

IS IT

ACROMEGALY

A PATIENT'S GUIDE



About the Australian Pituitary Foundation

The mission of the Australian Pituitary Foundation Ltd (APF) is to provide support for those who have experienced pituitary gland conditions. We promote awareness and share information among the medical community, public, pituitary patients and their families. We welcome anyone with an interest in pituitary disorders to join us by becoming a member.

About this booklet

This booklet aims to provide information for people going through the process of determining if they have acromegaly. It describes the tests you may have while you are being diagnosed. The booklet also includes tips from APF members who have been through this process before.

Disclaimer

This book is intended as a general introduction to the topic and in no way should be seen as a substitute for your own doctor's or health professional's advice. All care has been taken to ensure the information contained in this book is free from error and/or omission; however, no responsibility can be accepted by the Australian Pituitary Foundation, author, editor, or any person involved in the preparation of this book for loss occasioned to any person acting or refraining from action as a result of material in this book. Before commencing any health treatment, always consult your doctor.

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ACROMEGALY: AN OVERVIEW

Acromegaly is a condition caused by too much growth hormone (GH) being released by the pituitary gland. In most cases, this is due to a benign (non-cancerous) tumour on the pituitary gland (called a pituitary adenoma). In other patients, the cause is a tumour somewhere else in the body that makes and releases growth hormone.

Under normal conditions, growth hormone signals to the liver to produce another hormone called insulin-like growth factor (IGF-1). Together, growth hormone and IGF-1 play an important role in controlling bone and muscle mass.

In acromegaly, the very high levels of GH and IGF-1 cause overgrowth of all body tissues, including bones, muscle and skin. This can lead to enlargement of the hands, feet, nose and chin. These physical changes occur slowly so it can take a long time, often five to ten years, or even more, between when the condition starts and when it is diagnosed.

- Symptoms and signs**
- Enlargement of the hands, feet and facial features (e.g. nose, jaw, chin, tongue)
 - Splaying of the front teeth
 - Excessive sweating and oily skin
 - Skin tags
 - Joint pain
 - Fatigue
 - Snoring
- Large tumours that press on surrounding tissue may cause headaches and visual problems.

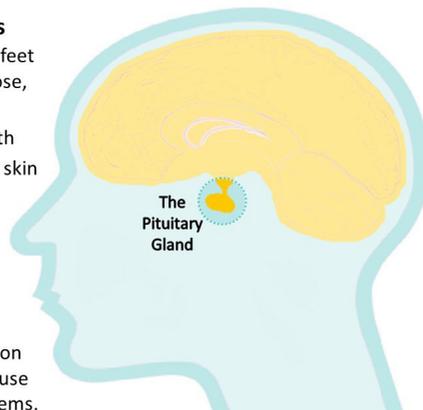


Figure One: Possible symptoms of acromegaly

DIAGNOSING ACROMEGALY

Once acromegaly is suspected, tests are needed to confirm the diagnosis and then tailor the treatment for you. The following sections describe some of these tests.

The exact details of each procedure may vary from centre to centre, but the following descriptions aim to give you an idea of what to expect. It is important, however, that you follow your doctor's or pathology centre's instructions rather than relying on information in this booklet.

For all tests, the Australian Pituitary Foundation recommends you request a copy of your results, which you can keep in a folder at home. This information can be valuable when visiting specialists whom you have not seen before. Having all your results together in one place is extremely useful to help doctors get a clear picture of your condition.

TESTING FOR ACROMEGALY

You may not need all the tests listed in this booklet. The exact tests you have will depend on the results of your previous tests.

PHYSICAL EXAMINATION

The first thing your doctor will do is check for signs and symptoms of acromegaly (Table 1). If you have several signs or symptoms, your doctor may suspect acromegaly and order some tests.

