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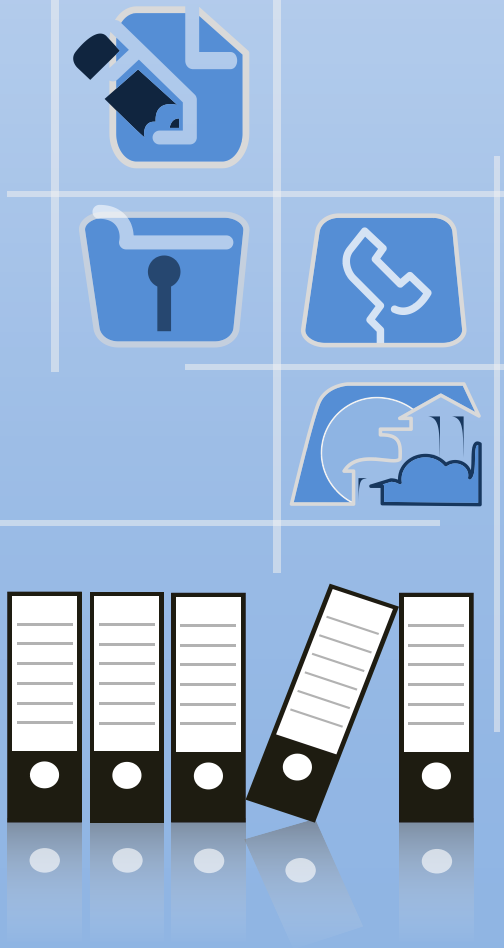
Diabetes Insipidus



Histiocytosis UK

Introduction

Despite the misery it causes, Histiocytosis is too rare a disease to have generated substantial research in medical circles. Unfortunately, for every child or adult fighting for his or her life, the pain and suffering are just as severe for children and adults afflicted with other better known disorders receiving funding.

For the children and adults battling these illnesses, there is now reason to hope. To ensure the research continues, we ask for your help, to complete the funding puzzle.

Our awareness and research programmes provide a beacon of hope for the many children and adults battling Histiocytosis, to ensure this research continues we ask you to pledge your support.

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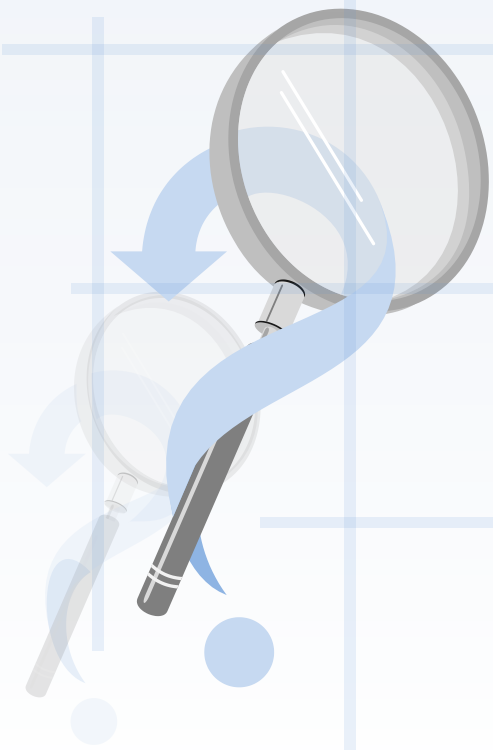

What Is Histiocytosis

What is Histiocytosis?

Histiocytosis is an umbrella term applied to a group of rare diseases, characterised by increased numbers of white blood cells called histiocytes in the blood and tissues. In all forms of histiocytosis, these cells, which are part of the protective immune system, begin to attack the body, targeting many organs of the body including the bone marrow, liver, spleen, lungs, skin, bone and brain.

The prognosis for patients varies greatly depending on the form of histiocytosis.

Please be advised that all the information you read in this document is not a replacement for the advice you will get from your consultant and their team.

Who we are?

Histiocytosis UK is a registered charity dedicated to promoting and funding scientific research into uncovering not only the causes of all histiocytic diseases, which include Langerhans Cell Histiocytosis and Haemophagocytic Lymphohistiocytosis, but also ensuring early diagnosis, effective treatment and a cure.

