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## Addison's disease

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### Summary

- Addison's disease or 'primary adrenal insufficiency' is a condition where the adrenal glands are unable to produce enough 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.
  - A sudden, acute worsening of symptoms is known as an Addisonian crisis.
  - An Addisonian crisis can be fatal unless treated quickly and appropriately.
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Addison's disease (primary adrenal insufficiency) is a rare and progressive (worsening) disorder that affects one in every 10,000 people. It affects people of 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 your body cope with different events and stresses. Addison's disease is caused by the inability of the adrenal glands to make enough regulating hormones.

Adrenaline is the best known of the hormones that are made by the adrenal glands in the adrenal medulla (the central part of the gland). The adrenal cortex (the outer part) also makes important hormones, the corticosteroids. They include cortisol, aldosterone and supplementary sex hormones. If you have Addison's disease, it is mainly your adrenal cortex that is affected. You cannot produce enough cortisol and, usually, also cannot produce enough aldosterone.

### 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 – 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 over years. Many symptoms can mimic other diseases, so diagnosis can be delayed.

### The hormone cortisol

Cortisol is produced by the outer layer of your adrenal gland, called the adrenal cortex. The amount of cortisol released by your adrenal glands is closely monitored by the master gland of your endocrine system, the pituitary, which is located below the brain in the base of your skull. The workings of the pituitary are governed by a brain structure called the hypothalamus.

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When cortisol levels are too low, the pituitary secretes the stimulating hormone adrenocorticotropin (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 your body manage physical and emotional stress
- increases and influences your blood sugar levels
- works with the hormone insulin to maintain constant blood sugar levels
- reduces inflammation
- helps your body maintain a constant blood pressure
- helps the workings of your immune system.

### **The hormone aldosterone**

Aldosterone is a mineralocorticoid, produced by the adrenal cortex. The amount of aldosterone in your body is monitored by your 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 your kidneys excrete too much sodium, you will also lose a considerable amount of body fluid. This reduces your blood volume and makes your blood pressure drop. Too much or too little potassium can affect the way your heart functions.

### **Causes of Addison's disease**

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. This is known as '**primary adrenal insufficiency**' (or primary Addison's disease).

Other causes of Addison's disease include:

- infection of the adrenal glands
- spread of cancer into the adrenal glands
- surgical removal of particular tumours in 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**. There are two types of polyendocrine deficiency syndrome – Type I and Type II. Both types tend to run in families.

**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**, also known as 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 adrenal deficiency are:

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

Treatment for primary adrenal deficiency is with glucocorticoid (cortisol) and mineralocorticoid (aldosterone) replacement, for life.

### **Secondary adrenal insufficiency**

Secondary adrenal insufficiency is different from primary adrenal insufficiency (Addison's disease). It is caused

when the pituitary gland is unable to produce enough ACTH (adrenocorticotropic hormone), which means the adrenal glands aren't prompted to secrete cortisol.

Hypothalamic disease may also lead to adrenal insufficiency. This is known as tertiary adrenal insufficiency, although often pituitary and hypothalamic causes are sometimes referred to together as secondary adrenal insufficiency.

Causes of secondary adrenal insufficiency may include:

- **glucocorticoid 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 ACTH-producing tumour of the pituitary gland. This results in too much cortisol being produced. Treatment requires surgical removal of the tumour. The pituitary gland may take months to begin production of ACTH once the tumour has been removed
- **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 (cortisol) replacement only.

### **Addisonian crisis**

A sudden, acute worsening of symptoms of Addison's disease is known as an Addisonian crisis. The risk of Addisonian crisis in people with Addison's disease or most cases of secondary adrenal insufficiency is six to eight per cent per year.

If untreated, an Addisonian crisis can be fatal. It may be caused by:

- illness – especially fever or gastroenteritis
- surgery
- interruption of adrenal hormone replacement medications.

The symptoms of Addisonian crisis include:

- nausea, vomiting or diarrhoea
- dizziness when standing
- palpitations (the feeling of having a fast-beating, fluttering or pounding heart)
- pain in the abdomen, back and legs
- mental confusion and loss of consciousness.

Not all symptoms occur on all occasions, but nausea, fatigue and dizziness are common early symptoms.

**If you experience symptoms of Addisonian crisis, call 000 for an ambulance or go immediately to the emergency department of your nearest hospital. You will need emergency treatment, including intravenous fluids, increased steroid medication and saline.** Blood tests may show low sodium, high potassium or low glucose.

If you have Addison's disease it may be helpful to wear a medical alert bracelet or pendant with identification and information about your condition, and to carry a hydrocortisone injectable for use in emergencies. Note: even if you use a hydrocortisone injection for Addisonian crisis, you will still need hospitalisation and ongoing monitoring.

### **Diagnosis of Addison's disease**

Diagnosis may involve:

- a complete detailed medical history which, among other symptoms, may reveal recent onset of excessive pigmentation in sun exposed areas, skin creases, scars and inside the mouth (this is common in Addison's disease but not secondary adrenal insufficiency)
- biochemical tests, which measure cortisol levels before and after a challenge injection of synthetic ACTH,

known as a 'short synacthen test'. Synacthen tests will show your baseline level of cortisol production and your body's response to an increased need for cortisol. If you have Addison's disease this test will show a flat or reduced response

- blood electrolyte and plasma renin tests, which will show if you need mineralocorticoid (aldosterone) replacement
- anti-adrenal antibody test – if the result is positive, it is very likely that you have autoimmune (primary) Addison's disease. However, even if you do not have these antibodies, you may still have Addison's disease
- x-rays, ultrasounds and CAT scans of your abdominal region to check for visual signs of damage and the size of your adrenal glands.

### **Treatment of Addison's disease**

Treatment aims to correct the levels of hormones that your body is not producing. Whether you have primary or secondary adrenal insufficiency, you will need hormone replacement for life.

Cortisol is replaced orally with hydrocortisone tablets, taken once or twice a day. If you are also deficient in aldosterone, it is replaced with oral doses of a mineralocorticoid called fludrocortisone acetate, taken once a day.

Treatment needs to be tailored to each person as adrenal hormone replacement requirements vary between individuals.

Your doctor will advise you on how to reduce the risk of Addisonian crisis, including how to adjust your medication if your body is stressed (for example due to an operation or illness) and how to use injectable hydrocortisone if you are vomiting and unable to keep down oral medications.

They will also advise you on how to let emergency medical personnel know what kind of care you need. This may include:

- wearing a MedicAlert bracelet or pendant
- carrying a card that tells emergency responders:
  - that you need a cortisol injection if you are found severely injured or unable to answer questions
  - your doctor's name and telephone number
  - who to notify in an emergency.

### **Where to get help**

- Your **GP (doctor)**
- **Australian Addison's Disease Association Inc** Tel. **0455 534 472**
- **Australian Pituitary Foundation** Tel. **1300 331 807**

**This page has been produced in consultation with and approved by:**

Australian Addisons Disease Association

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