Acromegaly

What is acromegaly?

Acromegaly is a disease of the pituitary gland. Your pituitary gland is a small gland at the base of your brain behind the bridge of your nose. It controls the functions of the other endocrine glands in the body. Sometimes it can be affected by a tumour (adenoma). These are almost invariably benign rather than cancerous and therefore do not spread anywhere else. Acromegaly is caused when your pituitary gland has a tumour which produces too much growth hormone.

Acromegaly is a very rare condition and only four new cases per million of population are diagnosed each year. It can develop at any age. If acromegaly develops before you have stopped growing (which usually occurs around 15 to 17 years of age), it causes gigantism because growth hormone promotes growth of legs and arms.

What are the symptoms of acromegaly?

The symptoms of acromegaly can be very distinctive and unpleasant, but can mostly be improved by treatment. Possible symptoms include:

- Change in appearance
- Increase in shoe size as feet enlarge in width
- Increase in size of hands - rings on fingers become very tight
- Lower jaw expands outwards causing trouble with the teeth and clicking while eating
- Nose and tongue enlarge with sinus problems
- Profound tiredness, falling asleep at any time
- Thick, greasy skin
- Excessive sweating
- Deepening voice
- Muscle weakness
- Joint pain
- Tingling sensation especially in hands, due to trapped nerves
- Headaches

About a third of patients develop diabetes mellitus (sugar diabetes).

The enlarged pituitary gland may press on the eye nerves that pass just above it, and this may cause you problems with your eyesight. In addition, the rest of the pituitary may become underactive. Because this gland controls the function of your ovaries or testes as

well as the thyroid and adrenal glands, some symptoms may relate to these. The commonest problems here are impotence, infertility, and irregular periods.
All of these symptoms tend to develop gradually and the changes may not be noticed for some time either by your or by your family.

How is acromegaly diagnosed?

You will have a test that involves drinking a sugar drink after which your blood will be measured for glucose and growth hormone over a period of about two hours (glucose tolerance test). If you have acromegaly your growth hormone levels will remain high throughout the test.
It will usually be about a month before you get the results. You will also have other tests to see if any hormones are missing. If so, these will be replaced. In addition, you will have a special magnetic scan called an MRI scan to look at the exact size of your pituitary gland. You may be given an injection before the scan to improve the results. A minority of patients are allergic to this injection, so do tell the specialist if you have asthma or any allergies. The scan does not hurt but it does involve being in the scanner for around half an hour. If you think this will make you claustrophobic or nervous, tell the doctor who will give you something to keep you calm.
You may also need to have your field of vision checked to assess whether the adenoma is having a pressure effect on the major nerve carrying information from the eye to the brain that enables you to see. This nerve passes very close to the pituitary gland.

Why do acromegalic patients need treatment?

Studies show that untreated acromegalic patients are more likely to suffer from high blood pressure, diabetes, stroke, and heart attack, with reduced life expectancy compared to the normal population. This can be improved after the successful treatment of acromegaly.
The growth of the tumour can cause pressure effects on the surrounding structures with symptoms such as the visual troubles mentioned above which can be relieved by treatment.
Effects of the excess growth hormone on the different body tissue can cause many symptoms as mentioned above, which treatment relieves.

How is acromegaly treated?

Acromegaly may be treated by operating on the pituitary gland, by radiotherapy, by drug treatment, or a combination of these. Nowadays, for most patients the first treatment is an operation, although this does depend on individual circumstances. The aim of all treatments is to reduce growth hormone levels to normal, as this is associated with the disappearance of your symptoms and improvement of your general wellbeing.

Surgery

The operation is usually carried out by making a small cut in front of the upper teeth behind the upper lip, or occasionally through the nose. This is called *transsphenoidal adenectomy* (TSA). Most patients can be treated very successfully this way, although

results are usually better if your tumour is small. The operation takes about an hour and you will normally be in hospital for about seven days. In some situations where the pituitary tumour is too big to remove by TSA an actual opening of the skull called a craniotomy may be required.

Surgery will normally lower your growth hormone levels considerably, but in some instances, the acromegaly is not cured. In such cases, consideration will be given to treating you further with radiotherapy and/or drug therapy.

Radiotherapy

Radiotherapy (treatment with radiation) might be needed if your surgery has not been completely successful or if it was not possible for you to have an operation. If you are given radiotherapy, it will be planned and carried out with extreme care. The treatment itself usually comprises four weeks of daily treatment (Monday to Friday as an outpatient). 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.

Drug treatment

The main type of drugs used for treating acromegaly are called *somatostatin analogues*. They prevent growth hormone (GH) release from the tumour. There are two somatostatin analogues available for the treatment of acromegaly:

- Octreotide (brand name Sandostatin). Octreotide has to be injected, either three times a day under the skin (subcutaneously) or once a month deep into muscle (Sandostatin LAR, intramuscularly). 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.
- Lanreotide (brand name Somatuline). Lanreotide (Somatuline LA, Ipsen) is available as an injection deep into skin (deep subcutaneous) which needs to be given every month.

Octreotide and Lanreotide help the symptoms of acromegaly by reducing growth hormone levels into the target range (of less than 5 mU/l) in more than half of patients. 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 patients find that stomach problems improve if they do not inject for about two hours after eating. In the longer term Octreotide 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. The injection into muscle needs to be given by a nurse.

How is my progress monitored?

Once treatment is successful your headaches will improve and so should any visual disturbance that you may have had. If you were suffering from excess sweating this should decrease and, if you had diabetes, it should improve or disappear altogether. You will also notice that the soft tissues of your hands and feet decrease in size and patients often notice that their facial features gradually return towards normal although this may take some time. Life expectancy after the successful treatment of early acromegaly is the same as normal population.

The success of the treatment of your acromegaly needs to be monitored. This is done by assessing the problems acromegaly causes you and by measuring the relevant hormones. Growth hormones can be measured several times during a single day, and in addition your IGF-I levels should be measured. The aim of treatment is to lower your average growth hormone level to less than 5 mU/l and have your IGF-I level in the normal range for your age. These will need repeating when there has been any change in your treatment, in order to determine its effect.

Where can I learn more?

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