

Addison's disease

Overview

Addison's disease, also known as primary adrenal insufficiency or hypoadrenalism, is a rare disorder of the adrenal glands.

The adrenal glands are 2 small glands that sit on top of the kidneys. They produce 2 essential hormones: cortisol and aldosterone.

The adrenal gland is damaged in Addison's disease, so it does not produce enough cortisol or aldosterone.

About 9,000 people in the UK have Addison's disease, with over 300 new cases diagnosed each year.

It can affect people of any age, although it's most common between the ages of 30 and 50. It's also more common in women than men.

Symptoms of Addison's disease

Early-stage symptoms of Addison's disease are similar to other more common health conditions, such as clinical depression or flu.

You may experience:

lack of energy or motivation (fatigue)

muscle weakness

low mood

loss of appetite and unintentional weight loss

increased thirst

Over time, these problems may become more severe and you may experience further symptoms, such as dizziness, fainting, cramps and exhaustion.

You may also develop small areas of darkened skin, or darkened lips or gums.

Although these symptoms are not always caused by Addison's disease, you should see a GP so they can be investigated.

Why it happens

Addison's disease is usually the result of a problem with the immune system, which causes it to attack the outer layer of the adrenal gland (the adrenal cortex), disrupting the production of the steroid hormones aldosterone and cortisol.

It's not clear why this happens, but it's responsible for 70% to 90% of cases in the UK.

Other potential causes include conditions that can damage the adrenal glands, such as tuberculosis (TB), although this is uncommon in the UK.

Treating Addison's disease

Addison's disease is treated with medicine to replace the missing hormones. You'll need to take it for the rest of your life.

With treatment, symptoms of Addison's disease can largely be controlled. Most people with the condition have a normal lifespan and are able to live an active life with few limitations.

But many people with Addison's disease also find they must learn to manage bouts of fatigue, and there may be associated health conditions, such as diabetes or an underactive thyroid (hypothyroidism).

Adrenal crisis

People with Addison's disease must be constantly aware of the risk of a sudden worsening of symptoms, called an adrenal crisis.

This can happen when the levels of cortisol in your body fall significantly.

An adrenal crisis is a medical emergency. If left untreated, it can be fatal.

If you or someone you know has Addison's disease and is experiencing severe symptoms, they will need a hydrocortisone injection immediately, either injected by themselves or by a person who is with them.

Then call 999 for an ambulance, saying it's an "adrenal crisis" or "Addisonian crisis".

Information about you

If you have Addison's disease, your clinical team will pass information about you on to the National Congenital Anomaly and Rare Disease Registration Service (NCARDRS).

This helps scientists look for better ways to prevent and treat this condition. You can opt out of the register at any time.

Symptoms

Addison's disease can be difficult to detect at first because early symptoms are similar to those of many other health conditions.

Initial symptoms of Addison's disease can include:

- overwhelming exhaustion and lack of energy
- loss of appetite and unintentional weight loss
- dizziness on standing
- low blood pressure (hypotension)
- patches of skin that are darker than the surrounding skin (hyperpigmentation)

craving salty foods
feeling and being sick
muscle weakness with cramps
abdominal pain
abnormal drowsiness or tiredness (lethargy)
low mood (mild clinical depression) or irritability
a frequent need to pee
increased thirst
difficulty concentrating
headaches
a higher temperature than normal

If you have periods, you may get irregular periods or miss periods completely. You may also lose your pubic hair and the hair under your armpits.

Children with Addison's disease may go through stages of puberty later than usual.

Some people with Addison's disease also develop low blood sugar (hypoglycaemia). This can cause symptoms such as difficulty concentrating, confusion, anxiety and even unconsciousness (particularly in children).

If you're experiencing symptoms of Addison's disease, see a GP so they can diagnose or rule out the condition. These symptoms will usually improve with appropriate treatment.

[Read more about diagnosing Addison's disease and treating Addison's disease.](#)

Adrenal crisis

If the levels of hormones produced by the adrenal gland become too low it can lead to a life-threatening situation known as an adrenal or Addisonian crisis. This can be triggered by severe physical stress like having an operation or having another illness like an infection.

During an adrenal crisis, the symptoms of Addison's disease appear quickly and are severe. This could happen when you're already experiencing initial symptoms or without any symptoms at all.

Signs of an adrenal crisis include:

severe dehydration
pale, cold, clammy skin
sweating
rapid, shallow breathing
dizziness
low blood pressure (hypotension)
severe vomiting and diarrhoea
abdominal pain or pain in the side
fatigue and severe muscle weakness
headache
severe drowsiness or loss of consciousness

An adrenal crisis is a medical emergency. If left untreated, it can be fatal.

