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

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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.

During the last century, there has been an ongoing debate about whether LCH is a condition triggered by an inflammatory process or is a rare cancer.

Today, it is assumed that the underlying cell originates from the bone marrow and the detection of certain mutations of these cells suggests LCH as a rare cancer. However, the true primary cause of LCH still remains a mystery.

It has been estimated that LCH occurs in 1 to 2 per million individuals.

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.

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

Will LCH patients recover?

Most patients survive LCH. Some will remain symptom free, while others may develop life-long problems. Overall, physicians will be able to discuss each patient's likelihood of responding and doing well, but it is often difficult for doctors to make definite predictions since LCH has clearly shown itself to be an unpredictable disease.

For patients with PLCH, discontinuing smoking is an essential part of the treatment. In many cases this can lead to a stabilization of the symptoms and even an improvement. Despite consistent smoking cessation, chemotherapy or targeted therapies, some patients develop severe and debilitating lung diseases.

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/

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.

Most often an oncologist or hematologist takes the main role in treating LCH patients. Sometimes a team approach may be appropriate, and the main physician may enlist the help of various types of specialists such as surgeons, pulmonologists, dermatologists or endocrinologists.

How is LCH treated?

Treatment varies from patient to patient. There is no widely agreed-upon treatment. It depends upon the individual patient and extent of areas of involvement. Some patients may have limited involvement, which does not progress to other areas and may not need any further treatment after a diagnostic biopsy (e.g. isolated cervical lymph node LCH). Others may need surgery, steroids or irradiation for localized disease. Multisystem LCH usually requires systemic chemotherapy. If certain mutations of the LCH cell are detected, targeted therapies can be applied, although the significance of this treatment is currently still unclear and is the subject of ongoing studies.

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