Central Diabetes Insipidus

BC CHILDREN'S HOSPITAL
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Notes

Acknowledgements

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For some time, you have been watching your son or daughter cope with a constant thirst and the need to urinate (pee) maybe as often as every hour. Now the doctor has explained your child’s condition - Central Diabetes Insipidus (DI). Your health care team and this booklet will help your family to understand this diagnosis. You will feel less anxious when you know how to manage DI. Once it is under control, your child won’t have to keep stopping his activities to go to the toilet or get a drink.

Diabetes Insipidus is not Diabetes Mellitus

When most people hear the term “diabetes” they think of diabetes mellitus (also called sugar diabetes). But the two conditions **are not alike** even though

- both health conditions have two of the same symptoms - thirst and the need to urinate often.
- both have “diabetes” in their name.

DI will not develop into diabetes mellitus. The cause of each condition is different and the treatment is different.

“Diabetes Insipidus” comes from the Greek and Latin words diabainein (to pass through) and insipidus (having no flavour) and so it means to “pass clear urine.” DI is NOT related to diabetes mellitus – sugar diabetes. It is a totally different condition.

What is Diabetes Insipidus (DI)?

DI is a condition in which the body cannot keep the correct amount of water in its system. This is because the kidneys are not holding in as much of the water coming through as they normally would. The child loses too much water in the urine, and then drinks large amounts to try to balance things out. But drinking does not satisfy the water needs of the body because the water does not stay in the body. The child remains thirsty.

What is the cause of Central DI?

A hormone called “vasopressin” also known as the “anti-diuretic hormone” (ADH) controls the amount of water in the body. ADH acts in the kidneys making them keep the right amount of water in the body. People with Central DI do not have enough (or any) of this hormone. The water they drink flushes through and out through the bladder.

The kidneys filter and then flush waste and excess water out through the bladder.

For pricing and ordering information

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Websites

NIH Patient Info
www.cc.nih.gov/ccc/patient_education/pepubs/di.pdf

Diabetes Insipidus Foundation
www.diabetesinsipidus.org

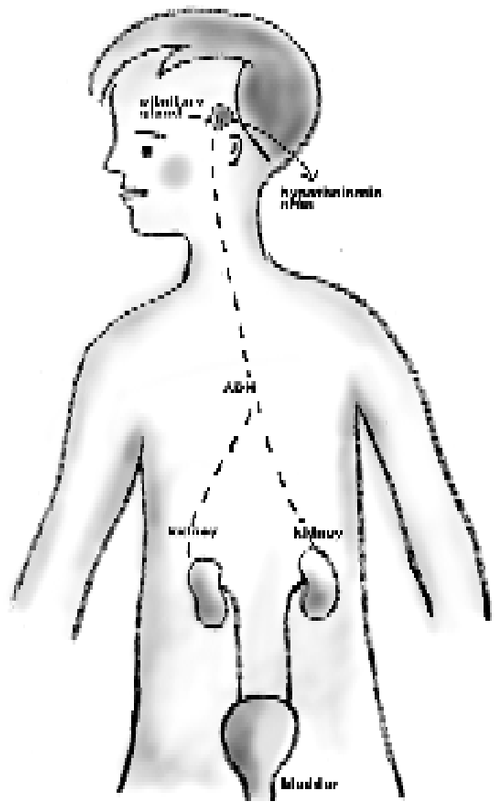
Medic Alert Canada
www.medicalert.ca

More links available from:

The Department of Endocrinology at BC Children's Hospital
http://endodiab.bcchildrens.ca

Why is there no anti-diuretic hormone (ADH)?

ADH is made in the hypothalamus (an area of the brain) and stored in the pea-sized pituitary gland (in the centre of the brain). If the cells in either of these areas are damaged, they cannot supply the hormone.



If a child has surgery or treatment near the pituitary gland, the health care team will watch for the symptoms of DI.

A tumour, surgery, radiation, brain injury, or disease such as Langerhans cell histiocytosis can all cause damage. In a few cases, the damage happens before birth when the brain is forming. Rarely this may be a condition that occurs in the family.

Note: Another type of DI is Nephrogenic DI. It is a much rarer condition. The problem is not in the tissues of the hypothalamus or pituitary gland. The problem is that the tissues of the kidneys do not respond to the hormone as they should.

How is DI diagnosed?

Most people with DI prefer water to other drinks.

The signs of DI are:

- Drinking large amounts of water. This is called polydypsia.
- Urinating large amounts. (Urine has no colour or smell.) This is called polyuria. Some children wet the bed.
- Poor growth and poor weight gain.
- More fevers than normal.
- Sickness from dehydration (lack of water in the body cells).