If you think you or someone you know with Addison's disease is having an adrenal crisis, they will need a hydrocortisone injection immediately, either injected by themselves or by a person who is with them.

Then call 999 for an ambulance, saying it's an "adrenal crisis" or "Addisonian crisis".

If an adrenal crisis is not treated, it can lead to a coma and death. There's also a risk your brain will not get enough oxygen if treatment is delayed, which can cause permanent disability.

Causes

Addison's disease develops when the outer layer of your adrenal glands (adrenal cortex) is damaged, reducing the levels of hormones it produces.

Problems with the immune system

A problem with the immune system is the most common cause of Addison's disease in the UK.

The immune system is your body's defence against infection and disease. If you're ill, your immune system produces antibodies – a special type of protein that destroys disease-carrying organisms and toxins. These antibodies attack the cause of the illness.

However, if you develop a problem with your immune system, it can start to attack your own healthy tissues and organs. This is called an autoimmune disorder.

Addison's disease can develop if your immune system attacks your adrenal glands and severely damages your adrenal cortex.

When 90% of the adrenal cortex is destroyed, your adrenal glands will not be able to produce enough of the steroid hormones cortisol and aldosterone. Once levels of these start decreasing, you'll experience symptoms of Addison's disease.

It's not clear why some people develop this problem with their immune system, although it can run in families.

Genetics

Research has shown that some people with certain genes are more likely to develop autoimmune disorders.

It's not clear how these genes lead to Addison's disease and similar conditions, but it does mean your risk of developing Addison's disease is increased if you or a close family member have another autoimmune condition, such as:

vitiligo – a long-term condition that causes pale, white patches to develop on the skin
type 1 diabetes – a long-term condition caused by your body not producing insulin

underactive thyroid (hypothyroidism)

Other causes

Tuberculosis (TB) is the most common cause of Addison's disease worldwide, but it's rare in the UK.

TB is a bacterial infection that mostly affects the lungs but can also spread to other parts of your body. It can cause Addison's disease if it damages your adrenal glands.

Other possible causes of Addison's disease include:

infections – such as those linked to AIDS, or fungal infections

a haemorrhage – very heavy bleeding into the adrenal glands, sometimes associated with meningitis or other types of severe sepsis

cancer – if cancer cells from elsewhere in your body spread to your adrenal glands

amyloidosis – a disease where amyloid, a protein produced by your bone marrow cells, builds up in your adrenal glands and damages them

surgical removal of both adrenal glands (adrenalectomy) – for example, to remove a tumour

adrenoleukodystrophy (ALD) – a rare, life-limiting inherited condition that affects the adrenal glands and nerve cells in the brain, and is mostly seen in young boys

certain treatments needed for Cushing's syndrome – a collection of symptoms caused by very high levels of cortisol in the body

Secondary adrenal insufficiency

The production of hormones from the adrenal gland can also be affected by damage to the pituitary gland – a pea-sized gland located below the brain that produces a hormone that stimulates the adrenal gland. This is called secondary adrenal insufficiency and is a separate condition to Addison's disease.

Secondary adrenal insufficiency can occur if your pituitary gland becomes damaged – for example, because of a tumour on the pituitary gland (pituitary adenoma).

Diagnosis

To help diagnose Addison's disease, a GP will first ask about your symptoms and review your medical history.

They're also likely to ask if anyone in your family has an autoimmune disorder (a condition caused by a problem with their immune system).

The GP will examine your skin for any evidence of brownish discolouration (hyperpigmentation), particularly in certain areas, such as:

the creases on your palm

your elbow crease

on any scars

your lips and gums

However, hyperpigmentation does not occur in all cases of Addison's disease.

You'll also be tested for low blood pressure (hypotension) while you're lying down and again shortly after you stand up. This is to see whether you have low blood pressure when you change position (postural or orthostatic hypotension).

Blood tests

If Addison's disease is suspected, blood tests will be carried out to measure the levels of sodium, potassium and cortisol in your body. A low sodium, high potassium or low cortisol level may indicate Addison's disease.

You may need to see a hospital hormone specialist (endocrinologist) for your blood to be tested for the following:

- a low level of the hormone aldosterone

- a high level of adrenocorticotrophic hormone (ACTH)

- a low level of glucose (sugar used for energy)

- positive adrenal antibodies (antibodies designed to attack the adrenal gland)

Any of the above could be a sign of Addison's disease.

Synacthen stimulation test

If cortisol in your blood is low or your symptoms strongly suggest Addison's disease, you'll need to have a synacthen stimulation test to confirm the diagnosis.

The GP may refer you to an endocrinology unit (a unit that specialises in the study of hormones) for the test. How urgent the referral is will depend on how severe your symptoms are.

