Condition guide

Acromegaly

Acromegaly is a condition caused by too much growth hormone. It can cause a range of symptoms. This booklet explains how you get diagnosed with acromegaly and some of the treatment options.

The Pituitary Foundation
For hormones • For health • For life

Contents

An explanation of hormones

What is the pituitary gland?

The pituitary gland secretes hormones. These are chemicals that carry messages from one cell to another through the bloodstream. The pituitary controls several hormone glands in the body, including the thyroid, adrenals, ovaries and testes, so is often described as the master gland. If the pituitary gland is not producing sufficient amounts of hormones this is called hypopituitarism. If hormones are being over-produced, then this can cause problems depending on the hormone creating concern.

What is acromegaly?

Acromegaly is a hormonal disorder that results from too much growth hormone (GH) in the body. The pituitary, a small gland situated behind the bridge of your nose and at the base of the brain, makes GH. Acromegaly is caused by a benign (non-cancerous) tumour of the pituitary gland, which is called an adenoma; this causes the pituitary to produce too much GH. The name "Acromegaly" comes from the Greek words for "extremities" (acro) and "great" (megaly).

Growth hormone-releasing hormone (GHRH) is made by the hypothalamus, a gland in the brain situated just above the pituitary, which stimulates the pituitary gland to produce GH. Secretion of GH by the pituitary into the bloodstream stimulates the liver to produce another hormone called **Insulin-like Growth Factor 1 (IGF-1)**. IGF-1 is what actually causes tissue growth in the body. Growth hormone has effects on many different parts of the body; in adults it is important to maintain normal energy levels and to keep body tissues, such as muscle and bone, healthy. In children it is essential to reach normal growth.

Acromegaly usually develops in adults between the ages of 30 and 50, but symptoms can appear at any age. If acromegaly develops before you have stopped growing (which usually occurs between the ages of 15 to 17) it can cause gigantism. This is where people can become much taller than usual, because GH promotes growth of legs and arms. As the growth of bones stops after puberty, excessive GH in adults won't result in increased height, but may cause various other changes outlined on the next page. Acromegaly is a rare condition with only around 4-6 new cases per million of the population being diagnosed each year.

Causes of acromegaly

Acromegaly can be caused by a tumour on the pituitary gland. This is non-cancerous. The tumour can cause excess GH production which causes acromegaly.

Very rarely, a tumour elsewhere in the body may produce GH releasing hormone which stimulates the pituitary to produce excessive GH.

I began noticing changes in my body and overall health.
I was an extremely active endurance athlete; Ironman races, long distance cycling, CrossFit and anything outdoors were my fun. But I soon started experiencing extreme weight gain, an inability to recover after workouts, changes in eating habits, severe headaches, to name only a few. I discussed these and the other symptoms with my Primary Care Physician time and time again, pleading for some insight and hoping for solutions. I didn't feel heard,
it was as if the doctor didn't believe me.

Risa, who lives with acromegaly.

Symptoms of acromegaly

Symptoms of acromegaly can vary and not all mentioned in this booklet may apply to everyone. Early symptoms may involve tiredness and sleep disturbances, headaches and swelling of the hands and feet. You may notice a change in ring or shoe size, and particularly the width of your feet. Gradually, bone changes alter your facial features with the brow and lower jaw protruding.

The nasal bone enlarges and lower teeth may be spaced out. An overgrowth of bone and cartilage can lead to arthritis and when tissue thickens it may trap nerves, causing carpal tunnel syndrome which results in weakness, numbness or pain in the hands. Organs in the body, such as the heart may enlarge.

Other symptoms of acromegaly can include:

- Enlarged lips, nose and tongue
- Deepening of the voice due to enlarged vocal cords and sinuses
- Thicker, coarse, oily skin
- Joint aches
- Excessive sweating and skin odour
- Skin tags – tiny flesh-coloured finger-like projections on the skin
- Loss, or lack of libido
- Erectile dysfunction
- Abnormalities of the menstrual cycle and sometimes breast discharge
- Headaches
- Fatigue and weakness
- Impaired vision
- Sleep apnoea – breaks in breathing during sleep due to obstruction of the airway
- High blood pressure

Diagnosis of acromegaly

The diagnosis is often delayed due to several reasons:

- the symptoms develop gradually over time
- the symptoms can be caused by other conditions
- the symptoms might not be linked together
- you and your friends/families may not notice the changes

You may have had acromegaly for several years before the condition is recognised.

