Australian Pituitary Foundation

GP & HEALTH PROFESSIONAL FACT FILE

UNDERSTANDING PITUITARY CONDITIONS

GP & HEALTH PROFESSIONAL FACT FILE

CONTENTS

Australian Pituitary Foundation

This fact file has been adapted for an Australian audience from the original produced by The Pituitary Foundation (used with their kind permission). Our grateful thanks go to the original authors, reviewed and editors.

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PITUITARY TUMOURS
AN OVERVIEW FOR GP'S

Australian Pituitary Foundation

WHAT ARE PITUITARY TUMOURS?

Pituitary tumours may affect any part of the pituitary, therefore the symptoms are wide-ranging and may not be recognised by the patient for some years. However, certain clusters of symptoms can give an indication of possible pituitary dysfunction.

Patients are often aged between 30–60 years and may have chronic headache and visual symptoms, particularly deteriorating peripheral vision. There may be symptoms of primary hormone hypersecretion, such as hyperprolactinaemia, acromegaly or Cushing's disease. However, symptoms may be much more general, including fatigue, amenorrhoea, loss of libido and erectile dysfunction associated with pituitary hypofunction. More rarely, a patient may present with polyuria as a result of diabetes insipidus.

INCIDENCE AND PREVALENCE

4 new cases per 100,000 population per year[1] (approximately 910 Australians every year).
78–94 cases per 100,000 population[2,3] (approximately 17,600–21,300 Australians affected at any given time)

WHAT ARE THE SIGNS AND SYMPTOMS?

- Pressure effects – headache and visual disturbance (e.g. loss of temporal vision)
- Inappropriate hormone secretion – e.g. PRL (hyperprolactinaemia), ACTH (Cushing's disease), GH (acromegaly)
- Hormone hypersecretion from a functioning tumour; or hyposecretion due to compression of the pituitary, hypothalamus or pituitary stalk
- Amenorrhoea and/or loss of libido caused by disturbance of FSH, LH, PRL or GnRH secretion

DIAGNOSIS

Pituitary tumours are often discovered:

- **Incidentally** – after a scan for another health issue
- **Visual disturbances** – if the patient presents with vision problems (tunnel vision, blurred vision, double vision) and headaches
- **Hormonal issues** – if the patients has symptoms associated with too little or too much hormone.

Tests to diagnose a pituitary tumour include:

- **Blood tests** – to measure hormone levels.
- **Scans** – a magnetic resonance imaging (MRI) or computed tomography (CT) scan can study the pituitary gland
- **Vision testing** – to check visual field

TREATMENT

Objectives of treatment:

- **Relief of pressure symptoms**
- **Restoration of appropriate hormone levels**
- **Prevention of further tumour growth**

Treatment options:

- **Surgery** – Surgery is usually transsphenoidal and typically involves a 4–5 day stay in hospital. Rarely, if the tumour is large, other approaches such as subfrontal surgery may be required. It is recommended that surgery be undertaken in a centre for pituitary diseases, or by a neurosurgeon with pituitary surgery expertise. Surgical treatment may be followed by other treatments, such as radiotherapy or medical intervention. Removal or reduction of the tumour may result in temporary or permanent damage to the normal pituitary tissue, resulting in hypopituitarism. Diabetes insipidus may also occur after surgery, but this is likely to be transient.
- **Radiotherapy**– Radiotherapy reduces the rate of recurrence of tumours and may be carried out alone or in conjunction with other treatments. In some cases, but not all, the principal long-term side effect of radiotherapy is gradual loss of pituitary function, ultimately resulting in the need for permanent hormone replacement therapy. Regular hormonal assessment is required in the long term to detect hormone deficiencies promptly.
- **Medical treatment** – For prolactinomas, drugs such as cabergoline, bromocriptine or quinagulide are the first-line treatments and often shrink the tumour as well as decrease hormone secretion. For other tumour types, medications are typically used as an adjunct to surgical treatment. Hormone replacement is needed in many cases. Analogues of somatostatin can block GH secretion in acromegaly.

QUESTIONS PATIENTS MAY ASK

Have I got cancer?

No – in this case tumour just means a benign 'lump'. Pituitary tumours only very rarely spread to other parts of the body. If left untreated, they may gradually increase in size.

Will I need treatment?

Not necessarily. Up to one in five people have a small pituitary tumour. If a small tumour is discovered accidentally, it may not be necessary to have any treatment at all. Treatment is only required if the tumour is causing you problems. Small tumours will be monitored to make sure they do not get larger.

Regular follow- up appointments with your endocrinologist are essential.

Will it go away when I'm treated?

It is often not possible to remove the tumour entirely. In most cases, however, its growth and activity can be controlled.

Will I have to take tablets in the long term?

Drugs given to suppress high hormone levels produced by the tumour may not be needed long term if other treatment has been given. If the tumour, or the treatment for it, affects the function of the normal pituitary gland, then long-term replacement therapy will be necessary – most patients find this readily acceptable.

Will I still be able to have a family?

Yes, if the reason for infertility is pituitary disease. In many cases, the effects of pituitary tumours on fertility can be treated.

ABBREVIATIONS

- **ACTH:** adrenocorticotrophic hormone
- **FSH:** follicle-stimulating hormone
- **GH:** growth hormone
- **GnRH:** gonadotropin-releasing hormone
- **LH:** luteinising hormone
- **MRI:** magnetic resonance imaging
- **OT:** oxytocin
- **PRL:** prolactin
- **TSH:** thyroid-stimulating hormone

MORE INFORMATION

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SYMPTOMS OF PITUITARY DISORDERS

Australian Pituitary Foundation

COMMON SYMPTOMS RELATED TO HIGH OR LOW LEVELS OF HYPOTHALAMIC–PITUITARY HORMONES

Tumour in the pituitary or hypothalamus	Weakness, peripheral vision loss, double vision, headache, low sex drive, tiredness
Growth hormone: Too much (acromegaly)	Increased height in children, tightness of rings on fingers, increasing shoe size, arthritis, facial changes (such as larger lips, nose, jaw and brow), separation of teeth, sweating and oily skin, diabetes, hypertension and sleep apnoea
Growth hormone: Too little	Failure to grow in childhood; decreased muscle mass, increased fat mass, severe fatigue and lack of motivation in adulthood
Prolactin: Too much (prolactinoma)	Irregular or absent periods, breast tenderness, milky discharge from the breasts in females; decreased sex drive and infertility in both genders
Prolactin: Too little	No symptoms in men; no breast milk production in women
Adrenocorticotrophin (ACTH): Too much (resulting in too much cortisol. Cushing's syndrome)	Weight gain around the stomach/hips, thin arms/legs, round flushed face, skin bruises easily, large purple stretch marks, bones break easily, high blood pressure and diabetes, mood swings, irregular periods

SYMPTOMS OF PITUITARY DISORDERS

Australian Pituitary Foundation

Adrenocorticotrophin (ACTH): Too little (resulting in low cortisol)	Weight loss, fatigue, nausea, arthralgias, low blood sugar levels (hypoglycaemia), low blood pressure, low sodium (hyponatraemia)
Thyroid-stimulating hormone (TSH): Too much	Thyroid enlargement, fatigue, tremors, sweating, unusually strong or odd heartbeat, heat intolerance, weight loss, nervousness, irregular or absent periods
Thyroid-stimulating hormone (TSH): Too little	Fatigue, weight gain, cold intolerance, coarse skin, irregular and heavy periods
Luteinizing hormone (LH) and follicular stimulating hormone (FSH): Too much (very rare)	Usually no symptoms; but possible symptoms may include visual field disturbance due to pituitary tumour size, enlarged testicles, high or low testosterone in men, enlarged ovaries in women, raised oestrogen level
Luteinizing hormone (LH) and follicular stimulating hormone (FSH): Too little	Delayed puberty, irregular or absent periods, decreased sex drive and fertility
Anti-diuretic hormone (ADH): Too much (Syndrome of inappropriate antidiuretic hormone secretion-SIADH)	Hyponotraemia resulting in fatigue, confusion and drowsiness
Anti-diuretic hormone (ADH): Too little (AVP deficiency, previously known as Diabetes Insipidus)	Frequent urination (including during the night), increased thirst, weakness – may lead to severe dehydration and raised sodium (hypernatraemia)

A GUIDE TO REFERRAL PATHWAYS

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Referral pathways for pituitary conditions depend on the diagnosis and clinical presentation of the pituitary condition. Most pituitary conditions are non-urgent and can follow a systematic, step-by-step process to decide the most appropriate referral pathway. There are certain situations whereby the condition is urgent and this necessitates immediate referral to either the emergency department, or to contact a pituitary neurosurgeon or endocrinologist directly by phone.

