

Hypopituitarism

What is hypopituitarism?

Hypopituitarism is a term used to describe a condition in which the **pituitary gland** is not able to produce one or more of the pituitary hormones. The lack of **ALL** pituitary hormones is known as **panhypopituitarism** ("pan" means all, and "hypo" means less than usual). The pituitary is a small, pea-sized gland located at the base of the brain and is controlled by the **hypothalamus**. It is often referred to as the "**master gland**", because its chemical messengers or **hormones** signal other endocrine glands to produce their own hormones. The pituitary is divided in two parts, the anterior- (the front) and the posterior- (the back) pituitary. The **anterior pituitary** produces hormones that are needed for normal body functions. It includes: **growth hormone (GH)**, **adrenocorticotrophic hormone (ACTH)**, **thyroid stimulating hormone (TSH)**, and **puberty hormones (gonadotropins called LH and FSH)**. The posterior pituitary produces the **antidiuretic hormone (ADH)** and **oxytocin**.

What causes hypopituitarism?

Hypopituitarism can be caused by anything that disrupts the normal functioning of the hypothalamus or the pituitary. **Congenital hypopituitarism** refers to the condition being present at birth, which means that either the pituitary or the hypothalamus did not develop normally prior to birth. **Acquired hypopituitarism** refers to damage to the pituitary or hypothalamus during or after birth. This can be caused by a severe head injury, surgical removal of the pituitary gland, radiation therapy, a brain tumor, or an inflammation or infection in the brain. In cases where the cause cannot be determined, it is often referred to as "idiopathic", which means "of unknown cause".

What are the possible effects of hypopituitarism?

Children with hypopituitarism may be unable to make one, two, or possibly all of their pituitary hormones. As a result, the body will not get these important hormonal messages, and therefore will be unable to respond to the message. The effects on the body vary depending on which hormone is missing. Following is a summary of the hormones and deficiencies there of:

Growth hormone deficiency

For children to grow to their full height potential, their bodies need adequate amounts of **growth hormone**. Growth hormone affects the growth

of bone and body tissues, as well as, muscle, fat, and sugar metabolism. When the pituitary does not produce enough growth hormone, a child may experience symptoms of low blood sugar (hypoglycemia). Without growth hormone a child will be normally proportioned, but grow very slowly. The child will be smaller and younger-looking than their peers.

Thyroid Stimulating Hormone (TSH) deficiency

TSH stimulates the **thyroid gland**, located in the front of the neck, to produce and release thyroid hormones, particularly **thyroxine (T4)**. T4 is important for brain development, growth and metabolic balance. Low levels of thyroid hormones, also referred to as "hypothyroidism", may include some or all of the following symptoms:

- Tiredness or sleeping more than usual.
- Cool, dry skin
- Cold intolerance
- Weight gain, slow growth
- Poor appetite
- Constipation
- Coarse, dry, thinning hair
- Irregular periods

Adrenocorticotrophic hormone (ACTH) deficiency

ACTH stimulates the **adrenal glands** (located on top of the kidneys) to produce **cortisol**. Cortisol keeps the body's blood sugar at a normal level and helps the body deal with physical stress, such as a fever or injury. If the pituitary is unable to produce normal amounts of ACTH, the adrenal glands will not produce enough cortisol. Symptoms of ACTH deficiency may include some or all of the following symptoms:

- Tiredness, weakness
- Dizziness, confusion
- Weight loss
- Diarrhea/nausea/vomiting
- Dehydration
- Frequent illness, such as ear infections
- Lower than normal body temperature
- Fast pulse, fast breathing
- Seizures from low blood sugars

Gonadotropin (LH and FSH) Deficiency

LH and FSH stimulate the sex organs to make **puberty hormones**. In females they stimulate the **ovaries** to make **estrogen and progesterone**, which is necessary for breast development, the menstrual cycle (periods), adolescent growth, and fat distribution. In males, they stimulate the **testes** to make **testosterone**, which is responsible for adolescent growth spurts, voice changes, increase in the size of the penis and testes, increased muscle mass and facial hair (beard). Lack of gonadotropins will result in delayed sexual development. Male infants may present with a small penis (micropenis).

Antidiuretic hormone (ADH) deficiency

ADH helps the body maintain the water and salt balance by acting on the **kidneys** to save more water when necessary. If ADH is decreased or lacking, the kidneys are unable to concentrate the urine and keep the body sufficiently hydrated. The child will drink a lot, but also urinate frequently, which causes the body to dehydrate. ADH deficiency is referred to as **diabetes insipidus** or DI. (This has nothing to do with diabetes mellitus, where people have problems with their blood sugars).

How is hypopituitarism diagnosed?

First the doctor will obtain a detailed history and a thorough **physical examination**. **Blood tests** will indicate hormone levels of pituitary hormones and target gland hormones. In some cases **hormone stimulation tests** are necessary to fully evaluate the functioning of the pituitary gland. A **urine sample** and **electrolytes** in the blood may be done, if ADH deficiency is suspected. An **MRI** (magnetic resonance imaging) can detect tumors or other structural abnormalities in the brain. An **X-ray** of the left hand and wrist will determine the child's bone age, which shows the maturity of the child's bones.

Treatment of hypopituitarism

The treatment of hypopituitarism involves **replacing the hormones** normally made by the target glands.

Growth hormone is replaced with a synthetic form of growth hormone. It is given through daily injections under the skin.

Thyroid hormone is replaced with a synthetic form of thyroxine called **levo-thyroxine** (also known as **Synthroid** or **Levoxyl**). It is available in pill form and needs to be taken daily.

Adrenocorticotropin deficiency is treated by replacing cortisol in form of **hydrocortisone (Cortef)**. This is available in pill form, liquid, or as injection, to be taken 2-3 times a day. The dose needs to be increased during illness, fever, or injury (please see the Emergency treatment sheet for further instructions).

Gonadotropin deficiency treatment involves beginning of replacement of **estrogen** or **testosterone** at the appropriate time to begin puberty. Estrogen is taken daily and progesterone is added after several months. These hormones mimic the way the ovaries would produce hormones. There will be a menstrual period at the end of each cycle. Estrogen and progesterone come in pill form and are also available in skin patches. Males are given testosterone replacement by monthly intramuscular (IM) injections.

ADH is replaced with **desmopressin** or **DDAVP**. It will be given once or twice a day, depending on the condition of the child and the required dose. This medicine is most commonly given by nasal spray. It is also available by nasal tube, by pill form, or by injection. If too much DDAVP is given, water retention can occur. If too little DDAVP is given, dehydration and electrolyte imbalance can occur. Both of these conditions can be **life-threatening** if left untreated.

All children with hypopituitarism should wear medical identification in form of a medic-alert bracelet or necklace.

When will the doctor need to see my child?

The extent of the pituitary deficiency will determine how often your child needs to see the pediatric endocrinologist at our office. Replacement therapy needs to be monitored and adjusted. It is also important to check if other pituitary hormones have been affected. Communication with the doctor about your child's condition is important for optimal treatment. Remember that you will also need to follow up with your child's primary care physician for routine health care.

If you have any further questions or concerns please call us at (303)783-3883.