

PEDIATRIC ADRENAL INSUFFICIENCY

THE FACTS YOU NEED TO KNOW

WHAT IS ADRENAL INSUFFICIENCY?

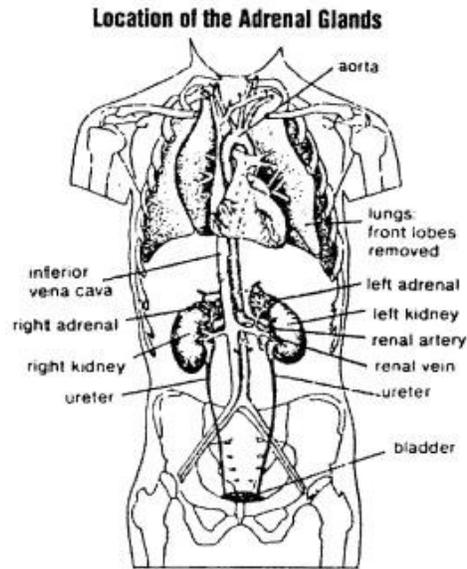
Adrenal Insufficiency is an uncommon disorder that occurs when the cortex of adrenal glands does not produce (or does not produce enough) of the vital steroid hormone, cortisol. Often, production of aldosterone is also affected. Cortisol mobilizes nutrients, modifies the body's response to inflammation and stress, stimulates the liver to raise the blood sugar, and helps to control the amount of water in the body.

Aldosterone regulates salt and water levels which affects blood volume and blood pressure.

There are two types of adrenal insufficiency:

- *Primary adrenal insufficiency (PAI)*, caused by disease intrinsic to the adrenal glands. PAI may result in the deficiency of both cortisol and aldosterone.
- *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 congenital adrenal hyperplasia (CAH), a genetic disorder caused by a mutation in the gene encoding 21-hydroxylase enzyme. As a result of the mutation, the adrenal cortex cannot produce cortisol and also, in most cases, aldosterone, yet may produce excess testosterone and related male-like hormones. This is called classic CAH. This genetic disorder is the most common cause of PAI and is usually diagnosed in the first two weeks of life. CAH can be severe (classic) and diagnosed in the newborn.



WHAT ARE THE CAUSES OF ADRENAL INSUFFICIENCY IN CHILDREN?

Adrenal insufficiency may be caused by:

- Genetic diseases such as congenital adrenal hyperplasia (CAH), adrenoleukodystrophy (ALD) or adrenomyeloneuropathy (AMN).
- Autoimmune injury when the body's immune system mistakenly attacks and damages the adrenal glands. This is called Addison's disease.
- Certain infections that can damage the adrenal gland.
- Hemorrhage into the adrenals.
- Surgical removal of both adrenal glands.
- A decrease in ACTH, a hormone that regulates cortisol secretion, due to genetic conditions, tumors or infections around the pituitary gland in the brain.
- Long-term steroid therapy, such as hydrocortisone, prednisone or dexamethasone, which are used to treat rheumatoid arthritis, severe asthma and immune conditions. These steroids can suppress the hypothalamic-pituitary-adrenal (HPA) axis.

WHAT ARE THE SYMPTOMS OF ADRENAL INSUFFICIENCY?

Children who present with adrenal insufficiency in early life can have signs of glucocorticoid deficiency (hyperpigmentation, hypoglycemia, prolonged jaundice, poor weight gain), mineralocorticoid deficiency (hypotension, salt loss, collapse), adrenal androgen excess (atypical genitalia in the case of virilizing CAH due to 21-hydroxylase deficiency), or associated features linked to a specific underlying condition.

- Slow weight gain
- Fatigue and generalized weakness
- Headache
- Nausea or vomiting
- Dehydration
- Low blood pressure
- Salt cravings
- May appear unusually tanned or have darkened skin creases, gums and scars

HOW IS ADRENAL INSUFFICIENCY IN CHILDREN DIAGNOSED?

Adrenal insufficiency (AI) is a potentially life-threatening condition that can be difficult to diagnose. It is an important diagnosis to consider in any sick newborn infant and prompt investigation and treatment are essential.

A definitive diagnosis of adrenal insufficiency requires that tests be carried out.

- Measure amount of cortisol and aldosterone in the blood.
- Document a lack of the normal increase in the levels of cortisol after administration of ACTH given by injection.
- Your child's doctor may also order imaging tests, such as ultrasound or CT of the adrenal gland, or an MRI of the brain.

Other tests may be required to rule out other causes of adrenal insufficiency such as CAH:

- Genetic tests
- A physical examination
- A family history

Sometimes, when there is a known family history of CAH, a fetus is diagnosed before birth.

HOW IS ADRENAL INSUFFICIENCY IN CHILDREN TREATED?

Children and adults alike are treated orally administered by hormone replacement. Oral hydrocortisone is the most common medication and is used to replace cortisol by taking 2 to 3 times a day. Prednisone may also be used. Children with aldosterone deficiency usually take fludrocortisone to help maintain salt balance.

Hydrocortisone granules can be prescribed for the treatment of adrenal insufficiency in infants, children, and adolescents. This new treatment is shown to be effective for childhood adrenal insufficiency, by providing the ability to accurately prescribe pediatric-appropriate doses.

STRESS DOSING AND EMERGENCIES

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 high 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 (e.g., Solu-Cortef, hydrocortisone sodium succinate) can be used; an emergency hydrocortisone injection kit or intramuscular injections should be available for such situations. Parents should learn how and when to administer intramuscular hydrocortisone injections. See YouTube video for instructions: <https://bit.ly/3ql2ysD>.

Persistence of these signs requires immediate treatment in an Emergency Room with intravenous saline (salt water) and hydrocortisone.



FOLLOW-UP CARE

Children with adrenal insufficiency need regular follow up and blood testing. Monitoring is necessary to avoid symptoms of adrenal insufficiency and ensure proper growth and development.

OUTCOME

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

SOURCES

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American Academy of Pediatrics and Pediatric Endocrine Society: <https://pedsendo.org/patient-resource/adrenal-insufficiency/>

Children's Hospital of Philadelphia: www.chop.edu/conditions-diseases/adrenal-insufficiency

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