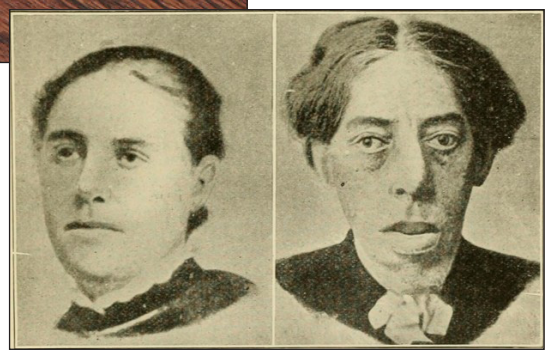
Enlarged hands and feets are common presentations of acromegaly but are not universal, so health professionals need to be alert to other combinations of symptoms.



BELOW: Images from a 1918 textbook *Physiology and biochemistry in modern medicine* show the change in facial features after 17 years of untreated acromegaly. With modern treatments, the extreme changes shown in the right-hand picture are now rarely seen.

PHOTOS: WIKIMEDIA COMMONS

Acromegaly – better awareness needed

Health professionals need greater awareness of acromegaly – a condition caused by excessive growth hormone – so sufferers can be diagnosed earlier and treated more successfully.

By Wayne Holtham

The name acromegaly literally means *"great or large extremities"*. This condition is caused by a pituitary gland tumour that secretes too much growth hormone (GH). While large extremities suggest a person has the illness, not all those with acromegaly present in this way. A sharp-eyed and informed health professional should be able to spot the collection of symptoms that indicate acromegaly, thus leading to correct diagnosis and treatment.

Nurses see patients just as often as doctors do, especially in general practice. I have written this article, and the New Zealand Acromegaly Society asks that nurses read it, in the hope of improving awareness of this condition. Greater awareness should lead to earlier diagnosis and, with it, earlier and more successful treatment.

The most common misconception about acromegaly is that it always manifests as the stereotypical extreme – the changes in physical features and height as a result of delayed diagnosis or non-treatment.

Worst-case scenario

A quick search on the internet will bring up such images – but they represent the worst-case scenario. Not all sufferers show all symptoms and modern medicine means most acromegaly patients are now diagnosed with much more subtle physical changes.

In the majority of cases, by the time of diagnosis, the tumour would have been active for five to 10 years or more, with gradual physical and hormonal changes over that time. Acromegaly usually develops in adults between the ages of 30 and 50, but symptoms can appear at any age.

The illness is traditionally considered a rare disease, with a prevalence of 40-60 per million population; however, more recent estimates brings it closer to 100-150 patients per million. This increase is probably due to an increased awareness of the condition, leading to earlier diagnosis and higher diagnosis rates. The ratio of male to female sufferers is approximately 50/50, with about 680 New Zealanders having the illness at any one time (diagnosed or undiagnosed).

Symptoms of acromegaly may include:

- **Facial changes:** Enlarged jaw and brow, increased spacing and misalignment of teeth, enlarged ears and nose.
- **Other physical changes:** Enlarged hands and feet, with rings feeling tighter

on fingers, and increased shoe size; thickened skin, skin tags and keloid scarring; excessive sweating, deeper voice; and soft-tissue swelling often described as "puffiness".

▶ **Other changes:** Snoring, sleep apnoea, diabetes, difficult to control hypertension, headaches, painful joints, enlarged organs such as cardiomegaly (enlarged heart), colonic polyps, weight gain and inability to control weight, peripheral vision loss, carpal tunnel syndrome, period changes in females, and general fatigue.

Gigantism in young people

If left undetected and untreated, it will lead to reduced quality of life, and increased morbidity and mortality. Acromegaly in childhood or adolescence may lead to rapid height gain and is generally called gigantism.

Symptoms are wide and varied, and many are unspecific. Therefore, it is often difficult to diagnose from looking at individual symptoms. When there are two symptoms together, one should suspect acromegaly; place multiple symptoms together and acromegaly is highly likely. A simple blood test specifically for IGF-1 (insulin-like growth factor 1) can be done to confirm or rule out acromegaly. This can be ordered by most GPs and specialists in the country.

One of the main goals of the Acromegaly Society, as a support organisation, is earlier diagnosis than sufferers currently experience. A lot of members report being referred to many different specialists over many years before finally being diagnosed. For example, a person with snoring, sleep apnoea and speech issues may be referred to an ear nose and throat specialist, and for carpal tunnel problems to an orthopaedic surgeon.

However, health professionals who see sleep apnoea and carpal tunnel syndrome together, for example, need to suspect acromegaly. Add in, say, headaches or significant sweating and you have very strong indicators for acromegaly. However many of these symptoms are very gradual, and present late in the development of the illness, making early diagnosis very difficult. The "she'll be right" attitude of many New Zealanders when it comes to health can also stretch out the time before a person is diagnosed.

The physical changes shown in the images on the opposite page occur slowly over decades. With advances in modern medicine, we almost never see the severe end of the disease (the woman's face on the right) anymore. By the severe stage, the disease is difficult to treat and many changes become irreversible. We need to move away from seeing these extreme but stererotypical physical changes as representing the average patient, and patients need to be offered IGF-1 screening blood test much earlier in the disease progression.

A lot of members report being referred to many different specialists over many years before finally being diagnosed.

Treatment

The usual first-line treatment is transsphenoidal surgery through the nose to remove the pituitary tumour. This provides immediate relief from many symptoms. In some cases, especially if diagnosed early and the pituitary tumour is still small, surgery results in a cure with no further treatment. However, the majority of patients will require other treatments, which include medical therapies and/or radiotherapy. Some sufferers have serious ongoing medical issues which limit their lifestyle, while others live somewhat normally. The larger the pituitary tumour, the less likely is successful treatment and full disease remission.

A simple blood test specifically for IGF-1 (insulin-like growth factor 1) can be done to confirm or rule out acromegaly. This can be ordered by most GPs and specialists in the country.

Immediately post-surgery, health professionals should be aware of the risk of onset of diabetes insipidus (DI), which is different from the commonly known diabetes mellitus. Mellitus means "sweet" in Latin, while "insipidus" means insipid, tasteless urine. DI is also known as ADH (anti-diuretic hormone) deficiency, which can result from damage to the posterior pituitary gland during pituitary surgery or radiotherapy. Without treatment, patients with DI can become dehydrated and hypernatraemic very quickly. This condition can be fatal if left unrecognised or untreated.

It is important to restate that not all patients with acromegaly have all the symptoms. In my own case, I did not have any foot growth, but presented with about 20 of the other symptoms listed in this article.

Our society's aim is to increase the awareness of acromegaly as a disease that can be identified by nurses, both in general practice and hospital environments. We refer to our dealings with acromegaly as a "journey" and we thank New Zealand nurses, wherever they may practise, for helping patients with that journey. Recently we have placed literature about acromegaly in hospitals and general practices, and are happy to supply or re-supply such literature for distribution to patients.

The Acromegaly Society welcomes new members, be they patients, family/support people, or health professionals with an interest in the disease. Please visit the society's website for more information (www.acromegaly.org.nz or www.facebook.com/acromegalysociety). •

Wayne Holtham is a national committee member of the New Zealand Acromegaly Society.