy and dental issues.

If acromegaly does not go into remission following surgery, further treatment may be necessary. These include considering further surgery (if it is safe), medication therapy or radiotherapy. Your endocrinologist will discuss these to identify the best treatment for you.

COMMON QUESTIONS

Why can't I get my wedding ring on?
Excess growth hormone causes enlargement of bones and swelling of soft tissue (muscles, tendons) in the hands. Successful treatment can reverse the swelling.

Why do my hands hurt?
Acromegaly can cause the nerves and soft tissue inside the wrist to swell. This puts pressure on the nerves of the hand, causing the hands to become painful or numb.

Why do I wake up tired?
Excess GH can enlarge the tissue around the nose, throat and tongue. This can obstruct the airway leading to poor sleep. This condition is called sleep apnoea.

Why am I getting headaches?
Headaches may be caused by a large pituitary tumour pressing on surrounding brain tissue or by a raised IGF-1 level.

Why does my jaw ache when I eat?
Growth of the bones in the jaw can disturb the teeth and put strain on the joints in the jaw, causing pain near the ear.

Why do I have backache?
Swelling of soft tissue (including muscle, cartilage and ligaments) can cause joint problems and pain in the back, which should reduce with treatment.

Why do I sweat so much?
The reason is not known, but when GH levels return to normal, the excess sweating stops.

Is acromegaly inherited?
Very rarely. Research to identify a genetic cause is ongoing.

MORE INFORMATION

The Australian Pituitary Foundation provides social support for patients and carers, and has published a range of patient resources on pituitary conditions and treatments. For more information, please visit our website: www.pituitary.asn.au

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Phone: 1300 331 807

REFERENCES

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