Cushing's

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For hormones • For health • For life

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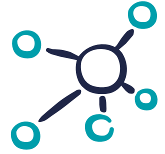
An explanation of hormones



What is the pituitary gland?

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

What is cortisol?

Cortisol is a hormone made by the adrenal glands (two small glands which lie just above each kidney), and it is vital for life. It has several functions, which include helping to:

- regulate blood pressure
- regulate the immune system
- regulate metabolism
- assist the body in responding to stress

What is Cushing's?

The condition is named after Harvey Cushing, an American neurosurgeon, who first described people with this condition in 1912.

Cushing's syndrome occurs, when there is too much of the hormone cortisol (the body's natural glucocorticoid steroid hormone) produced in the body (this is called 'endogenous Cushing's'). It can also be caused by long-term glucocorticoid treatment (e.g., when taking a steroid such as prednisolone for conditions such as asthma, rheumatoid arthritis or ulcerative colitis).

The most common cause of endogenous Cushing's is Cushing's disease (around 70% of the cases). This is when a benign tumour (growth) of the pituitary gland produces the hormone called ACTH (Adrenocorticotrophic Hormone), which goes through the blood stream to the adrenal glands. This in turn causes the adrenal glands to release too much cortisol.

Another cause of Cushing's syndrome is when usually one tumour in the adrenal gland produces and secretes too much cortisol in the blood. Lastly, there could be a small growth in another part of your body which is having the same effect; this is called ectopic Cushing's.

Cushing's disease is diagnosed in around 1.3 – 1.5 people per million population per year, but it is now being found more frequently when it is specifically investigated. This condition largely affects more women than men, but the reason for this is currently unknown. It is most commonly diagnosed between the ages of 30 to 40; however, children may also be diagnosed. Cushing's has no known environmental triggers.

What are the signs and symptoms of Cushing's?

The signs and symptoms of having Cushing's are very varied, and usually several are present at once. Symptoms can include:

- Excessive and rather quick (or at times more gradual) onset of weight gain around your trunk (your arms and legs may remain unchanged and can appear quite thin compared to your body).
- Weak muscles, especially in your legs.
- A rounder and redder than usual face (a classic feature of Cushing's known as 'moon face') and you may have developed acne.
- Weaker bones (an X-ray may show a fractured rib for instance) due to steroid-induced osteoporosis (thinning and brittleness of bones); therefore, you have an increased risk of fracture.
- High blood pressure (hypertension) or diabetes mellitus.
- A tendency to bruise easily and have deep red/purple stretch marks (striae) usually appearing on the abdomen, similar to those which occur during pregnancy, but are more pronounced.
- Irregular periods or stop having them altogether.
- Excessive hair growth on parts of the body and often on the face in women.
- Reduction or absence of sex drive (libido).
- Reduced fertility.
- Generally unwell with low energy and be more susceptible to infections.
- Mood swings, which can include being more irritable, feeling depressed or anxious. In some cases, psychological problems can be severe, and may result in a diagnosis of a mental health crisis.
- In children, it may show itself by growth stopping and weight increasing.

Cushing's affects many functions of the body, both mentally and physically, and affects different people in different ways. Because Cushing's progresses slowly and gradually, in most cases, it can go unrecognised for quite some time, sometimes resulting in depression. Many people realise looking back that they experienced symptoms of the condition two or more years before they were referred to an endocrinologist. However, as they didn't know about Cushing's at the time, they were unaware that they were displaying symptoms of the condition.

Cyclical Cushing's

Rarely, the amount of cortisol causing the condition can fluctuate significantly. This is called 'Cyclical Cushing's' as symptoms vary often over days, weeks, months or even years, depending on your cortisol levels. This can cause difficulty and delays in diagnosis. Occasionally, your endocrinologist will need to repeat testing to assess whether you have this more uncommon type of Cushing's.

How is Cushing's diagnosed?

The signs and symptoms of Cushing's can vary widely and as a result, it can be difficult to diagnose.