If your doctor suspects acromegaly, the growth hormone (GH) and IGF-1 levels in your blood would be measured. However, a single blood test of an elevated GH level may not be sufficient to diagnose acromegaly, as GH is secreted by the pituitary in spurts and results can vary widely from minute to minute.

An oral glucose tolerance test can be used to help diagnose acromegaly. This involves drinking a glucose solution which in healthy people should lower GH levels, but in those with acromegaly this suppression of GH levels does not occur.

Once the CT was done, life changed. It was decided I had a Pituitary macroadenoma and over the next few months I had what felt like every test the NHS knew how to do: Nuclear scans, MRI's, CT's, blood tests, OGTT's. The list goes on, but eventually it was decided the tumour had given me Acromegaly. The normal range of IGF-1 for my age is 14.9-32.4, mine at that time was 131.2.

Sam, who was diagnosed with acromegaly after having unexplained symptoms for many years.

Treatment of acromegaly

Treatment options include surgery, medical therapy and radiotherapy. The goals of treatment are to:

- Reduce excess GH to normal levels
- Relieve any pressure that the growing tumour may be exerting
- Preserve normal pituitary function, or to treat any hormone deficiencies
- Improve the symptoms of acromegaly

Studies show that people with untreated acromegaly are more likely to suffer from diabetes, high blood pressure and heart problems, with a reduced life expectancy compared to the normal population. However, life expectancy can be improved and normalised after the successful treatment of acromegaly.

Drug treatment

There are three types of drugs for treating acromegaly – Somatostatin Analogues, Dopamine Agonists, and Pegvisomant – and each acts by different mechanisms.

Somatostatin Analogues

- Somostatin Analogues work on specialist markers (somatostatin receptors) to inhibit GH release from the tumour.

There are two somatostatin analogues available for the treatment of acromegaly:

- Octreotide
- Lanreotide

Somatostatin analogues require injections. Octreotide can be given once a month deep into muscle (intramuscularly) for the long-acting preparation or occasionally three times a day just under the skin (subcutaneously). The short-acting preparation of octreotide is provided as ampoules or vials, which can be used for several doses. It is normally kept in the fridge but must be removed and come naturally to room temperature before use. The long-acting preparation is usually given by a nurse at your GP surgery. Octreotide can interact with other medicines: make sure your doctor knows you are taking octreotide.

Lanreotide is available as a once per month depot injection. A depot injection means it is a slow-release medication injected. You may be trained to give this injection yourself.

Both of these preparations can be left out of the fridge for up to 2 weeks, provided they have been kept away from direct sunlight and excessive heat. However, you should keep them in the fridge as much as possible.

When you first start taking **somatostatin analogues**, it may give you stomach colic or diarrhoea, but these effects usually wear off within a few days. Some people find that stomach problems improve if they wait for two hours after eating before injecting. In the longer term both may cause gallstones, but these very seldom cause problems.

In general, the effectiveness and side-effects of long-acting somatostatin preparations are similar to the three times daily octreotide but you may prefer the convenience of only having to have an injection once a fortnight or once a month.

Dopamine Agonists

There are several dopamine agonists available, all are taken by mouth but only two are widely used:

- Cabergoline
- Bromocriptine

Cabergoline, a long-acting dopamine agonist that causes fewer side-effects than **bromocriptine**, appears to be more effective in lowering GH and IGF-I levels, and usually needs to be taken only twice weekly. It is not licensed for the treatment of acromegaly but is an accepted and widely used treatment. Dopamine agonists used in very high doses to treat Parkinson's disease have been shown to cause heart valve changes in some people but studies in people receiving cabergoline for acromegaly have been reassuring in this respect.