URGENT REFERRALS

VISUAL LOSS

Symptoms: Classic "Tunnel-vision" potentially progressing to blindness

Immediate Action:
Ophthalmologist for Visual Field assessment + MRI Brain

Referral:
Urgent referral to a neurosurgeon

PITUITARY APOPLEXY

Symptoms: Sudden onset headache, potential sudden blindness, and collapse

Immediate Action:
If presented to GP, immediate referral to emergency department

Referral:
Immediate referral to an emergency department via ambulance for pituitary blood tests + CT Brain

NON-URGENT REFERRALS

PITUITARY HORMONE DYSFUNCTION

Symptoms: Fatigue, general malaise, loss of libido, weight fluctuation

Action:
Pituitary blood tests: Prolactin, 8am cortisol, growth hormone, IGF-1, TSH, free T4, FSH/LH, estradiol (women) or testosterone (men)

Referral:
Referral to a Pituitary Endocrinologist

INCIDENTAL FINDINGS

Situation: Pituitary lesion (cyst/tumour) found incidentally on MRI or CT brain

Action:
Pituitary blood tests: Prolactin, 8am cortisol, growth hormone, IGF-1, TSH, free T4, FSH/LH, estradiol (women) or testosterone (men)

Referral:
Pituitary Neurosurgeon or Endocrinologist, who will further refer as part of multi-disciplinary management if necessary

A GUIDE TO ACROMEGALY

Australian Pituitary Foundation

WHAT IS ACROMEGALY?

Acromegaly occurs when your pituitary gland makes too much growth hormone due to a pituitary tumour. These tumours don't spread outside of the pituitary gland in the vast majority of cases. Growth hormone is released into the bloodstream and the liver makes insulin-like growth factor-1 (IGF-1) as a result. Both hormones play a role in causing bone and soft tissue growth as well as other metabolic abnormalities, such as high blood pressure or diabetes, that are characteristic of acromegaly. Overgrowth of facial features, hands and feet is common, while, in children and adolescents, excess growth hormone causes tall stature as well (called gigantism). These changes often happen slowly, over many years, making recognition difficult.

Acromegaly is most often found in adults aged 40 to 50 years. Without treatment, it may lead to health issues like type 2 diabetes and heart disease. Treatment helps to lower your risk of chronic health problems and improve your symptoms.

HOW COMMON IS ACROMEGALY?

Around 715 to 3,500 Australians are living with acromegaly. Each year, between 50 and 280 people are diagnosed.[1]

WHAT ARE THE SIGNS AND SYMPTOMS?

The signs often appear very slowly. You may have signs for five to 10 years before a diagnosis.

PHYSICAL SIGNS:

- Large hands, feet and facial features (nose, jaw, chin, tongue)
- Skin tags
- Excess sweat and oily skin
- Splaying of the front teeth

GENERAL SIGNS:

- Joint pain
- Tiredness
- Snoring
- Headaches or eye problems

COMMON COMPLICATIONS:

- Carpal tunnel syndrome
- Osteoarthritis
- Sleep apnoea
- Depression
- High blood pressure
- Heart disease
- Type 2 diabetes
- Impaired glucose tolerance
- Spine fracture, scoliosis, kyphosis
- Colon polyps (tissue growths in the colon)
- Erectile dysfunction
- Enlarged heart and liver
- Risk of some cancers
- Multinodular goiter
- Decreased quality of life

DIAGNOSIS

Acromegaly is hard to diagnose in its early stages as changes occur over many years. If you have signs, you will need tests to confirm the cause. Common tests include:

- **Blood tests** – to check growth hormone and IGF-1 levels in the blood
- **Growth hormone suppression test** – this test measures growth hormone levels in the blood before and after drinking a sweet drink with 75g of glucose; levels of GH should fall if the gland is working as normal
- **Scans** – a magnetic resonance imaging (MRI) or computerised tomography (CT) scan can find the size and site of the tumour.

Other tests

Your doctor may order tests to check for common health issues[2], such as:

- **Eye field testing** – if the tumour is pressing on the optic nerves joining the eyes to the brain
- **Echocardiogram** – an ultrasound of your heart
- **A sleep study** – an all-night sleep test
- **Colonoscopy** – a bowel exam.

After Diagnosis

After diagnosis, it is essential to see:

- **An endocrinologist** with experience in managing pituitary diseases, and
- **A neurosurgeon** with pituitary expertise.

It is also common to seek second opinions.

TREATMENT

Treatment helps to ease your symptoms and reduce swelling, but it can't reverse the size of your bones. Treatment will depend on the size of your tumour, your age and your health. Some people need several treatments.

Options include:

- **Surgery to remove or reduce the size of the tumour** – transsphenoidal surgery (through the nose) is the first option for most people. Surgery can remove the tumour in 50-80% of cases. If the tumour is too large and can't be removed, you may need medication or radiation to control your growth hormone levels.
- **Medications to reduce or block growth hormone** – medications are an option (see table below), but they can cause side effects such as diarrhoea, bloating, nausea, gallstones, fatigue and headaches.
- **Radiation to destroy tumour cells** – radiation may help if medication is ineffective or anticipated to be required for the long term. Radiation can take many years, even up to a decade to control GH levels.

MEDICATIONS

MEDICATION	HOW IT WORKS	DELIVERY METHOD
Somatostatin analogues Lanreotide, Octreotide, Pasireotide	• Acts like the hormone somatostatin, which naturally reduces growth hormone release • Reduces growth hormone and IGF-1 levels • Reduces tumour size	Monthly injections
Growth hormone receptor antagonist Pegvisomant	• Blocks growth hormone effects • Reduces IGF-1 levels • Does not reduce growth hormone levels or tumour size	Daily injections
Dopamine agonists Cabergoline, Bromocriptine (usually only used in mild disease)	• Reduces tumour growth hormone levels • Reduces IGF-1 levels	Tablets

ONGOING TESTS

If your condition resolves after surgery, you will still need yearly health checks and blood tests. Growth hormone and IGF-1 levels can rise without any signs, and this can cause health problems. Keeping your hormones within the normal range lessens this risk.

Most people will have an MRI after surgery. You may need further scans if:

- Surgery could not remove the whole tumour
- Your symptoms return
- Your hormone levels rise during a future test.

Some people may have low levels of other hormones controlled by the pituitary (eg thyroid hormone, cortisol) at the time of tumour diagnosis or after surgery or radiotherapy. This most often requires long-term hormonal replacement therapy. You can learn more about this from the Australian Pituitary Foundation (hypopituitarism).

If surgery does not resolve your signs and symptoms, you may need more treatment. Options include:

- More surgery
- Medications
- Radiotherapy.

Your doctor will discuss these options and find the best treatment for you.

HEALTH ISSUES

See your doctor each year to check for health issues. Common issues include:

- Depression
- Headaches
- Carpal tunnel syndrome
- Osteoarthritis or pain
- High blood pressure
- Heart health
- Erectile dysfunction
- Glucose intolerance or diabetes
- Sleep problems (sleep apnoea)
- Colonoscopy
- Dental issues.

Why can't I get my wedding ring on?

Excess growth hormone causes enlarged bones and swelling of muscles and tendons in the hands. Treatment can reverse the swelling.

Why do my hands hurt?

High growth hormone levels can cause the nerves and soft tissue inside the wrist to swell. This puts pressure on the nerves of the hand, causing the hands to become painful or numb.

Why do I wake up tired?

Excess growth hormone can enlarge the tissue around the nose, throat and tongue. This can obstruct the airway leading to poor sleep (sleep apnoea).

Why am I getting headaches?

Raised IGF-1 levels, or a tumour pressing on nearby brain tissue, can cause headaches.

Why does my jaw hurt when I eat?

Growth of the bones in the jaw can disturb the teeth. This strains the joints in the jaw, causing pain near the ear.

Why does my back hurt?

Swelling of soft tissues can cause joint problems and pain in the back. Treatment can reduce these symptoms.

Why do I sweat so much?

The reason is unknown, but the sweating stops when GH levels return to normal.

Is acromegaly inherited?

In about 5% of cases there may be a genetic cause that contributed to the development of the tumour. Your doctor may discuss genetic testing if you are under the age of 30 or have other members of the family affected by pituitary or other endocrine tumours.

MORE INFORMATION

The Australian Pituitary Foundation provides social support for patients and carers, and has published a range of patient resources on pituitary conditions and treatments.

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A GUIDE TO ARGININE VASOPRESSIN DEFICIENCY

Australian Pituitary Foundation

WHAT IS ARGININE VASOPRESSIN DEFICIENCY (AVP-D)?

Arginine vasopressin deficiency (AVP-D) is a condition that causes you to produce large amounts of urine and feel very thirsty all the time. AVP-D was previously referred to as Diabetes Insipidus.