The Charity aims to support patients and their families by means of information and awareness as well as raise public and professional awareness of histiocytic disorders. Its team of Trustees include the UK's leading paediatric LCH and HLH specialists.

Diabetes Insipidus

What is Diabetes Insipidus?

Diabetes insipidus (DI) is a rare disorder that can occur as a consequence of histiocytosis involving the pituitary gland. It should not be confused with the more common diabetes mellitus, also known as sugar diabetes, which results from too much sugar in the blood. Although both disorders have similar symptoms, in every other way including the cause and treatment, they are completely unrelated diseases. The rate of occurrence for DI is not known, because there has been no organized method to count the number of patients.

Diabetes insipidus 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 (increased urination), which then leads to increased thirst.

The connection between histiocytosis and diabetes insipidus was first reported in the late 1800s. Since then, DI has been recognized as a characteristic feature of LCH. It is known to also occur in other histiocytic disorders, such as Rosai Dorfman and JXG.

It is believed that approximately between 5% and 50% of LCH patients develop DI depending on the extent of disease. The risk of developing DI in patients with multisystem LCH is 4 to 6 times more than those with single-system disease. Patients with skull, facial, and/or eye bone lesions are at much higher risk of developing DI. This risk is increased further if LCH remains active for a longer period or if it recurs.

Diabetes insipidus is recognized by a great increase in the amount of urine passed (often several gallons per day) and an increased thirst. Any patient with known LCH with an increase in drinking habits or passing large amounts of urine should be tested for DI.

Diabetes insipidus is diagnosed with a water deprivation test, which measures changes in body weight, blood values, urine output, and urine composition when fluids are withheld over a several-hour period. It is very important that this test be supervised by a knowledgeable physician in a medical setting. An x-ray test called an MRI scan is sometimes performed to see if there is change in the brain and pituitary area, but this test alone cannot diagnose DI.

Diabetes insipidus is usually a permanent, lifelong condition and cannot be cured. However, the symptoms of constant thirst and urination can be well controlled with treatment with DDAVP, a synthetic kind of vasopressin, and normal, symptom-free quality of life can be restored.

FAQ - DI

1. What is the difference in diabetes insipidus and diabetes mellitus?
Two different types of hormones are involved: diabetes insipidus due to impaired production by the pituitary gland of a hormone called antidiuretic hormone and may occur as a consequence of histiocytosis. Diabetes mellitus, on the other hand, results from too much sugar in the blood, due to impaired insulin production by the pancreas. Although both disorders have similar symptoms of increased thirst and urination, in every other way including the cause and treatment, they are completely unrelated diseases.

2. What are the chances my child will develop diabetes insipidus?
DI occurs in as many as 25% of all patients and as many as 50% of patients with multisystem LCH.

3. Why is it important that the testing for DI be done in a clinic/hospital?
The water-deprivation test is a complicated procedure that requires highly trained medical professionals to perform specialized measurements. The body's water balance must be carefully monitored during the procedure to prevent rapid and dangerous dehydration.

4. How is a water deprivation test done?
This test includes timed measurements (some done every hour and others done every other hour) of blood pressure, pulse, weight, urine, and blood. Fluid is withheld during testing. The test may take up to 8 hours to complete, but it may be stopped sooner, depending on lab results. Further information and instructions will be provided by your physician.

5. Can diabetes insipidus be reversed?
Once DI has been diagnosed, the chance of reversal is uncommon. However, it has been reported in some cases where treatment was started within a few days of symptom onset.

6. Can diabetes insipidus occur before the diagnosis of LCH?
DI can be the first presenting symptom, although one-half of these patients develop LCH lesions within 1 year after the onset of diabetes insipidus.

7. Can diabetes insipidus due to LCH occur when there is no known involvement anywhere else?
Yes. It is believed that this occurs in less than 10% of patients. The diagnosis is made from biopsy of the tumor in the pituitary stalk.

8. Can diabetes insipidus in LCH be prevented?
There is evidence that a rapid start of chemotherapy after onset of multisystem LCH may prevent DI.