Synacthen is a manmade (synthetic) copy of the adrenocorticotrophic hormone (ACTH). ACTH is naturally produced by the pituitary gland (a pea-sized gland below the brain) to encourage the adrenal glands to release cortisol and aldosterone.

When synacthen is given, the adrenal glands should respond in the same way as they would to ACTH by releasing cortisol and other steroid hormones into the blood.

A blood sample will be taken and tested for cortisol before an injection of synacthen is given into your arm. Further blood samples will be taken to measure cortisol after 30 minutes and after 60 minutes.

If the ACTH level is high but the cortisol and aldosterone levels are low, it's usually confirmation of Addison's disease.

Thyroid function test

Your thyroid gland may also be tested to see if it's working properly.

Your thyroid gland is found in your neck. It produces hormones that control your body's growth and metabolism.

People with Addison's disease often have an underactive thyroid (hypothyroidism), where the thyroid gland does not produce enough hormones.

By testing the levels of certain hormones in your blood, your endocrinologist (a specialist in hormone conditions) can determine whether you have hypothyroidism.

Scans

In some cases, your specialist may refer you for a scan of your adrenal glands – this could be a CT scan or an MRI scan.

Diagnosis during an adrenal crisis

If Addison's disease is left untreated, it eventually leads to an adrenal crisis. This is where the symptoms of Addison's disease appear quickly and severely.

During an adrenal crisis, there's not enough time to perform a synacthen stimulation test to confirm Addison's disease.

If possible, blood will be taken and tested for any abnormalities. While you're waiting for the results, treatment may be started with steroid injections, and fluids containing salt and glucose.

Read more about treating Addison's disease.

Driving

If you're diagnosed with Addison's disease and have a bus, coach or lorry licence, it's your legal obligation to inform the Driver and Vehicle Licensing Agency (DVLA).

Treatment

If you have Addison's disease, you'll need to take daily medicine to replace the lost hormones. This should help you to live an active life, although many people find they still need to manage their fatigue.

Sometimes, the underlying causes of Addison's disease can be treated. For example, tuberculosis (TB) is treated with a course of antibiotics over a period of at least 6 months.

However, most cases are caused by a problem with the immune system that cannot be cured.

Medicine for Addison's disease

Treatment usually involves corticosteroid (steroid) replacement therapy for life.

Corticosteroid medicine is used to replace the hormones cortisol and aldosterone that your body no longer produces. It's usually taken in tablet form 2 or 3 times a day.

A medicine called hydrocortisone is usually used to replace the cortisol. Other possible medicines are prednisolone or dexamethasone, although these are less commonly used.

Aldosterone is replaced with a medicine called fludrocortisone. Your GP may also ask you to add extra salt to your daily diet, although if you're taking enough fludrocortisone medicine

this may not be necessary. Unlike most people, if you feel the urge to eat something salty, then you should eat it.

Some people take dehydroepiandrosterone (DHEA) to improve their stamina or libido (sex drive). DHEA is not available on the NHS but you may be able to get a private prescription for it and you can buy it from some pharmacies.

If you experience symptoms that you believe may benefit from DHEA, speak to your GP or specialist endocrinologist. They can arrange blood tests and recommend the correct dose for you.

In general, the medicines used for Addison's disease do not have side effects, unless your dose is too high. If you take a higher dose than necessary for a long time, there's a risk of problems such as weakened bones (osteoporosis), mood swings and difficulty sleeping (insomnia).

Living with Addison's disease

Many people with Addison's disease find that taking their medicine enables them to continue with their normal diet and exercise routines.

However, bouts of fatigue are also common, and it can take some time to learn how to manage these periods of low energy.

Some people find that needing to take regular doses of medicine is restrictive and affects their daily life or emotional wellbeing. Missing a dose of medicine, or taking it late, can also lead to exhaustion or insomnia.

Some people can develop associated health conditions, such as diabetes or an underactive thyroid (hypothyroidism), which require extra treatment and management.

You'll usually need to have appointments with an endocrinologist every 6 to 12 months so they can review your progress and adjust your medicine dose, if necessary. Your GP can provide support and repeat prescriptions in between these visits.

Failing to take your medicine could lead to a serious condition called an adrenal crisis, so you must:

- remember to collect your repeat prescriptions

- keep spare medicine as necessary – for example, in the car or at work, and always carry some spare medicine with you

- take your medicine every day at the right time

- pack extra medicine if you're going away – usually double what you would normally need, plus your injection kit (see below)

- carry your medicine in your hand luggage if you're travelling by plane, with a note from your doctor explaining why it is necessary

- carry a steroid emergency card or download one onto your mobile phone

You could also tell close friends or colleagues about your condition. Tell them about the signs of adrenal crisis and what they should do if you have one.

