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Acromegaly

Acromegaly is a rare, progressive condition in which you make too much growth hormone. This causes a range of symptoms that develop gradually, often over several years.

The most noticeable symptoms are that your hands and feet become larger and features of your face may become more prominent.

The cause is usually a small non-cancerous growth (tumour) in the pituitary gland. Treatment options include surgery to remove the growth and medicines to block the release or effects of growth hormone.

What is acromegaly?

Acromegaly is a condition where you make too much growth hormone. This excess hormone stimulates the growth of bones and tissues in the body, leading to an abnormal increase in the size of various body parts, particularly the hands, feet, and facial features. Without proper treatment, acromegaly can lead to serious health problems and can shorten life expectancy. It is crucial to recognise and treat this disease early to prevent serious health issues.

What causes acromegaly?

In nearly everyone with acromegaly, the excess hormone comes from a small growth (tumour) in the pituitary gland (a pea-sized gland below the brain). This is a non-cancerous (benign) growth called a pituitary adenoma. The adenoma may grow up to 1-2 cm across. However, as it is benign, it does not spread to other parts of the body.

If the adenoma is tiny, measuring less than 1 cm across, it is called a microadenoma. Pituitary adenomas and microadenomas usually develop due to a random change in a cell of the gland. This change leads to uncontrolled growth of the cells, forming the tumour. The abnormal cells in the adenoma then make a lot of growth hormones.

In a few rare cases, acromegaly runs in families due to genetic reasons. See separate leaflet called Genetic Testing for more information.

Rarely, acromegaly is caused by an overproduction of a hormone called growth hormone-releasing hormone (GHRH), which comes from the hypothalamus in the brain. This stimulates the cells in the pituitary gland to produce too much growth hormone. Very rarely, tumours in other parts of the body, such as the lungs, pancreas and ovaries, can make growth hormones.

Who develops acromegaly?

Acromegaly is rare. About four to six people in a million develop acromegaly each year in the UK. It mainly develops in adults between the ages of 30 and 50. Men and women are equally affected, but men tend to be younger at the time of diagnosis.

Rarely, it affects children. If it develops in a child, it causes a condition called gigantism. This is because growth hormone stimulates the growth of bones, and in children, bones are still in their growing phase until the end of puberty (usually between 15 to 17 years).

Note: the rest of this leaflet is about acromegaly in adults.

Acromegaly symptoms

Acromegaly comes from the Greek words akros (meaning extremity) and megas (meaning large) referring to the condition's characteristic enlarged hands and feet. This is a typical feature but there are many other symptoms. The symptoms develop gradually. Therefore, diagnosis is often delayed, sometimes by as much as ten years. This is because the early physical changes can be subtle and may go unnoticed by the patients, their families and even healthcare practitioners.

It is useful to divide the symptoms into two types: those caused by too much growth hormone and those caused by the enlarging tumour (adenoma) in the pituitary gland inside your skull.

Symptoms caused by too much growth hormone

The excess of growth hormone in your bloodstream can affect various tissues in your body which can make them thicken or grow.

So, over time one or more of the following may develop:

- Your hands and feet may grow larger and broader, leading to an increase in glove and shoe sizes over time. It might become difficult to remove a wedding ring.
- Your skin may thicken particularly on the face, and become more greasy and sweaty.
- Changes to your face may include: thickened lips and nose, thickening of your scalp, your jaw becoming more prominent. These changes develop very gradually so may not be noticed by your family or friends. However, comparing old photographs to your current appearance may suggest your facial appearance has changed.
- Your dentist might notice that your teeth are spacing out or that they no longer align correctly when you bite down.
- Your vocal cords thicken which may cause your voice to deepen.
- Your tongue may become enlarged, which can lead to frequently biting it.
- Thickening of cartilage may cause joint pain and arthritis in various joints.
- Thickening of the nasal passages can make you snore loudly and may cause obstruction in airflow when you are asleep (sleep apnoea). This can make you have a poor night's sleep and make you drowsy during the day.
- You may develop carpal tunnel syndrome, which occurs when a nerve in the wrist is squeezed by thickened tissue. This can cause pain, tingling and weakness in parts of the hands or arms.

- Women may experience irregular or missed periods.

Other effects of too much growth hormone may include:

- General tiredness.
- Some muscle weakness.
- About half of people with acromegaly also develop diabetes because growth hormone stops insulin from working properly.
- High blood pressure. This develops in about one in three people.
- Increased risk of heart disease and stroke. This is probably because of the increased risk of developing high blood pressure and diabetes.
- People with acromegaly have an increased chance of developing small benign growths (polyps) in the bowel and a slightly increased chance of developing bowel cancer.
- People with acromegaly also have a slightly increased chance of developing thyroid cancer.
- If you have acromegaly you will be offered screening for bowel and thyroid cancers (see below).

