This information is about a condition called Langerhans cell histiocytosis (LCH) in children. LCH is classified as a cancer and sometimes requires treatment with chemotherapy. About 50 children in the UK develop LCH each year. It can affect children of any age, and is more common in boys than in girls.

More children than ever are surviving childhood cancer. There are new and better drugs and treatments, and we can now also work to reduce the after-effects of having had cancer in the past.

It is devastating to hear that your child has cancer, and at times it can feel overwhelming but there are many healthcare professionals and support organisations to help you through this difficult time.

Understanding more about the cancer your child has and the treatments that may be used can often help parents to cope. We hope you find the information here helpful. Your child’s specialist will give you more detailed information, and if you have any questions it is important to ask the specialist doctor or nurse who knows your child’s individual situation.

This factsheet is published in conjunction with CCLG’s booklet entitled: ‘Children and Young People with Cancer: A Parent’s Guide’.

Langerhans cell histiocytosis (LCH)

LCH is an unusual condition. It has some characteristics of cancer but, unlike almost every other cancer, it may spontaneously resolve in some patients while being life-threatening in others. LCH is classified as a cancer and sometimes requires treatment with chemotherapy. LCH patients are therefore usually treated by children’s cancer specialists (paediatric oncologists/haematologists).

Langerhans refers to Dr Paul Langerhans, who first described the cells in the skin which are similar to the cells found in LCH lesions.

Histiocytosis refers to histiocytes which are cells that are part of the immune system, and are found in many parts of the body. There are two types of histiocytes:
- macrophage/monocyte cells - these destroy harmful proteins, viruses and bacteria in the body
- dendritic cells - these stimulate the immune system

Langerhans cells are dendritic cells and are normally only found in the skin and major airways. In LCH, the abnormal dendritic cells that look similar to Langerhans cells may be found in different parts of the body, including the bone marrow, skin, lungs, liver, lymph glands, spleen and pituitary gland. When these abnormal dendritic cells accumulate in these tissues, they may cause damage.

LCH is divided into two groups:
- single-system LCH - when the disease affects only one part of the body, for example the skin or the bone
- multi-system LCH - when it affects more than one part of the body

Causes

The cause of LCH is unknown. It cannot be caught from other people and is not passed on in families.

Signs and symptoms

The symptoms of LCH will depend on which part of the body is affected and whether the disease is affecting more than one part of the body. The lymph glands may be enlarged, and the child may be irritable and have a poor appetite. Pain in the bone and/or swelling and lumps on the skull can occur if LCH is affecting the bone. A skin rash such as cradle cap or nappy rash may occur if the skin is affected. A discharge from the ear or hearing problems can occur if the ear is affected. The child may have breathing difficulties if LCH affects the lungs or chest. Tummy problems such as diarrhoea and liver problems including jaundice can occur if LCH is within the abdomen.
In 10–20% of patients with multi-system disease, the pituitary gland at the base of the brain is affected, causing hormonal problems. This can lead to the child passing larger amounts of urine and being very thirsty. This is called diabetes insipidus, which is different from sugar diabetes and can be well-controlled with specific medicines. Occasionally, other pituitary hormones may be affected, causing poor growth or delayed puberty, which can also be treated.

**How LCH is diagnosed**

A variety of tests and investigations may be needed to diagnose LCH. Tests are likely to include the removal of a sample of cells from an affected part of the body (a biopsy). This is usually done in an operation under a general anaesthetic. The cells are then examined under a microscope. X-rays are taken of the bones, the skull and the lungs. Blood and urine tests will also be done. Additional scans and tests may be required depending on which parts of your child’s body is affected. These tests help the doctors decide whether the disease is a single-system or multi system type.

When your child is having the tests, they may need to stay in hospital. Any tests and investigations that your child needs will be explained to you.

**Treatment**

Single-system LCH may disappear on its own without any treatment. In some children, treatment such as surgery and corticosteroids (such as prednisolone) may be used. Multi-system LCH is usually treated with chemotherapy and corticosteroids. The length of treatment varies from child to child.

