

Understanding Adrenal Insufficiency

Adrenal insufficiency is a disorder that affects the adrenal glands, small glands that sit on top of the kidneys. This handout will tell you about adrenal insufficiency and its symptoms and causes. It will also tell you about how we test for adrenal insufficiency and how we treat it. Finally, it will tell you about adrenal crisis, a serious condition.

If you're concerned or have questions, call the Pediatric Endocrine Unit at (617) 726-2909. We want to hear from you!

WHAT IS ADRENAL INSUFFICIENCY?

Adrenal insufficiency is a condition in which the adrenal glands do not produce enough of a hormone called cortisol. Cortisol has many important jobs in the body, including preventing low blood pressure and low blood sugar. Cortisol is also important for helping the body respond to stresses such as illness, injury, and surgery.

In some cases of adrenal insufficiency, the adrenal glands also do not make enough aldosterone. Aldosterone is a hormone that helps the body keep the right balance of water and the salts sodium and potassium.

WHAT ARE THE SYMPTOMS OF ADRENAL INSUFFICIENCY?

In the early stages, you might not notice the symptoms of adrenal insufficiency because they're often subtle. Your child might have these symptoms:

- Feeling tired more often than usual (fatigue)
- Stomach pain or nausea
- Muscle aches
- Feeling light-headed
- Darkening of the skin, often most noticeable on the gums, over the knuckles or in the lines of the hands

WHAT CAUSES ADRENAL INSUFFICIENCY?

There are many causes of adrenal insufficiency, which can be divided into 2 categories. These categories are **primary adrenal insufficiency** and **central adrenal insufficiency**.

Primary adrenal insufficiency is when the adrenal glands do not work the way they should. Causes of primary adrenal insufficiency include:

- Genetic causes
- Autoimmune diseases or disorders that affect the adrenal glands
- Serious infections of the adrenal glands
- Having surgery to remove the adrenal glands

Important Words to Know

Adrenal glands
The adrenal glands are located next to the kidneys. They make many hormones, including cortisol and aldosterone.

Aldosterone
Aldosterone is a hormone that helps your body keep the right balance of water and salts.

Cortisol
Cortisol is a hormone that helps prevent low blood pressure and low blood sugar. It also helps your body respond to illness, injury and surgery. It is the most common glucocorticoid in humans.

Glucocorticoid
Glucocorticoids are hormones with many jobs, including keeping blood sugar levels within a normal range. Artificial forms of glucocorticoids can be given to people whose bodies have trouble making enough of their own glucocorticoid.

Stress-dosing
Stress-dosing is when you give your child extra doses of a medication called glucocorticoid. You must give your child stress doses when he or she is sick, injured or having surgery.

Central adrenal insufficiency is when the pituitary gland (a small gland underneath the brain that controls other glands) does not make enough adrenocorticotrophic hormone (ACTH). ACTH is a hormone that helps control how much cortisol your adrenal glands release. Causes of central adrenal insufficiency include:

- When the pituitary gland does not develop properly
- Injury to the pituitary gland, like trauma or radiation
- Tumors or other growths near the pituitary gland
- Having surgery to remove the pituitary gland
- Long-term use of high-dose glucocorticoid medications, like dexamethasone, prednisone, prednisolone or certain inhaled glucocorticoids

HOW DO WE DIAGNOSE ADRENAL INSUFFICIENCY?

We diagnose adrenal insufficiency through a blood test called an **adrenocorticotropic hormone (ACTH) stimulation test**. The ACTH stimulation test measures how well your child's adrenal glands respond to a medication called **cosyntropin**, which is an artificial form of ACTH.

For this test, we give your child an injection of cosyntropin. Then, we measure your child's cortisol levels in his or her blood 30 minutes later and again 60 minutes (1 hour) later.

HOW DO WE TREAT ADRENAL INSUFFICIENCY?

The good news is that cortisol and aldosterone can both be replaced by oral medications (medications taken by mouth). **It is extremely important that your child takes these medications every day or he or she might develop adrenal crisis, a life-threatening condition.**

To replace cortisol, your child's doctor will prescribe a medication called a **glucocorticoid**. There are many choices of glucocorticoids including hydrocortisone (Cortef®), prednisone, and dexamethasone. If your child's aldosterone levels are also low, your child's doctor will prescribe **fludrocortisone** (Florinef®).

HOW SHOULD I TREAT MY CHILD IF HE OR SHE IS SICK?

When your child is sick, injured or having surgery, he or she will need more replacement glucocorticoid than the amount you usually give. This means you should increase the dose of your child's glucocorticoid.

When you increase the dose of your child's glucocorticoid, it is called stress dosing. The doses are usually 2-3 times higher than your child's regular dose. Your doctor will go over how you should increase your child's dose.

You should increase the dose if your child has any of the following:

- Fever (over 100° F)
- Flu-like symptoms
- Any illness where he or she is too sick to go to school
- Injuries such as a broken bone
- Surgery in which he or she will need anesthesia ("going to sleep")

If your child is vomiting or is too sick to take anything by mouth, he or she will need an injection (shot) of hydrocortisone (Solu-Cortef). It is extremely important to make sure that glucocorticoid levels in your child's body are high enough to prevent serious illness. Your child's endocrine team will teach you how to do the injections at home.

If your child is sick, it is never wrong to give stress doses of glucocorticoid or to give an injection. If you are unsure whether your child needs stress-dosing or an injection, please call our on-call pediatric endocrinologist, available 24 hours a day at 617-726-2909. We would rather hear from you than have you worry alone!

WHAT IS THE LONG-TERM OUTLOOK FOR MY CHILD'S ADRENAL INSUFFICIENCY?

Your child will do very well as long as he or she is treated properly with the right medications and doses. Right now, there is no cure for adrenal insufficiency. It is a life-long disease.

What is Adrenal Crisis?

Adrenal crisis is a life-threatening condition when your child's blood sugar and blood pressure are too low and his or her salt levels are off-balance. Contact your doctor right away if he or she shows these signs and symptoms:

- Headache
- Stomach pain or nausea
- Feeling lightheaded
- Might look pale
- Might be confused
- Might be drowsy or difficult to arouse

Did you know?

It's very important for your child to wear a medical ID, like a medical alert bracelet, that states he or she has adrenal insufficiency. If there's ever an emergency, this will help make sure your child receives the right treatment by the medical team.

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For more information please call

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