Table 1: Signs and symptoms of acromegaly

Physical appearance	General symptoms
<ul style="list-style-type: none">• Enlargement of the hands, feet and facial features (e.g. nose, jaw, chin, tongue)• Skin tags• Excessive sweating• Oily skin• Gaps forming between teeth in the lower jaw	<ul style="list-style-type: none">• Joint pain• Fatigue• Snoring• Numbness or tingling in the hands or feet• Low sex drive• Headaches• Visual problems

Some other medical conditions are often seen in patients with acromegaly. These can include:

- Carpal tunnel syndrome
- Osteoarthritis
- Sleep apnoea
- Depression
- Cardiovascular (heart) disease
- High blood pressure
- Type 2 diabetes
- Impaired glucose tolerance
- Vertebral (spine) fracture
- Colon cancer
- Erectile dysfunction

Sometimes it is easiest to track the changes in your body by looking back over old photos. It can be helpful to take these to the doctor to help them understand how your body has changed over time and why you are concerned about it.



Figure Two: Enlarged hands, feet and jaw can be seen in Daniel, APF member with acromegaly

BLOOD TESTS

The first tests you are likely to have are baseline blood tests. These will include standard tests, such as a full blood count and blood sugar levels, along with tests for liver and kidney function. Separate tubes of blood will be needed for some of these tests. You are also likely to be tested for levels of all pituitary hormones to give a picture of what's happening with your pituitary function.

For acromegaly, the most important results are your GH and IGF-1 levels.

GH and IGF-1 levels

Purpose: To check the level of GH and IGF-1 in your blood. People with acromegaly produce high levels of GH, which stimulates the liver to produce high levels of IGF-1.

Procedure: This is like an ordinary blood test, where you attend a pathology centre and a nurse takes blood from a vein in your arm. Staying hydrated by drinking water can help the procedure go more smoothly.

A sterile needle is inserted in your arm and blood is withdrawn into a vial. The procedure only takes a few minutes. It can take over a week for your results to reach your doctor, as most pathology centres do not run these tests often.

Meaning of results: In acromegaly, IGF-1 levels are much higher than usual. There is no exact value used to define what level of IGF-1 is normal or high, as different laboratories use different methods for measuring IGF-1. This means normal ranges can vary. Check with your doctor or your laboratory results to see whether you are in the laboratory's normal range.

The average level of GH is also much higher than usual in people with acromegaly. A single GH measurement, however, may not accurately reflect your daily average, as GH changes throughout the day. IGF-1 is far more stable, so a high IGF-1 level is a more reliable method for diagnosis.

These blood tests will need to be repeated over time to check your body's response to treatment.

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ORAL GLOCOSE TOLERANCE TEST

Purpose: To check whether GH can be reduced by glucose. If the pituitary gland is functioning normally, glucose should cause GH levels to fall.

Procedure: This test may be done at a pathology centre or at a hospital outpatient facility.

Before the test, you will need to fast (eat nothing) from midnight, and not eat or drink anything except water until the test is over.

A staff member will place a very thin tube (cannula) in a vein in your arm before the test. This will be used to take blood samples without the need for multiple needles. A baseline blood sample will then be collected, and you may also be asked to give a urine sample.

You will be given a sweet drink that contains glucose. Blood samples will then be collected at regular intervals over the next 2 to 3 hours. The blood samples are tested for GH and glucose levels. After the final blood sample is taken, you can eat, drink and resume your normal activities.

Most people experience no problems with this test, although it can make some people feel nauseas, or give them a stomach-ache or headache. This normally disappears shortly after the test when you have eaten something.

Meaning of results: After drinking the glucose solution, GH levels should fall to very low levels if the pituitary gland is functioning normally. If GH levels do not fall, acromegaly may be the cause.

Other conditions that can cause lack of GH reduction during the oral glucose tolerance test include diabetes mellitus, liver or kidney disease and anorexia nervosa.

“This one was not a big deal. Just a case of drinking the mixture then reading magazines while testing occurs!”

“Take a book (or something to occupy yourself with) as you will be sitting around for several hours and it will help to have something to do”.

GH DAY CURVE

Purpose: To check your GH levels over a day to determine your average level. This can give more information than a single measurement, as GH levels change across the day.

Procedure: This day procedure can be done at a pathology centre or at a hospital outpatient facility. The test will take all day, as blood samples need to be collected every 2-3 hours.

Before the tests starts, a staff member will place a very thin tube (cannula) in a vein in your arm. This will be used to collect blood samples without needing multiple needles. These samples will be sent to the laboratory to measure GH levels.

