

Diabetes Insipidus

What is diabetes insipidus (DI)?

Diabetes insipidus is a rare disorder in which a person urinates an excessive amount, is more thirsty than usual, and drinks an excessive amount of fluid. This can be caused by a hormone deficiency (anti-diuretic hormone or ADH) or a decreased ability of the kidney to respond to this hormone.

It is important to differentiate diabetes insipidus from diabetes mellitus. The term “diabetes” was first used to describe a condition with frequent urination. Diabetes mellitus is much more common and is typically the disease one thinks of when they hear the word ‘diabetes’. Diabetes mellitus is a disorder of the processing of blood sugar in the body. When the blood sugar gets too high in the bloodstream, some of it is filtered out by the kidneys into the urine. This causes the person to urinate frequently and also drink a lot of fluids to replace what is being lost in the urine. As you can see, the symptoms for diabetes insipidus and diabetes mellitus are very similar. However, the underlying changes within the body to cause the individual diseases are very different.

What are the symptoms of diabetes insipidus?

- *Increased thirst (polydipsia)
- *Increased urination (polyuria) – urine is often clear or a very faint yellow in color
- *It is often hard to gauge as a parent what is increased or excessive. Your child’s pediatrician or endocrinologist can help you determine this amount based on the child’s age and size.
- *Getting up to go the bathroom several times at night (nocturia)
- *Signs of dehydration such as elevated heart rate, dry mouth, sunken eyes or dizziness going from sitting to standing
- *Problems gaining weight and growing because they are drinking such an excessive amount of fluid, they feel too full to eat food

What are the types of diabetes insipidus?

Central DI: This type of DI is caused by decreased production of a hormone called antidiuretic hormone or ADH. It’s also called arginine vasopressin or AVP. ADH is released by the pituitary gland in the brain and signals to the kidneys to keep fluid in the body rather than releasing it into the urine. For example, if your child is playing outside on a hot summer day they become slightly dehydrated. The pituitary gland will release ADH which signals to the kidneys to not make as much urine because the child’s body needs that fluid to stay in the bloodstream. If your child’s brain does not produce enough of this hormone, then the kidneys do not receive this signal and the child keeps urinating. This causes the body to be dehydrated and as a result the child is very thirsty.

Nephrogenic DI: In this type of DI, the pituitary gland makes plenty of ADH but the kidneys are resistant or unresponsive to the ADH. Children with nephrogenic DI will also have increased urination and thirst.

What causes central and nephrogenic DI?

Central DI can develop in a person that has any type of damage to the area of the brain called the hypothalamus or the pituitary gland. This can be from surgery, trauma, or a tumor in the brain. Children can be born with a small or abnormally shaped pituitary gland that results in decreased production of ADH and possibly other hormones as well. There are cases of central DI that we don't know the cause for, this is called idiopathic DI. Sometimes an auto-immune process can harm the cells in the brain that make ADH which causes central DI.

Nephrogenic DI is typically inherited, meaning it is passed down through genes from one family member to another. Certain mutations mostly affect boys while others can affect boys and girls. There are certain medications that can cause nephrogenic DI (lithium and certain drugs that treat HIV are the most common). Finally, if a person has long standing kidney disease they can also develop nephrogenic DI.

How is diabetes insipidus diagnosed?

Under the guidance of a pediatric endocrinologist a child typically undergoes a "water deprivation test" to help make the diagnosis of DI. During this test, the child is not eating or drinking anything for a specified period of time. This test is designed to purposefully cause a dehydrated state in the body and see how the body responds. Sometimes this test is done in the hospital and other times it is done outpatient. Blood and urine samples are taken at various times during the testing. The child is also monitored for signs of dehydration including a decrease in weight or an increase in heart rate. The duration or length of the test is determined by your child's doctor. If the doctor determines the urine and blood samples are consistent with DI, often a small dose of vasopressin (synthetic ADH) is given to see how your child responds to it. This can help distinguish between central and nephrogenic DI.

How is DI treated?

Central DI is typically treated by replacing the ADH hormone that is missing. The medication that is generally used is called desmopressin or DDAVP. It is available as a pill, a nasal spray or subcutaneous (under the skin) injection that can be given at home. Sometimes your child will be asked to drink more or less water depending on their sodium level and level of dehydration.

Nephrogenic DI is typically treated in a variety of ways, often with a combination of therapies. Your doctor may make recommendations to decrease the salt or protein content of your child's diet. Sometimes a class of drugs called diuretics are given. The treatment for nephrogenic DI varies greatly on the child's age, weight and other presence of other health issues.