The tests used to diagnose Cushing's are complicated and may take some time; they may also need to be repeated on several occasions. This is because if doctors don't gather all the right information about your case, then this can lead to the wrong diagnosis or the wrong treatment. You may have tests as an inpatient or an outpatient. The first tests conducted are to establish that Cushing's is present. This is because there can be various other reasons for the symptoms caused by Cushing's. If Cushing's is likely, then further tests establish the location.

There are two main tests needed to identify Cushing's. These are the dexamethasone test and the 24 hour urine test. You will be given a tablet called dexamethasone. In people who do not have Cushing's, taking this tablet will completely suppress the production of the hormone cortisol.

You may also have a series of blood tests, urine tests and/or saliva tests. The urine test involves collecting all the urine you pass during 24 hours (for example, between eight o'clock one morning and eight o'clock the next morning). The hospital will provide a special container for this, and you will be told how to take accurate and clean samples.

If these initial tests indicate that Cushing's is likely, you will then need further tests to find its location. You may be admitted to hospital for these, and it is likely that you may be referred to a hospital where they are very familiar with Cushing's. Tests include a higher dose of dexamethasone and measuring hormones in the blood coming from the pituitary gland. At the end of all these blood tests your arms might be quite bruised.

You will also have your pituitary and/or adrenal glands scanned, using a type of magnetic scan called an MRI scan, or by a form of X-Ray called a CT scan. You may be given an injection during the scan to improve the results. This test is often called the MRI with Contrast test. A minority of people are allergic to this injection, so do tell the specialist if you have asthma or any allergies. The scan does not hurt, but the MRI machine can be very noisy and it may involve being inside the scanner for around half an hour. If you think this will make you claustrophobic or nervous, tell your GP who may give you something to help you relax. If ectopic Cushing's is a possibility, you may be scanned from 'head to toe' to look for the cause.

"I first noticed that something was wrong when I started to gain weight. I used to play football on a Monday, did spin classes on Tuesday, Wednesday, Thursday, Metafit on Friday and golf on Saturdays – Sundays were a rest day. I only ate on chicken, eggs and rice, yet I kept getting heavier. The doctors kept saying it was down to me so I continued this diet for 3 years.

I was then diagnosed with diabetes, and also started to experience high blood pressure and a round, red face. I was still told that it was due to my lifestyle, event though I tried to explain the amount of exercise that I was doing.

After about 6 years of these unexplained symptoms, one morning I was getting ready for work and I passed out in the living room. My wife found me around an hour later when she got up from work. The doctor I was seeing for diabetes, at Russells Hall Hospital in Dudley, suggested that I might have Cushing's disease. I was sent for an MRI and they found a tumour on my pituitary gland."

Steven, who was diagnosed with Cushing's after unexplained weight gain.

How is Cushing's treated?

Your endocrinologist may decide to treat you whilst you wait for a full diagnosis with drugs to reduce the amount of cortisol produced by your adrenal glands. If so, you may have to spend two or three days in hospital to assess your response to the tablets or attend regularly as an outpatient.

It is likely that you will have surgery to remove the tumour, and hormone replacement therapy after surgery.

Surgery

If your Cushing's is caused by a pituitary tumour (Cushing's disease), you will usually need an operation. It is carried out under a general anaesthetic through the nose. This is called transsphenoidal surgery. By going behind the nose in this way, the surgeon can see your pituitary gland without having to operate on the main part of your head. Sometimes the back of the nose needs patching with tissue taken from under the skin of the thigh or abdomen which will leave a small scar there.

Alternatively, if your Cushing's is caused by a small growth in the adrenal glands, then you may need an operation to remove one or both of them (if you have a growth in both adrenal glands).

