

Fact Sheet

Arginine Vasopressin Deficiency

What is Arginine Vasopressin Deficiency (Diabetes Insipidus)?

Arginine Vasopressin Deficiency (AVP-D), formerly known as Diabetes Insipidus, is a rare disorder that can occur as a consequence of histiocytosis involving the pituitary gland.

In 2022, to deter any confusion of diabetes insipidus and diabetes mellitus in treatment, an international effort was led by endocrinology and pediatric societies proposing changing the name of “diabetes insipidus” to “arginine vasopressin deficiency (AVP-D)” for central etiologies (originating in the hypothalamus/pituitary gland), and “arginine vasopressin resistance (AVP-R)” for nephrogenic etiologies (originating in the kidney). AVP-D or Central Diabetes Insipidus, can occur in histiocytic disorders.

AVP-D is a result of damage to the pituitary gland, a small gland at the base of the brain which stores and releases a hormone called ADH (antidiuretic hormone), also known as vasopressin. This hormone normally causes the kidney to control the amount of water released as urine from the body. When the pituitary is damaged, the kidneys lose too much water resulting in increased urination, which then leads to increased thirst.

Among all individuals with diabetes insipidus/AVP-D/AVP-R, histiocytosis (LCH or ECD) can be the underlying cause in 10% of cases. Although the total rate of occurrence is unknown.

The connection between histiocytosis and AVP-D was first reported in the late 1800s. Since then, AVP-D has been recognized as a characteristic feature of Langerhans cell histiocytosis, Erdheim-Chester Disease, and less commonly, Rosai Dorfman Disease and xanthogranuloma.

What are the symptoms of Arginine Vasopressin Deficiency (Diabetes Insipidus)?

- » Dehydration
- » Sticky mouth or reduced tears
- » Change in appetite
- » Fatigue/sleepiness
- » Low body temperature
- » Rapid heart rate
- » Low blood pressure/shock
- » Extreme thirst and frequent urination

What is the treatment for Arginine Vasopressin Deficiency (Diabetes Insipidus)?

AVP-D (DI) that is undiagnosed and untreated can dramatically hurt a patient’s everyday life. Because of the extreme urination and thirst, activities of daily living can be greatly affected. Sleeping through the night is often not possible and travel is difficult. Patients may feel the need to reduce fluid intake; however, this can be dangerous and even life-threatening. Once diagnosed and treated with synthetic vasopressin called DDAVP, symptoms quickly improve, and a better quality of life is restored.

People with AVP-D (DI) should consider educating healthcare professionals when hospitalized about concerns related to fluid restriction and the importance of desmopressin for their health. Rapid changes in blood sodium levels can be dangerous, and in severe cases may cause brain damage. Different protocols for treating hypo/hypermnatremia should be discussed. Please see attached Emergency Care Guide for people with AVP-D (DI), which can be distributed to healthcare providers.

Participation in research into histiocytosis and related conditions, including AVP-D (DI), is important and one way that people impacted by histiocytic disorders can get involved and give back. Visit www.histio.org/research/clinical-trials or contact us at info@histio.org to learn more.

Histiocytosis Association

The Histiocytosis Association is a global nonprofit organization dedicated to addressing the unique needs of patients and families dealing with the effects of histiocytic disorders while leading the search for a cure. It is the only organization of its kind, connecting the patient and medical communities to: *grow and share knowledge of histiocytic disorders, provide critical emotional and educational support to patients and families, and identify and fund key research initiatives that will lead to a world free of histiocytic disorders.*

Histiocytic disorders affect fewer than 200,000 people. It is thus considered an “orphan disease” and, as such, does not receive substantial government funding for research. The Association relies on contributions from corporations, foundations and individual donors to fund critical research, build awareness and conduct community outreach initiatives.

For more information, contact:

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