AVP-D occurs because the body can't make enough antidiuretic hormone (ADH, also called arginine vasopressin). ADH and AVP are terms that can both be used and refer to the same hormone. ADH is the pituitary hormone that regulates the water level in your body by controlling the amount of urine your kidneys produce.

If you have AVP-D, ADH doesn't regulate the water level in your body, and you produce too much urine. Symptoms include producing more than three litres of diluted urine per day and constant thirst, even after drinking lots of water.

Healthy adults produce one to two litres of urine per day. A person with severe AVP-D who drinks a lot of fluids can produce up to 19 litres of urine daily.

COMMON CAUSES:

- Tumours affecting the hypothalamus or pituitary stalk
- Complication following surgery on the pituitary
- Brain injuries that damage hypothalamus or pituitary
- Infections or inflammatory conditions that involve the pituitary or hypothalamus
- Autoimmune conditions that damage ADH cells

LESS COMMON CAUSES:

- Cancer that spreads to the brain
- Wolfram syndrome, a rare genetic disorder that also causes vision loss
- Experiencing a loss of oxygen that causes brain damage (eg stroke or near-drowning)
- Can be associated with congenital brain malformations.

Sometimes, there is no apparent cause.

AVP-D can also occur if the body produces the right amount of ADH, but the kidneys do not respond. This type is called nephrogenic diabetes insipidus.

HOW COMMON IS AVP-D?

AVP-D affects only about 1 in 25,000 people.

There is a rare form of AVP-D called adipsic AVP-D. This condition typically occurs after damage to the hypothalamus, resulting in the loss of thirst sensation in affected patients. Managing this condition poses significant challenges as it often leads to rapid and substantial changes in blood sodium levels. Patients with adipsic AVP-D are commonly prescribed a daily fluid intake and are required to regularly measure their body weight. Additionally, frequent blood tests are necessary to closely monitor their condition.

WHAT ARE THE SIGNS AND SYMPTOMS?

- Urinating more than three litres in 24 hours
- Diluted urine
- Constant thirst
- Excessive drinking (polydipsia)
- Preferring cold drinks

Children and babies may also have these signs:

- Heavy, wet nappies
- Bed-wetting
- Trouble sleeping
- Delayed growth
- Vomiting or constipation
- Weight loss

DIAGNOSIS

AVP-D can be diagnosed with the following tests:

- **Blood tests** – these can measure sodium levels and a hormone called copeptin
- **Water-deprivation test** – in this test, you are deprived of fluid for up to 8 hours; the test records your urine volume and weight changes
- **Arginine stimulated copeptin test** - in this test you are given arginine which increases the secretion of vasopressin and copeptin from the posterior pituitary gland (where the antidiuretic hormone is secreted from)
- **Hypertonic sodium infusion test** – in this test you are given extra sodium in order to measure the hormone response to this using copeptin measurement
- **Scans** – a magnetic resonance imaging (MRI) or computerised tomography (CT) scan can confirm or exclude the presence of pituitary disease.

After Diagnosis

After diagnosis, it is essential to see:

- **An endocrinologist** with experience in managing pituitary diseases

TREATMENT

There is no cure for AVP-D, but treatments can relieve symptoms and improve your quality of life.

Treating AVP-D involves taking a medication called desmopressin to replace AVP. Desmopressin can help to relieve excess thirst, decrease urine output and prevent dehydration.

Generally, you take the medication two to three times daily as a nasal spray or as tablets. Taking too much of this medication too often can cause hyponatraemia (low sodium). If you miss a dose, it is best not to take another dose, as that can increase the risk of hyponatraemia. Some doctors recommend leaving out one dose of desmopressin each week as a precaution to minimise the risk of hypotranaemia. Your doctor can give you more information about medications and doses. Symptoms of low sodium include lethargy, headaches and confusion.

ONGOING MANAGEMENT

Once you have your treatment plan, you can manage your condition successfully at home. It is essential to see your doctor every six to 12 months. Your doctor will take a general health exam and look for signs of the following:

- Fluid retention
- Hyponatraemia
- Frequent and excess urination.

During times of illness (such as fevers, vomiting and diarrhoea), people with AVP-D can easily develop problems with taking their medication and problems with their sodium level. Thus it is vital to discuss what to do during illnesses with your medications with your specialist and have a "sick day management plan". During acute illnesses, some people have to get to hospital sooner than someone without AVP-D as the sodium level can become abnormal due to the hormonal deficiency of AVP.

MORE INFORMATION

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A GUIDE TO CRANIOPHARYNGIOMAS

Australian Pituitary Foundation

WHAT ARE CRANIOPHARYNGIOMAS?

A craniopharyngioma is a rare, benign (non-cancerous) brain tumour. They can be solid or cystic (fluid-filled sacs). These tumours begin near the pituitary gland, the small pea-sized gland that sits near the base of the brain. The pituitary gland releases hormones that control many body functions.

Craniopharyngiomas can occur at any age but are most common in children aged five to 15 and older adults over 50.

These tumours can affect the function of the pituitary gland and other nearby structures in the brain. Symptoms include gradual changes in vision, tiredness, excessive urination and headaches. Children with craniopharyngiomas may grow slowly and be smaller than expected.

HOW COMMON ARE CRANIOPHARYNGIOMAS?

Around 450 Australians are living with craniopharyngiomas. Each year, about 30 people are diagnosed.[1]

WHAT ARE THE SIGNS AND SYMPTOMS?

COMMON SIGNS

- Headaches (sometimes accompanied by vomiting)
- Diabetes insipidus (excessive thirst and urination)
- Vision disturbance
- Disturbed sleep patterns
- Slow growth
- Behavioural problems
- Early or delayed puberty
- Increased sensitivity to cold or heat
- Tiredness
- Frequent infections
- Hypopituitarism (in children, this causes impaired growth, delayed puberty and other symptoms related to the other hormone deficiencies that may be present).

If the tumour affects the hypothalamus, signs can include:

- Increased or decreased appetite (resulting in weight problems)
- Increased thirst
- Mood swings
- Sleep disturbance
- Reduced concentration
- Short-term memory loss

EARLY-CHILDHOOD SYMPTOMS

In early childhood, tumours can be fast-growing and more aggressive

Symptoms are often present for a number of years before a diagnosis is made.

Because these tumours may be associated with raised intracranial pressure, symptoms can also include headaches, nausea and vision problems.

DIAGNOSIS

Standard tests to diagnose a craniopharyngioma include:

- **Clinical assessment** – a medical history and neurological exam
- **Blood tests** – these can identify changes in hormone levels that suggest a tumour is affecting the pituitary gland
- **Scans** – a magnetic resonance imaging (MRI) or computerised tomography (CT) scan can find the tumour's size and position

After Diagnosis

After diagnosis, it is essential to see:

- **An endocrinologist** with experience in managing pituitary diseases, and
- **A neurosurgeon** with pituitary expertise.

TREATMENT

Surgery - Surgery is the preferred treatment. The type of surgery depends on the size, location and position of the tumour. Surgery options include:

- Minimally invasive (transsphenoidal) – the surgeon reaches the tumour through the nose
- Open surgery (craniotomy) – this involves opening the skull, most commonly in the centre of your scalp, to gain access to the tumour.

Surgery may not be able to remove the tumour altogether because the tumour sticks to surrounding structures. If some tumour cells remain, you may need radiotherapy or chemotherapy.

Sometimes, you may need a cerebrospinal fluid shunt to drain the fluid before surgery or if the tumour regrows. Your neurosurgeon can talk to you more about this.

Radiotherapy - Radiotherapy is often needed after surgery – especially if there is residual tumour after surgery. Radiotherapy uses X-rays and protons to kill tumour cells.

Chemotherapy - Chemotherapy is a drug treatment that destroys tumour cells.

Medication - Surgery may cause new hormonal imbalances or hypopituitarism. After surgery, you may need medication replacing cortisol, growth hormone, thyroid hormone and sex hormones. It is important to have regular checks with your endocrinologist on hormone levels following surgery.

ONGOING MANAGEMENT

After the surgery, you will need long-term follow-up to:

- **Monitor for tumour regrowth** – especially important in the first three years after surgery
- **Treat side effects** – related to pituitary function, such as pituitary hormone deficiencies and Arginine Vasopressin Deficiency (AVP-D)
- **Monitor emotional health** – surgery can affect the person's emotions and cause depression
- **Monitor steroid therapy** – and how to adjust the dose when stressed or ill.

It is common for the tumour to reoccur; however, it is less likely if the person has surgery and radiotherapy.

When to go to the hospital

Seek urgent medical care if:

- **Your vision gets worse or becomes impaired** – this can indicate that cysts are enlarging rapidly
- **You notice clear fluid dripping down the back of the throat or through the nose soon after surgery** – this may indicate a cerebrospinal fluid leak.

Is it cancer?