No specific preparation is needed for this test. You can eat and drink as usual. If you take regular medications, these can usually be taken as normal, unless your doctor tells you differently.

Meaning of results: In people with acromegaly, the average level of GH across the day is much higher than usual. If treatment is successful, GH levels should be within the normal range.

MAGNETIC RESONANCE IMAGING (MRI)

Purpose: To image the brain to check the pituitary gland for a tumour. Knowing the size and location of a tumour is essential for planning treatment.

Procedure: MRI is an advanced scanning method that uses powerful magnets and radio waves to produce a detailed image of a body part – in this case, the brain and pituitary gland. The images are produced without use of x-rays, and the procedure is safe and painless. It takes approximately 15 to 60 minutes to take a full picture of your brain.

During the procedure, you will lie on a platform that slides into the MRI machine. The machine makes a lot of noise. Ear plugs or music headphones are often available to help you manage this noise.

You will attend a scanning centre or a hospital for this procedure. Due to the strong magnetic fields produced by the machine, it is very dangerous to have anything metal in the room. For this reason, you must change into a robe to make sure there are no metal buttons or zips to cause problems. You will be asked if you have a pacemaker or any metal implants (such as a hip replacement), as if you have these you cannot have an MRI. You need to take off all jewellery and take out any piercings. Take this seriously as the magnetic fields involved are very strong.

The staff will put a cannula in your arm so they can inject a contrast solution later during the scan. When you get into the room, you will be asked to lie down on the MRI platform. Your whole body will slide into the machine, which makes some people feel claustrophobic. The staff will give you a button to hold so that if you feel you need to stop the test at any time, you can tell them immediately.

Your head is placed in a headrest to limit your movement. When the table retracts into the machine, you may need to squeeze your arms together a little in order to fit into the machine comfortably.

Members of staff talk to you through a speaker at times throughout the test to tell you how long each scan takes and check if you are okay. They may also position a mirror to reflect images of the staff in the booth. This enables you to see them if you look up, which can be comforting. The table may shift a little between scans to position you correctly for the next scan.

After the initial scans are complete, you will be moved out of the tunnel a little, so the contrast dye can be injected (usually into the cannula placed in your hand/arm) for the second set of scans. You may feel a cold sensation travelling up your arm as the dye is injected. The dye (usually gadolinium) helps to outline the tumour and distinguish it from normal pituitary tissue.

Some people are worried about moving during a scan. You will need to keep as still as possible, but small movements like blinking and swallowing are okay. Make sure you ask the technicians about any specific concerns you have before the scan – remember, no question is a dumb question, the technician is there to make this process as easy for you as possible.

After the procedure is over, you should sit up slowly and take your time getting up to prevent dizziness. Staff will remove the cannula from your arm and bandage the area.

Meaning of results: If you have a pituitary tumour, the MRI should show where and how big it is. This information helps with planning your treatment.

Top tips for having an MRI from APF members

Know what you are there for. Understand that this is part of finding out information that is going to help with your diagnosis.

Make sure you go to the toilet beforehand as the whole scanning procedure may last an hour or so.

Take the offer of music if it is given. It does help.

If you are claustrophobic in any way, it is best to let people know before, not during. It is preferable to do this when you make the appointment, so that staff can discuss options to assist you.

Wearing an eye mask alleviated my knowing how close the top of the machine was to my face.

Breathe deeply and relax! I used it as an enforced break to actually try and have a rest; it was almost like meditating.

I found it best to close my eyes and pretend you are trying to go to sleep in your own bed – the thumping after a while becomes rhythmic. The music is drowned out, but it is good to have it in the background. You are given a button, which is on a long lead, to push if you get into trouble, that is comforting.

I have a plan in my head before I go in the machine to plan my next holiday away, right from the initial planning, to the packing of the suitcase and the journey itself! It keeps my mind

Remember, it doesn't hurt at all

OTHER TESTS

For some patients, a few other tests may be needed.

VISUAL FIELD TEST

Purpose: To check for blind spots or loss of peripheral vision caused by a pituitary tumour pressing on your optic nerve. Vision can be affected in some people with a pituitary tumour larger than 1cm. This is because the pituitary tumour presses on the optic nerves that connect the eye to the brain.