If a child has some of these signs, the doctor may suggest a *water deprivation test* to confirm the diagnosis.

Q. My daughter is on Desmopressin 0.05 mL morning and evening. She is having chemotherapy right now and gets a lot of IV fluid. Does the Desmopressin dose need to change?

A. Large amounts of IV fluids are given to flush the chemotherapy toxins out of her body. If you give the regular dose of Desmopressin the IV fluids will be held in the body causing water intoxication and also flushing may not happen. Toxins may not clear quickly. Ask your specialists (endocrinologist and oncologist) about this. They may decide <u>not</u> to give the regular dose of Desmopressin while the IV is running.

Anytime people with DI are asked to drink more or less to prepare for a medical test or treatment, they should check with the endocrinologist and make a plan to adjust the Desmopressin dose. Their body cannot adjust to fluid changes in the normal way. Drinking a lot more than usual, or not drinking at all, can be a risk to their health.

Q. My son needs surgery next month. What will happen with the Desmopressin?

A. It is very important for the surgeon and anesthetist to consult the endocrinologist about the best way to manage the DI before, during and after surgery.

Conclusion

Central DI can be a challenge for child and family. For a while it may seem that you "never get it right". In time you, and your child, will get to know how his body responds to Desmopressin. You will help him balance his dose of Desmopressin and his fluid intake. Life does return to normal – there are fewer trips to the bathroom and fewer glasses of water and fewer sleepless nights – for you too.

Q. ***I'm 18 and am going to my Grade 12 Grad next month. I want to party with my friends. My mother says it is dangerous for me to drink alcohol. Is she just trying to scare me?***

A. Drinking a lot of anything is not a good idea for people taking Desmopressin your body will have to hold that extra fluid until the Desmopressin wears off. All the fluid held in your system will cause the salt level in your body to fall and you will end up with a headache – not much fun at a grad party! Your beer drinking pals have kidneys that will keep them urinating quite a bit and control their salt levels. Your kidneys are not going to be able to help your body out in this way. Even a few beers (or non-alcoholic drinks) can cause you a lot of trouble – you could become sick enough to miss the whole show. So, really, the answer is that a small amount of alcohol is not the concern. The problem is drinking gets out of hand at times like Grad. Do yourself (and your friends) a service – party and have a good time without the drinks.

Note: Alcohol can also interfere with the potency of the Desmopressin.

Q. I'm worried that my teenager may be experimenting with "recreational drugs." How will this interact with the desmopressin?

A. It is not recommended that any recreational drug is used in combination with medications because there are many unknown interactions. One drug we know is a dangerous combination with desmopressin is ecstasy. When desmopressin and ecstasy are taken together the action of the desmopressin is increased, this can lead to dangerously low sodium levels.

The aim of the test is to answer these questions:

1. Is it really DI? Can the kidneys hold water in the body and put only the right amount of water into the urine?

   To answer this question, the child is not allowed to drink or eat (because there is some fluid in food) for 8 or more hours. Samples of urine and blood are measured regularly during the test. If the urine **does** become more concentrated with less water, it suggests that the body does have the ability to supply ADH. In this case, the diagnosis may not be DI.

   If the urine **does not** become more concentrated, it means that either
   - there is not enough ADH being made – the most likely diagnosis is Central DI or
   - *the kidneys cannot respond to ADH – the most likely diagnosis is Nephrogenic DI.*

Note: When water is lost and not replaced (by drinking) the amount of natural sodium (salt) in the blood slowly rises. The level of sodium in the child's blood is measured often during the test. The sodium level shows whether the body is holding in the right amount of water.

2. What type of DI is it?

Once the test results confirm DI, your child is given a dose of synthetic (made by a drug company) ADH called *Desmopressin*. If the kidneys can use

the synthetic hormone and work as they should, the diagnosis is confirmed as **central diabetes insipidus**.

Note: People become very, very thirsty during this test. It seems unkind and unfair to put children through it. Sadly, there is no other way to find out if your child has DI.

How is Central DI treated?

Central DI is treated with Desmopressin. It replaces the natural hormone ADH that causes the kidneys to hold in water.