No. A craniopharyngioma is a rare benign (not cancerous) brain tumour. These tumours rarely spread to other parts of the body.

What causes craniopharyngiomas?

These tumours likely grow from leftover pieces of tissue that remain in early pregnancy when the baby's head, face and brain are forming.

Who gets craniopharyngiomas?

They can affect people of any age. Cases occur most commonly in children aged 5 to 14 but also adults between 50-70.

Are there any new treatment options?

You or your child may be a candidate for a clinical trial. Talk to your doctor about the options.

Are they inherited?

We still don't know. Research to find a genetic cause is ongoing.

MORE INFORMATION

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A GUIDE TO CUSHING'S DISEASE

Australian Pituitary Foundation

WHAT IS CUSHING'S DISEASE?

Cushing's disease occurs when your body is exposed to high levels of the steroid hormone cortisol. Causes include:

- A pituitary tumour that causes your body to make excess adrenocorticotropic hormone (ACTH), which in turn leads to your adrenal glands producing too much cortisol (called Cushing's disease).
- A tumour from somewhere else in the body e.g. in the lung, that also can produce excess ACTH (called ectopic Cushing's syndrome).
- An adrenal tumour that produces too much cortisol.
- Using steroid medications for a long time.

Cortisol is the main hormone that helps your body deal with physical stress (such as injury or infection), controls blood sugar levels and blood pressure and reduces inflammation.

HOW COMMON IS CUSHING'S DISEASE?

Around 300 to 1,600 Australians are living with Cushing's disease. Each year, about 41 people receive a diagnosis..

WHAT ARE THE SIGNS AND SYMPTOMS?

Cushing's disease can be hard to diagnose as many signs are common in other health issues. Symptoms tend to come on slowly over time.

Physical Signs

- Thin, fragile skin that bruises easily
- Red and purple streaks across the skin
- Muscle loss and weakness
- Weight gain, often around the waist
- Increased back fat between the shoulders (a "buffalo" hump)
- Rounded face
- Irregular or no periods
- Hirsutism in women (excess growth of facial and body hair)
- Reduced growth in children
- Infertility
- Poor wound healing

General Signs

- Fatigue/lethargy
- Poor quality of life
- Poor school/work performance

Emotional changes

- Depression
- Anxiety
- Unable to think clearly
- Mood and behaviour changes
- Increased irritability
- Decreased libido

Other conditions[4]

- Osteoporosis
- Fractures
- High blood pressure
- High blood glucose/diabetes
- Infections e.g skin fungal

DIAGNOSIS

If you have signs of Cushing's disease, you will need tests to confirm a diagnosis. No single test can diagnose Cushing's disease. Common tests include:

- **Blood tests** – to measure hormone levels
- **24-hour urinary free cortisol tests** – to measure daily cortisol production
- **Late night or midnight salivary cortisol tests** – to check for loss of daily cortisol rhythm
- **Overnight dexamethasone suppression tests** – to see if your body can suppress cortisol production normally
- **Scans** – a magnetic resonance imaging (MRI) or computerised tomography (CT) scan can see the pituitary and/or adrenal glands.

Other tests you may need include:

- **Inferior petrosal sinus sampling (IPSS)** – to check whether ACTH is being released from the pituitary gland or elsewhere
- **Visual field test** – to check for vision loss
- **Bone mineral density (BMD) test** – to check your bone health.

If the test results suggest Cushing's disease, your doctor may order more tests to find the reason for high cortisol levels.

Around one in six people with Cushing's disease have cyclical Cushing's syndrome, meaning they have periods of normal or even low cortisol levels and high cortisol production.

If you have cyclical Cushing's syndrome, your results can appear normal, depending on when you have the tests. You may need further tests to catch a period of high cortisol production.

TREATMENT

Treatment aims to restore your cortisol to normal levels, and improve your symptoms and your quality of life. Your medical team will need to tailor your treatment depending on factors such as:

- Age
- Sex
- Tumour size
- Hormone levels
- Fertility concerns
- Medications
- Other medical conditions
- Side effects.

Treatment options include:

- **Surgery** – to remove or reduce the size of the tumour
- **Radiotherapy** – to reduce cortisol levels if surgery does not help
- **Medication** – to control cortisol levels
- **Removal of adrenal glands (bilateral adrenalectomy)** – used when cortisol levels are unable to be controlled adequately by surgery, radiotherapy, medication or all 3; and sometimes if pregnancy is desired when surgery has not been successful.

Medications can reduce high cortisol levels before surgery, between surgery and radiotherapy, and while waiting for radiotherapy to take effect. They

- **Reduce the production of cortisol from the adrenal glands** – osilodrostat, ketoconazole, metyrapone, mitotane
- **Prevent the pituitary gland from releasing ACTH** – cabergoline and pasireotide
- **Block the action of cortisol around the body** – mifepristone.

WHAT HAPPENS AFTER SURGERY?

- **Cortisol levels can remain high (hypercortisolaemia)** – your medical team will determine your next steps
- **Cortisol levels return to normal (eucortisolaemia)** – your doctor will monitor your cortisol levels as they may change over time
- **Cortisol levels can be low (hypocortisolaemia)**, indicating a successful surgery; however, you may need steroid therapy for six to 12 months or longer.

Hormone changes after surgery may cause new symptoms, such as nausea, fatigue, depression and anxiety. You can notice signs even if you are taking cortisol medication. You may need to allow up to a year or more to recover fully. Your medical team will give you information about cortisol replacement medications and how to adjust the dose when stressed or ill. It is also essential to wear a medical alert tag and carry instructions for emergency steroid treatment.

WHAT IS THE LONG TERM PLAN?

There is a high recurrence rate in people with Cushing's disease. Life-long monitoring is essential and therefore you must:

- Check your hormone levels stay within the healthy range (including cortisol and other pituitary hormones)
- Check for tumour regrowth
- Adjust any medication as needed.

Surgery and radiotherapy can cause hypopituitarism in some people, where the pituitary gland can't produce other pituitary hormones. If this occurs, you may need long-term hormone replacement therapy.

COMMON QUESTIONS

Why am I putting on weight?

Cortisol converts proteins, carbs and fats into energy. Your body stores the energy you don't need as fat. High cortisol levels can lead to long-term fat accumulation.

Why am I getting facial hair?

ACTH stimulates the adrenal gland to produce androgens, which promote facial hair growth.

Why do I bruise so easily?

Cortisol increases the breakdown of tissue proteins, causing the tiny blood vessels near the skin (capillaries) to become weak, break easily and cause bruising.

Does Cushing's disease run in families?

Most cases of Cushing's disease are not inherited.

Why do I have stretch marks?

Cortisol causes an increased breakdown of proteins in the skin, which makes the skin more fragile.

Why do I feel so weak?

Muscle wasting, which mainly affects muscles in the upper arms and thighs, can also occur with the breakdown of proteins. You may find it difficult to climb stairs or stand after sitting. Extra weight can also strain your muscles, causing pain and fatigue.

Why am I so moody?

The excess hormone produced by the tumour affects your brain and can cause dramatic mood swings.

Will I be cured after treatment?

Cushing's syndrome can be controlled or put in remission in most cases using either surgery, radiotherapy, medication or a combination. However, Cushing's disease can recur or cortisol levels rise again even after years of followup. Regular follow- up appointments with your endocrinologist are essential.

MORE INFORMATION

The Australian Pituitary Foundation provides social support for patients and carers, and has published a range of patient resources on pituitary conditions and treatments.

For more information, please visit our website: www.pituitary.asn.au

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A GUIDE TO GROWTH HORMONE DEFICIENCY

Australian Pituitary Foundation

WHAT IS GROWTH HORMONE DEFICIENCY?

Growth hormone deficiency is a condition that occurs if your pituitary gland doesn't produce enough growth hormone. Growth hormone is a hormone that promotes growth in children and helps maintain typical body structure in adults. It also plays a role in metabolism.

The pea-sized pituitary gland sits at the base of the brain and is responsible for releasing hormones that control many different body processes. If the pituitary gland is damaged, some or all of these hormones, including growth hormone, can be deficient.

Damage to the pituitary gland can occur due to the following:

- Pituitary tumours
- Tumours of the hypothalamus (a part of the brain that helps control the release of hormones from the pituitary gland)
- Damage to the pituitary or hypothalamus after surgery or radiotherapy
- Brain injury
- A bleed in the brain
- Infections in the brain or nervous system.

Growth hormone deficiency can be diagnosed in childhood or start in adulthood.

In children, it can be due to:

- Unable to produce or respond to growth hormone from birth
- Abnormal development of the pituitary gland
- Structural brain or skull defects that are present since birth.

Treatment options include growth hormone medication to restore normal levels.

HOW COMMON IS GROWTH HORMONE DEFICIENCY?

