

A GUIDE TO ARGININE VASOPRESSIN DEFICIENCY



WHAT IS ARGININE VASOPRESSIN DEFICIENCY (AVP-D)?

Arginine vasopressin deficiency (AVP-D) is a condition that causes you to produce large amounts of urine and feel very thirsty all the time. AVP-D was previously referred to as Diabetes Insipidus.

AVP-D occurs because the body can't make enough antidiuretic hormone (ADH, also called arginine vasopressin). ADH and AVP are terms that can both be used and refer to the same hormone. ADH is the pituitary hormone that regulates the water level in your body by controlling the amount of urine your kidneys produce.

If you have AVP-D, ADH doesn't regulate the water level in your body, and you produce too much urine. Symptoms include producing more than three litres of diluted urine per day and constant thirst, even after drinking lots of water.

Healthy adults produce one to two litres of urine per day. A person with severe AVP-D who drinks a lot of fluids can produce up to 19 litres of urine daily.

COMMON CAUSES:

- Tumours affecting the hypothalamus or pituitary stalk
- Complication following surgery on the pituitary
- Brain injuries that damage hypothalamus or pituitary
- Infections or inflammatory conditions that involve the pituitary or hypothalamus
- Autoimmune conditions that damage ADH cells

LESS COMMON CAUSES:

- Cancer that spreads to the brain
- Wolfram syndrome, a rare genetic disorder that also causes vision loss
- Experiencing a loss of oxygen that causes brain damage (eg stroke or near-drowning)
- Can be associated with congenital brain malformations.

Sometimes, there is no apparent cause.

AVP-D can also occur if the body produces the right amount of ADH, but the kidneys do not respond. This type is called nephrogenic diabetes insipidus.

HOW COMMON IS AVP-D?

AVP-D affects only about 1 in 25,000 people.

There is a rare form of AVP-D called adipsic AVP-D. This condition typically occurs after damage to the hypothalamus, resulting in the loss of thirst sensation in affected patients. Managing this condition poses significant challenges as it often leads to rapid and substantial changes in blood sodium levels. Patients with adipsic AVP-D are commonly prescribed a daily fluid intake and are required to regularly measure their body weight. Additionally, frequent blood tests are necessary to closely monitor their condition.

WHAT ARE THE SIGNS AND SYMPTOMS?

- Urinating more than three litres in 24 hours
- Diluted urine
- Constant thirst
- Excessive drinking (polydipsia)
- Preferring cold drinks

Children and babies may also have these signs:

- Heavy, wet nappies
- Bed-wetting
- Trouble sleeping
- Delayed growth
- Vomiting or constipation
- Weight loss

DIAGNOSIS

AVP-D can be diagnosed with the following tests:

- **Blood tests** – these can measure sodium levels and a hormone called copeptin
- **Water-deprivation test** – in this test, you are deprived of fluid for up to 8 hours; the test records your urine volume and weight changes
- **Arginine stimulated copeptin test** - in this test you are given arginine which increases the secretion of vasopressin and copeptin from the posterior pituitary gland (where the antidiuretic hormone is secreted from)
- **Hypertonic sodium infusion test** – in this test you are given extra sodium in order to measure the hormone response to this using copeptin measurement
- **Scans** – a magnetic resonance imaging (MRI) or computerised tomography (CT) scan can confirm or exclude the presence of pituitary disease.

After Diagnosis

After diagnosis, it is essential to see:

- **An endocrinologist** with experience in managing pituitary diseases

TREATMENT

There is no cure for AVP-D, but treatments can relieve symptoms and improve your quality of life.

Treating AVP-D involves taking a medication called desmopressin to replace AVP. Desmopressin can help to relieve excess thirst, decrease urine output and prevent dehydration.

Generally, you take the medication two to three times daily as a nasal spray or as tablets. Taking too much of this medication too often can cause hyponatraemia (low sodium). If you miss a dose, it is best not to take another dose, as that can increase the risk of hyponatraemia. Some doctors recommend leaving out one dose of desmopressin each week as a precaution to minimise the risk of hypotranemia. Your doctor can give you more information about medications and doses. Symptoms of low sodium include lethargy, headaches and confusion.

ONGOING MANAGEMENT

Once you have your treatment plan, you can manage your condition successfully at home. It is essential to see your doctor every six to 12 months. Your doctor will take a general health exam and look for signs of the following:

- Fluid retention
- Hyponatraemia
- Frequent and excess urination.

During times of illness (such as fevers, vomiting and diarrhoea), people with AVP-D can easily develop problems with taking their medication and problems with their sodium level. Thus it is vital to discuss what to do during illnesses with your medications with your specialist and have a "sick day management plan". During acute illnesses, some people have to get to hospital sooner than someone without AVP-D as the sodium level can become abnormal due to the hormonal deficiency of AVP.

MORE INFORMATION

The Australian Pituitary Foundation provides social support for patients and carers, and has published a range of patient resources on pituitary conditions and treatments.

For more information, please visit our website: www.pituitary.asn.au

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REFERENCES

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