

What is the Histiocytosis Research Trust?

The Histiocytosis Research Trust was set up as a registered charity in 1991 by Dr Jon Pritchard and Paul Kontoyannis and is dedicated to promoting and funding scientific research into uncovering not only the causes of histiocytosis diseases, but also ensuring early diagnosis, effective treatment and a cure.

The H R Trust also aims to support patients and their families as well as raise public and professional awareness about histiocytic disorders. It has strong links with The Nikolas Symposium, The Artemis Association, The Histiocyte Society, Euro-Histio-Net, and the Histiocytosis Association of America.

How You Can Help

Research into LCH has been very limited due to lack of funds and all our research to date has been the result of donations and fundraising. In order to achieve our objective to find a cure as well provide practical support for both patients and their families, we need to continue raising as much money as we can. To achieve this, we need your help.

By raising money and working together, our chances of securing a cure will be even greater.

LCH is indeed rare but for the few it affects, it causes devastation, fear and sometimes, death. But together we can find a cure.



Langerhan's Cell Histiocytosis in Children

Together we'll find a cure

Tel: +44 (0)7850 740241
www.hrtrust.org
Email: info@hrtrust.org
PO Box 435, Leeds LS17 1GE
United Kingdom
Registered in England & Wales
Charity No. 1004546

Langerhan's Cell Histiocytosis (LCH) is a rare disease – about 50 children each year are diagnosed in the UK. Doctors and scientists often call it an 'orphan' disease because it is so rare. It presents with many different symptoms and as a result can be difficult to diagnose.

Children suffering from LCH have too many histiocytes – a histiocyte is a type of white cell which normally helps to fight infection in the skin. However, in those suffering from LCH, the histiocytes gather together in large numbers causing damage to healthy parts of the body, such as the bone marrow, skin, lungs, liver, lymph glands, spleen, pituitary gland and brain. Why this happens, we do not know.

LCH has had several different names over the years - Eosinophilic Granuloma, Hand-Schüller-Christian disease, Abt-Letterer-Siwe disease and Histiocytosis X.

We don't know what causes LCH – it is *not* hereditary and *not* infectious.

LCH is divided into two main groups – single system and multi-system.

When LCH is described as a 'single-system' disease, it means that it only affects one system in the body – for example skin, bone or an organ. If it is only in one place in that particular system, it is single site and if in more than one, multi-site or multi-focal. So a child with several affected areas in the bones but no disease elsewhere, is considered to have 'multi-focal, single-system' disease.

When LCH is found in more than one 'system', for example in both the skin and bones, it is described as 'multi-system' disease. Children with 'multi-system' disease affecting the liver, spleen, lung or bone marrow can have a more serious form of LCH. This is then described as multi-system disease with “risk organ” involvement and may require more intensive treatment.

It is important to remember that the vast majority of children will recover completely from LCH with a 90% survival rate. Some children, however, may be left with life-long problems and in a small number of multi-system cases the disease can be life-threatening.

Sometimes the disease comes back, but unlike cancer, treatments for LCH that have worked before may be used again.

Symptoms of LCH

LCH varies from patient to patient and can affect one or several parts of the body, for example:

Bone – bumps or indentations, broken bones or sore limbs, headaches

Skin – persistent cradle cap, nappy rash or a rash in the folds of the skin

Lung, liver, spleen, bone marrow – breathing problems, jaundice, anaemia

Eye – bulging of the eye or rarely vision problems

Teeth and gums – loose/lost teeth, swollen gums

Intestines – diarrhoea, sickness and poor growth

Pituitary Gland – hormonal problems which can cause excessive thirst and urination or poor growth

Brain, spinal cord – learning difficulties, unsteadiness, lack of co-ordination

Children may also have general symptoms such as fevers, weakness or failure to gain weight. Some children can seem very well, which can make the diagnosis even harder to take in.

Diagnosis and Treatment

Doctors may carry out a number of tests in order to diagnose LCH. These tests are needed to see if the disease is 'single-system' or 'multi-system' and to determine the best form of treatment.

These include blood tests, x-rays, ultrasound scan, CT scan, MRI scan, biopsy of the tissue and hormone tests.

All of these tests can take either a few days or a couple of weeks and may involve a stay in hospital.

LCH is different to cancer because it usually burns itself out – although we don't know how or why this happens. In some cases, the disease will go away by itself without any treatment. In other cases, treatment is needed to keep the disease under control and prevent too much damage. The treatment is tailored to the extent of the disease. Treatment options can include surgery, steroids and chemotherapy.