Growth hormone deficiency affects around two to three per 10,000 people.

WHAT ARE THE SIGNS AND SYMPTOMS?

Growth hormone deficiency affects adults and children differently.

Growth Hormone Deficiency in Adults:

- Increased body fat
- Increased risk of heart disease
- Reduced muscle mass and strength
- Reduced bone mineralisation
- Low energy/lethargy
- Decreased sweating
- Anxiety and depression
- Poor concentration and memory
- Sleep problems
- Thin, dry skin

Growth Hormone Deficiency in Children:

The main sign is slow height growth (around 3.5cm) each year after the child's third birthday. This results in short stature (when a child is below the fifth percentile compared to other children of the same age and sex).

Children may also have:

- A young-looking face for their age
- Impaired hair growth
- Delayed puberty
- Headaches.

Children can develop hypopituitarism later in life.

DIAGNOSIS

Growth hormone deficiency is usually diagnosed with a combination of the following investigations listed below. A paediatric or adult endocrinologist will need to be involved in making the diagnosis:

- **Physical exam** – to measure height, weight, arm and leg lengths
- **Blood tests –** to measure levels of other hormones related to growth hormone levels as well as other pituitary hormones
- **Stimulation testing –** these specialised tests take a few hours to perform but are necessary to establish a diagnosis for purposes of obtaining growth hormone replacement therapy on PBS. Most commonly these include one of the following tests: Glucagon Stimulation Test or Insulin Tolerance Test. Both tests are designed to stress the pituitary and cause an increase in growth hormone levels (in normal individuals).
- **Scans** – a magnetic resonance imaging (MRI) or computerised tomography (CT) scan can help find any underlying disorder by revealing abnormalities of the hypothalamus or the pituitary glands.

TREATMENT

Treatment aims to:

- Replace growth hormone
- Reduce physical symptoms
- Improve the person's quality of life
- Improve the person's metabolic health.

Treatment for growth hormone deficiency involves either a daily or weekly injection of synthetic growth hormone. You typically inject the hormone into the layer of fat under the skin on the stomach.

Your specific dose will depend on your unique health needs, age, sex and other medications. Your endocrinologist is the best person to talk to about this.

In Australia, there are strict rules about who is eligible for ongoing access to growth hormone therapy through the Pharmaceutical Benefits Scheme (PBS). This medication is for children with growth hormone deficiency and adults with severe growth hormone deficiency.

Usually, your specialist needs to submit a written application demonstrating you need this medication. You may also need specific tests to prove your eligibility.

You may also need other hormone replacement medications if you have other hormone deficiencies. Your doctor can guide you on this.

ONGOING MANAGEMENT

Your doctor will then monitor your hormone levels and adjust your dose when needed to keep the levels in a normal range and avoid side effects. Side effects are uncommon when growth hormone is replaced within a normal range. Excessive growth hormone replacement can lead to:

- Swelling
- Joint or muscle pain
- Increased blood pressure
- Carpal tunnel syndrome (numb, weak or painful hands or wrists)

Often, these side effects will reduce when you reduce your dose. Your doctor will see you every six to 12 months to monitor your symptoms and hormone levels.

How long do I need to take growth hormone therapy?

Everyone is different. Some children may only need to take medication until they reach their full height potential. Some adults may need to take it for life if they feel they are getting benefits from it. Your doctor is the best person to talk to about this.

How quickly will my symptoms improve?

Usually, you can notice improvements a few weeks to months after starting medication. It can take a year to see the full benefits.

Are there any other forms of growth hormone therapy besides injections?

Other forms, such as tablet forms, are not currently available.

Is growth hormone deficiency inherited?

We still don't know. Research to find a genetic cause is ongoing.

MORE INFORMATION

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A GUIDE TO HYPOPITUITARISM

Australian Pituitary Foundation

WHAT IS HYPOPITUITARISM?

Hypopituitarism occurs when your pituitary gland doesn't make enough of one or more pituitary hormones.

Your pituitary gland is the size of a pea and sits at the bottom of the brain. This gland releases hormones that control or play a role in many of your body's functions. It also stimulates other glands to release hormones. If your body doesn't make enough pituitary hormones, other glands that rely on these hormones are also affected.

WHAT CAUSES HYPOPITUITARISM?

There are many causes, such as[2]:

- **Tumours** – a tumour in or near the pituitary gland can cause pressure on normal tissue and affect hormone levels
- **Surgery** – removing a pituitary tumour can cause temporary or permanent hypopituitarism
- **Radiotherapy** – sometimes, radiotherapy treatment for a brain tumour can damage the pituitary gland.
- **Congenital** - a condition or malformation of the pituitary gland present from birth

Less common causes include:

- Head or brain injury
- Severe bleeding in the brain
- Severe blood loss during childbirth (Sheehan's Syndrome)
- Rare diseases or infections (meningitis, tuberculosis)
- Certain conditions present at birth (congenital)
- Inflammation processes involving the pituitary gland.
- Certain medications e.g long term use of opioids

HOW COMMON IS IT?

Approximately 11,600 Australians currently live with hypopituitarism. Around 1,070 people are diagnosed every year.

WHAT ARE THE SIGNS AND SYMPTOMS?

Signs and symptoms develop gradually and worsen over time. They can be subtle and overlooked for months or years. In some people, signs develop quickly.

Signs and symptoms vary depending on the hormones affected. Having multiple hormones affected can mask the signs of the first deficiency.

COMMON SYMPTOMS (AND TARGET ORGAN AFFECTED)

Growth hormone deficiency (whole body)

- Lack of growth and sexual development (in children)
- Excessive tiredness
- Muscle weakness
- Decreased bone density
- Increased body fat
- Poor quality of life

Adreno-corticotrophic hormone deficiency (adrenal glands)

- Pale appearance
- Low blood pressure
- Dizziness
- Tiredness
- Weight loss
- Stomach pain
- Depression
- Low stress tolerance
- Poor quality of life
- Hypoglaecemia at birth

Thyroid-stimulating hormone deficiency (thyroid)

- Weight gain
- Decrease energy
- Sensitivity to cold
- Constipation
- Dry skin
- Hair loss
- Concentration difficulties
- Prolonged jaundice in babies

Follicle-stimulating hormone / Luteinising hormone deficiency in females (ovaries)

- Oestrogen and progesterone
- Irregular or loss of periods
- Low libido (sex drive)
- Hot flushes
- Loss of body hair
- Vaginal dryness (pain during sex)
- Sleep disturbance

Follicle-stimulating hormone / Luteinising hormone deficiency in males (testes)

- Erectile dysfunction
- Low libido (sex drive)
- Low sperm count
- Infertility
- Loss of facial and body hair

Prolactin deficiency (breast)

- Inability to produce breast milk

Antidiuretic hormone deficiency (kidneys)

- Extreme thirst
- Frequent urination and a large volume of urine (polyuria)
- Signs of Arginine Vasopressin Deficiency (formerly known as diabetes insipidus)

DIAGNOSIS

Tests to confirm a diagnosis include one or a combination of the following tests:

- **Blood tests** – to measure hormone levels
- **Stimulation tests** – to assess the body's response to certain hormones
- **Scans** – a magnetic resonance imaging (MRI) or computed tomography (CT) scan can examine the pituitary gland
- **Vision testing** – to check for eye problems, as tumours can affect the optic nerve.

You may need further tests, depending on your symptoms and results.

Other tests may check your:

- **Adrenal function** – tests include the insulin tolerance test, glucagon stimulation test, short synacthen test and early morning cortisol
- **Growth hormone levels** – a glucagon stimulation or insulin tolerance tests are commonly used to assess adequacy of growth hormone levels
- **Sex hormones** – a blood test to measure sex hormone levels, menstrual history and semen analysis
- **Thyroid function** – a blood test can check thyroid hormone levels
- **Risk of arginine vasopressin deficiency** – urine volume, urine and blood sodium levels, hypertonic saline stimulation test and water deprivation tests.

TREATMENT

Hormone replacement therapy is often the first treatment step. Your dose will match the amount your body would typically produce. The medications you need depend on the specific hormones that need replacing.

Sometimes, treating the condition that causes low hormone levels can correct hypopituitarism.

