HISTIOCYTOSIS ASSOCIATION™
A Rare Community

Fact Sheet

Langerhans Cell Histiocytosis (LCH)

What is Langerhans cell histiocytosis?

Langerhans cell histiocytosis (LCH) occurs in patients when the body accumulates too many immature Langerhans cells, a subset of the larger family of cells known as histiocytes. A Langerhans cell is a type of white blood cell that normally helps the body fight infection. In LCH, too many Langerhans cells are produced and then build up in certain parts of the body where they can form tumors or damage organs.

The cause of this disease is unknown, although many possibilities have been explored, including viruses, exposure to toxins in the environment, family history and geography. LCH is not caused by a known infection. It is not contagious, nor is it believed to be inherited. Scientific discussions on the definition of LCH continue to be debated in terms of its classification as either an immune dysfunction or a rare cancer (neoplastic and malignant or not malignant). There remain differing opinions among experts as to whether it is definitively a cancer or not.

LCH is believed to affect less than one in 200,000 children, but any age group can be affected. It occurs most often between the ages of 1 to 3-years-old. It is, however, believed to be under-diagnosed. Some patients may have no symptoms at all, while others have symptoms that are mistaken for injury or other conditions/diseases.

LCH was first described in medical literature in the mid to late 1800's. Through the years it has been known by various names, such as histiocytosis-X, eosinophilic granuloma, Abt-Letterer-Siwe disease, Hashimoto-Pritzger disease and Hand-Schuller-Christian syndrome. In 1973, the name Langerhans cell histiocytosis was introduced. This name was agreed upon to recognize the central role of the Langerhans cell.

What are the symptoms of Langerhans cell histiocytosis?

Symptoms of LCH may include:

- Skin (rash)
- Bone (single or multiple lesions)
- Lung, liver, spleen (dysfunction)
- Pituitary gland - causing diabetes insipidus (characterized by excessive thirst and urination)
- Ear (chronic infections or discharge)
- Loose or lost teeth/swollen gums

What is the treatment for Langerhans cell histiocytosis?

Some cases of LCH with limited involvement may not require treatment. For patients with more extensive disease, chemotherapy may be necessary. Treatment depends upon the individual patient and is planned after thorough testing to determine the extent of the disease. Primarily, hematologists and oncologists treat children with LCH.

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:

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