About the Association

A Rare Community

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
- Identify and fund key research initiatives that will lead to a world free of histiocytic disorders

Partnership for a Cure

The Histiocytosis Association works closely with the Histiocyte Society, an international organization of over 240 physicians and researchers, which is dedicated to studying the histiocytic disorders. Through this partnership, understanding of this disease has greatly increased, and survival rates and quality of life continue to improve.

Community Outreach

While the search for more effective treatments and a cure continues, the Histiocytosis Association is dedicated to supporting and empowering the patients and families who live with these diseases every day.

Funding the Association

Histiocytic disorders are considered "orphan diseases." An orphan disease is one that affects less than 200,000 individuals in the United States.* Subsequently, these disorders do not receive a high priority for government-funded research. The Association relies on contributions from corporations, foundations and individual donors to fund critical research, build awareness and conduct community outreach initiatives.

Rare Disease Act of 2002

You are not Alone

While the search for more effective treatments, and ultimately a cure continues, the Histiocytosis Association is dedicated to informing and empowering those who live with histiocytic diseases every day.

Whether newly diagnosed and learning to navigate this rare diagnosis or searching for strength and support while caring for a loved one, you can turn to the Histiocytosis Association to connect you with a community who understands what you are going through.

The Histiocytosis Association invites you to become a part of this our rare community. We encourage you to explore our online resources by visiting our website:

www.histio.org

Making a Donation

All donations are tax deductible (Federal Tax ID # 22-2827069). Visit www.histio.org/donate to join us in the pursuit of a cure.

HISTIOCYTOSIS ASSOCIATION
A Rare Community

Histiocytosis Association
332 North Broadway | Pitman, NJ 08071 USA

Phone: +1 856-589-6606 | Fax: +1 856-589-6614
www.histio.org | info@histio.org
Federal Tax ID # 22-2827069

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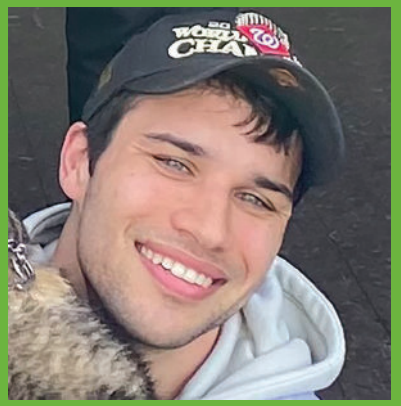

LANGERHANS CELL HISTIOCYTOSIS IN ADULTS

HISTIOCYTOSIS™ ASSOCIATION
A Rare Community™

What is Langerhans cell histiocytosis?

Langerhans cell histiocytosis (LCH) in adults is a rare disorder that occurs when the body produces too many Langerhans cells (histiocytes), which is a type of white blood cell that helps fight infection. While Langerhans cells are found in normal, healthy people, there is an over-production and build-up of these cells which can lead to organ damage in adults with LCH.

LCH is caused by somatic mutations in genes that control how histiocytes, or LCH cells, grow. Cells from LCH tumors (lesions) are found to have cancer-causing DNA changes (mutations); these include mutations of the BRAF, MAP2K1, and ARAF genes. These mutations may cause LCH cells to grow inappropriately in areas where they are not usually present, and damage tissues or form lesions (tumors). Previously thought to be an auto-immune disorder, LCH was classified as a blood cancer in 2008 by the World Health Organization.

It has been estimated that LCH occurs in 1 to 2 per million individuals. Among adults, the average of age of diagnosis of LCH is around 40 years.

Isolated lung involvement is referred to as pulmonary LCH (PLCH), which can be observed almost exclusively in smokers. If organs such as bones, skin or lymph nodes, etc. are affected individually, this is referred to as single system LCH. The involvement of multiple organ systems is called multisystem LCH.

Recently, it has been discovered that about 60% of patients have a BRAF gene mutation, which may provide the opportunity for targeted therapies.

Will LCH patients recover?

The prognosis of LCH has continued to improve with introduction of newer treatments and early diagnosis. As they may be at higher risk, it is important for patients to continue to follow up with their healthcare provider to monitor for second cancers or side effects of treatment

For patients with PLCH, discontinuing smoking is an essential part of the treatment. Despite consistent smoking cessation, chemotherapy or targeted therapies, some patients develop severe and debilitating lung diseases.

What tests are done to diagnose LCH?

Diagnosis of LCH is made following a biopsy and microscopic evaluation of the affected tissue. Other tests that may be carried out to determine the extent of the disease include blood and urine tests, PET-CT and/or MRI scans. Other testing may be done depending upon symptoms.

Some patients may develop a condition called diabetes insipidus, more recently known as Arginine Vasopressin-Deficiency. This condition is characterized by excessive thirst and urination.

Symptoms of LCH

Symptoms of LCH in adults vary greatly from patient to patient. It is also possible to have disease in a particular location without noticable symptoms. Possible site involvement include:

- Skin (rash)
- Bone (single or multiple lesions)
- Lung, liver, spleen (dysfunction)
- Teeth and gums (loose/lost teeth, swollen gums)
- Ear (chronic infections or discharge)
- Central nervous system issues (problems balancing,depression, learning issues)
- Pituitary gland - causing diabetes insipidus *(characterized by excessive thirst and urination)*

Pain has been experienced by many adults with LCH. While pain can be caused by bone lesions or bone defects that do not heal completely with therapy, it has also been observed that some patients have pain when there is no active disease seen on imaging. The cause of this pain is not understood by scientists.

It should be emphasized that not all patients have all symptoms. A more detailed list is available at www.histio.org/langerhans-cell-histiocytosis-in-adults/

How is LCH treated?

Treatment of LCH depends upon the individual patient and is planned after thorough testing to determine the extent of the disease. It may include no treatment with the lesion being watched for signs of remission or minimal types of treatment with surgery or low-dose chemotherapy. In patients with more extensive disease, such as when multiple organ systems are involved, systemic chemotherapy is most often recommended. The treatment approach to LCH has evolved significantly due to discovery of gene mutations leading to the successful use of targeted therapies in most cases. A team of health care providers should plan treatment for LCH.

To learn more about the Association and its Scientific Initiatives, Outreach Efforts and Research Program visit www.histio.org.

The Histiocytosis Association is dedicated to raising awareness about histiocytic disorders, providing educational and emotional support, and funding research leading to better treatments and a cure.

A world free of histiocytic disorders.

www.histio.org