Addison's disease
Summary

- Addison's disease is a progressive disorder where the adrenal glands are unable to produce sufficient hormones.
- Causes can include infection, damage, and an autoimmune response that prompts the immune system to attack and destroy the adrenal glands.
- Treatment includes steroid replacement therapy that must be managed for life.
- An Addisonian crisis can be fatal unless treated quickly and appropriately.

Addison's disease (chronic adrenal insufficiency) is a rare and progressive disorder that affects between one and six in every 100,000 people. It affects people of both sexes and all ages.

The human body has two adrenal glands, one on top of each kidney. These glands form part of the endocrine system, which works with the nervous system and the immune system to help the body cope with different events and stresses. Addison's disease is caused by the inability of the adrenal glands to make sufficient amounts of regulating hormones.

Adrenaline is the best known of the hormones that are secreted by the adrenal glands in the adrenal medulla (the central part of the gland). The adrenal cortex (the outer part) also produces important hormones, the corticosteroids. They include cortisol, aldosterone and supplementary sex hormones.

In a person with Addison's disease, only the adrenal cortex is affected. The person cannot produce enough glucocorticoid or cortisol and, occasionally, also fails to produce sufficient mineralocorticoid. Levels of aldosterone are nearly always low in people with Addison’s disease.

Causes of Addison's disease

Most cases of Addison's disease are caused by an autoimmune response that attacks and damages the adrenal glands over time.

Other causes include:
- infection
- cancer
- surgical removal of particular tumours in the adrenal or pituitary glands or the hypothalamus.

Symptoms of Addison's disease

The symptoms of Addison's disease can include any or all of the following:
- loss of appetite and weight
- nausea, vomiting or diarrhoea
- muscle weakness
- chronic, worsening fatigue
- low blood pressure
- salt cravings
- dehydration
- hypoglycaemia, or low blood sugar levels (especially in children)
- increased pigmentation of the skin, particularly around scars and bony areas
- irregular or no menstrual periods in women
- mood swings, mental confusion or loss of consciousness.

These symptoms can develop quickly (especially in children and teenagers), or progress slowly for 20 years or more. Many symptoms can mimic other diseases, so diagnosis can be delayed.

The hormone cortisol

Cortisol is produced by the outer layer of the adrenal gland, called the adrenal cortex. The quantities of cortisol released by the adrenal glands are closely monitored by the master gland of the endocrine system, the pituitary, which is located in the brain.

The workings of the pituitary are governed by another brain structure, the hypothalamus. When cortisol levels are too low, the pituitary secretes the stimulating hormone adrenocorticotropic (ACTH). On the other hand, high levels of cortisol cause the pituitary gland to decrease ACTH secretion, which slows cortisol production.

Cortisol plays many vital roles and is essential to many body functions because it:
- works with adrenaline to help the body manage physical and emotional stress
- converts protein into glucose to boost flagging blood sugar levels
- works in tandem with the hormone insulin to maintain constant blood sugar levels
- reduces inflammation
- helps the body maintain a constant blood pressure
- helps the workings of the immune system.

The hormone aldosterone

Aldosterone is a mineralocorticoid, also produced by the adrenal cortex. The amount of aldosterone in the body is monitored by the kidneys, which secrete hormones to increase or decrease aldosterone production. Aldosterone regulates electrolytes (such as sodium and potassium) in the blood. This helps to maintain blood pressure and heart function.

If too much sodium is excreted by the kidneys, a considerable amount of body fluid is also lost. This reduces blood volume and drops blood pressure. Too much or too little potassium can affect the way the heart functions.

Primary adrenal insufficiency

Addison's disease can occur gradually, and is defined when approximately 90 per cent of the adrenal gland(s) is damaged. This is known as primary adrenal insufficiency. Around
seven out of 10 cases of Addison's disease are caused by an autoimmune response, where the body's own immune cells attack and destroy the adrenal glands. In some cases, other glands of the endocrine system are affected by an autoimmune response, in a condition called polyendocrine deficiency syndrome.