Procedure: An optometrist or ophthalmologist will perform this test. The process is similar to a normal eye exam. This is usually very straightforward and is not difficult or stressful. This test takes around 10 minutes for each eye.

Meaning of results: Any loss of vision or blind spots may be due to the pituitary tumour putting pressure on the optic nerves that connect your eye to your brain.

You should be aware that results of visual field testing can impact on your life. For example, if you are found to have severely affected peripheral (side) vision, you may not be allowed to drive. If treatment corrects the problem, you may be able to resume driving after the necessary medical and road authority checks have been completed.

COMPUTED TOMOGRAPHY (CT) SCANS

Purpose: To locate a pituitary tumour, CT scans are used instead of MRI if you have metal implants in your body (such as a pacemaker, or hip or knee replacement).

Procedure: You will attend a hospital radiology department or an imaging centre, change into a gown and lie down on the CT scanner bed. You will be asked to lie still while the bed moves through a doughnut-shaped scanner. This takes a series of detailed x-rays from different angles. A dye may be injected into a vein or swallowed to improve the outline of the tissues.

“I had a CT before an MRI and in comparison, I found it much less stressful.... Most of my body was outside the scanner so I didn’t feel claustrophobic, and the scans were quite short, so it was easy to stay still.

A CT scan is rather straightforward and no more intrusive than an x-ray. Not at all intimidating”.

Meaning of results: The radiologist will examine the scans and report if a tumour is visible. If a tumour is found, information about its location and size will be determined for planning treatment.

COLONOSCOPY

Purpose: To look for polyps, tumours, ulcers and areas of inflammation or bleeding in your colon (bowel). A polyp is a benign (non-cancerous) growth which may have the potential to become cancerous. Patients with acromegaly have an increased risk of developing polyps in the colon.

Procedure: Before this test, you will need to take special solutions to clean out your colon (called 'colon prep'). This can take 1 to 2 days, depending on the type of prep.

During this time, you will need to stay home so that you can use the bathroom frequently. This is because the prep causes diarrhoea in order to clean out your bowel. If your bowel is not prepared properly it can make the test more difficult or may even mean the test cannot be performed and you have to come back another time.

Colonoscopies are carried out in a hospital or a day facility. When you arrive, you will change into a gown and may be given a small enema.

The doctors at the centre will explain your particular procedure to you, but the following description aims to give you an idea of what to expect.

You will be given a sedative or a light anaesthetic to keep you comfortable and help you relax. If you are sedated, you may sleep during the procedure (which usually takes around 30 to 60 minutes), although you may be aware of being moved, or having your colon inflated with air (to allow the doctor to get a better view).

During the procedure, the doctor inserts a long, thin, flexible tube with a camera on the end (colonoscope) into your anus and moves it through the rectum into the bowel.

The doctor looks for any abnormal areas such as polyps or tumours. If they find a polyp, it can be removed during the colonoscopy (a simple process called a polypectomy). If they find a tumour (lump of tissue), they can take a biopsy to determine if it is benign or cancerous.

When the procedure is over, you will rest for an hour or so until the sedative wears off. During this time, you may pass a lot of air out of your anus. This is the air that was used to inflate your colon. Someone needs to be available to drive you home after the procedure.

“I think the hardest part is the preparation and drinking what seems to be litres and litres of liquid and goo.”

“The actual procedure is quite easy – I don’t seem to be able to remember anything about it, but some people tell me that they talk to the doctor while having the procedure!”

“Afterwards, you just recover from the light anaesthetic and get the digestive system going again. I always experience gas pain in the abdomen and under the front of the shoulders”.

“Stick to the procedure of drinking of the liquids. Otherwise, if you are not completely cleaned out a repeat test might have to be done. Start eating light food for a day or so until everything is back to normal.”

A variation on this procedure is a computed tomography (CT) colonography (also known as a virtual colonoscopy). This is like a CT scan, where your colon is scanned using x-rays and a computer compiles the results into a three-dimensional image.

This technology may not be available, and even if it is, polyps cannot be removed at the same time as the colonoscopy. As technology and accuracy of detection improve, this procedure may be increasingly available.

