

BI-LO Charities Children's Cancer Center Langerhans Cell Histiocytosis

Langerhans cell histiocytosis (LCH) occurs when the body accumulates too many immature Langerhans cells—a type of white blood cell that normally helps the body fight infection. The excess cells can form tumors or damage organs.

The cause of LCH is unknown, but it is believed to occur in one out of every 200,000 children. Any age group can be affected, but LCH occurs most often between the ages of 1 and 3. It may appear as a single lesion or it can affect many body systems, including skin, bone, lymph glands, liver, lung, spleen, brain, pituitary gland and bone marrow.

Symptoms depend on the location and severity of the condition. It usually is diagnosed with a tissue biopsy in addition to other testing such as X-rays and blood studies. A biopsy of an involved site is necessary to make a definitive diagnosis.

While some cases of histiocytosis may not require treatment, patients with more extensive disease may need chemotherapy.

LCH in the skin, lymph nodes or pituitary gland usually improves with treatment and is referred to as low risk. LCH that involves the spleen, liver, bone marrow or lung is classified as high risk and may be more difficult to treat. Some cases, such as those in the bones, can be either low or high risk. Most patients with LCH survive the disease.

Some patients may develop long-term side effects such as diabetes insipidus, stunted growth, loss of teeth, bone defects, hearing loss or neurologic problems, while other patients do not experience side effects.

Factors affecting the chance of recovery and options for treatment include the extent of the disease, whether "risk organs" (liver, spleen, lung, bone marrow) are involved and how quickly the disease responds to initial treatment.

Patients with LCH usually need long-term follow-up care to detect late complications of the disease or treatment. These complications may include problems of skeletal deformity or function, liver or lung problems, endocrine abnormalities, dental issues, or neurological and neurocognitive dysfunction.