**Polyendocrine deficiency syndrome**

There are two types of primary adrenal insufficiency and both types tend to run in families. They are:

- **Type I** – is more common in children. Symptoms include underactive parathyroid, pernicious anaemia, recurring candida infections, chronic active hepatitis and slow sexual development.
- **Type II** (Schmidt's syndrome) is more common in younger adults. Symptoms include underactive thyroid, type 1 diabetes and, less commonly, vitiligo (a skin condition).

Other conditions related to primary Addison's disease are:

- **Adrenomyeloneuropathy (AMN)** – which can occur in some adults. It affects the spine and is degenerative over time.
- **Adrenoleukodystrophy (ALD)** – occurs (rarely) in some children (one in 100,000), especially males. It can cause brain damage and can be fatal. Survivors often develop AMN.

Treatment for primary Addison's disease is with glucocorticoid and mineralocorticoid replacement for life.

**Secondary adrenal insufficiency**

Sometimes, Addison's disease is caused by the pituitary gland's inability to produce sufficient amounts of ACTH, which means the adrenal glands aren't prompted to secrete cortisol. This is known as secondary adrenal insufficiency.

Causes of secondary adrenal insufficiency may include:

- **Some medications** – inflammatory disorders such as rheumatoid arthritis and asthma are often treated with prolonged or high-dose steroids (glucocorticoid replacements). If the dose is suddenly stopped, or not reduced by appropriate tapering measures, the pituitary gland may respond by failing to produce enough ACTH. This situation can sometimes be reversed.
- **Cushing's disease** – a benign tumour of the pituitary gland that produces ACTH. This results in too much cortisol being produced. Treatment requires surgical removal of the tumour and, in some cases, removal of damaged adrenal gland(s).
- **Other causes** – infections, reduced blood flow, radiotherapy and some neurosurgery can damage the pituitary gland or hypothalamus, and decrease the ability to produce ACTH.

Treatment for secondary Addison's disease is with glucocorticoid replacement only.

**Addisonian crisis**

A sudden, acute worsening of symptoms is known as an Addisonian crisis. If untreated, an Addisonian crisis can be fatal.

It can be caused by:

- **extreme stress** – an accident, excessive heat or physical exertion
- **severe illness** – especially dehydration from vomiting or diarrhoea
- **sudden shock** – for example, the death of a significant person.

The symptoms of Addisonian crisis include:

- violent pain in the abdomen, back and legs
- nausea, vomiting or diarrhoea
- low blood pressure, low blood sugar, high potassium, low sodium and a rapid heart rate
- possible mental confusion and loss of consciousness.

Prompt emergency hospital treatment must be sought, including intravenous fluids, increased steroid medication and saline. Many people who suffer from Addison’s wear a medical alert bracelet or pendant with information and identification, and carry a hydrocortisone injectable for use in emergencies. They will still need hospitalisation and ongoing monitoring.

**Diagnosis of Addison's disease**

Diagnosis may involve:

- a complete detailed family history, with special attention to any other endocrine disorders
- biochemical tests, which measure cortisol levels before and after a challenge injection of synthetic ACTH, known as a 'short synacthen test'. Synacthen tests will indicate the person's baseline level of cortisol production and their response to an increased need for cortisol in the body. An person who suffers from Addison’s may show a flat or reduced response
- blood electrolyte and plasma renin tests, which will indicate if there is a need for mineralocorticoid replacement
- **anti-adrenal antibody test** – if the result is positive, primary Addison's disease is definitively diagnosed. However, even if these antibodies do not exist, the person may still have Addison's disease
- **x-rays, ultrasounds and CAT scans** of the abdominal region to check for visual signs of damage and the size of adrenal glands.

**Treatment of Addison's disease**

Treatment aims to boost or replace insufficient or absent steroid components. Glucocorticoid replacement is essential for people suffering from primary or secondary Addison's, and must be taken for life.

Treatment should:

- be tailored to each person over the course of their life
- be altered, in consultation with a doctor, during illness or other stressful events
- allow for the different needs of children and young adults.