You can find out more about the operation in our 'Surgery and Radiotherapy' booklet, or on our website: ***www.pituitary.org.uk/surgery***

Most people are up and about and eating normally the following day and are back at home within a few days. Recovery can be challenging as your body has to get used to lower levels of cortisol. You should plan to be away from work for four to six weeks, however this may be longer. Recovery times can vary significantly depending on your particular job and circumstances. You will need to avoid blowing your nose for three weeks or more while it heals. You may also lose your sense of smell for weeks or months - although this usually returns to normal.

For a few days after the operation, some people feel very thirsty and pass urine more than normal. This condition is called AVP Deficiency (previously diabetes insipidus) and is usually temporary but occasionally can become permanent. It can be treated by using a drug called desmopressin. Occasionally, it is necessary to carry out a second operation if the first is not completely successful. This can sometimes be done within seven to ten days.

You will need further hormone tests, either immediately and/or four to six weeks after the operation. Again, this may involve a hospital stay. These tests are designed to show whether or not the operation has been a success, and whether you have developed deficiencies of other pituitary hormones, known as hypopituitarism, which may need replacement.

After surgery you may feel worse for some months, or often up to a year, before you begin to feel better. Relapse is possible, even after many years, so you should be kept on long-term monitoring. Over time, your strength and mood will eventually improve, and other symptoms will gradually diminish.

Hormone Treatment

After surgery you may have to have replacement for the cortisol (which is called 'hydrocortisone' when it is in tablet form) or another steroid tablet such as prednisolone. This compensates for a temporary reduction in your body's ACTH production. This occurs because the normal control mechanisms are 'switched off' after being exposed to too much cortisol for so long.

Hydrocortisone is taken in the form of tablets, usually two or three times a day. If prescribed and you don't take the tablets (or when they are deliberately not given immediately after the operation or during re-assessment tests) you may feel generally weak, tired and 'ill'. However, you will feel better after you begin to take the tablets again.

Going into remission

Many people will be completely cured – their cortisol levels are no longer high – after pituitary surgery but your doctor may prefer to be cautious and call this ‘remission’. If you had Cushing’s for a while before starting treatment, you may have lost some bone content. Bone scans may be given to check for osteoporosis; and treatment can both improve osteoporosis and limit the consequences. However, bone may not completely return back to normal.

Muscle strength often eventually returns to normal, but it may not always return back to full strength; if this was affected prior to treatment, a return to normal may happen gradually, over a period of months or even a couple of years.

Cushing’s is also a major cause of high blood pressure and diabetes – both of which are improved after successful treatment. In many cases, these conditions disappear and do not need treatment; in others, better control will usually be achieved on much less medication than before. Your periods should return soon after surgery and you should be able to have children. Similarly, if your fertility has been affected by the treatment, you can be given hormones to restore your fertility and enable you to have children.

If the treatment of your pituitary gland is not fully effective, there are other solutions. You may need to have both adrenal glands removed; this is called an adrenalectomy. This is usually performed as keyhole surgery, so recovery from the operation is quick and you should only need to stay for a few days. After an adrenalectomy you will need to take replacement cortisol, and an additional salt-retaining hormone tablet (called fludrocortisone). You will need to take both of these tablets for the rest of your life. You can find out more about this in our booklet ‘Adrenal Insufficiency’. To prevent any recurring problems in the future, or if pituitary surgery is only partially successful, you may also be given pituitary radiotherapy. We have more information about radiotherapy in our booklet ‘Surgery and Radiotherapy’ or on our website.

There are other medications that can reduce cortisol levels if the surgery is unsuccessful. After unsuccessful pituitary surgery – where not all of the tumour can be removed initially and the cortisol levels remain high, the choice between adrenalectomy, pituitary radiotherapy and ongoing drug treatment for Cushing's is a very complicated one. Make sure you discuss this fully with your endocrinologist and understand the reasons why a particular treatment has been recommended.

In non-pituitary cases of Cushing's syndrome (see 'What is Cushing's?' above), treatment depends on the cause.

