

Congenital Adrenal Hyperplasia (CAH)

- Sha-Tiah Sampson

Introduction

Congenital Adrenal Hyperplasia is a condition that affects the endocrine system. It is an inherited disorder of the adrenal glands. The adrenal glands which are located on top of the kidneys produce three types of hormones: Cortisol, Aldosterone, and Androgens. The life essential hormones are Cortisol and Aldosterone.

Cortisol

Cortisol is an energy and blood sugar regulating hormone. These effects include controlling the body's blood sugar levels and thus regulating metabolism, acting as an anti-inflammatory, influencing memory formation, controlling salt and water balance, influencing blood pressure and helping development of the foetus

Aldosterone

Aldosterone affects the body's ability to regulate blood pressure. It sends the signal to organs, like the kidney and colon, that can increase the amount of sodium the body sends into the bloodstream or the amount of potassium released in the urine. The hormone also causes the bloodstream to re-absorb water with the sodium to increase blood volume. All of these actions are integral to increasing and lowering blood vessels. Indirectly, the hormone also helps maintain the blood's pH and electrolyte levels. This hormone plays an important role in cardiovascular system.

Androgens

Androgens are “masculine hormones that cause pubic hair growth and masculine changes in puberty, called virilization. Additionally, these hormones are believed to regulate the function of many organs, including the reproductive tract, bone, kidneys, liver and muscle. In adult women, androgens are necessary for estrogen synthesis and have been shown to play a key role in the prevention of bone loss, as well as sexual desire and satisfaction.

Congenital Adrenal Hyperplasia

Normally there is an enzyme that is necessary for the body to make cortisol and aldosterone. In CAH the enzyme is missing, partially inactive, or totally inactive, which leads to a “road block” in the “hormone production highway”.if the enzyme is missing or severely inactive, both cortisol and aldosterone will not be produced. If the enzyme is is partially inactive, the cortisol produced will be smaller than needed amounts.

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In addition the , other hormones which are in the same or different “highway” to make cortisol or aldosterone are also affected and may be abnormally low, or abnormally high. ACTH produced from the pituitary gland , which is located at the base of the brain, stimulates the adrenals to make cortisol. When the cortisol is low the pituitary gland will send more ACTH to the adrenals in an attempt to make more cortisol, which results in a “backup” or over production of the other adrenal hormones just before the “roadblock” occurs.

The different types of CAH

There are several form of CAH , depending on which enzyme is deficient . The treatment and symptoms will vary according to the type of CAH. The most common type of enzyme deficiency that causes CAH is 21 hydroxylase(21OH deficiency). Other adrenal enzyme deficiency that less frequently cause CAH are steroidgenic.

What causes CAH

It is an inherited disorder. This means that the person is born with it. The chromosome for the missing or inactive enzyme was passed on to the child from both parents. In general both parents of children with CAH are healthy, but each parent carried the partial trait for CAH.

What does it affect ?

It affects the natural flow of the bodies ability to maintain meiosis. Children who have this condition sometimes go through precocious puberty in which they start puberty early on in life, and are stunted at growth. In some cases they are born with more masculinizing traits, or feminizing traits and may need to repair and complete the sexual anatomy development. They can go through an adrenal crisis at any given time.

Adrenal crisis

It is the onset of shock. A person going through this can present with pale, cool clammy skin. The shock could be triggered by a fever, a broken bone, or anything that stresses the body beyond its capacity to cope.

(CAH)

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