Desmopressin comes in several forms:

- Intranasal **solution** (nose drops) given by a delivery tube (rhinyle) rather than a dropper, 0.1milligrams/milliliter (mg/mL)
- Intranasal **spray** (nose spray), 10 micrograms/dose (10 mcg/dose or 0.01mg/dose)
- **Tablets** (to swallow) 0.1 mg or 0.2 mg
- **Melts** 60 mcg or 120 mcg melted on the tongue
- For very small doses, the pharmacy can make a dilute solution, to be given by mouth

Most clinicians suggest starting with the Desmopressin in the intranasal solution form. It allows the person to take small, precise doses. Instead of using a dropper, the exact dose is loaded into a **rhinyle** (a thin plastic tube). One end goes into the child's nose. The child, or an adult, gently blows the other end, blowing the dose into the child's nose. The Desmopressin enters the blood through the lining of the nose.

In hospital, a nurse may give the Desmopressin by attaching a small syringe (no needle) to the rhinyle, and give a gentle puff of air to push the medicine into the nostril.

Q. ***My foster daughter is six, and she has some behaviour problems. It seems to us that she uses drinks as a way to get our attention or to distract from tasks she does not want to do. I find it difficult to know how to respond because I don't always know when it is "break through" and when it is behaviour.***

A. This is a problem. It is made more difficult by the fact that some children with DI also have an abnormal sense of thirst. The request for a drink may come from this rather than a behaviour issue. Either way, you and your child can agree on a plan. Make it a little difficult to get the drink – s/he has to interrupt her play, go to the kitchen table, sit down, and drink her water (not pop or juice).

If you think that your child has an abnormal thirst mechanism (s/he is still wanting drinks even though s/he had her Desmopressin just an hour ago) you will need to discuss a plan with your health care professional, to be sure s/he is getting enough fluid, but not too much. The child's intake may have to be actually measured and limited, based on regular blood sodium levels. If you decide the situation is a behaviour problem, try to get some help with this while not totally denying her water either, because this is her only way to maintain normal balance in her body.

Q. ***What should I do if my child is sick with vomiting and diarrhea?***

A. Your child can become dehydrated very quickly. Call your doctor or take your child to the emergency department.

Watch for

- the action pattern of Desmopressin. It will not change much from one day to the next. It doesn't usually last 10 hours one day, and only 4 hours the next day.
- the urine amounts and colour.

Q. ***Everyone says that it is healthy to drink lots of water to flush out your system. Is this true for my son with DI?***

A. No. Your son should drink only when he is thirsty. The body's **natural** ADH hormone supply can adjust to different amounts of water. Desmopressin cannot adjust like this. So, your son's body can't keep the balance between water coming in and water going out. How much goes out depends on how much Desmopressin there is in the body. If he drinks more than his body needs, the extra fluid will not flush through. Instead it may cause the blood to be diluted (called water intoxication) and cause problems like headaches, nausea, seizures and coma. Thirst is a sign that the body needs water. Your son should "listen to his body" and drink only when he is thirsty.

Some forms of Desmopressin are refrigerated; some are not. Read your package insert for directions

Q. ***The Desmopressin doesn't seem to be working as well as it used to. What's wrong?***

A. Maybe ….

- the lining of the nose is not absorbing all the Desmopressin. If your child has a stuffy nose, make sure she blows it before you give the hormone. When the person has a cold, you may need to increase the dose by 25%. Check this with your health care professional first.
- the Desmopressin may have lost strength. It may be old or you may have left it out of the fridge too long. If so, try a new bottle.
- your child may need a dose increase.

The rhinyle may seem strange but the children and teens get used to it very quickly. The effect of the Desmopressin makes life so much better that they don't mind taking the dose this way.

How does one decide on the right dose?

Each person's dose is decided by a trial that starts with a small amount once each day.
The child will feel less thirst and less need to urinate starting about 20 minutes after taking the dose. Then you will wait to see how long the dose lasts. When the dose is "used up" – called **breakthrough** - signs of DI will start up again. The signs are:

- urinating large amounts of urine every half hour or more often
- pale almost colourless urine
- great thirst for water.

"When the Desmopressin wears off my daughter can gulp down an 8-ounce glass of water and ask for another right away. Breakthrough is as clear as that!"

The child takes another dose at breakthrough. If that comes only a few hours after the first dose, this suggests that the dose is not enough for the child. Some children do well with one small dose at bedtime. Most children need a dose about every 12 hours.

It is important to wait for breakthrough to give the next dose. Too much ADH will make the kidneys hold too much water in the body.

"When the Desmopressin wears off my daughter can gulp down an 8-ounce glass of water and ask for another right away. Breakthrough is as clear as that!"

