

ADDISON'S DISEASE

Patient Information Fact Sheet

What is Addison's Disease?

Addison's disease, also known as primary or chronic adrenal insufficiency, is a relatively rare disorder in which the adrenal glands (located just above the kidneys) are unable to produce sufficient hormones. Cortisol is the hormone that is usually lacking, but sometimes another hormone called aldosterone is also affected.

Cortisol plays a vital role in many bodily functions, the most important being that of helping the body to respond to stress. It also aids in other processes, including maintaining blood pressure, slowing the immune system's inflammatory response, balancing the effect of insulin breaking down sugar, and regulating the metabolism of proteins, carbohydrates, and fats. The amount of cortisol produced by the adrenal glands is usually in fine balance, but in Addison's disease this process is disrupted and lower levels are produced. Cortisol production is controlled by two glands: the adrenal gland and the pituitary gland. Aldosterone acts on the kidney and regulates the excretion of sodium and potassium in the urine. Addison's disease is thought to affect 1 to 4 people per 100,000.

What are the symptoms of Addison's disease?

The symptoms of Addison's disease usually appear gradually. Chronic fatigue and muscle weakness, loss of appetite, and weight loss are all symptoms of the disease. In around half of people, nausea, diarrhea and vomiting occur. If aldosterone levels are low, blood pressure will drop and may cause dizziness or fainting. In addition, the skin may become pigmented (darkened) in skin folds, elbows, knees, knuckles, and joints. The pigmentation may be more noticeable on areas of scarring or on the lips. A craving for salty foods may occur as a result of salt loss, and irritability and depression may also be present. In women, menstrual periods may become irregular or stop altogether. Hypoglycemia (low blood sugar) may also occur. This may be more severe in children than in adults. An Addisonian crisis may be triggered by a stressful event such as an illness or an accident. In about 25% of people with Addison's disease, a crisis is the first appearance of symptoms; however, in the remainder of people, the symptoms are usually severe enough to cause them to seek medical advice before a crisis occurs. The symptoms of an Addisonian crisis are exaggerated symptoms of Addison's disease and include pain in the lower back, abdomen or legs, and severe vomiting and diarrhea. This is followed by dehydration, low blood pressure, and loss of consciousness, all of which may occur suddenly. If left untreated, an Addisonian crisis can be fatal.

What causes Addison's disease?

Addison's disease may be caused by a disorder of the adrenal glands (primary adrenal insufficiency). The disease may also be caused if the pituitary gland stops producing



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enough adrenal stimulating hormone (secondary adrenal insufficiency). In most cases, primary adrenal insufficiency occurs as a result of the body's own immune system slowly destroying the outer layer of the adrenal glands. This form of Addison's is what is known as an autoimmune disease and accounts for about 80% of cases. In developed countries, tuberculosis (TB) accounts for about 20% of cases of primary adrenal insufficiency when the adrenal glands are destroyed by the infection.

When Dr. Thomas Addison first identified the disease in 1849, TB was the most common cause of the disease. Other less common causes of primary adrenal insufficiency are chronic infections, cancer cells that spread to the adrenal glands from other parts of the body, and surgical removal of the adrenal glands. Secondary adrenal insufficiency is caused by a reduction in the amount of adrenal stimulating hormone (ACTH) produced by the pituitary gland. In most cases, secondary adrenal insufficiency is caused by a pituitary tumor. It may also occur when high doses of steroids are used for long periods to treat other diseases such as asthma, causing temporary or permanent loss of adrenal function.

What tests confirm a diagnosis of Addison's disease?

An ACTH stimulation test is used to diagnose Addison's disease. For this test, levels of cortisol in the blood and/or urine are measured before and after an intravenous injection (into a vein) of a synthetic form of adrenal stimulating hormone. Cortisol levels are measured 30–60 minutes after the injection. Levels should rise during this time but, if there is adrenal insufficiency, there may be little or no rise in cortisol levels. If the results of this test are abnormal, a longer test will be carried out. Your doctor will explain these tests to you. Another test that may be used is an insulin-induced hypoglycemia test. For this test, blood glucose and cortisol levels are measured, and an injection of a fast-acting insulin is then administered. The levels are measured again at 30, 45, and 90 minutes following the injection. The normal response is for blood glucose levels to fall and cortisol levels to rise. Secondary adrenal insufficiency may be caused by a pituitary problem so a CT scan of the pituitary gland may be ordered.

How is Addison's disease treated?

Treatment of Addison's disease involves therapy to replace the hormones that are not being produced by the adrenal glands. Cortisol is replaced with tablets containing a synthetic steroid called **hydrocortisone** (Cortef). If there is also a deficiency of aldosterone, tablets containing another steroid such as **fludrocortisone acetate** can be given. People with Addison's disease should always carry some identification stating their medical condition so that a cortisol injection can be given in an emergency. Some people wear a bracelet or neck chain containing relevant information. When traveling, it is advisable for those with Addison's disease to carry an emergency syringe and an injectable form of cortisol. Increase your medication for Addison's disease during periods of stress or mild respiratory infections to prevent complications of the disease.



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If severe infections occur, or if diarrhea and vomiting occur (meaning that the oral tablets are not being absorbed properly), medical attention must be sought promptly.

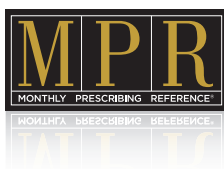
Further Information

National Adrenal Diseases Foundation: <http://www.nadf.us/diseases/addisons.htm>

National Institutes of Health: <http://www.ncbi.nlm.nih.gov/pubmedhealth/PMH0001416/>

National Endocrine and Metabolic Diseases Information Service: <http://endocrine.niddk.nih.gov/pubs/addison/addison.aspx>

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