

LANGERHANS CELL HISTIOCYTOSIS (LCH)

A PARENT'S GUIDE

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INTRODUCTION

This booklet provides information and advice on the condition Langerhans Cell Histiocytosis (LCH) and how to deal with the diagnosis and subsequent treatment. It is not a replacement for the information you will get from your consultant and his or her team.

LCH varies a lot, so you need to focus on your child and how the disease is affecting him or her. Your consultant will be able to help with the specific questions about your child's particular disease and treatment. It is a good idea to write down things you want to know, so that you remember everything you would like to ask when you see your medical team. It is also useful to have a notebook to write down what you are told, as it can be difficult to take everything in at the time. There are a few extra pages at the back of this booklet for this purpose.

This booklet has been divided into three sections:

- INFORMATION ABOUT LCH
- HOW TO COPE
- WHERE TO GET SUPPORT

Many of the terms you will come across may be unfamiliar, at least initially. There is a detailed glossary at the end of the booklet.



Medical Illustration Leicester

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LCH is so complicated, it is difficult to explain it briefly to casual friends like other mums at the school – I just say that it is a disease treated with chemo.

THE DISEASE



What is Langerhans Cell Histiocytosis (LCH)?

Langerhans Cell Histiocytosis is a rare disease – about 1 in 200,000 children become ill with it and there are about 50 new cases each year in the UK. Adults can also be affected. Doctors and scientists often call it an 'orphan' disease because it is so rare. As a result, research into why it happens and how it can best be treated has been limited.

LCH is one of several diseases known as the 'histiocytoses' or 'histiocytic disorders' as they involve cells in the body known as histiocytes. A histiocyte is a type of white blood cell which normally helps to fight infection. 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, skin, lymph nodes, lungs, liver, spleen, bone marrow, and pituitary gland. Why this happens, we do not yet know.

Is LCH a cancer?

This question has been the subject of debate and scientific study for a long time. There is now evidence that the histiocytes in most LCH lesions have genetic abnormalities, similar to the abnormalities seen in some cancer cells. These findings support the notion that LCH is a cancer. However, the way LCH affects people ranges from being very aggressive and resistant to treatment, to mild forms of the disease which may resolve spontaneously. This is unlike almost all cancers. LCH is classified as a cancer but more research is being undertaken to improve our understanding of the origin and mechanism of this disease.

THE DISEASE CONTINUED

How does LCH affect patients?

Some patients have no symptoms at all and the disease is found 'by accident' when they are investigated for something unrelated. More often however, they have symptoms caused by the LCH affecting a particular part of the body. The most common 'system' affected is the skeleton and this is the case in about 80% of patients. Bone LCH usually causes pain and/or swelling and may affect one or several bones. The bones of the skull, spine and limbs are most commonly affected. Skin involvement is present in about a third of patients and the skin is the 2nd most commonly involved organ. Skin LCH can mimic numerous skin conditions including 'cradle cap' and 'nappy rash'. Less commonly LCH may affect the ears, lungs, liver, spleen, brain, lymph nodes ('glands') and rarely the gut. Some patients develop symptoms due to a lower production of certain hormones (e.g. excessive thirst and excessive fluid intake due to diabetes insipidus) and in a small number of patients LCH causes a reduction in the number of blood cells (haematological dysfunction). Lower numbers of blood cells may make them feel tired, appear pale and result in them being more susceptible to bruising/bleeding and infection. In addition to symptoms caused directly by the effect of LCH on specific organs/parts of the body, general symptoms such as fever, weakness or failure to gain weight may also be present.

Although it is possible for LCH to affect people in many different ways it is not uncommon for LCH patients to appear well - this can make the diagnosis even harder to take in.

What happens to patients after they had LCH?

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, most 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) and may need treatment for these late effects.