NHS steroid emergency card

If you have Addison's disease, you'll need to take steroids on a long-term basis. Because of this, you'll need to carry a steroid emergency card (for adults) or an adrenal insufficiency card (for children) with you at all times.

The card will make healthcare professionals aware you're taking steroids if you're admitted to hospital in an emergency. This means they'll be able to make sure you get any treatment you need appropriately and quickly.

The card describes the steps that need to be taken in an emergency. It also has a QR code that links to further specialist advice. You can download and carry it with you, or you could use the image of the card as a screensaver on your mobile phone to show healthcare teams in an emergency.

Find out more about steroid emergency cards for adults on the Addison's Disease Self-Help Group (ADSHG) website.

Find out more about the adrenal insufficiency card for children on the British Society for Paediatric Endocrinology website.

Medical alert bracelets

It's also a good idea to wear a medical alert bracelet or necklace that informs people you have Addison's disease.

After a serious accident, such as a car crash, a healthy person produces more cortisol. This helps you cope with the stressful situation and additional strain on your body that results from serious injury. As your body cannot produce cortisol, you'll need a hydrocortisone injection to replace it and prevent an adrenal crisis.

Wearing a medical alert bracelet will inform any medical staff treating you about your condition and what medicine you need.

Medical alert bracelets or necklaces are pieces of jewellery engraved with your medical condition and an emergency contact number. They're available from a number of retailers. Ask your GP if there's one they recommend, or go to the MedicAlert website.

If you need to stay in hospital, the healthcare professionals responsible for your care will also need to know you need steroid replacement medicine throughout your stay. It's important to note that this advice still applies even if you're not supposed to eat or drink (nil by mouth) for any reason.

Adjusting your medicine

At certain times, your medicine may need to be adjusted to account for any additional strain on your body. For example, you may need to increase your medicine dosage if you experience:

an illness or infection – particularly if you have a high temperature

an accident, such as a car crash
an operation, dental or medical procedure – such as a tooth filling or endoscopy
strenuous exercise that's not usually part of your daily life
severe emotional or psychological stress, such as grief
This will help your body cope with the additional stress. Your endocrinologist will monitor your dosage and advise about any changes.

Over time, as you get used to the condition and learn what can trigger your symptoms, you may learn how to adjust your medicine yourself. However, always consult your GP or specialist if you're unsure.

Sick day rules

Your endocrinologist can advise you on "sick day rules". This is when you increase your medicine dose to keep you well during periods of stress, illness or injury. If you do not increase your dose at these times, you may experience an adrenal crisis. Extra doses help your body to respond.

The ADSHG advises you always to have 3 months' supply available of your essential steroid medicine. This is in case you need to increase your dose to follow your sick day rules, or to cover if there is a shortage of your medicine.

Emergency treatment

You and a partner or those you live with should be trained to inject hydrocortisone in an emergency. Your GP or endocrinology team can prescribe the medicine needed for an emergency hydrocortisone injection kit.

This could be necessary if you go into shock after an injury, or if you experience vomiting or diarrhoea and are unable to keep down oral medicine. This may happen if you're pregnant and have morning sickness. Your endocrinologist will discuss with you when an injection might be necessary.

If you need to administer emergency hydrocortisone, always call your GP immediately afterwards. Check what NHS out-of-hours services are available in your local area, in case the emergency is outside normal working hours.

You can also register yourself with your local ambulance service, so they have a record of your requirement for a steroid injection or tablets, if you need their assistance.

Treating adrenal crisis

Adrenal crisis, or Addisonian crisis, needs urgent medical attention.

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Then call 999 for an ambulance, saying it's an "adrenal crisis" or "Addisonian crisis".

Signs of an adrenal crisis include:

severe dehydration
pale, cold, clammy skin
sweating
rapid, shallow breathing
dizziness
low blood pressure (hypotension)
severe diarrhoea and vomiting
abdominal pain or pain in the side
fatigue and severe muscle weakness
headaches
severe drowsiness or loss of consciousness

In hospital, you'll be given lots of fluid through a vein in your arm to rehydrate you. This will contain a mixture of salts and sugars (sodium, glucose and dextrose) to replace what your body is lacking. You'll also be injected with hydrocortisone to replace the missing cortisol hormone.

Any underlying causes of the adrenal crisis, such as an infection, will also be treated.

The Society for Endocrinology website has more information about adrenal crisis

Free prescriptions

If you're receiving treatment for Addison's disease, you're entitled to a medical exemption certificate. This means you do not have to pay for any prescriptions you need.

Check if you can get free prescriptions

Support

Addison's disease will change your life. It needs daily management and support.

It may help you to join patient support groups, such as the Addison's Disease Self-Help Group and The Pituitary Foundation.