**Where to get help**

References

- Badash M, Addison's Disease (Adrenal insufficiency, adrenocortical hypofunction, chronic adrenocortical insufficiency, hypoadrenalinism), FindLaw, Nucleus Medical Media Inc, USA.
- Margulies P, North American Survey of Individuals with Addison’s Disease (1997), National Adrenal Diseases Foundation, USA.
- Adrenal insufficiency and Addison’s disease, National Institute of Diabetes and Digestive and Kidney Diseases (NIDDK), National Institutes of Health, USA.
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More information

Immune system

The following content is displayed as Tabs. Once you have activated a link navigate to the end of the list to view its associated content. The activated link is defined as Active Tab

- Immune system explained
- Lymphatic system
- Spleen
- Lupus
- Autoimmune disorders

Immune system explained

- Immune system explained
  The immune system remembers every germ it has ever overcome.
- Vaccines
  Vaccines trick the body into building immunity against infectious diseases without causing the actual disease.

Lymphatic system

- Fluid retention (oedema)
  Fluid retention (oedema) occurs when fluid isn’t removed from the body tissues, including the skin. Causes include the body's reaction to hot weather, a high salt intake, and the hormones associated.
- Lymphatic system
  The lymphatic system manages fluid levels in the body, filters out bacteria and houses types of white blood cells.
- Lymphoedema
  Women who have undergone treatment of breast cancer are particularly susceptible to lymphoedema of the arm.
- Lymphoma
  Lymphoma is a general term for a cancer that begins in the lymphatic system.

Spleen

- Spleen
  Surgically removing a diseased or damaged spleen is possible without causing any serious harm to the person.
- Splenomegaly
  Any conditions that cause a rapid breakdown of blood cells can place great strain on the spleen and make it enlarge.

Lupus

- Lupus
  Lupus can be mild or life-threatening, depending on which tissues are affected.
- Lupus and infections
  The most common infections for people with lupus include those of the respiratory tract, skin and urinary system.
- Lupus and medication
  Lupus most commonly appears in women of childbearing age.
- Lupus and pregnancy
  Lupus can be controlled with medications, so the majority of affected women are able to have children.

Autoimmune disorders

- Addison’s disease
  Most cases of Addison’s disease are caused by an autoimmune response that attacks and damages the adrenal glands over time.
- Autoimmune disorders
There is generally no cure for an autoimmune disorder, but the symptoms can be managed...

- **Diabetes type 1**
  Type 1 diabetes can affect anyone of any age, but is more common in people under 30 years...

- **Guillain-Barré syndrome**
  Most people with Guillain-Barré syndrome experienced some form of viral or bacterial infection before the onset of symptoms...

- **Henoch-Schonlein purpura**
  Henoch-Schonlein purpura causes a purple spotted skin rash which lasts around one to four weeks, and is often marked by relapses...

- **HIV**
  In Australia, HIV is most commonly spread when having sex without a condom and when sharing needles and other injecting equipment...

- **Hughes syndrome**
  Hughes syndrome is thickening of the blood caused by abnormal immune system cells...

- **Idiopathic thrombocytopenic purpura (ITP)**
  Idiopathic thrombocytopenic purpura (ITP) is a rare autoimmune disorder in which a person's immune system destroys the platelets that help their blood to clot...

- **Lipoedema**
  Lipoedema is a painful, chronic, symmetrical swelling in the legs, thighs, buttocks and sometimes arms due to the accumulation of fat in the subcutaneous tissues. The onset often occurs during puberty...

- **Myasthenia gravis**
  Myasthenia gravis is an autoimmune disease that causes muscle weakness...

- **Polymerosis**
  Polymerosis is hard to diagnose and may be mistaken for muscular dystrophy...

- **Raynaud's phenomenon**
  Raynaud's phenomenon can be a sign of a more serious underlying condition, so see your doctor if you experience it...

- **Reactive arthritis**
  Reactive arthritis is a form of arthritis that occurs as a result of some bacterial infections...

- **Retroperitoneal fibrosis**
  Retroperitoneal fibrosis is the abnormal growth of tissue on and around abdominal structures, including blood vessels and ureters...

- **Rheumatoid arthritis**
  Early treatment of rheumatoid arthritis is important in helping you manage the condition more effectively...

- **Scleroderma**
  The most common symptom of scleroderma is a thickening and hardening of the skin, particularly of the hands and face...

