This fact sheet is about a condition called Langerhans' cell histiocytosis (LCH) in children. We hope it answers your questions. If you have any further questions, you can ask a nurse or doctor involved in your child's treatment.

If you'd like to discuss this information with our cancer support specialists, call the Macmillan Support Line free on 0808 808 00 00, Monday–Friday, 9am–8pm. If you're hard of hearing you can use textphone 0808 808 0121, or Text Relay. For non-English speakers, interpreters are available. Alternatively, visit macmillan.org.uk

Includes the following information

- Langerhans' cell histiocytosis (LCH)
- Causes of LCH
- Signs and symptoms
- How LCH is diagnosed
- Treatment
- Side effects of treatment
- Late side effects
- Clinical trials
- Follow-up
- Your feelings
- Useful organisations
- Related Macmillan information

Langerhans' cell histiocytosis (LCH)

LCH is not strictly a cancer, but rather a cancer-like condition that may be treated with chemotherapy. In its more serious forms, LCH can behave like a cancer and is therefore usually treated by children's cancer specialists (paediatric oncologists).

Langerhans' refers to Dr Paul Langerhans, who first described the cells in the skin from which LCH develops.

Histiocytes are cells which are part of the immune system and are found in many parts of the body. There are two types of histiocytes: macrophage monocyte cells, which destroy harmful proteins, viruses and...
bacteria in the body; and dendritic cells, which stimulate the immune system.

Langerhans' cells are dendritic cells and are normally only found in the skin and major airways. In LCH, the Langerhans' cells are abnormal and spread via the bloodstream into many parts of the body, including the bone marrow, skin, lungs, liver, lymph glands, spleen and pituitary gland. When Langerhans' cells are present 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.

About 50 children in the UK develop Langerhans' cell histiocytosis (LCH) each year. It can affect children of any age, and is more common in boys than in girls.

Causes of LCH
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.

If LCH affects the bone, this can cause pain in the bone and/or swelling/lumps on the skull. If the skin is affected, a skin rash, such as cradle cap and nappy rash, may occur. A discharge from the ear or hearing problems can occur if the ear is affected. If LCH affects the lungs or chest, the child may have breathing difficulties. LCH within the abdomen may cause tummy problems, such as diarrhoea, and liver problems, including jaundice. The lymph glands may be enlarged. Children may be irritable and have a poor appetite.

In 2–3 out of every 10 children 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 again can be treated.

How LCH is diagnosed
A variety of tests and investigations may be needed to diagnose LCH. X-rays will often be taken of the bones, the skull, and the lungs. Blood tests will also be taken. These tests help the doctors to decide if the disease is a single-system or multi-system type.

Tests are likely to include the removal of a sample of cells (biopsy), and this is usually done in an operation, under a general anaesthetic. The cells are examined under a microscope. An MRI (magnetic resonance imaging) scan of the brain may also be carried out.

Any tests and investigations that your child needs will be explained to you. The booklet A parent’s guide to children’s cancers, gives details of what the tests and scans involve.

Treatment
Single-system LCH may disappear on its own without any treatment. This may occur following a biopsy. In some children, treatment such as surgery and corticosteroids (such as prednisolone) may be used.

Multi-system disease is usually treated with chemotherapy and corticosteroids. The length of treatment varies from child to child.

Chemotherapy is the use of anti-cancer (cytotoxic) drugs to destroy cancer cells, and corticosteroids are hormonal
substances which 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 upon the particular treatment being used and the part of the body that is being treated.

Side effects of chemotherapy can include: feeling sick (nausea) and vomiting, hair loss, an increased risk of bruising and bleeding, and tiredness. Corticosteroids may cause a rounding of the face (sometimes called a moon face), mood changes, and an increased appetite, which can lead to weight gain. All of these side effects are reversible when treatment stops.

**Late side effects**

Some children may develop after-effects of the disease. These effects can happen within months or years of the diagnosis and are more likely to be due to the disease itself than to the treatment given. However, a few children may develop late effects from the drugs or the treatment they receive.

Possible late side effects include reduction in bone growth, delayed puberty, hearing problems, and lung problems.

**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; hospitals that carry out trials do however have access to good equipment and follow precise guidelines when giving treatment.

The 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 if it's right for your child.

**Follow-up**

Between eight and nine of every 10 children who develop LCH will recover from it. Sometimes the disease can come back, and so children will have regular check-ups in the outpatients clinic.

Follow-up is also important to look out for late side effects. 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 a serious illness is one of the worst situations that you can be faced with. You may have many different 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's not possible to address in this fact sheet all of the feelings you may have. However, the Macmillan/CCLG booklet *A parent’s guide to children’s cancers* talks about the emotional impact of caring for an unwell child and suggests sources of help and support.
Your child may have a range of powerful emotions throughout their experience of illness. The parent's guide discusses this further and talks about how you can support your child.

Our booklet Peppermint Ward is a storybook for younger children with cancer. It looks at the issues they and their family may face, and helps them explore their feelings.

Useful organisations

Children's Cancer and Leukaemia Group (CCLG)
University of Leicester, 3rd Floor, Hearts of Oak House, 9 Princess Road West,
Leicester LE1 6TH
Tel 0116 249 4460 (main office)
Email info@cclg.org.uk
www.cclg.org.uk
Coordinates research and care for children and their parents. There are 21 CCLG specialist centres for the treatment of childhood cancer and leukaemia, covering all areas of the UK and Ireland (there's a map of the centres on the website). Has information about the CCLG, childhood cancer and leukaemia.

CLIC Sargent
Griffin House, 161 Hammersmith Road,
London W6 8SG
Tel 0300 330 0803
Email info@clicsargent.org.uk
www.clicsargent.org.uk
Offers practical support to children and young people with cancer or leukaemia, and to their families.

The Histiocytosis Association
www.histio.org
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.

Related Macmillan information

- A parent's guide to children's cancers
- Katie's Garden (a storybook for primary school-age children about a girl's experience of cancer)
- Peppermint Ward – a story about children with cancer
- Understanding cancer research trials (clinical trials)
- Understanding chemotherapy
- Understanding radiotherapy

For copies of this related information call free on 0808 808 00 00, or see it online at macmillan.org.uk

Our website click4tic.org.uk has cancer information written specifically for young people. Get advice and support, and connect with other young people affected by cancer.

This fact sheet has been written, revised and edited by Macmillan Cancer Support's Cancer Information Development team. It has been approved by our medical editor, Dr Terry Priestman, Consultant Clinical Oncologist.

With thanks to: Dr Kevin Windebank, Senior Lecturer in child health & Consultant paediatric and adolescent oncologist; and the people affected by cancer who reviewed this edition.
This fact sheet has been compiled using information from a number of reliable sources, including:


This fact sheet was revised in 2011. The next edition will be available in 2013.