- With adrenal tumours causing Cushing's, the offending adrenal gland(s) is usually removed.
- In ectopic Cushing's the underlying tumour may be removed during an operation or treated with radiotherapy or even chemotherapy. If the cause cannot be clearly identified, then an adrenalectomy may be recommended.

"It's learning to know you've got good days and bad days and not feel guilty when you do have bad days and need to rest. It is frustrating but there is nothing you can do about that. You've just got to take the better days when they come and make the best of them as much as you can."

Nadine, who was diagnosed with Cushing's in 2003.

Cushing's continuing care

People who have had Cushing's always require long-term monitoring and this will be shared between your endocrinologist and GP. Because pituitary conditions are relatively rare, you might find that you will be the only person with Cushing's that your GP is treating, and they might find it helpful to have a copy of our 'Pituitary Disease Fact File for General Practitioners'.

Psychological aspects

Cortisol is linked to mood so people with Cushing's can have depression. Usually, this depression is atypical in presentation: gaining weight and being restless or agitated are common signs and symptoms. Memory and sleep patterns can also be affected. The degree of depression can be linked to the level of cortisol.

Cushing's-induced depression is usually relieved by treatment of the Cushing's but often takes some months or more to recede. There is no convincing evidence of a link between stress causing Cushing's.

Loss of libido, infertility & relationships

You may suffer from a low sex drive, impotence or lack of self-esteem due to the imbalance of hormones and, in some cases, physical changes. This, in turn, may cause a strain on your relationship. There is also a possibility that you may have problems conceiving. It may help to talk to your partner about how you are both feeling and to consult your GP/endocrinologist.

Living with Cushing's

You may be entitled to free prescriptions. You should ask your GP, pharmacist or endocrine clinic for help with this.

If you are taking steroid replacement medication, or you have AVP deficiency (diabetes insipidus), it is a good idea to wear a medical information bracelet or equivalent as the information will help doctors if you have an accident and are unconscious.

There are various medical emblems available; our website includes contact details for several organisations.

Employment

For your stay in hospital if you have had surgery, the ward staff will give you a certificate for your employer and advise you how long you will be expected to remain off work. Your GP can issue further certificates if you require these. If you are experiencing any difficulties in retaining or returning to your employment, at any stage of your pituitary condition you should contact your local Citizens Advice. If you need extra employment support because of a disability your local Jobcentre Plus can put you in touch with one of their Disability Employment Advisers.

Alcohol and replacement hormones

There is no interaction between alcohol and most replacement hormones, and you are allowed to drink in moderation, such as the national guidelines – it is safest not to drink more than 14 units a week on a regular basis.

Common questions

Q: Will I be able to wear my dentures after the pituitary operation?

A: You may find that your dentures do not fit very well until the swelling goes down. This should only take a few days.

Q: When will my excess facial hair improve?

A: Remission of Cushing's usually results in a gradual improvement of excess facial hair in women. In many cases this will resolve completely over a few months. However, excess facial hair is also a very common problem in women without Cushing's, and if you have had a tendency towards this in the past, then remission of the Cushing's may not completely resolve the problem. In this case, there are a variety of treatments available which you should discuss with your endocrinologist.

Q: I have heard that hydrocortisone is a steroid. Will it be bad for me?

A: You will only be given the amount that your body would normally make, so you should not have any of the problems normally associated with steroids.

Q: Will my body go back to how I used to look?

A: You should usually get back to your usual self in time, but if you have had Cushing's for a long time before the diagnosis was made, this may take a few years and will require a healthy eating plan, careful dieting and exercise (gently at first if you are not used to it).

Q: Is a pituitary tumour hereditary?

A: Only in very exceptional cases.

More information

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

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

Endocrine Nurse Helpline

Our specialist endocrine nurses can provide medical guidance.

Information and Support Helpline

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

Telephone Buddy

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

Support Groups

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

Events

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

About
The Pituitary Foundation

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

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

Become a member and support our work

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

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

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

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

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

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

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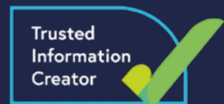

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