Therefore, people using **Cabergoline** for pituitary conditions will need to have regular echocardiograms (heart scans). Your endocrinologist or GP will arrange this for you. **Bromocriptine** usually needs to be taken two or three times a day. Unfortunately, although **dopamine agonists** are taken by mouth, they are often less effective than **somatostatin analogues** which have to be injected. Cabergoline may cause you to be constipated, although this can be alleviated by increasing the fibre in your diet. At the beginning of treatment, you may also suffer nausea or dizziness when you first stand up. These effects also tend to wear off with time.

Pegvisomant

Pegvisomant is a drug which works differently. It does not try to inhibit the release of GH from the pituitary into the blood but instead stops the production of IGF-1.

This should block all the unwanted effects of GH and studies in people with acromegaly suggest it is very effective. It is given as a daily subcutaneous (under the skin) injection. It is currently used for people where the more traditional treatments have not been successful.

Radiotherapy

Radiotherapy (treatment with radiation) might be needed if your surgery has not been completely successful (not all the tumour could be removed), or if it was not possible for you to have an operation. The most common reason for incomplete tumour removal is proximity to the important blood vessel supplying the brain (internal carotid artery). In this situation, the surgeon may decide that it is safer to leave some of the tumour behind and rely on other ways of lowering GH. If you are given radiotherapy, it will be planned and carried out with extreme care. Using the images from your MRI or CT scans; the radiotherapy team will spend time planning exactly where the X-rays will be aimed.

You will attend the clinic on two or three occasions to have a special mask or fixation device made. This is a clear plastic mask or device which is used to hold your head still and ensure the radiation beams are correctly aligned. Conventional radiotherapy treatment involves low doses of radiation usually given through three parts of the head - one on top of your scalp, and one area just beside each ear. The treatment itself usually comprises 5 - 6 weeks of daily treatment (Monday to Friday). Stereotactic radiotherapy is more finely focused high dose radiotherapy and may be completed in a single session. You would be given specialist advice about the appropriate form of radiotherapy treatment for your pituitary adenoma. It may take several months, or even years, after the treatment for the effects of radiotherapy to be complete.

While you are waiting for this, you may be given drug treatment to improve control of GH levels. Levels of other hormones will also be monitored during this time to assess your pituitary gland function.

For more information about radiotherapy, see our booklet entitled **Pituitary Surgery and Radiotherapy.**

What effects will I see from my treatment?

Once treatment is successful, you should notice that the soft tissue in your hands and feet decrease in size and people often notice that their facial features gradually return towards normal. This may take some time which can be frustrating.

If you were suffering from excess sweating this should decrease, and if you had diabetes mellitus it should improve or disappear altogether. Your headaches will usually improve and so will any visual disturbances that may have been present beforehand. If you snored, it should be improved. Areas of increased bone growth (for example: hands, feet and jaw) will remain the same.

You should be looked after by a specialist (an endocrinologist). Ideally, your endocrinologist will have access to specialist nurses and radiology plus access to a neurosurgeon specialising in pituitary disease.

The success of the treatment of your acromegaly needs to be monitored. The aim of treatment is to lower your average GH level to less than 1 mcg/L and have your IGF-1 level in the normal range for your age.

Your endocrinologist will ask you how your symptoms have changed, in particular whether there has been any change in your ring size and facial appearance. It is also important for you to have regular blood tests to check your GH and IGF-1 levels, as well as the function of the rest of the pituitary gland. GH production can also be assessed using a glucose tolerance test (see above) or by taking several blood tests over a few hours or a day. In order to determine its effect, these blood tests will need repeating when there has been any change in your treatment.

Long term care

You will almost always require long-term monitoring with regular blood tests from your endocrinologist and GP.

People with acromegaly have an increased chance of developing bowel polyps (small benign growths) and bowel cancer. You will normally be offered a routine colonoscopy, and usually this will be repeated every 5 years if you are over 40. A colonoscopy is a test where a doctor looks into your colon (large bowel) using a flexible telescope; this test can diagnose bowel problems and is carried out to ensure earliest detection, even before symptoms develop, so that the chance of a complete cure is high. However, always tell your doctor if you develop any new symptoms from your bowel, such as a change in your usual bowel habit, passing blood, persistent abdominal pain or unintentional weight loss.

