Pediatric Endocrinology Fact Sheet

# Adrenal Insufficiency: A Guide for Families

## What is adrenal insufficiency?

The adrenal gland is located on top of the kidney and makes 3 types of hormones: corticosteroids or glucocorticoids (the main hormone is cortisol, which is also known as *hydrocortisone*); mineralocorticoids (the main hormone is aldosterone); and weak male-type sex steroid hormones known as the *adrenal androgens. Cortisol* is a hormone that helps to maintain blood sugar levels and helps in metabolism of fat, protein, and carbohydrates. Cortisol is especially important in times of stress. *Aldosterone* controls salt balance in the body through its effect on the kidney. *Adrenal androgens* are the hormones that are responsible for the development of pubic and underarm hair.

Production of cortisol by the adrenal gland is controlled by the pituitary gland hormone called *adrenocorticotropic hormone* (ACTH), which, in turn, is controlled by a brain hormone called *corticotropin-releasing hormone* (CRH).

There are 2 kinds of adrenal insufficiency. One form is *primary adrenal insufficiency,* in which the adrenal gland cannot produce enough cortisol or aldosterone. This form is also called *Addison disease.* The other form is *secondary* or *central adrenal insufficiency,* in which ACTH or CRH fails to signal to the adrenal gland, leading to decreased cortisol levels.

Babies can be born with adrenal insufficiency (congenital adrenal insufficiency) or can develop adrenal insufficiency during childhood or adolescence for many reasons (acquired adrenal insufficiency). Children may also develop temporary adrenal insufficiency after being treated with high-dose steroids for a medical condition. These children need to be closely monitored by a pediatrician when their steroid dose is being decreased.

## What are the symptoms of adrenal insufficiency?

The symptoms of adrenal insufficiency include fatigue, muscle weakness, decreased appetite, and weight loss.

Infants may fail to regain their birth weight and have trouble feeding. Some individuals experience nausea, vomiting, and diarrhea. In older children, symptoms can include dizziness, sweating, low blood sugar, and low blood pressure. Individuals with primary adrenal insufficiency may have salt craving and darkening of the skin.

## What causes adrenal insufficiency?

The most frequent cause of acquired primary adrenal insufficiency is autoimmune and is associated with the presence of antibodies that are associated with damage to the adrenal gland. Genetic disorders can also cause primary adrenal insufficiency. Other causes include infections, abnormal bleeding into the adrenal gland, adrenal tumors, and surgical removal of the adrenal gland.

Babies can be born with congenital primary adrenal insufficiency because of the inability of the adrenal gland to make enough cortisol and/or aldosterone. Many of the genetic disorders that can cause primary adrenal insufficiency are inherited. Sometimes, production of both cortisol and aldosterone is decreased. In other genetic disorders, only the production of cortisol is reduced. In some children, aberrant development of the external genitalia or excessive bone maturation may be noted.

Abnormalities of the brain and/or pituitary gland can prevent production of ACTH or CRH. Such disorders can also lead to adrenal insufficiency.

## How is adrenal insufficiency diagnosed?

The most common way to diagnose primary adrenal deficiency is to obtain a fasting blood sample early in the morning to check both cortisol and ACTH levels.

In primary adrenal insufficiency, the cortisol level will be low with an elevated ACTH level. In secondary adrenal insufficiency, the cortisol level is low with an ACTH level that is low or normal but not high. Sometimes, an ACTH stimulation test will be needed to confirm the diagnosis.

Additional blood work can include measurement of blood sodium, potassium, glucose, and plasma renin activity. In some instances, imaging studies, such as ultrasound, magnetic resonance imaging (MR imaging), or computed tomography (CT) scans, may be helpful.

## How is adrenal insufficiency treated?

The disorder is treated by hormone replacement. Oral hydrocortisone or other similar medications are used to replace cortisol and need to be taken 2 to 3 times a day. Patients with aldosterone deficiency usually take a pill called *fludrocortisone* to help maintain salt balance.

The hydrocortisone dose will usually need to be increased at times of significant body stress because your child's body cannot make more hydrocortisone. This is called *stress dosing.* Examples of stress include fever, severe diarrhea, severe vomiting, severe trauma, or surgery. It is best to ask your child's doctor for specific instructions on stress dosing. If a child is not able to take oral medications because of vomiting or being unconscious, hydrocortisone injections (eg, Solu-Cortef, Hydrocortisone sodium succinate) can be used; an emergency hydrocortisone injection kit for intramuscular injections should be available for such situations. Parents should learn how and when to administer intramuscular hydrocortisone injections.

With appropriate treatment, children with adrenal insufficiency can lead a normal life and have a normal life span.

## Can adrenal insufficiency crises be prevented?

Once the diagnosis of adrenal insufficiency has been confirmed, parents and patients need to learn how and when to administer daily medication and the higher cortisol doses for stress situations. All patients should wear medical alert identification badges.

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