Chemotherapy is anti-cancer (cytotoxic) medication that can destroy cancer cells, and corticosteroids are hormonal substances that are naturally produced in the body. Both chemotherapy and corticosteroids can help to destroy the LCH cells.

**Side effects of treatment**

Chemotherapy and corticosteroids used to treat LCH may cause side effects, and your child’s doctor will discuss these with you before treatment starts. Any possible side effects will depend on the particular treatment being used.

Side effects of chemotherapy can include feeling sick (nausea) and vomiting, hair loss, an increased risk of infection, bruising and bleeding, and tiredness. Side effects of corticosteroids can include mood changes and an increased appetite, which can lead to weight gain. All of these side effects should get better when treatment stops.

**Clinical trials**

Many children have their treatment as part of a clinical research trial. Trials aim to improve our understanding of the best way to treat an illness, usually by comparing the standard treatment with a new or modified version. Research has found that people receiving treatment within clinical trials tend to do as well as, or better than, similar patients treated outside clinical trials. This does not mean that trial treatments are always better. However, hospitals that carry out trials do have access to good equipment and, like all hospitals, they follow precise guidelines when giving treatment.

Most treatment trials for LCH are set up and organised by The Histiocyte Society, a group of international specialists in histiocytosis. If appropriate, your child’s medical team will talk to you about taking part in a clinical trial and will answer any questions you may have. Written information is often provided to help explain things.

Taking part in a research trial is completely voluntary, and you’ll be given time to decide whether it’s right for your child.

**Follow-up care**

It is important to know that the vast majority of children will recover completely from LCH. Some children however, are left with persistent/recurring problems and for a very small number of patients with multi-system LCH, it can be a life-threatening condition. After successfully completing treatment, patients will have follow-up clinic appointments. LCH sometimes comes back (‘reactivates’) and may need treatment again. If this happens, treatments for LCH that have worked for them before may be effective again. The same or different treatment may then be required. Patients are also monitored for possible permanent consequences of the disease (e.g. a low production of certain hormones, hearing problems, lung problems) and may need treatment for these conditions.

If you have specific concerns about your child’s condition and treatment, it’s best to discuss them with your child’s specialist medical team who know the situation in detail.
Your feelings

As a parent, the fact that your child has cancer is one of the worst situations you can be faced with. You may have many emotions, such as fear, guilt, sadness, anger and uncertainty. These are all normal reactions and are part of the process that many parents go through at such a difficult time. It is not possible to address in this factsheet all of the feelings you may have. However, the CCLG booklet ‘Children & Young People’s Cancer: A Parent’s Guide’, talks about the emotional impact of caring for a child with cancer and suggests sources of help and support.

Your child may have a variety of powerful emotions throughout their experience of cancer. The Parent’s Guide discusses these further and talks about how you can support your child.

Useful organisations

Children’s Cancer and Leukaemia Group (CCLG)
www.cclg.org.uk
CCLG supports the 1,700 children who develop cancer each year in the UK. It gives support to healthcare professionals involved in caring for children with cancer and is instrumental in the development of high standards of care.

CLIC Sargent
www.clicsargent.org
CLIC Sargent provides clinical, practical, financial and emotional support to children with cancer.

The Histiocytosis Association
www.histio.org
The Histiocytosis Association is an international partnership of patients, families, physicians and friends, which aims to promote scientific research and education related to histiocytosis. It aims to provide solutions to some of the problems that are specific to patients suffering from this disease, and to offer support to patients and their families.

Histiocytosis Research Trust
www.histiocytosis.org
The Histiocytosis Research Trust is a group of patients, families, doctors and scientists dedicated to supporting patients and their families, raising awareness of histiocytic disorders and funding research to find a cure. They are in touch with many families around the UK and overseas. It also publishes the ‘Langerhans Cell Histiocytosis (LCH): A Parent’s Guide’.

References

This factsheet has been compiled using information from a number of reliable sources, including:


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