- **Sjogren’s syndrome**
  Sjogren’s syndrome can be managed with medications and products such as artificial tears and saliva...

- **Thyroid - Hashimoto’s disease**
  Hashimoto's disease progresses very slowly over many years, so the symptoms may go unnoticed...

- **Thyroid - hyperthyroidism**
  Hyperthyroidism can be diagnosed with a simple blood test that measures thyroid hormone levels...

**Related Information**

- **Cushing's syndrome**
  Some people have Cushing's syndrome symptoms when they take glucocorticoid hormones to treat inflammatory conditions such as asthma, lupus or rheumatoid arthritis...

- **Sjogren’s syndrome**
  Sjogren’s syndrome can be managed with medications and products such as artificial tears and saliva...

- **Retroperitoneal fibrosis**
  Retroperitoneal fibrosis is the abnormal growth of tissue on and around abdominal structures, including blood vessels and ureters...

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Hormones – cortisol and corticosteroids

Cortisol helps to maintain blood pressure, immune function and the body’s anti-inflammatory processes...

Related information on other websites

- Australian Addison's Disease

Content Partner

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How to cut down on salt

1. Many of us are eating almost twice the amount of salt (sodium) that we need for good health, largely because it’s hidden in many of the foods we eat. A whopping three-quarters of the salt we eat comes from processed foods. Too much salt can lead to high blood pressure, which puts us at risk of stroke, heart disease and chronic kidney disease. Try these simple tips and cut down your intake.
2. **Limit bought food**

Food eaten at restaurants, cafes and from takeaway outlets can be much higher in salt than food prepared at home. The best bet is to make them occasional treats rather than part of your regular diet. They’re also high in kilojoules, saturated fat and sugars. And if you think that sweets are immune from high levels of salt, think again – muffins, donuts and pancakes with maple syrup contain more than 400 mg, which is above ‘safe’ levels. Don’t be fooled by healthier options either – a Caesar salad bought over the counter may contain more salt than the average cheeseburger!

3. **Read food labels**

The amount of salt in foods can vary considerably between brands. Look for products with ‘no-added salt’, or at least ‘reduced’ or ‘low salt’. The best choices are those with less than 120 mg of sodium per 100 g. If you have kids, watch how much they’re consuming – too much salt in childhood can lead to a lifetime of health risks. If you need help choosing foods, check out the government’s Health Star Rating – the more stars a product has the healthier it is.

Hint: when using canned vegetables, choose ones with ‘no-added salt’.
4. **Ditch the salt shaker**

Try to break the habit of automatically salting your meals at the table. And when cooking, don’t add salt. A better option is to add [herbs](#), a splash of [olive oil](#), citrus juice or zest to enhance flavours. Spices such as pepper or cumin are great alternatives too.

Hint: grow some [thyme](#) in a pot it adds flavour to soups, stews, and meat.

5. **Get clever in the kitchen**

Don’t add salt to food when cooking. Instead, give your meals a boost with [herbs](#) (e.g. basil, coriander), spices (e.g. cumin, pepper), fresh ginger, [garlic](#), [chilli](#), [vinegar](#) and [lemon](#) juice or zest. If it’s hard to cut out salt altogether, do it slowly and introduce new flavours gradually. Seasonings are high in salt too – use low or reduced-salt stocks, stock powder and gravy powder and dilute them more than the instructions recommend. When cooking pasta, noodles, rice or vegetables don’t add salt to water.

Hint: don’t add the amount of salt stated in recipes – try adding half or less. If a recipe includes other salty ingredients like stock powder, Asian sauces, olives, capers, anchovies, bacon, ham or smoked salmon, then you shouldn’t need extra salt.
Think you can taste it?

As consumers, we put a lot of trust in businesses who make our food. But when it comes to salt, can you trust your tastebuds? Foods don’t have to taste salty to be high in salt. Take control and get into the habit of reading labels.