If you are taking hydrocortisone for adrenal insufficiency, it is important to tell the doctor or colonoscopy nurses before the colonoscopy procedure. Double your usual dose as soon as the preparatory laxatives take effect and for the duration of the bowel preparation. You will be given an 100mg hydrocortisone injection 30 minutes before the procedure. Some centres may want to admit you to hospital the night before to give the bowel preparation and provide hydrocortisone cover. Following the procedure, take double your usual hydrocortisone dose for the rest of the day and the next 24 hours, then resume your usual hydrocortisone treatment.

Coping with acromegaly

In addition to physical changes in people with acromegaly, many find their condition to be emotionally challenging – particularly if they were unwell for some time before a correct diagnosis was made. There may be stress caused either by specific physical aspects (for example: loss of libido, exhaustion, joint pain) or by factors such as changes to your face and bodily appearance, and anxiety. In addition, both the fear of anticipated surgery so close to the brain and the fact that for some people recovery can take quite a while, can be stressful. All of these issues can also be very difficult for the family and close friends of people with acromegaly.

Use our website to find out what additional support is available.

Common questions

Q: What should I do if I can't remember whether I have taken my medication?

A: Don't risk taking a double dose, except in the case of any hydrocortisone replacement. Wait until your next dose is due and carry on as normal.

Q: Can I drink alcohol?

A: Alcohol intake within standard NHS recommendations should not cause problems, but you should speak to your consultant for advice.

Q: My lifestyle means that it is inconvenient for me to take octreotide/lanreotide at the prescribed times. What can I do?

A: You should not alter the times that you take drugs, or their dose, without speaking to your consultant first. If you would like further advice about issues you may have due to work or lifestyle, please call The Pituitary Foundation Helpline.

Q: After I have injected my octreotide/lanreotide, I tend to get a stinging feeling and redness around the injection area for about 15 minutes. Can anything be done about this?

A: If you take care to ensure your injection has reached room temperature before injecting, this effect should be minimised.

Q: How do I obtain syringes and needles and how do I dispose of them?

A: This varies from area-to-area. Your endocrine specialist nurse will be able to advise further.

More information

We have a full range of booklets to support people with their pituitary conditions, as well as information across our website. You can find this at **www.pituitary.org.uk**.

If you would like more support then we have a range of services that may be suitable:

Endocrine Nurse Helpline

Our specialist endocrine nurses can provide medical guidance.

Information and Support Helpline

Our volunteer and staff run helpline allows you to speak to others with pituitary conditions, and ask practical questions about living with a pituitary condition.

Telephone Buddy

This service provides one to one support with someone with a similar pituitary journey as you. For example someone with the same condition, or a parent of someone with a condition.

Support Groups

We have a number of volunteer-led support groups across the UK, which host meetings with endocrinologists and peer support for patients.

Events

We host online and in-person events with endocrinologists on specific conditions/topics. These give people the opportunity to hear from professionals and ask questions.

About
The Pituitary Foundation

We're a dedicated team offering practical, emotional and peer support to everyone living with or impacted by a pituitary condition, to feel empowered and live with a greater sense of wellbeing.

For over 30 years, we've been amplifying voices and striving towards positive developments for the pituitary community. We work alongside healthcare professionals, clinical research teams and specialist organisations to raise the profile of pituitary conditions, finding better solutions for everyone affected by these life-changing illnesses now and in the future.

Become a member and support our work

Becoming a member is an excellent way to show your commitment to our work at The Pituitary Foundation.

As members you'll enjoy a range of benefits including free copies of Pituitary Life magazine – full of great articles from endocrinologists and inspiring stories from people living with pituitary conditions. You'll also be able to have a say on how the charity is run, and get early access to our fantastic events.

A yearly donation of £25 allows us to continue our work now and in the future.

You can become a member at: **www.pituitary.org.uk/membership**

All information in this guide is general. If you have any concern about your treatment or any side effects please read the Patient Information booklet enclosed with your medication, or consult your GP or endocrinologist.

Our publications are supported financially by several pharmaceutical companies, in line with ABPI guidelines, including Ipsen, Merck, Sandoz and Sparrow Pharmaceuticals.

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Oct 2024 | Review Oct 2027