While you and your doctor are finding the correct dose, the doctor may ask you to keep a record of the "ins and outs". That means writing down the **time** and measured **amounts** each time your child **drinks** ("ins") or **urinates** ("outs"). You and your doctor will use this information to adjust the dose of Desmopressin.

What are the side effects of Desmopressin?

Some effects are the result of the kidneys holding back too much water. The child can have a headache and nausea if the dose is too high, or is taken before breakthrough occurs. If your child has these effects, she should not have anything to drink before the breakthrough.

Other side effects include irritation (redness and soreness) of the lining of the nose, and abdominal (stomach) cramps. Tell your doctor if your child has any of these problems.

Questions from parents and teens:

Q. Will this condition go away?
A. Almost always, the condition is life-long. If the condition is the result of swelling in the pituitary area after an injury or surgery, the condition may disappear when the swelling goes down.

Q. My daughter is 10, and had brain surgery to remove a tumour last week. Now she has diabetes insipidus, and is on Desmopressin. She has suddenly started wetting the bed. She is really upset by this. Will she be a bed-wetter now?
A. Bedwetting happens to quite a few children as they are working on finding the "just right" balance of timing and dose for Desmopressin. This stage can take a few weeks but bedwetting will stop once she finds a dose and timing that works for her.

Q. Our child is not taking the dose during school time. Do we need to tell the school about this condition?
A. We suggest that you tell your child's teachers about DI. It is very likely that your child will have a few "breakthroughs" at school. The teacher should know that the need to go to the bathroom as often as every half-hour is real, as is the need to drink water when desired. It helps if the teacher understands what these signs mean and how they are treated. Ask if the school wants a letter from the health care professional to confirm the diagnosis.

Your child should wear a medical alert bracelet at all times. Ask the nurse how to get one or check the Medic Alert website.

How to measure the dose of Desmopressin?

The nurse will show you how to use the directions in the Desmopressin package to measure the dose into the rhinyle. At first, many people find it easier to use a needless syringe to draw up the dose into the rhinyle.

When the dose gets to 10 micrograms, you may want to try the nasal spray. It is easier to use.

Important: The spray must coat the inside of the nostril, not the back of the throat. The person should take a breath and then **hold it while the Desmopressin is sprayed**. Do not breathe in during the spraying.

Mark's story – an example of finding the dose that works best.

I'm 16 years old and I had surgery in the fall for a brain tumour. Soon after that I developed central DI, and started on Desmopressin. My usual dose of Desmopressin is 0.1 mL (10 mcg) by rhinyle before bed and that lasts until 3 pm the next day. Then I take 1 spray (10 mcg) of Desmopressin. That is the same number of micrograms, but it only lasts me 6 hours, and that's perfect, because then it is time for my nighttime dose again. It took me about 2 months to get this routine just right. For a while I was taking a dose in the middle of the night because I had to wait for breakthrough. I can tell when the Desmopressin is wearing off, even before I have to start going to the washroom. I just have a sense of it that I can't describe.

Special notes for parents of infants:

Converting weight to volume:
1 gram of water (or urine) is 1 ml in volume.

For example, your baby's wet diaper weighs 50 gm. You will note 50 ml output on your record.

Note: the weight of a dry diaper must be subtracted.

Your doctor needs a lot of accurate information to decide on the right dose of Desmopressin for a baby who can't tell you about thirst.

The doctor may ask you to measure and record:

- the amount of fluid that baby takes in – milk, water, juice and food. The dietitian can show you how to measure the amount of fluid in baby foods. A breastfeeding baby can be weighed before and after each feeding on a precise scale.

- the amount of fluid that baby puts out. The nurse will show you how to measure by weighing diapers on a kitchen scale available at large pharmacies or department stores, $20.00 to $50.00.

- the urine specific gravity using Multistix. The nurse will show you how to do this, using cotton balls in the diaper.

Your doctor may also ask you to take your baby each week to your local laboratory to measure the *blood sodium (salt) level*.

The normal sodium in the blood is 135-145 mmol/L.

A **low** sodium level means that too much water is being kept in the body - the Desmopressin dose may be too high.

A **high** sodium level means that too much water is being lost- the Desmopressin dose may be too low. This information is important to prevent dehydration (too little water in the body). Babies can become seriously ill very quickly if they lose too much water.

Giving Desmopressin to your baby:

- the intranasal dose is very small so using the rhinyl for nasal Desmopressin is not common
- the pharmacy can make a special dilution to be given by mouth
- the doctor may recommend the tablet form of Desmopressin
- the doctor may recommend a thiazide diuretic and a special formula or breast milk. This is a different approach to D.I. management and you will receive specific home instructions