Convenience a no go

In this busy world, a lot of us are looking for ways to spend less time in the kitchen and that’s why convenience products and packaged foods are so popular. Most of these foods are high in salt and best avoided. Cut back on things like – deli meats, flavoured instant pasta, instant noodles, savoury snack foods and crackers, instant cup-a-soups, dehydrated soup mixes and ‘heat and eat’ frozen or refrigerated meals. Use the government’s Health Star Rating to choose lower sodium products. Even better, cut down the amount of these products you’re consuming and go fresh – it really is best!

All salt is the same
Don't be fooled by trendy claims – all salt is the same. Whether it's pink, rock or vegetable, it's still salt, and has the same effect on our health. Any salt contains sodium, and is just the same as ordinary table salt.

9. **How much is too much?**

We need sodium to regulate fluids in our body – most of us can get it naturally in our diet without it being added. We only need a small amount each day but many of us are eating way more. **Adults should eat less than 2,000 mg (or 5 g) of sodium – about a teaspoon of salt a day. Children need less than one quarter of a teaspoon of salt per day.** The adequate intake of sodium for children starts at around 0.5 g salt (1–3 year olds) to 2 g salt (9–13 year olds).

Hint: got **high blood pressure** – reduce your salt intake to 2,000 mg (5 g) per day. And don’t forget to **talk with your doctor or health practitioner** about what’s right for you.

10. **It's in the sauce**

Most simmer sauces, gravies and condiments are stacked with salt to enhance their flavour. A squirt of sauce, a drizzle of dressing on a salad can really start to add up. Popular products that contain loads of salt include soy sauce, tomato sauce, relish, mustard, stir-fry sauces, marinades, stock, gravy, mayonnaise and salad dressings. Anything pickled such as olives, gherkins and capers is high in salt too.

Hint: try to make your own with fresh ingredients when you can. Start with **our homemade tomato sauce** using fresh tomatoes – make up a batch and freeze. It’s great with
11. Natural? Think again

Even though a label may say ‘natural’ doesn’t mean food is healthy - it could contain loads of salt. Download a free app like FoodSwitch, which scans barcodes and gives the nutritional value of loads of foods. If you have kids, get them to help with the shopping. They can search for healthy alternatives using your phone - it’s great for starting healthy habits early in their lives. The best way to ensure we’re eating ‘natural’ foods is to eat plenty of fruit, vegetables and wholegrains.

12. Don’t listen to those cravings

Salt has a craving effect on our bodies – the more we eat, the more we want. And, for some of us, it makes us eat more too. Try to cut down slowly, so you're less likely to crave ‘bad things’, such as junk and fast foods! Don’t be disheartened, foods might taste different from the start, but our taste buds will adjust – it just takes time. So stick with it. If you find you are craving excessively, see a doctor – it could be a sign of an underlying medical condition, such as Addison’s disease.
Surely not breakfast!

Believe it or not, breakfast cereals can be high in salt, and that toast we’re eating could be too. When shopping, read the nutritional information and make sure to look for low sodium products. Your best bet is to choose cereals that are less likely to contain added salt such as oats or unsweetened muesli. If you have time, make your own Bircher muesli - it’s cheaper, healthier and filling!

Time for brunch or lunch?

A popular lunch staple is the humble sandwich or roll filled with deli meats. Even if you make your own lunch you may not know exactly how much salt you’re actually consuming. Limit your intake of deli meats that are high in salt such as salami, ham, corned beef, and chicken loaf. Go easy on those Sunday brunches too – smoked salmon, sausages, bacon and prosciutto are high in salt – just one rash of bacon alone is a third of our maximum daily intake! Be on the lookout for brands that are lower in sodium.

Hint: vary fillings in lunches – such as tuna in spring water (it’s less salty than brine), hard-boiled eggs, or patties and dips made with legumes (e.g. falafels or hummus.)
15. **Converting sodium into salt**

If you’re still confused by food labels, try this simple calculation to work out how much salt is in a product:

Multiply the amount of sodium (in mg) by 2.5

*For example: 400 mg sodium in a serve of food X 2.5 = 1,000 mg (or 1 gram) of salt*

16. **Cramps don’t need salt**

Contrary to what we might believe, if your muscles are exercising, they need water not salt. Cramps are a sign that our body is dehydrated and needs replenishing. Drink plenty of water before, during and after exercise, and especially on hot days.
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