What is Langerhans Cell Histiocytosis (LCH)?

Langerhans Cell Histiocytosis is a rare disease, which can affect children of any age. The name of the disease is derived from the type of body cell involved, the histiocyte, - osis meaning increased numbers. Langerhans refers to the doctor who first described normal Langerhans cells.

A histiocyte is a normal white blood cell, which is part of the immune system and is found in many parts of the body. It is found mostly in the bone marrow, the blood stream, the skin, the liver, the lungs, the lymph glands and the spleen. Its job is to help destroy certain foreign materials and fight infection.

In LCH there are an increased number of histiocytes. Instead of helping to protect the body, these cells in increased numbers can cause tissue damage.

LCH is divided into two groups:

• Single system LCH, when the disease affects only one part of the body
• Multi system LCH, when it affects more than one part of the body

It is also possible for single system to progress to multi system.

The cause of LCH is unknown.

What are the signs and symptoms?

Symptoms may vary. Besides the more common symptoms such as fever, weight loss and fatigue, the following symptoms might develop, isolated or in combination.

Skin
• severe scalp rash like cradle cap
• groin rash like severe nappy rash

Bone
• painful swelling in a bone (lump) commonly skull or limb
• loose teeth if it affects the jaw bones
• ear infection or a discharge from the ear if it affects skull bones near the ear

Lymph glands
• swollen glands

Liver and spleen
• enlarged liver which can cause jaundice (yellowing of the skin and eyes) or abdominal swelling
• enlarged spleen which can cause abdominal swelling

Lungs
• shortness of breath and cough

Eye
• vision problems

Endocrine glands
• Diabetes Insipidus is the most common hormone abnormality, particularly in multi system LCH and is characterised by excessive thirst and urination.

How is Langerhans Cell Histiocytosis diagnosed?
Diagnosis is usually made by taking a small sample (a biopsy) of the tissue involved for examination in a laboratory.

What other tests are done?
To find out the extent of the disease, several other tests may be required. These include blood tests, X-rays, CT scan (computer tomography, a computer-assisted x-ray), biopsy of the liver or bone marrow.
How can Langerhans Cell Histiocytosis be treated?

Treatment is planned after thorough evaluation of your child, to find out the extent of the disease. In some cases the disease will disappear without any treatment at all. In others a combination of treatment is used.

Your child's doctor will talk with you about the best treatment for your child, and both you and the doctor will make the decision about treatment.

Treatments used are:

- Chemotherapy (cancer drugs). Pamphlets with specific information will be given to you for each drug.
- Steroids i.e. Prednisolone which plays a role in suppressing the body’s immune system. Common side effects include stomach irritation, nausea and vomiting, increased appetite, weight gain and mood swings. More detailed information is available in a pamphlet; please ask nursing or medical staff.
- Small doses of radiation may be used to treat diabetes insipidus but is considered carefully due to its side effects.
- Limited surgery

The goal of an overall treatment plan is to use as little treatment as possible to keep the disease under control, allowing the body to heal by itself.

Is it cancer?

No. Over the years LCH has been treated with cancer drugs (chemotherapy). Therefore, oncologists, specialists who treat cancer, also treat children with LCH. However, the disease is not a cancer, but it is a chronic illness that requires ongoing medical support.

How long will my child's treatment last?

The length of treatment depends on the number of parts of the body affected by the disease. In some cases the disease will disappear without any treatment at all. In others it might need 12 months of chemotherapy.

The doctor will talk to you about your child's individual treatment plan and the length of treatment involved.
Will my child recover from the disease?

Most children who develop LCH will recover from it. The older the child, the higher the chance of recovery. Sometimes the disease can come back (recur) and so the doctor will regularly follow up your child.

Long-term side effects can possibly develop depending on the treatment given. The doctor will be able to discuss this with you.

References:

- Histiocytosis Association of America
  www.histio.org/association/library

- Histiocyte Society
  www.histio.org/society

- Sydney Children's Hospital - Centre for Cancer & Blood Disorders
  www.kids-cancer.org/langerhans.htm

- Harvard Medical School's Consumer Health Information
  www.intelihealth.com

Please refer to section 7 “Sources of More Information” or check the parent’s library in McGuiness & McDermott children’s clinic for further information.