MISSING PITUITARY HORMONE	MEDICATION
Growth hormone	**Growth hormone injections into the fat under the skin**: Replaces growth hormone (you will need tests and an application to prove you need it)
Adrenocorticotrophic hormone	**Hydrocortisone, cortisone acetate or prednisolone tablets:** Replaces cortisol (steroid therapy)
Follicle-stimulating hormone / Luteinising hormone	**Oestrogen (patches, gel or tablets) with progesterone (patches, tablets or IUD) (women)**: Replaces ooestrogen or progesterone. **In women without a uterus progesterone replacement is not required.** **Testosterone muscle injection, patches or gel applied to the skin, or tablets (men):** Replaces testosterone **Gonadotropin (men and women):** Induces ovulation or sperm production if you want to get pregnant
Thyroid-stimulating hormone	**Levothyroxine tablets:** Replace thyroid hormone levels
Antidiuretic hormone	**Desmopressin taken by tablets or melt-wafer:** Treats arginine vasopressin deficiency

WHAT YOU NEED TO KNOW ABOUT CORTISOL REPLACEMENT THERAPY

Your doctor will work with you to find the most suitable dose. You will take higher amounts in the morning to match your body's natural cycle. If you are sick or stressed, you may need to change your dose to match the natural increase in cortisol that would normally occur.

Your doctor will closely monitor your medications to maintain the right balance. Pituitary hormones interact between themselves and other hormones. Finding a medication regime that works for you can take up to two years.

You may also need to adjust your medications, depending on your symptoms and situation, such as:

- Illness
- Pregnancy and breastfeeding
- Surgery
- Stress
- Trauma
- Weight changes.

After a serious accident or medical emergency, you may need a hydrocortisone injection. Your endocrinologist can give you more advice on this. If you are vomiting or have diarrhoea, seek urgent medical attention. You will not absorb your medication and may need a cortisone injection.

Wearing a medical alert bracelet or tag and carrying instructions for emergency steroid treatment is essential. Low cortisol levels can be a life-threatening emergency.

ONGOING MANAGEMENT

You will need regular check-ups to monitor your:

- Hormone levels
- Medications
- Physical health and wellbeing
- Mental health
- Heart and bone health
- Tumour growth.

COMMON QUESTIONS

If I take hormone replacement therapy, will I feel the same as before my hypopituitarism developed?

Hormone replacement therapy replaces hormones to levels that would occur if your pituitary gland was working normally. However, it is hard to mimic the natural hormone changes that occur in response to everyday experiences.

Everyone is different, so getting the balance right can take a while.

Medications also cause side effects in some people. Always talk to your doctor before changing any medication doses.

Will I still be able to have a family?

In some cases, treatment can restore your sex hormones to normal levels and fertility. Talk to your doctor about your desire to have a family.

How long will I need to take medication?

If you have a tumour or treatment that has affected the pituitary gland function, and your function does not recover, you may need to take medications for life.

You may need a single medication or a combination of tablets, patches, gels or injections.

What are the long-term implications of having hypopituitarism?

Many people need to take daily medication for life to reduce the risk of health conditions caused by missing hormones, like osteoporosis, heart disease and stroke.

Regular visits to your doctor will help find any changes needed to your medication and support your overall health and wellbeing.

MORE INFORMATION

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A GUIDE TO NON-FUNCTIONING PITUITARY TUMOURS

Australian Pituitary Foundation

WHAT ARE NON-FUNCTIONING PITUITARY TUMOURS?

A non-functioning pituitary tumour is a benign (non-cancerous) tumour that develops in the pituitary gland and does not secrete a hormone into the blood stream. A functioning pituitary tumour, on the other hand, does secrete one or more pituitary hormones into the blood.

The pituitary gland sits in the skull, below the brain and above the nasal passages. This gland plays a vital role in many body functions and processes.

Pituitary tumours can exist for years without causing symptoms. Sometimes, they are found by chance when you have a brain scan for another reason. Most are small tumours, less than one centimetre.

The most common symptoms of a non-functioning pituitary tumour are headaches and vision problems due to pressure on the optic nerve.

The tumour can also damage the pituitary gland resulting in hypopituitarism (where your body does not produce enough of one or more pituitary hormones). One pituitary hormone, prolactin, may be raised because of pressure on the pituitary stalk.

HOW COMMON ARE NON-FUNCTIONING PITUITARY TUMOURS?

Non-functioning pituitary tumours make up about 30% of all pituitary tumours[2].

WHAT ARE THE SIGNS AND SYMPTOMS?

Common signs include:

- Visual disturbances
- Infrequent periods
- No periods
- Reduced libido and potency in men
- Headaches.

DIAGNOSIS

A non-functioning pituitary tumour can be diagnosed through the following:

- **Blood tests** – to check the pituitary function and hormone levels
- **Scans** – a magnetic resonance imaging (MRI) or computerised tomography (CT) scan can find the size and site of the tumour
- **Visual field tests** – to see if your vision is affected

After Diagnosis

After diagnosis, it is essential to see:

- **An endocrinologist** with experience in managing pituitary diseases, and
- **A neurosurgeon** with pituitary expertise.

TREATMENT

Treatment options for non-functioning pituitary tumours include monitoring (no treatment), surgery and radiotherapy. Medical therapy may need to include hormone replacement if there is hypopituitarism.

Monitoring (no treatment)

You may not need treatment for your tumour if you don't have any signs or symptoms.

Your doctor will monitor your tumour for growth using MRI scans. You may need scans every six to 12 months and then every two to five years.

Surgery

A larger tumour (more than 1cm) along with symptoms such as headaches or visual defects may need surgery. The most common type of surgery is minimally invasive, also known as transsphenoidal. The surgeon reaches the tumour through the nose instead of making a large cut, as in open surgery.

If the tumour is large, your surgeon may be unable to remove all the tumour. Subsequent treatment may include monitoring the residual, further surgery or radiotherapy

After your surgery, you may need to take hormone replacement therapy medications if your hormones are affected by the tumour. Your doctor can give you more information about this.

If your tumour continues to grow after surgery, you may need radiotherapy.

After surgery, your vision will likely have stabilised or improved. Some people find that they have more headaches. Other issues that can occur immediately after surgery include:

- Cerebrospinal fluid (CSF) leak
- Meningitis
- Changes in blood salt levels as a result of AVP-deficiency (diabetes insipidus) or excess AVP (SIADH causing hyponatraemia).

Your medical team will monitor you in the hospital and treat any complications. Most people need to allow four to six weeks to recover and return to their usual routines.

Other complications, like cerebrospinal fluid leaks, are rare. Your neurosurgeon can discuss the risks and treatments for complications.

Radiotherapy

Radiotherapy uses radiation (X-rays or Gamma rays) to inactivate tumour cells. In most cases, you will only need radiotherapy if your tumour has grown after your surgery.

Your neurosurgeon may be able to remove more of the tumour through surgery if the tumour's location makes it possible. But if you cannot have more surgery, radiotherapy can help stop the tumour from growing.

Radiotherapy can cause hypopituitarism, so it is essential to have pituitary function tests regularly.

DIAGNOSIS

It is essential to see your doctor regularly for vision tests and scans. It is usual to have a scan 3 months following surgery and then commonly each year for 2-3 years and thereafter less depending on whether any tumour is present. If you take steroids and become very ill or stressed, you need to increase your medication dose. Your doctor can talk to you more about this and add specific advice to your ongoing treatment plan.

Seek urgent medical care if:

- Your vision gets worse or becomes impaired – this can indicate that tumours are enlarging rapidly
- You notice clear fluid dripping down the back of the throat or through the nose soon after surgery – this may indicate a cerebrospinal fluid leak.

Do I have cancer?

No. A tumour is a lump of abnormal tissue. If left untreated, your tumour may get bigger. Pituitary tumours very rarely spread to other parts of the body

Will I need treatment?

Small tumours may not need treatment. Most tumours are small and don't need treatment. You only need treatment if your tumour is growing or causing problems.

Will it go away after my treatment?

Surgery may not be able to remove the entire tumour. It is common for some of the tumour cells to remain after surgery. In most cases, you can control future tumour growth and manage your symptoms.

Will I have to take tablets in the long term?

Everyone is different. Some people may need to take medications in the long term. You might not need medication if you had surgery or radiotherapy. If your tumour stops your pituitary gland from producing enough hormones, you may need long-term hormone replacement therapy.

Will I still be able to have a family?

You can still have a family if treatment reverses your pituitary tumour's fertility effects. If your tumour or treatment has affected your natural menstrual cycle you may require hormone therapy to become pregnant.

What causes a pituitary tumour?

Doctors still aren't sure what causes pituitary tumours, but research is ongoing.

Does it run in families?

It is rare to find patterns of pituitary tumours in families.

MORE INFORMATION

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A GUIDE TO PITUITARY TUMOURS

Australian Pituitary Foundation

WHAT ARE PITUITARY TUMOURS?

A pituitary tumour is an abnormal growth that develops in the pituitary gland. It is also known as an adenoma. The pituitary gland can be found at the base of the brain in line with the top of your nose. Most pituitary tumours are benign (non-cancerous) and less than one centimetre.

