Langerhans’ cell histiocytosis (LCH)

This information sheet from Great Ormond Street Hospital explains the causes, symptoms and treatment of Langerhans’ cell histiocytosis and where to get help.

Langerhans’ cell histiocytosis (LCH) is a rare condition. Histiocyte cells normally help protect the skin, but sometimes the body has too many of them and they move around the body, causing damage.

LCH can affect bones or organs and the symptoms present in a number of different ways. These can range from a skin rash and lumps on the skull to a swollen tummy, breathing difficulties and diarrhoea.

About 50 children in the UK are diagnosed each year. It can affect children of any age. It is more common in boys than in girls.

What causes Langerhans’ cell histiocytosis?

The cause of LCH is unknown. It is not hereditary. It is not cancer and the good news is that there is a high survival rate.

Children with LCH are more likely to get lung disease triggered by smoking so all family members of children diagnosed with LCH must not smoke. The child or teenager diagnosed with LCH should never smoke themselves either.

The immune system contains cells called histiocytes. Langerhans’ cells are a specific type of histiocyte that help fight infection in the skin. When a child has LCH, these cells spread through the bloodstream to other healthy parts of the body where they can cause damage.

LCH can be divided into two groups. Single-system LCH is when only one part of the body is affected. Multi-system LCH is when multiple parts of the body are affected.

What are the signs and symptoms of Langerhans’ cell histiocytosis?

The symptoms of LCH depend on many factors, including which part of the body is affected and whether it is single-system or multi-system.

LCH often takes the form of a single lesion in the bone, but it can involve many organs and sometimes in infants it presents is a similar way to leukaemia.

The symptoms can be quite general:

- LCH in the bone can cause pain in the bone and maybe lumps on the skull
- LCH in the skin can cause persistent nappy rash and cradle cap
LCH in the abdomen can cause fever, swollen tummy, diarrhoea and maybe jaundice.
LCH in the lungs may cause breathing difficulties.

For a few children diagnosed with LCH, the pituitary gland can be affected. This gland sits in the brain and is responsible for hormones in the body. Most children present with symptoms of wanting to drink all the time and need to urinate frequently. In rare cases, growth can also be affected.

How is Langerhans’ cell histiocytosis normally diagnosed?
As the symptoms for LCH are quite general and it is very rare, sometimes many doctors are visited before the diagnosis is suspected. A biopsy is needed to confirm the diagnosis. Then other tests are done to see which organs are involved these include x-rays, scans, blood tests and urine tests.

How is Langerhans’ cell histiocytosis normally treated?
In some cases, the biopsy may trigger healing itself and treatment is not needed.

Although not cancer, the condition is usually treated by oncologists (cancer specialists), this is because they work with nurses trained to give chemotherapy and chemotherapy works well in the treatment of LCH.

Usually the treatment is mild and can be given on day care. Rarely young children become very sick and need inpatient treatment.

Chemotherapy can cause side-effects such as nausea, vomiting, hair loss and tiredness. The child’s doctor will discuss this with parents before treatment begins. It is important to remember that the side-effects last only as long as the treatment.

What happens next?
Most children are cured from LCH. There is a 90 per cent survival rate. About a quarter of children with multi-system disease have the disease reactivate after stopping treatment and need to be treated again. Treatment is effective even if it has to be started again.

Children can be left with ongoing problems as a result of the condition but this depends on which organs were affected. Rarely the liver is damaged to the extent that the children require liver transplant. Children with pituitary disease require ongoing life long hormone therapy. Children who have very damaged lungs may require a lung transplant.

Regular check-ups are recommended in case the condition returns. Follow-ups can also identify any side effects that may appear over time.
Further help and advice
Talk to the child’s doctor or health visitor.

Contact the Histiocytosis Research Trust for advice and support:

Histiocytosis Research Trust
Tel: 07850 740 241
Email: info@hrtrust.org
Website: www.hrtrust.org

Notes

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