

ADDISON'S DISEASE

• THE FACTS YOU NEED TO KNOW •

Provided as a service by:

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Diseases Foundation, Inc.**

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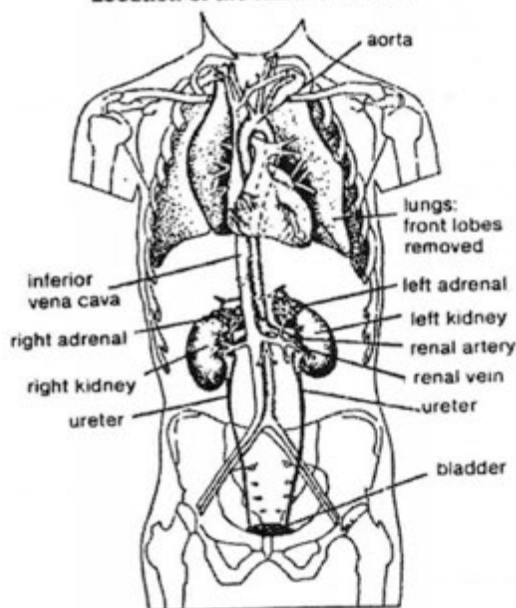
National Adrenal Diseases Foundation

WHAT IS ADDISON'S DISEASE?

Addison's disease is a severe or total deficiency of the hormones made in the adrenal cortex, caused by its destruction. There are normally two adrenal glands, located one above each kidney. The adrenal glands are really two endocrine (ductless or hormone producing) glands in one. The inner part of the adrenal gland (called the medulla) produces epinephrine (also called adrenaline) which is produced at times of stress and helps the body respond to "fight or flight" situations by raising the pulse rate, adjusting blood flow, and raising blood sugar. However, the absence of the adrenal medulla and epinephrine does not cause disease.

In contrast, the outer portion of the adrenal gland, the cortex, is more critical. The adrenal cortex makes two important steroid hormones, cortisol and aldosterone. Cortisol mobilizes nutrients, modifies the body's response to inflammation, stimulates the liver to raise the blood sugar, and also helps to control the amount of water in the body. Aldosterone regulates salt and water levels which affects blood volume and blood pressure. Cortisol production is regulated by another hormone, adrenocorticotrophic hormone (ACTH), made in the pituitary gland which is located just below the brain. Classical Addison's disease results from a loss of both cortisol and aldosterone secretion due to the near total or total destruction of both adrenal glands. This condition is also called primary adrenal insufficiency. If ACTH is deficient, there will not be enough cortisol produced, although aldosterone may remain adequate. This is secondary adrenal insufficiency, which is distinctly different, but similar to Addison's disease, since both include a loss of cortisol secretion.

Location of the Adrenal Glands



WHAT CAUSES ADDISON'S DISEASE?

When Dr. Thomas Addison first described this disease in London in 1855, the most common cause was tuberculosis. This remained the leading cause until the middle of the twentieth century when antibiotics progressively reduced TB's incidence. Since then, the major cause of Addison's disease results from an auto-immune reaction in which the body's immune system erroneously makes antibodies against the cells of the adrenal cortex and slowly destroys them. That process takes months to years. There are also several less common causes of Addison's disease: other chronic infections besides tuberculosis, especially certain fungal infections, invasion of the adrenal by cancer cells that have spread from another part of the body, especially the breast; CMV virus in association with AIDS; rarely, hemorrhage into the adrenals during shock; and the surgical removal of both adrenals.

HOW IS ADDISON'S DISEASE TREATED?

Since all of the manifestations of Addison's disease are caused by the lack of cortisol and aldosterone, the treatment is to replace these with similar steroids. Cortisol is usually replaced orally by hydrocortisone or cortisone acetate, less often with prednisone tablets, divided into morning and afternoon doses. Aldosterone is replaced by an aldosterone-like synthetic steroid, fludrocortisone (Florinef) tablets given once daily. The doses of each of these medications are adjusted for the individual's size and any co-existing medical conditions. In emergencies or during surgery, hydrocortisone must be given intravenously. Patients with Addison's disease should be taught to treat minor illnesses with extra salt, fluids and extra hydrocortisone. This is especially important if fever, vomiting or diarrhea is present. Persistence of these signs requires immediate treatment in an emergency room with intravenous saline (salt water) and hydrocortisone. Since Addison's disease is a chronic condition, daily replacement medication can never be stopped.