Meaning of results: If no problems are found, you will need follow-up colonoscopies approximately every five years to check for new growths. If polyps are found and removed, you will need regular follow-up colonoscopies around every three years to check for new polyps. If a tumour is found, you will be referred for further specialist management.

ECHOCARDIOGRAM

Purpose: To check the size and appearance of your heart. The high levels of GH in acromegaly can cause swelling of the heart, which can damage the heart and lead to serious heart conditions.

Procedure: Echocardiograms are ultrasounds of the heart performed by ultrasound or heart specialists (cardiologists). This procedure uses sound wave echoes to image your heart.

There are usually no special preparations before the test. You can eat and drink normally and take your regular medications unless your doctor tells you otherwise.

At the clinic, you will be asked to remove your clothing from the waist up and lie on the echo table (bed). Sensors will be stuck to your chest to monitor your heart. The technician will then apply a gel to your chest and move a probe (transducer) around on your skin to record images of

your heart. You may be asked to roll onto your side or hold your breath at times during this procedure.

These images will be recorded and the results sent to your doctor. The whole procedure normally takes around 45 minutes.

Meaning of results: If your heart is normal, no further testing may be required. If changes to your heart are found, you will be referred to a cardiologist (heart specialist) for follow-up. The echocardiogram will need to be repeated over time to monitor your heart's response to treatment.

SLEEP STUDY

Purpose: To determine if you have sleep apnoea. This is a condition where the airway closes during sleep, cutting off breathing and causing oxygen levels in the blood to fall. Too much GH can cause the tongue to grow and the soft tissue in the throat to swell, increasing the likelihood of sleep apnoea.

Procedure: This overnight procedure is performed in a special sleep clinic. The sleep clinic staff will contact you before your study to give you instructions about what to expect and to bring with you.

Before the test, you can eat and drink normally, but may be advised to avoid caffeinated drinks on the day of the study.

When you arrive, you will be taken to your bedroom for the night. A sleep technician will then 'wire you up' for the study. This involves attaching small sensors (electrodes) to your head, face, chest and legs. These sensors measure your brain waves, muscle activity, heart rate, breathing and leg movements while you sleep.

To get good measurements, these sensors are taped or glued onto clean skin. Sometimes this involves rubbing the skin first with a light abrasive material, which can be a little uncomfortable. Once all the sensors are in place you can move about normally.

When you go to bed, the sensors will be attached with wires to a computer to record your sleep. It can feel strange to be connected to all the wires in bed. Many people do not sleep as well as they do at home – this is completely normal and generally does not affect the results.

A low light video camera will allow the staff to see you from a nearby room. If you need to use the bathroom during the night, the sleep technician will help by unplugging the wires to make a bathroom visit possible.

In the morning, the sleep technician will remove all the sensors and wires and you may be asked to fill out a questionnaire. You are then free to start your day as usual.

Meaning of results: If your results indicate you stopped breathing and your oxygen levels fell while you were sleeping, you may have sleep apnoea. Your specialist will discuss the next steps with you, such as using a special mask connected to a continuous positive airway pressure machine while you sleep to keep your airways open.

Top tips for having a sleep study from APF members

Do not drink any coffee on the day of your study.

Make sure you are tired and get as comfortable as you can before the study commences.

Take your own pillow from home.

Make sure you go to the loo before going to bed.

If possible, lie on your stomach so that your head can easily turn so pressure is not put on all the contact points.

Don't expect a peaceful night's sleep. You only need a few hours to get good results – a full night's sleep is not necessary.

WHAT HAPPENS NEXT?

Treatment for acromegaly aims to lower your GH level to within the normal range, which can improve symptoms and prevent other health complications from the disease.

Treatment will be individualised for you but will often include one or more of the following approaches.

Surgery

This is usually the first line of treatment to remove the pituitary tumour causing the excess GH production. This surgery is most often performed through the nasal passages (called transsphenoidal surgery) to minimise damage and scarring. It is important that this is carried out by an experienced neurosurgeon with a special interest in pituitary surgery.

Surgery does not always remove the entire tumour as this is not always possible.

If surgery is successful, swelling of soft tissue (e.g. muscle and cartilage) will improve within a few days and GH levels will return to normal within hours. Bone changes though, are often not reversible.