Also, in about one in three cases, the adenoma also makes too much of another hormone called prolactin. This can cause sexual and menstrual problems and a milky discharge from the nipple. Many men with acromegaly also develop erectile dysfunction (impotence) and loss of sex drive.

Symptoms caused by the growing tumour

In many cases, the tumour is very small and does not cause symptoms just by its size. However, in some cases, the tumour grows enough to press on the nearby tissues inside the skull. This can lead to:

- Headaches.
- Problems with vision. The tumour may press on the optic nerves (the nerves going from the eyes to the brain) which are just next to the pituitary gland.

- Other normal cells in the pituitary gland may become squashed and damaged. As a result, the pituitary gland may stop being able to produce some other hormones. This can cause an underactive thyroid gland and/or an underactive adrenal gland (small glands above each kidney), which can cause various other symptoms.

How is acromegaly diagnosed?

- A blood test to measure the level of insulin-like growth factor 1 (IGF-1) (see below) is used if acromegaly is suspected. This may also be used as a way of monitoring the disease to see how well treatment is working.
- The diagnosis of acromegaly is made by a glucose tolerance test. In this test, you drink a sugary solution containing 75 grams of glucose. You then have a series of blood tests over two hours. Normally, glucose should lower the growth hormone level in your blood. However, if you have acromegaly, the growth hormone level stays high.
- A blood test can measure the level of growth hormone but this is not a reliable test. This is because the levels of growth hormone in the body fluctuate a lot throughout the day in everybody.
- A magnetic resonance imaging (MRI) scan can show the size of any tumour.
- Eye and visual tests can assess if the tumour is pressing on the optic nerve.
- If you are confirmed as having acromegaly, other tests will be needed to see if the tumour is causing a lack or excess of other hormones made by the pituitary gland.
- Other recommended blood tests include those for thyroid function, plasma cortisol and prolactin.
- Other tests may include chest X-ray, electrocardiogram (ECG), echocardiogram, Positron Emission Tomography (PET) scan and CT scan.

What are the treatments for acromegaly?

The goal of the treatment is to lower growth hormone and IGF-1 levels to normal, shrink an enlarged tumour, and treat any hormone deficiencies. Most symptoms and characteristics of acromegaly can be improved or reversed with successful treatment. However, any excess bone growth that has already occurred is permanent.

Most people with acromegaly have surgery to remove the pituitary adenoma (tumour). Medication and radiotherapy might be required after surgery, or in some cases, as an alternative to surgery.

Surgical treatment

The most common treatment is to remove the adenoma by surgery. This is done using very fine instruments. There are two different ways to operate on the pituitary gland:

The first method is known as endonasal trans-sphenoidal surgery. In this procedure, the surgeon accesses your pituitary gland through a small cut in the inside of one of your nostrils, often using keyhole surgery techniques.

In another method, the surgeon approaches the pituitary gland through a small incision behind your upper lip, just above your front teeth. The instruments are then passed through the base of your skull - the sphenoid bone. The aim is to remove the adenoma but to leave the rest of the pituitary gland intact.

In about nine out of ten cases with smaller tumours, the operation is successful without needing further treatment. However, the operation is less successful in those with larger tumours. Sometimes it is not possible to remove all the tumour cells. If any remain and your growth hormone level stays high after surgery, the other treatments listed below are likely to work.

Your surgeon will advise on the possible complications which can sometimes occur. For example, sometimes the operation may damage some other parts of the pituitary gland. This may cause a reduced production of some other hormones. If this occurs, you will need to take replacement hormone therapy.

Medication

Medication can be used if surgery is not possible or not wanted. It is also used when surgery fails to completely remove the tumour, and the level of growth hormone remains high.

- **Somatostatin analogues** (such as octreotide, lanreotide and pasireotide) reduce the level of growth hormone to normal and reduce the tumour size in over half of cases. Symptom improvement is achieved in eight out of ten cases. However, currently most of these medicines need to be given as an injection. These treatments need to be taken for life and can be expensive. They work in a similar way to somatostatin. Side-effects are usually mild, with some people developing abdominal pains and diarrhoea but these usually wear off with time. Gallstones can also occur but rarely cause problems. Pasireotide can also raise the likelihood of having high blood sugar and developing diabetes.
- **Dopamine agonists** (such as cabergoline, bromocriptine and quinagolide) can be taken as tablets. They work by preventing the release of growth hormone from tumour cells. However, they work in a small proportion of people. Side-effects such as feeling sick and dizzy are also quite common.
- **Pegvisomant** (Somavert®) is taken as a daily injection. However, unlike the other medicines listed above, it does not act directly on the pituitary gland. Pegvisomant works by blocking the action of growth hormone on your body's cells. Therefore, while many of the symptoms caused by excess growth hormone can be eased, this does not reduce the size of the adenoma, meaning any headaches may not be relieved.

