A GUIDE TO ACROMEGALY

Australian Pituitary Foundation

WHAT IS ACROMEGALY?

Acromegaly occurs when your pituitary gland makes too much growth hormone due to a pituitary tumour. These tumours don't spread outside of the pituitary gland in the vast majority of cases. Growth hormone is released into the bloodstream and the liver makes insulin-like growth factor-1 (IGF-1) as a result. Both hormones play a role in causing bone and soft tissue growth as well as other metabolic abnormalities, such as high blood pressure or diabetes, that are characteristic of acromegaly. Overgrowth of facial features, hands and feet is common, while, in children and adolescents, excess growth hormone causes tall stature as well (called gigantism). These changes often happen slowly, over many years, making recognition difficult.

Acromegaly is most often found in adults aged 40 to 50 years. Without treatment, it may lead to health issues like type 2 diabetes and heart disease. Treatment helps to lower your risk of chronic health problems and improve your symptoms.

HOW COMMON IS ACROMEGALY?

Around 715 to 3,500 Australians are living with acromegaly. Each year, between 50 and 280 people are diagnosed.[1]

WHAT ARE THE SIGNS AND SYMPTOMS?

The signs often appear very slowly. You may have signs for five to 10 years before a diagnosis.

PHYSICAL SIGNS:

- Large hands, feet and facial features (nose, jaw, chin, tongue)
- Skin tags
- Excess sweat and oily skin
- Splaying of the front teeth

GENERAL SIGNS:

- Joint pain
- Tiredness
- Snoring
- Headaches or eye problems

COMMON COMPLICATIONS:

- Carpal tunnel syndrome
- Osteoarthritis
- Sleep apnoea
- Depression
- High blood pressure
- Heart disease
- Type 2 diabetes
- Impaired glucose tolerance
- Spine fracture, scoliosis, kyphosis
- Colon polyps (tissue growths in the colon)
- Erectile dysfunction
- Enlarged heart and liver
- Risk of some cancers
- Multinodular goiter
- Decreased quality of life

DIAGNOSIS

Acromegaly is hard to diagnose in its early stages as changes occur over many years. If you have signs, you will need tests to confirm the cause. Common tests include:

- **Blood tests** – to check growth hormone and IGF-1 levels in the blood
- **Growth hormone suppression test** – this test measures growth hormone levels in the blood before and after drinking a sweet drink with 75g of glucose; levels of GH should fall if the gland is working as normal
- **Scans** – a magnetic resonance imaging (MRI) or computerised tomography (CT) scan can find the size and site of the tumour.

Other tests

Your doctor may order tests to check for common health issues[2], such as:

- **Eye field testing** – if the tumour is pressing on the optic nerves joining the eyes to the brain
- **Echocardiogram** – an ultrasound of your heart
- **A sleep study** – an all-night sleep test
- **Colonoscopy** – a bowel exam.

After Diagnosis

After diagnosis, it is essential to see:

- **An endocrinologist** with experience in managing pituitary diseases, and
- **A neurosurgeon** with pituitary expertise.

It is also common to seek second opinions.

TREATMENT

Treatment helps to ease your symptoms and reduce swelling, but it can't reverse the size of your bones. Treatment will depend on the size of your tumour, your age and your health. Some people need several treatments.

Options include:

- **Surgery to remove or reduce the size of the tumour** – transsphenoidal surgery (through the nose) is the first option for most people. Surgery can remove the tumour in 50-80% of cases. If the tumour is too large and can't be removed, you may need medication or radiation to control your growth hormone levels.
- **Medications to reduce or block growth hormone** – medications are an option (see table below), but they can cause side effects such as diarrhoea, bloating, nausea, gallstones, fatigue and headaches.
- **Radiation to destroy tumour cells** – radiation may help if medication is ineffective or anticipated to be required for the long term. Radiation can take many years, even up to a decade to control GH levels.

MEDICATIONS

MEDICATION	HOW IT WORKS	DELIVERY METHOD
Somatostatin analogues Lanreotide, Octreotide, Pasireotide	• Acts like the hormone somatostatin, which naturally reduces growth hormone release • Reduces growth hormone and IGF-1 levels • Reduces tumour size	Monthly injections
Growth hormone receptor antagonist Pegvisomant	• Blocks growth hormone effects • Reduces IGF-1 levels • Does not reduce growth hormone levels or tumour size	Daily injections
Dopamine agonists Cabergoline, Bromocriptine (usually only used in mild disease)	• Reduces tumour growth hormone levels • Reduces IGF-1 levels	Tablets

ONGOING TESTS

If your condition resolves after surgery, you will still need yearly health checks and blood tests. Growth hormone and IGF-1 levels can rise without any signs, and this can cause health problems. Keeping your hormones within the normal range lessens this risk.

Most people will have an MRI after surgery. You may need further scans if:

- Surgery could not remove the whole tumour
- Your symptoms return
- Your hormone levels rise during a future test.

Some people may have low levels of other hormones controlled by the pituitary (eg thyroid hormone, cortisol) at the time of tumour diagnosis or after surgery or radiotherapy. This most often requires long-term hormonal replacement therapy. You can learn more about this from the Australian Pituitary Foundation (hypopituitarism).

If surgery does not resolve your signs and symptoms, you may need more treatment. Options include:

- More surgery
- Medications
- Radiotherapy.

Your doctor will discuss these options and find the best treatment for you.

HEALTH ISSUES

See your doctor each year to check for health issues. Common issues include:

- Depression
- Headaches
- Carpal tunnel syndrome
- Osteoarthritis or pain
- High blood pressure
- Heart health
- Erectile dysfunction
- Glucose intolerance or diabetes
- Sleep problems (sleep apnoea)
- Colonoscopy
- Dental issues.

COMMON QUESTIONS

Why can't I get my wedding ring on?

Excess growth hormone causes enlarged bones and swelling of muscles and tendons in the hands. Treatment can reverse the swelling.

Why do my hands hurt?

High growth hormone levels 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 growth hormone can enlarge the tissue around the nose, throat and tongue. This can obstruct the airway leading to poor sleep (sleep apnoea).

Why am I getting headaches?

Raised IGF-1 levels, or a tumour pressing on nearby brain tissue, can cause headaches.

Why does my jaw hurt when I eat?

Growth of the bones in the jaw can disturb the teeth. This strains the joints in the jaw, causing pain near the ear.

Why does my back hurt?

Swelling of soft tissues can cause joint problems and pain in the back. Treatment can reduce these symptoms.

Why do I sweat so much?

The reason is unknown, but the sweating stops when GH levels return to normal.

Is acromegaly inherited?

In about 5% of cases there may be a genetic cause that contributed to the development of the tumour. Your doctor may discuss genetic testing if you are under the age of 30 or have other members of the family affected by pituitary or other endocrine tumours.

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

Email: support@pituitary.asn.au

Phone: 1300 331 807

REFERENCES

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