There is always some risk the tumour may grow back, so you will need to have regular check-ups with your endocrinologist.



BEFORE SURGERY



DURING SURGERY



6 MONTHS AFTER SURGERY

Figure Three: After treatment, the soft tissue swelling caused by acromegaly usually goes away, but the changes in bone size are generally not reversible.

Medication

In Australia, there are three main types of medication available that act to reduce or block GH. These medications may be used before and/or after surgery to control GH and IGF-1 levels.

Somatostatin analogues are the main type of medication used to treat acromegaly. These are injectable drugs that act like the natural hormone, somatostatin, which stops GH release. This reduces GH and IGF-1 levels and may also reduce tumour size. This medication is available as lanreotide (Somatuline LA[®], Somatuline Autogel[®]), pasireotide (SigniforTM, Signifor LARTM) and ocreotide (Sandostatin[®], Sandostatin LAR[®]).

Growth hormone receptor antagonists block the effects of GH in the body by blocking the receptors on cells that bind to GH. This reduces IGF-1 levels, but does not reduce the level of GH or tumour size. These are taken as daily injections, and available in Australia as pegvisomant (Somavert[®]).

Dopamine agonists stop the release of GH from the tumour and can reduce levels of IGF-1 and tumour size. These are taken in tablet form and are available as cabergoline and bromocriptine.

Radiotherapy

Radiotherapy may be recommended if surgery is unable to remove your entire tumour. Radiotherapy does not make the tumour disappear, but it can help stop further growth. This can take months or years to achieve. A common side effect of radiotherapy is hypopituitarism, where the pituitary gland is unable to produce enough of other pituitary hormones. This can be treated with hormone replacement therapy.

MORE INFORMATION

The following is a list of trusted organisations that are an excellent source of information or support for people with acromegaly.

Australian Pituitary Foundation (APF)

The APF has information on acromegaly on its website (www.pituitary.asn.au) as well as information about treatment options and support. The APF also organises educational sessions across Australia and can put you in touch with other people with your condition. See below for contact details to get in touch or join the APF.

Pituitary associations world-wide

Other international pituitary associations have web pages and information on acromegaly. When viewing these pages, it is worth remembering that medical practise and treatments can differ between countries.

The Pituitary Foundation (UK)

- www.pituitary.org.uk
- Follow the links to Pituitary conditions, then 'acromegaly'

Pituitary Network Association (international association)

- www.pituitary.org
- Search their website for 'acromegaly'

Web resources

The government hosted Health Direct website contains links to reliable sources of information on many diseases. Use the search function to look for acromegaly information: www.healthdirect.gov.au

Acromunity is an online resource for acromegaly information: www.acromunity.com

Support and Networking

Support for you and your loved ones will be vital during your journey with acromegaly. The APF has a peer support line where you can talk to people who have been through similar experiences as you.

The APF has an exclusive patient support register where you can ask questions and share your experiences in a secure environment. The APF also organises social gatherings and educational sessions where you can meet and network with others. The APF also holds a collection of stories from people with pituitary conditions, including acromegaly. Please consider joining the APF to access these resources.

Join the APF

You can join the Australian Pituitary Foundation by filling in the membership form, available in the membership section of the APF website: www.pituitary.asn.au.

The APF's mission is to provide support to those who have experience pituitary gland conditions.

We promote awareness and disseminate information helpful to the medical community, public, pituitary patients and their families.

The APF's objectives are to:

- Provide a forum for the exchange of information and ideas and for the discussion of problems related to pituitary disorders
- Promote public awareness of pituitary disorders and the need for government support
- Act as a resource group providing support and disseminating information
- Encourage scientific research for the prevention, alleviation, care, treatment and cure of pituitary disorders.

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Acknowledgements

The APF gratefully acknowledges the contributions of Dr Carmela Caputo and members of the APF to the development of this booklet.

Pfizer Australia provided an educational grant to support the production of this material. Pfizer Australia have not reviewed or provided editorial input into its content.

This 2nd Edition was revised and updated by A/Professor Nicolette Hodyl (Vertex Health) for the Australian Pituitary Foundation in 2019. The 1st Edition was prepared by Jennifer Gan (Accuwrite Medical®; 2011).