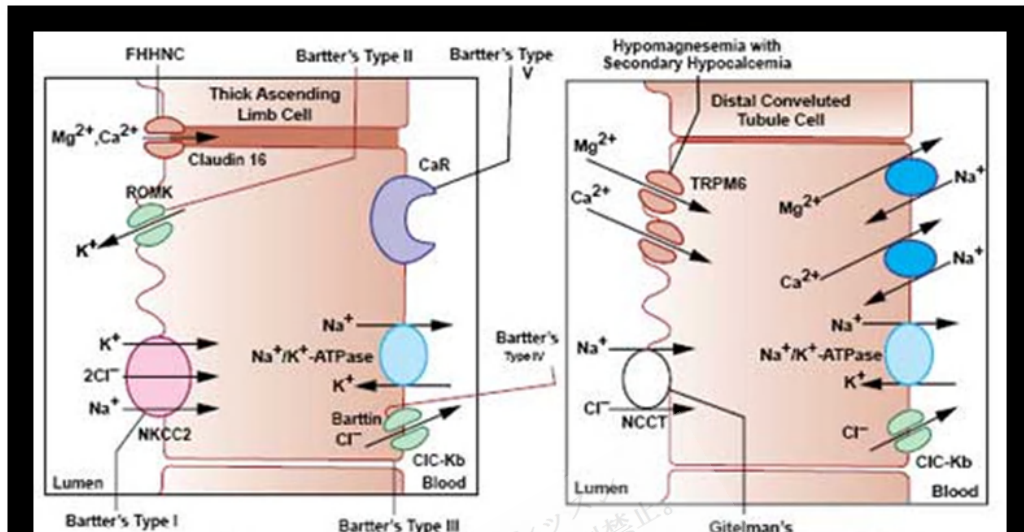


Bartter syndrome



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Causes

There are five gene defects known to be associated with Bartter syndrome. The condition is present from before birth (congenital).

The condition is caused by a defect in the kidney's ability to reabsorb sodium. Persons with Bartter syndrome lose too much sodium through the urine. This causes a rise in the level of the hormone aldosterone and makes the kidneys remove too much potassium from the body. This is known as potassium wasting.

The condition also results in an abnormal acid balance in the blood called hypokalemic alkalosis, which causes too much calcium in the urine.

Symptoms

This disease usually occurs in childhood. Symptoms include:

- Constipation
- Growth failure
- Increased frequency of urination
- Low blood pressure
- Kidney stone
- Muscle cramping and weakness

Exams and Tests

The diagnosis of Bartter syndrome is usually suspected when low levels of potassium are seen in the blood. The potassium level is usually less than 2.5 mEq/L. Unlike other forms of kidney disease, this condition does not cause high blood pressure and there is a tendency toward low blood pressure. Other signs of this syndrome include:

- High levels of potassium, calcium, and chloride in the urine
- High levels of the hormones renin and aldosterone in the blood
- Low blood chloride
- Metabolic alkalosis

These same signs and symptoms can also occur in people who have taken too many diuretics or laxatives. Urine tests can be done to rule out other causes.

In Bartter syndrome, a biopsy of the kidney typically shows too much growth of kidney cells called the juxtaglomerular apparatus. However, this is not found in all patients, especially in young children.

Treatment

Bartter syndrome is treated by keeping the blood potassium level above 3.5 mEq/L. This is done by following a diet rich in potassium or taking potassium supplements.

Many patients also need salt and magnesium supplements, as well as medicine that blocks the kidney's ability to get rid of potassium. High doses of nonsteroidal anti-inflammatory drugs (NSAIDs) may also be used.

Outlook (Prognosis)

The long-term outlook for patients with Bartter syndrome is not certain. Infants who have severe growth failure may grow normally with treatment. Although most patients remain well with ongoing treatment, some develop kidney failure.

Possible Complications

Kidney failure is a possible complication.

When to Contact a Medical Professional

Call your health care provider if your child is:

- Having muscle cramps
- Not growing well
- Urinating frequently

Reference

<http://www.nlm.nih.gov>

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