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

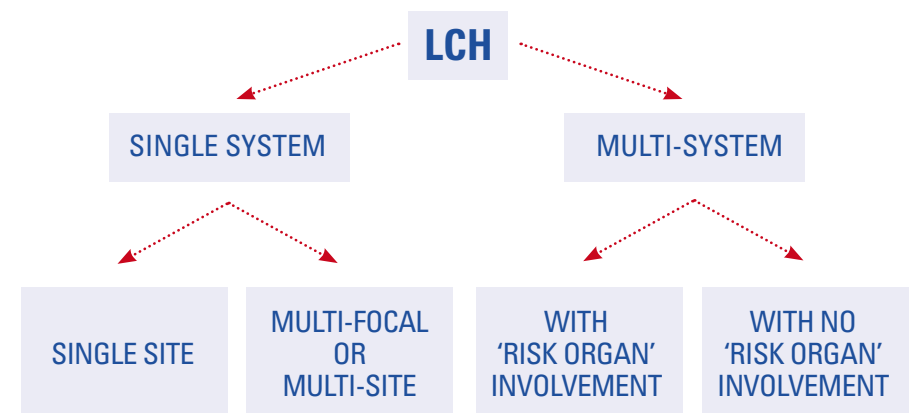
Single system disease

When LCH is described as a 'single system' disease, it means that it is only affecting one system in the body – for example, skin, bone or an organ. The majority (>70%) of patients have single system disease. If the LCH is only present in one place in that particular system, it is called single site and if in more than one place, it is called multi-site or multi-focal disease. Therefore a child with several affected bones, but no disease elsewhere, is considered to have 'single system, multi-focal' disease.

Multi-system disease

When LCH is found in more than one 'system', for example, in both the skin and bone, it is described as 'multi-system' disease. Children with 'multisystem' disease affecting the liver, spleen or bone marrow, are considered to 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.

This classification helps doctors to decide what treatment is required and for how long it should be given.



THE DISEASE CONTINUED

How is LCH diagnosed?

The diagnosis of LCH is usually made by performing a biopsy of an affected part of the body. A biopsy is the removal of a small piece of tissue while a patient is under an anaesthetic. This piece of tissue is then examined under a microscope.

Doctors will be carrying out a number of further tests. These tests are done to see how LCH affects that part of the body and if any other systems are involved (multi-system disease). This information helps the medical team to decide on the best treatment for your child.

Some of the following tests may be carried out on your child:

- **Blood tests** these are done to check how many of the of different types of blood cells there are and to assess how well the liver and kidneys are working
- **Urine test** to evaluate how concentrated/dilute the urine is
- **X-ray** pictures of the chest and of the bones (also known as a skeletal survey).
- **Ultrasound Scan** – sound waves are used to build up a picture of the inside of the body. A clear gel is spread over the skin and a microphone is passed over the body. The sound waves bounce off the organs inside the body and are picked up by the microphone. A computer turns the sound waves into pictures.
- **CT Scan** – A CT (Computerised Tomography) scan takes a number of x-ray pictures of the body from different angles and uses a computer to convert them into cross-sectional x-ray pictures or 'slices' of the body. Depending on how big a part of the body is being scanned this can take between a few seconds and a few minutes. CT scans are painless, but small children may need to be sedated or anaesthetised ('put to sleep as for an operation') to ensure they remain still while the scan takes place.



- **MRI Scan** – Magnetic Resonance Imaging (MRI) Scan uses magnetic and radio waves to take pictures of the body. It is painless, but usually takes longer than a CT scan and is quite noisy. As it is important to keep very still, small children are normally sedated or given an anaesthetic.
- **Biopsy** – the removal of a small piece of tissue from an organ or part of the body for microscopic examination. Most often the biopsy is done of affected bone or skin but biopsies may also need to be carried out on the liver, lung or bone marrow.
- **Water Deprivation Test** – if doctors suspect there is a hormone problem known as diabetes insipidus, your child may need to undergo a water deprivation test. This is a test to measure how much urine is made and how concentrated it becomes when no water is given to a patient for a certain amount of time (also called fluid deprivation test)

To complete all of these tests can take between a few days and a couple of weeks and may involve a stay in hospital.

THE DISEASE CONTINUED

How is LCH treated?

LCH is different to almost all cancers because in some cases it may 'burn itself out' without any treatment. We don't know how or why this happens.

In other cases, treatment is needed to get the disease under control and prevent too much damage. Treatment may involve an operation (surgery) and/or medication. The treatment will be tailored to your child and will depend on the extent and sites of their disease.

Main treatment options:

• **Surgery**

The diagnosis of LCH is made by taking a small sample of tissue from an affected part of the body (biopsy). If there is only a single bony lesion, the biopsy is sometimes combined with curettage (scraping out of the abnormal tissue in the lesion). These procedures may initiate a process of healing in the LCH lesion and sometimes this is all that is required.