Pituitary tumours can exist for years without causing symptoms. Sometimes, they are found by chance when you have a brain scan for another reason. Around one in five people (10-20%) have a pituitary tumour. Most are small tumours.[1,2]

Pituitary tumours can be:

- Functioning – develops in the pituitary gland and secretes one or more pituitary hormones into the blood stream
- Non-functioning – develops in the pituitary gland and does not secrete a hormone into the blood stream

HOW COMMON ARE PITUITARY TUMOURS?

Around 1 in 1000 individuals have a clinically significant pituitary tumour.

WHAT ARE THE SIGNS AND SYMPTOMS?

NON-FUNCTIONING

Non-functioning tumours can cause low hormone levels due to the pressure they apply on the pituitary gland. This can cause signs such as:

- Fatigue
- Irregular or loss of periods
- Loss of libido (sex drive)
- Erectile dysfunction.
- Vision impairnment

Because these symptoms are common and occur with many health issues, you can have a tumour for many years before knowing about it.

FUNCTIONING

Functioning tumours cause signs of the conditions based on the excess hormone made:

- Prolactinoma – caused by too much prolactin
- Cushing's disease – is caused by too much adrenocorticotrophic hormone (ACTH)
- Acromegaly – is caused by too much growth hormone.

A guide to each of these conditions is available from the Australian Pituitary Foundation.

PITUITARY APOPLEXY

Pituitary apoplexy is a rare event that occurs if a tumour bleeds and suddenly increases in size. It is a medical emergency. It may cause a sudden loss of hormones or vision due to pressure on the eye nerves. Signs include a sudden, severe headache, nausea, vomiting and aversion to light and sound. If you or someone you know develops these signs, go to your nearest emergency room immediately.

DIAGNOSIS

Pituitary tumours are often discovered:

- **Incidentally** – after a scan for another health issue
- **Visual disturbances** – if you have vision problems (tunnel vision, blurred vision, double vision) and headaches, your doctor may order tests
- **Hormonal issues** – if you have symptoms associated with too little or too much hormone.

Tests to diagnose a pituitary tumour include:

- **Blood tests** – to measure hormone levels.
- **Scans** – a magnetic resonance imaging (MRI) or computed tomography (CT) scan can study the pituitary gland
- **Vision testing** – to check your visual field

You may need more tests, depending on the results and type of hormones your tumour is producing.

After Diagnosis

After diagnosis, it is essential to see:

- **An endocrinologist** with experience in managing pituitary diseases, and
- **A neurosurgeon** with pituitary expertise.

TREATMENT

Treatment options include montioring (no treatment), medication, surgery and radiotherapy. Treatment aims to:

- Relieve or reverse symptoms
- Prevent vision loss
- Restore hormone levels
- Prevent tumour growth.

Monitoring (no treatment)

If your tumour has no symptoms, you may not need any treatment. Your doctor will monitor your tumour regularly to make sure it doesn't get any larger.

Medication

Medication can help to treat some tumours and conditions, such as prolactinomas.

Dopamine agonists, such as cabergoline and bromocriptine, are frequently used in prolactinomas to reduce prolactin levels and shrink the tumour, but occasionally used in other tumour types as well. Medications such as somatostatin analogues (lanreotide, octreotide, pasireotide) are most commonly used in acromegaly to control growth hormone levels if surgery has not removed all the tumour. Drugs reducing production of cortisol levels are used in Cushing's disease.

In all tumour types, hormone replacement therapy may also be needed to treat hormone deficiencies.

Surgery

If you have signs and symptoms, you may need surgery. The most common type of surgery is minimally invasive, also known as transsphenoidal. The surgeon reaches the tumour through the nose instead of making a large cut, as in open surgery.

If the tumour is large, your surgeon may be unable to remove all the cells. You may need another surgery several months later.

TREATMENT

After your surgery, you may need to take hormone replacement therapy medications if your hormones are affected by the tumour. Your doctor can give you more information about this.

If your tumour continues to grow after surgery, you may need radiotherapy.

After surgery, your vision will likely have stabilised or improved. Some people find that they have more headaches.

Other issues that can occur immediately after surgery include:

- Cerebrospinal fluid (CSF) leak
- Meningitis
- Changes in blood salt levels as a result of AVP-deficiency (diabetes insipidus) or excess AVP (SIADH causing hyponatraemia).

Your medical team will monitor you in the hospital and treat any complications. Most people need to allow four to six weeks to recover and return to their usual routines.

Other complications, like cerebrospinal fluid leaks, are rare. Your neurosurgeon can discuss the risks and treatments for complications.

Radiotherapy

Radiotherapy reduces the chance of your tumour returning. You may have radiotherapy alone or with other treatments. It is usually performed where surgery or medication can not successfully control the tumour or hormone levels.[4]

After radiotherapy, your doctor will monitor your hormone levels. Radiotherapy can sometimes cause a gradual loss of pituitary function and low levels of pituitary hormones. If this occurs, you will need permanent hormone replacement therapy.

COMMON QUESTIONS

Do I have cancer?

Pituitary tumours very rarely spread to other parts of the body.

Will I need treatment?

Small tumours may not need treatment. Most tumours are small and don't need treatment. You only need treatment if your tumour is growing or causing problems.

Will it go away after my treatment?

Surgery may not be able to remove the entire tumour. It is common for some of the tumour to remain after surgery. In most cases, you can control future tumour growth and manage your symptoms.

Will I have to take tablets in the long term?

Everyone is different. Some people may need to take medications in the long term. You might not need medication if you had surgery or radiotherapy. If your tumour stops your pituitary gland from producing enough hormones, you may need long-term hormone replacement therapy.

Will I still be able to have a family?

You can still have a family if treatment can reverse the fertility effects of your pituitary tumour. If your tumour or treatment has affected your natural menstrual cycle you may require hormone therapy to become pregnant.

What causes a pituitary tumour?

Doctors still aren't sure what causes pituitary tumours, but research is ongoing.

Does it run in families?

It is rare to find patterns of pituitary tumours in families.

MORE INFORMATION

The Australian Pituitary Foundation provides social support for patients and carers, and has published a range of patient resources on pituitary conditions and treatments.

For more information, please visit our website: www.pituitary.asn.au

Email: support@pituitary.asn.au

Phone: 1300 331 807

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Acknowledgement – We are grateful to the members of the Australian Pituitary Foundation for reviewing this information.

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A GUIDE TO PROLACTINOMA

Australian Pituitary Foundation

WHAT IS A PROLACTINOMA?

A prolactinoma is caused by a tumour of the pituitary gland, a small gland at the base of the brain. In the vast majority of cases these tumours do not spread outside of the pituitary gland. They usually grow slowly and some don't grow much at all. Prolactinomas cause the pituitary gland to produce too much of the hormone prolactin. This causes a decrease in the levels of some sex hormones namely oestrogen and testosterone.

Prolactin plays a role in milk production and breast tissue development. High levels can cause irregular periods, infertility, erectile dysfunction (via low testosterone levels) and galactorrhoea (breast milk secretion). If large they may cause headaches or visual changes.

There are two types:

- **Microprolactinoma** – less than 1cm and more common in women
- **Macroprolactinoma** – greater than 1cm and more common in men

HOW COMMON IS IT?

Between 500 and 1200 Australians are diagnosed with prolactinomas each year. About 6400 to 16000 Australians are currently living with a prolactinoma.

WHAT ARE THE SIGNS AND SYMPTOMS?

MEN

- Reduced libido
- Erectile dysfunction
- Infertility due to low sperm count

In men, the symptoms can be more subtle and develop slowly, resulting in a later diagnosis.

WOMEN

- Irregular or absent menstrual cycle
- Milk secretion from breasts
- Infertility
- Reduced libido
- Vaginal dryness or pain during sex

Prolactinomas are often found early in women, as symptoms are more evident.

BOTH MEN AND WOMEN

Large prolactinomas can press or invade surrounding brain tissue, causing:

- Some loss of peripheral vision
- Headaches
- Fatigue

Some people may develop hypopituitarism, where the body doesn't make enough pituitary hormones. More information about hypopituitarism is available from the Australian Pituitary Foundation.

DIAGNOSIS

Tests to diagnose a prolactinoma and work out a treatment plan include:

- **Blood tests** – to measure levels of prolactin and pituitary function (including sex hormones, thyroid function tests, steroid and growth hormones)
- **Scans** – magnetic resonance imaging (MRI) or computed tomography (CT) scan can see the pituitary gland and the site of tumour
- **Eye testing** – to check the visual field
- **Bone mineral density test** – to check bone health.

High levels of prolactin may also be due to another cause, including:

- Pregnancy or breastfeeding
- Stress
- Side effects of other medication (dopamine antagonists, antipsychotics, some antidepressants, opiates)
- Polycystic ovarian syndrome
- Hypothyroidism (low thyroid function)
- Pituitary stalk dysfunction
- Kidney failure

After Diagnosis

After diagnosis, it is essential to see:

- **An endocrinologist** with experience in managing pituitary diseases, and
- **A neurosurgeon** with pituitary expertise.

