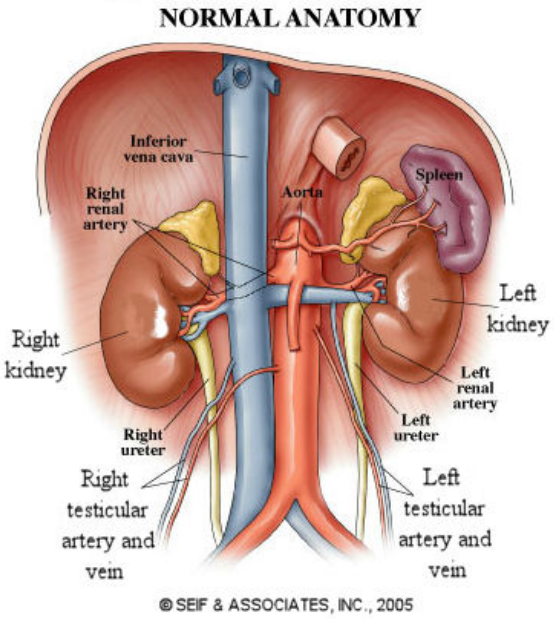
Addison's Disease, Adrenal Insufficiency

Addison's disease is a glandular (*endocrine*) or hormonal disorder. It is also called adrenal insufficiency or hypocortisolism. It affects about 1 in 100,000 people. It can affect men and women in all age groups. This disease occurs when the adrenal glands do not make enough of the hormone **cortisol**. In some cases, the adrenal glands also do not make the hormone **aldosterone**. Without the right levels of these hormones, your body cannot maintain critical life functions.



The adrenal glands are located just above the kidneys. Cortisol is a steroid hormone. Its most important job is to help the body respond to stress.

Cortisol also helps the body to:

- Maintain blood pressure and heart (*cardiovascular*) function.
- Slow the immune system's response to inflammation.
- Control the use of proteins, carbohydrates (sugars), and fats.
- Maintain a sense of well-being.

CAUSES

A lack of cortisol can happen for different reasons.

- Primary adrenal insufficiency is Addison's disease. The adrenal glands do not produce enough, or any, cortisol. Usually, aldosterone is also not produced.
- Secondary adrenal insufficiency. The pituitary gland may not make enough of a hormone called ACTH (adrenocorticotropin). This hormone causes the adrenal glands to produce cortisol. Usually, there is enough aldosterone.

Primary Adrenal Insufficiency can be caused by:

- Autoimmune disease. Your body can produce antibodies that attack its own organs (in this case, your adrenal glands). The reasons why this happens are not well understood at this time. Sometimes, other organs are also affected (the polyglandular autoimmune syndromes).
- An infection of the adrenal glands. Possible causes include tuberculosis, viruses (including HIV), and fungal infections.

Secondary Adrenal Insufficiency

This form is much more common than the primary form. It can be traced to a lack of ACTH. Without ACTH, the adrenal glands cannot make cortisol. Causes include:

- Diseases of the pituitary gland. This is a small gland in the brain that controls many important body functions. The pituitary gland produces ACTH, among other hormones. Pituitary gland tumors, injury, or surgery can cause inadequate ACTH production, which causes inadequate cortisol production.
- Medications:
  - Megestrol (used for cancer treatment and to stimulate appetite) and some pain medications can impair production of cortisol.
  - Use of cortisol medication (steroids) causes your adrenal glands to not produce cortisol. When you stop this medication, it can take time for the adrenal glands to start producing cortisol again. This can be a dangerous situation, requiring slowly reducing your cortisol medicine.

SYMPTOMS

Symptoms of adrenal insufficiency normally begin slowly. Problems seen with the disease are:

- Severe tiredness (*fatigue*).
- Muscle weakness.
- Loss of appetite.
- Weight loss.

About 50% of the time, the following symptoms occur:

- Nausea, vomiting and diarrhea.
- Drops in blood pressure.
- Dizziness or fainting.
- Darkening of the skin (with primary disease only).
- Being easily angered (*irritable*).
- Depression.
- Salt craving.
- Low blood sugar (*hypoglycemia*).
- Irregular or no menstrual periods.

The symptoms slowly get worse. They are often ignored until a stressful event like an illness or an accident occurs. This is called an Addisonian crisis, or acute adrenal insufficiency. Without treatment, an Addisonian crisis can cause death.

Symptoms of an Addisonian crisis include:

- Sudden, severe pain in the lower back, abdomen, or legs.
- Severe vomiting and diarrhea.
- Dehydration.
- Low blood pressure.
- Loss of consciousness.

DIAGNOSIS

In its early stages, adrenal insufficiency can be hard to diagnose. Your caregiver will need to:

- Review your medical history.
- Review your symptoms, such as dark tanning of the skin.
- Perform lab tests. Results will show if levels of cortisol are too low, and will help identify the cause. CT scan or MRI scan of the adrenal and pituitary glands may also be necessary.

TREATMENT

With proper treatment, you can live a normal life with Addison's disease or adrenal insufficiency.

- Missing hormones need to be replaced in order to treat Addison's disease. Cortisol is replaced with hydrocortisone tablets taken by mouth. Other forms of cortisol may also be used. Since cortisone levels normally are higher in the morning and lower in the evening, you may need different doses at different times of the day. Be sure to follow your instructions carefully.
- If your body cannot maintain the right levels of salt (*sodium*) and fluids because of too little aldosterone, you will also be given a medication called fludrocortisone (Florinef). This drug replaces aldosterone.

Important points about treatment:

- Any sudden (*acute*) illness can increase your body's need for cortisol. Surgery, or other stress on the body, can do the same. If you have Addison's disease and you are ill or having surgery, you will need an increase in hormone medication to prevent an Addisonian crisis. Untreated, an Addisonian crisis can cause death.
- If you are too ill to take your medication or you cannot keep it down, you must take medicine through a shot (*injection*). You or someone who lives with you will need to learn how to give you this injection. The shot will take the place of both hydrocortisone and fludrocortisone. If you find it necessary to give yourself injectable medication, call your caregiver right away, or go to the nearest hospital emergency room.

HOME CARE INSTRUCTIONS

- Always carry an identification card stating your condition in case of an emergency. The card should:
  - Alert emergency personnel about the need to inject 100 mg of cortisol if you are severely hurt or cannot respond.
  - Include your caregiver's name and phone number.

- Include the name and number of your closest relative to contact.
- When traveling, carry a needle, syringe, and an injectable form of cortisol for emergencies.
- Know how to increase medication during periods of stress or mild colds.
- Wear a warning bracelet or neck chain to alert emergency personnel. Many companies sell medical ID products.
- Take your medications as prescribed. **Do not** stop your medications without medical supervision.
- Learn about your condition. Ask your caregiver for further resources. This is a potentially dangerous, but easily managed illness.

SEEK MEDICAL CARE IF:

- You have Addison's disease and you are in need of surgery.
- You have Addison's disease and you have an acute illness.
- You have weakness, weight loss, or other unexplained symptoms.

SEEK IMMEDIATE MEDICAL CARE IF:

You have symptoms of crisis:

- Sudden, severe pain in the lower back, abdomen, or legs.
- Severe vomiting and diarrhea.
- Dehydration.
- Low blood pressure.
- Loss of consciousness.

You are experiencing severe:

- Infections or other illness.
- Vomiting.
- Diarrhea.

These conditions can bring on an Addisonian crisis.

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