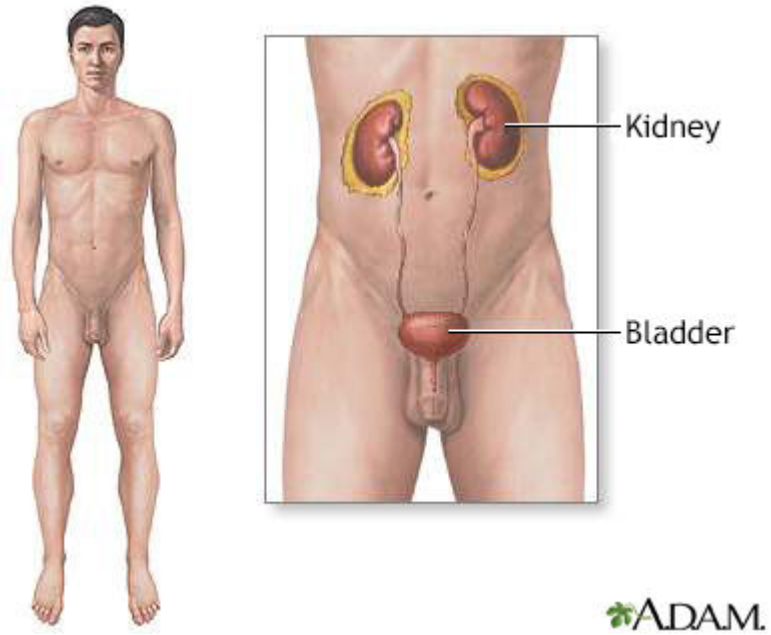
Nephrogenic diabetes insipidus

Definition

Nephrogenic diabetes insipidus (NDI) is a disorder in which a defect in the small tubes (tubules) in the kidneys causes a person to pass a large amount of urine and lose too much water.



Causes

Normally, the kidney tubules allow most water in the blood to be filtered and returned to the blood.

NDI occurs when the kidney tubules do not respond to a hormone in the body called antidiuretic hormone (ADH), also called vasopressin. ADH normally causes the kidneys to make the urine more concentrated.

As a result of not responding to the ADH signal, the kidneys release too much water into the urine. This causes the body to produce a large quantity of very dilute urine.

NDI is very rare. Congenital nephrogenic diabetes insipidus is present at birth. It is a result of a defect passed down through families. Men are usually affected, though women can pass this gene on to their children.

Most commonly, NDI develops because of other reasons. This is called an acquired disorder. Factors that can trigger the acquired form of this condition include:

- Blockage in the urinary tract
- High calcium levels
- Low potassium levels
- Use of certain drugs (lithium, demeclocycline, amphotericin B)

Symptoms

You may have intense or uncontrollable thirst, and crave ice water.

You will produce large amounts of urine, usually more than 3 liters, and up to 15 liters per day. The urine is very dilute and looks almost like water. You may need to urinate every hour or even more, even during the night when you are not eating or drinking as much.

If you do not drink enough fluids, dehydration can result. Symptoms may include:

- Dry mucous membranes
- Dry skin

- Sunken fontanelles (soft spot) in infants
- Changes in memory or balance

Other symptoms that can occur due to lack of fluids, causing dehydration, include:

- Fatigue, feeling weak
- Headache
- Irritability
- Low body temperature
- Muscle pain
- Rapid heart rate
- Weight loss
- A change in alertness, and even coma

Exams and Tests

The health care provider will examine you and ask about your or your child's symptoms.

A physical exam may reveal:

- Low blood pressure
- Rapid pulse
- Shock
- Signs of dehydration

Tests may reveal:

- High serum osmolality
- High urine output, regardless of how much fluid you drink
- Kidneys do not concentrate urine when you are given ADH (usually a medicine called desmopressin)
- Low urine osmolality
- Normal or high ADH levels

Other tests that may be done include:

- Sodium blood test
- Urine 24-hour volume
- Urine concentration test
- Urine specific gravity
- Supervised water deprivation test

Treatment

The goal of treatment is to control the body's fluid levels. A large amount of fluids will be given. The amount should be about equal to the amount of water being lost in the urine.

If the condition is due to a certain medicine, stopping the drug may improve symptoms. But, do NOT

Outlook (Prognosis)

If a person drinks enough water, this condition will not have much effect on the fluid or electrolyte balance of the body. Sometimes, passing a lot of urine for a long time can cause other electrolyte problems.

If the person does not drink enough fluids, high urine output may cause dehydration and high levels of sodium in the blood.

NDI that is present at birth is a long-term condition requiring lifelong treatment.

Possible Complications

Untreated, NDI may cause any of the following:

- Dilation of the ureters and bladder
- High blood sodium (hypernatremia)
- Severe dehydration
- Shock
- Coma

When to Contact a Medical Professional

Call your provider if you or your child has symptoms of this disorder.

Prevention

Congenital NDI cannot be prevented.

Treating the disorders that can lead to the acquired form of the condition may prevent it from developing in some cases.

Alternative Names

Nephrogenic diabetes insipidus; Acquired nephrogenic diabetes insipidus; Congenital nephrogenic diabetes insipidus; NDI

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