TREATMENT

Treatment aims to reduce symptoms by reducing prolactin levels. Options include:

- **Monitoring (no treatment)** – if there are no symptoms and prolactin levels are only slightly high, or after menopause
- **Medication** – called dopamine agonists, medications can reduce prolactin levels, return oestrogen or testosterone levels to normal and shrink the size of the tumour
- **Surgery** – to reduce tumour size or remove the tumour
- **Radiotherapy** – rarely used when other treatment options are not working to control the tumour.

Medication is often successful in reducing prolactin levels, decreasing tumour size and relieving symptoms in many people. If prolactin levels are difficult to return to normal, women may also need to take oestrogen and men may need to take testosterone. The most common medications that reduce prolactin production and tumour size include:

- **Bromocriptine** – a daily tablet
- **Cabergoline** – a tablet taken once or twice per week; more effective than bromocriptine[3]

Sometimes, the prolactinoma does not respond to medication, or the medication causes side effects. In these cases, you may need surgery or radiotherapy. Increasingly, these days, surgery may be an option for some patients with small tumours who wish to avoid long-term medication.

MEDICATION MANAGEMENT

Most people need to take medication in the long term. You may be able to try stopping after three to five years, depending on your prolactin levels, tumour size and location, and menopausal state. Medications can cause side effects in some people, including dizziness, nausea and headaches.

You can limit these side effects by:

- Taking your tablets before bed at night
- Starting with a low dose
- Increasing your dose gradually, as guided by your doctor.

Cabergoline or bromocriptine may sometimes cause behaviour changes which should also be discussed with your doctor. These include obsessive-compulsive behaviour (eg excessive gambling or video gaming), hypersexuality and increased anxiety or depression. It's essential to have regular check-ups with your doctor to monitor your symptoms and any side effects. Very rarely with high dose cabergoline or bromocriptine the heart valves can become leaky. Regular heart ultrasounds will be recommended if you are on long term/high dose cabergoline or bromocriptine.

PREGNANCY AND PROLACTINOMAS

Tell your doctor if you are planning to become pregnant or fall pregnant while taking medications. If you take bromocriptine or cabergoline, there is no evidence of an increased risk to the pregnancy or baby.[5] Women with microprolactinomas can generally stop taking medications after a pregnancy is confirmed and may only need to restart after finishing breastfeeding.

If you have a macroprolactinoma, you may need to keep taking your medication and have regular eye tests during pregnancy. It is safe to have MRIs in the second and third trimesters if needed. During pregnancy, there is a slight risk of tumour enlargement. If this occurs and causes a threat to your vision, medication may be restarted in pregnancy and occasionally surgery is required.

COMMON QUESTIONS

Why have my periods stopped?

High prolactin levels can stop the body from making hormones that regulate the menstrual cycle, causing irregular or missed periods.

Why am I no longer interested sexually in my partner?

High prolactin levels stop your body from making sex hormones, which can lead to low libido (sex drive).

Why is sex painful?

High prolactin levels cause oestrogen levels to fall. Low oestrogen can lead to thinning of the inner lining of the vagina. Treatment helps restore normal oestrogen levels.

Can a microprolactinoma grow into a macroprolactinoma?

In the majority of cases, no.

Why do I have a discharge from my breasts?

As prolactin increases to help women produce milk to breastfeed, high prolactin levels can cause the same effect.

How long will I need to take my medication?

Prolactin levels can return to normal a few weeks after you start taking your medication, but it is essential to keep taking medication until your doctor tells you otherwise. Some people may try stopping after three or five years or after surgery to see if their prolactin levels remain normal. If you stop taking the medication, your prolactin levels may rise, and the tumour size may increase. It is important to see your doctor for regular check-ups to monitor your prolactin levels and tumour size.

MORE INFORMATION

The Australian Pituitary Foundation provides social support for patients and carers, and has published a range of patient resources on pituitary conditions and treatments.

For more information, please visit our website: www.pituitary.asn.au

Email: support@pituitary.asn.au

Phone: 1300 331 807

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A GUIDE TO
RATHKE'S CLEFT CYST

Australian Pituitary Foundation

WHAT IS A RATHKE'S CLEFT CYST?

A Rathke's Cleft Cyst is congenital deformity that develops while the fetus is growing in the womb. It is a a benign (non-cancerous) collection of fluid that forms in a gap during the early development of the pituitary gland. This is called the Rathke's pouch. Usually, this gap closes before birth as the pituitary gland forms but in some people, the gap doesn't close and leaves a space where a cyst can form.

The pituitary gland is a pea-sized structure at the brain's base, releasing hormones that play a role in many body functions.

Most cysts don't cause symptoms and rarely get bigger. Cysts can cause symptoms when they press on the pituitary gland or an optic nerve. These cysts may disrupt hormones or cause vision problems. The average age of diagnosis in adults is 42, and cysts are more common in women than men. One of the signs is irregular periods due to a hormonal imbalance, so women may be more likely than men to notice signs.

HOW COMMON IS A RATHKE'S CLEFT CYST?

Rathke's cleft cysts affect around 2.5 million Australians at any given time.

WHAT ARE THE SIGNS AND SYMPTOMS?

GENERAL SIGNS

The vast majority of these cysts cause no symptoms. However, symptoms may include:

- Headaches – due to the cyst pressing on surrounding structures
- Vision changes – blurry vision, vision loss and loss of peripheral vision
- Hyperprolactinaemia in women – when the body makes too much of the hormone prolactin

HYPOPITUITARISM

Hypopituitarism is when the body doesn't make enough of one or more pituitary hormones. Symptoms can include:

- Appetite loss
- Low sex drive
- Loss of periods in women
- Loss of body or facial hair
- Growth changes
- Weight gain or loss
- Body temperature issues
- Tiredness and confusion.

DIAGNOSIS

Rathke's cleft cysts are diagnosed with the following:

- **Blood tests** – to check your pituitary function
- **Vision field tests** – to check if you have vision impairment
- **Scans** – a magnetic resonance imaging (MRI) or computerised tomography (CT) scan can find the size and site of the cyst.

TREATMENT

Monitoring (no treatment)

If your cyst is an incidental finding (found during a scan for an unrelated condition) and is not causing symptoms, you don't need to do anything except monitor it with an MRI scan every 12 months for the first three years.

It is rare for cysts found incidentally to get bigger.

Treatment

If your cyst is causing symptoms, the most common treatment is surgery which is most often performed through the nose (transsphenoidal surgery). The cyst is either drained but may also be fashioned to allow drainage of fluid back into the normal brain fluid.

ONGOING MANAGEMENT

After your surgery, symptoms such as headaches and visual issues should improve. However, some people develop hypopituitarism (when the body doesn't make enough of one or more pituitary hormones). Your doctor will test your pituitary function after your surgery and can give you hormone replacement medication if necessary.

Other complications of surgery can include:

- Cerebrospinal fluid (CSF) leak
- Meningitis
- Changes in blood salt levels as a result of AVP-deficiency (diabetes insipidus) or excess AVP (SIADH causing hyponatraemia).

Your neurosurgeon will discuss the risks and complications of surgery. If your cyst comes back, you can have a second surgery. Recurrences most often occur in the first five to six years after surgery.

It is usual to have a scan 3 months following surgery and then commonly each year for 2-3 years and thereafter less commonly depending on whether any cyst recurrence has been seen.

It's also essential for your doctor to assess your pituitary function, hormone deficiencies and cyst regrowth every six months to one year.

Surgery can be emotionally challenging. Ask your doctor about mental health support if you are struggling to cope.

Radiotherapy is not a standard treatment option.

If you take steroids and become very ill or stressed, you need to increase your medication dose. Your doctor can talk to you more about this and add specific advice to your ongoing treatment plan.

Seek urgent medical care if:

- Your vision gets worse or becomes impaired – this can indicate that cysts are enlarging rapidly
- You notice clear fluid dripping down the back of the throat or through the nose soon after surgery – this may indicate a cerebrospinal fluid leak.

MORE INFORMATION

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RESOURCES

Australian Pituitary Foundation

The Australian Pituitary Foundation provides support for patients with pituitary conditions.

Website: www.pituitary.asn.au

Email: support@pituitary.asn.au

Phone: 1300 331 807

The UK's Pituitary Foundation (www.pituitary.org.uk) and the USA's Pituitary Network Association (www.pituitary.org) are also excellent sources of information and interaction with others affected by pituitary conditions.

ORGANISATIONS

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Email: admin@impotenceaustralia.com.au
Web: www.impotenceaustralia.com.au

ACCESS Australia Infertility Network
PO Box 3605, Rhodes Waterside
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The Australian Thyroid Foundation Ltd
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