

# Adrenal Insufficiency: A Guide for Families

## What is Adrenal Insufficiency?

The adrenal glands are located on top of the kidney and make three types of hormones called corticosteroids (often referred as cortisol), mineralocorticoids (main hormone is aldosterone), and weak male hormones or androgens. Cortisol is a hormone that helps maintain blood vessel tone and blood sugar levels and is especially important in times of stress. Aldosterone controls salt balance in the body. Androgens are sex hormones that are responsible for pubic and underarm hair. The production of cortisol by the adrenal glands is controlled by the pituitary gland hormone called adrenocorticotropic hormone (abbreviated as ACTH), which in turn is controlled by a brain hormone called corticotropin-releasing hormone (abbreviated as CRH). There are, therefore, two kinds of adrenal insufficiency: one is called primary adrenal insufficiency, because the adrenal gland is damaged and cannot produce cortisol (the pituitary hormones work well) and aldosterone; the other is called central adrenal insufficiency, because the brain hormones ACTH or CRH are not present to stimulate the adrenal glands to produce cortisol.

A child can be born with adrenal insufficiency (congenital adrenal insufficiency) or can become adrenally insufficient anytime after birth for many reasons (acquired adrenal insufficiency).

## What Causes Adrenal Insufficiency?

The most common kind of acquired primary adrenal insufficiency is “autoimmune” and is caused by antibodies and white blood cells directed against a child’s own adrenal glands — normally, antibodies are made against agents that come from outside of the body, such as germs that infect the body.

Occasionally, the immune system makes antibodies to body parts instead. Over time, the antibodies directed against the adrenal glands damage them so they cannot produce cortisol any longer. There are several other causes for acquired primary adrenal insufficiency, such as: infections, abnormal bleeding (adrenal hemorrhage), or tumors destroying or replacing the adrenal gland.

There are several reasons a baby might be born with congenital primary adrenal insufficiency — most are due to the inability of the adrenal glands to make cortisol and/or aldosterone well, and many of these are inherited. In some cases, the production of both cortisol and aldosterone are reduced, and in others, only the production of cortisol is reduced. Some of these problems are also associated with abnormalities in development of the genitals and/or bone abnormalities.

The most common cause of secondary adrenal insufficiency is prolonged administration of steroids to treat medical conditions, such as severe asthma or kidney, joint, or skin problems. This is also called iatrogenic adrenal insufficiency. When a child receives steroid therapy for more than one week, the steroids levels build up in the blood and shut down the ACTH

production from the brain. This in turn shuts off the production of cortisol by the adrenals. However, a medical problem rarely develops until the treatment has gone on much longer. Less frequent causes are related to the inability of the pituitary to make ACTH or CRH because of the presence of tumors. Babies can also be born with lack of ACTH or CRH, because the part of the brain that is supposed to make them is underdeveloped. Very rarely, central adrenal insufficiency can be due to the fact that ACTH is made but the adrenal glands do not respond to ACTH stimulation.

Adrenal insufficiency is uncommon except for the kind associated with prolonged medical treatment with steroids.

## How is Adrenal Insufficiency Diagnosed?

The most common way to diagnose primary adrenal deficiency is to obtain a blood sample early in the morning to check a cortisol level and an ACTH level. In primary adrenal insufficiency, cortisol levels will be low with an elevated ACTH level. In secondary adrenal insufficiency, cortisol level is low with an ACTH level which is low or normal but not high.

## How is Adrenal Insufficiency Treated?

Treatment can be accomplished by giving hydrocortisone or other similar medications to replace the cortisol production that is impaired. It is important to know that at time of stress, such as times when a child may have fever or is vomiting, the hydrocortisone does need to be increased, tripled or more, because that is what the adrenals would do on their own if they were functioning properly. If a child is not able to keep down the hydrocortisone because of vomiting or illness, and injection of hydrocortisone works best in emergencies. With appropriate treatment, children with adrenal insufficiency can lead a normal life and have a normal life span.

## Can Adrenal Insufficiency be Prevented?

Adrenal insufficiency cannot be prevented unless it is caused by steroid treatment. Doctors should be careful in prescribing long-term steroids. When steroid therapy is no longer needed, doctors have to be careful to set up a plan whereby steroids are tapered so that the brain can “wake up” and resume stimulating the adrenals on its own.

*Kathleen Bethin, MD, PhD, FAAP, and Teresa Quattrin MD, FAAP, PES/AAP-SOEn Patient Education Committee*

American Academy of Pediatrics

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