Radiotherapy

Radiotherapy is an option to reduce the size of the tumour and hence reduce the production of growth hormone. Radiotherapy focuses high-intensity radiation on your pituitary tumour to destroy the abnormal cells. It may be used if you are not able to have surgery or if medicines have not worked.

There are two main types of radiotherapy used to treat acromegaly: stereotactic and conventional. Stereotactic radiotherapy uses a high-dose radiation beam and is more commonly used for adenomas because of its precision, reducing the likelihood of damage to nearby healthy cells. Conventional radiotherapy involves smaller doses spread over four to six weeks, as the radiation beam is wider and less precise.

It may take several years for the growth hormone levels to normalised after radiotherapy. You can take medication while waiting for the radiotherapy to work.

A possible side-effect of pituitary radiotherapy is damage to other normal pituitary cells. This can cause a reduced level of other hormones. However, if this occurs, you can take replacement hormone therapy. Radiotherapy may also have an effect on your fertility.

Acromegaly and cancer screening

As mentioned earlier, people with acromegaly have an increased chance of developing small benign growths (polyps) in the bowel and bowel cancer. Therefore, if you are diagnosed with acromegaly and are aged 40 years or more, you will normally be offered a routine colonoscopy every three to five years (You may only be offered one every ten years if your hormone levels are always normal.)

Colonoscopy

A colonoscopy is a test where an operator - a doctor or nurse - looks into your large bowel (colon) with a flexible telescope. It can diagnose bowel problems such as polyps and bowel cancer. The aim is to detect those people who develop cancer as early as possible (before symptoms develop) when the chance of a complete cure is high.

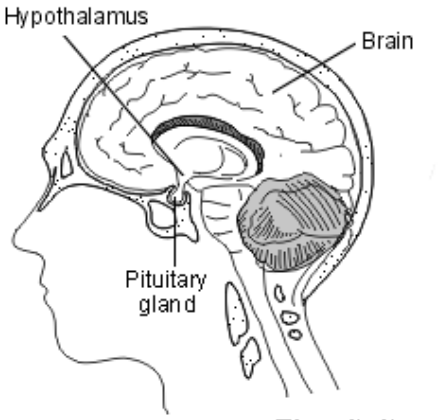
However, you should always tell your doctor if you develop any new symptoms from your bowel, such as persistent diarrhoea, passing mucus, passing blood, or tummy (abdominal) pain.

Ultrasound scan

People with acromegaly also have an increased chance of developing thyroid cancer. The thyroid is a small gland in the front of your neck. Your doctor will want to examine your thyroid gland every so often and may arrange for you to have an ultrasound scan of your neck if you have developed any lumps in your thyroid.

Acromegaly is **not** linked to any other cancers, such as breast cancer or prostate cancer.

Understanding the pituitary gland and growth hormone



The pituitary

The pituitary gland lies just below the brain. It makes several hormones, including growth hormone. (A hormone is a chemical that is made in one part of the body, passes into the bloodstream and then has effects on other parts of the body).

The amount of growth hormone that you make is partly controlled by other hormones which come from a small part of the brain called the hypothalamus. This is just above the pituitary gland and makes GHRH.

GHRH tells the pituitary gland to make growth hormone when the level of growth hormone in the blood is low. The hypothalamus also makes a hormone called somatostatin. Somatostatin stops the pituitary gland from making growth hormone when the level of growth hormone in the blood is high.

Growth hormone helps to stimulate growth and repair of various body tissues. It is needed in childhood to help children to grow. Growth hormone acts on some tissues directly. It also stimulates the liver to make another hormone called IGF-1.

Many of the effects of growth hormone are actually due to IGF-1 which acts on various cells in the body.

Further reading

- Guidelines for colorectal cancer screening and surveillance in moderate and high risk groups; British Society of Gastroenterology (May 2010)
- The Pituitary Foundation
- John A. Jane, Jr, MD, Michael P. Catalino, MD, MSc, and Edward R. Laws, Jr, MD.
- Katznelson L, Laws ER Jr, Melmed S, et al; Acromegaly: an endocrine society clinical practice guideline. J Clin Endocrinol Metab. 2014 Nov;99(11):3933-51. doi: 10.1210/jc.2014-2700. Epub 2014 Oct 30.
- Giustina A, Barkan A, Beckers A, et al; A Consensus on the Diagnosis and Treatment of Acromegaly Comorbidities: An Update. J Clin Endocrinol Metab. 2020 Apr 1;105(4). pii: 5586717. doi: 10.1210/clinem/dgz096.
- National Institute of Diabetes and Digestive and Kidney Diseases: acromegaly
- Fleseriu M, Biller BMK, Freda PU, et al; A Pituitary Society update to acromegaly management guidelines. Pituitary. 2021 Feb;24(1):1-13. doi: 10.1007/s11102-020-01091-7. Epub 2020 Oct 20.

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