Proper maintenance treatment requires regular visits to a physician for examinations, laboratory tests, and discussions about symptoms. Certain blood tests, including sodium, potassium, blood counts and plasma renin are very useful in monitoring the response to adjustments in dosage. There is no single blood or urine test that is perfect by itself.

WHY SHOULD ADDISONIANS CONSULT AN ENDOCRINOLOGIST?

Endocrinologists are specialists in hormonal diseases, including Addison's disease. Because of the rarity of the disease, an endocrinologist will have more training and experience in properly diagnosing and treating Addison's disease than most physicians.

ARE THERE RELATED DISEASES?

Secondary adrenal insufficiency, caused by a lack of ACTH, results in a deficiency of cortisol, but usually not aldosterone. The cause is either pituitary disease, such as a tumor, or the prolonged use of "steroid" medication that suppresses ACTH. The treatment is simply to replace cortisol, usually with the synthetic steroid prednisone, but sometimes hydrocortisone or cortisone acetate.

Auto-immune Addison's disease, the most common type, can be associated with other auto-immune diseases that similarly affect other endocrine glands. The most common one is the thyroid. If an underactive thyroid (hypothyroidism) coexists with Addison's disease, this is called Schmidt's syndrome. Less commonly associated auto-immune diseases are insulin-dependent diabetes mellitus, and insufficiencies of the parathyroid glands, gonads, and vitamin B12 absorption (pernicious anemia).

WHY IS IT CONSIDERED A RARE DISORDER?

There are no accurate statistics on the incidence of Addison's disease in the United States. A study in London showed thirty-nine cases per million population as of 1960. Twelve were due to tuberculosis. In the non-tuberculosis group, women were three times more likely to have Addison's disease. Extrapolation of these figures to the U.S. would give about 8,800 cases, but this is probably an underestimation.

WHAT ARE THE SYMPTOMS OF ADDISON'S DISEASE?

The slowly progressive loss of cortisol and aldosterone secretion usually produces a chronic, steadily worsening fatigue, a loss of appetite, and some weight loss. Blood pressure is low and falls further when a person is standing, producing lightheadedness. Nausea, sometimes with vomiting, and diarrhea are common. The muscles are weak and often go into spasm. There are often emotional changes, particularly irritability and depression. Because of salt loss, a craving for salty foods is common. Finally, the increase in ACTH due to the loss of cortisol will usually produce a darkening of the skin that may look like an inappropriate tan on a person who feels very sick. Unfortunately, the slowly progressive chronic symptoms are usually missed or ignored until a sudden event like a flu virus, an accident, or the need for surgery suddenly precipitates a dramatic change for the worse because of the deficient response from the adrenals to one of these stresses. This is referred to as an Addisonian crisis and is a medical emergency.

HOW IS ADDISON'S DISEASE DIAGNOSED?

A medical history of the symptoms mentioned above, especially hyperpigmentation of the skin or gums, is often enough to raise a strong suspicion, prompting the appropriate tests. Quite often, however, the first clue is from the abnormal results of routine tests done in a hospital or doctor's office. These may include an elevated blood level of potassium, a low blood level of sodium, a shift in the ratio of certain white blood cells, or surprising changes on an EKG or chest x-ray that are caused by high potassium or low blood volume. Other causes for these changes, particularly from medications, must be considered first. A definitive diagnosis of Addison's disease requires that definitive tests be carried out. These tests measure the amount of cortisol and aldosterone in the blood and urine, and document a lack of the normal increase in the levels of these two hormones after administration of ACTH given by injection. An elevated blood level of ACTH should also be found. If the patient is very sick and Addison's disease is suspected, treatment can be initiated while the diagnostic tests are being done. Once the diagnosis of Addison's disease is established, an effort should be made to find the cause by checking for tuberculosis and other infections through skin tests and x-rays. Antibodies to adrenal tissue, especially to 21-OH can now be measured, and are specific to autoimmune Addison's disease.



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HOW NORMAL IS AN ADDISONIAN'S LIFE?

As long as the proper dose of replacement medication is taken every day, an Addisonian can have a normal crisis-free life. There are no specific physical or occupational restrictions. Routine care includes regular physician visits, avoidance of dehydration, and the use of extra medication during illness. Pregnancy is possible, but will require extra monitoring of the replacement medication. Every Addisonian should wear an identification bracelet or preferably a necklace stating that he or she has the disease, to insure proper emergency treatment. An identification card outlining treatment is also suggested. Today, people with Addison's disease should have a normal life expectancy.

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