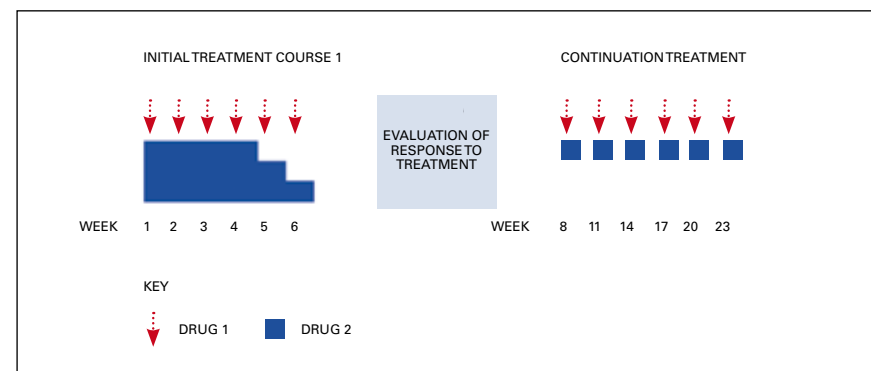
• **Medication**

Steroids – corticosteroids are medicines used to reduce inflammation and stop the body's immune system acting in the wrong way. Your child may be given prednisolone (a type of steroid) which is usually taken by mouth. Steroids can also be injected into an LCH lesion.

Chemotherapy – this is used to destroy LCH cells and is generally given intravenously. This means that the chemotherapy medicine is diluted in fluid and given straight into a vein (into the bloodstream). Some chemotherapy medicines may be taken by mouth.

Your oncologist or haematologist will decide on a treatment plan for your child once all the relevant tests are completed. A protocol is a special plan that will detail your child's treatment if medication is required. There are international treatment protocols, developed over many years through large clinical trials (clinical trials are explained in more detail on page 18).

The protocol will describe the frequency, timing and length of the different elements of treatment. An example of what a protocol flow sheet (summary) may look like is shown in the diagram below. This shows the different drugs, and the timing of their administration. It also shows the duration of the various elements of the treatment. Your child's treatment plan may change since it is dependent on how he or she responds to the treatment. You will be given a copy of your child's flow sheet so that you can follow progress throughout the treatment.



If your child needs chemotherapy, he or she will probably need a central line. This is a tube that is inserted into a large blood vessel and tunnelled under the skin to where it can be easily accessed. It allows blood samples to be taken and treatment, including chemotherapy, to be given easily and painlessly. The central line is put in under general anaesthesia and can stay in place until the end of your child's treatment. Your hospital will provide more information about the types of central lines they offer.

If your child is unwell or requires very intensive treatment he or she will need to be in hospital. Most children however, receive all their intravenous treatment by visiting their hospital's out-patient or day-care facility. This may only be available at your treating oncologist/haematologist's hospital or it may be available at your local hospital under a shared care arrangement with a local doctor. Some treatment, like that taken by mouth, can be given at home.

While on chemotherapy and for up to 6 months afterwards, your child will be susceptible to infections. This is known as being immunocompromised. If he or she develops a fever or becomes unwell you need to contact your treating team immediately for advice. Your child may need immediate hospital admission and treatment with antibiotics.

What are the side-effects of the medication?

The main drugs used to treat LCH and their common side effects are described below.

Different drugs cause different side effects. Everyone is different and will react to treatment in a different way. Some children may have very few side effects while others will have a lot. Almost all side effects are only short-term and will gradually disappear once the treatment has stopped.

“The steroids heightened his behaviour – it was like normal toddler behaviour but with the volume turned up.”

“We call them ‘roid rages’ – the tantrums she gets on steroids.”

- **Steroids** – *Prednisolone/Prednisone* - this is a type of corticosteroid (steroid) that can reduce inflammation and suppress the immune system. It is usually given by mouth, in the form of pills to swallow, pills that dissolve in water, or as a liquid medicine.

Possible side-effects include: irritation of the stomach lining (indigestion/discomfort/pain), increased appetite, weight gain, changes in behaviour (mood swings/difficulty in sleeping/anxiety/irritability), increase in blood sugar level (like someone with diabetes), high blood pressure, increased risk of infection due to suppression of the immune system, impaired wound healing, irregular or absent periods, and inflammation of the pancreas.

- **Chemotherapy** – Drugs used to treat cancer. These drugs work by killing cells that are in the process of multiplying (by dividing and forming new cells).

The main areas of the body that may be affected by chemotherapy are those where cells rapidly divide and grow. Examples include the lining of the mouth causing a sore mouth, the digestive system causing diarrhoea, skin and hair, causing hair loss, and the bone marrow (spongy material that fills the bones and produces new blood cells), causing low blood counts.

THE DISEASE CONTINUED

There are three main types of blood cells:

- Red blood cells which carry oxygen around the body
- White blood cells which fight infections
- Platelets which help the blood to clot to prevent bleeding and bruising

Chemotherapy reduces the production, and therefore, the number of blood cells in the body. Too few red blood cells cause anaemia, and the person becomes tired and pale. If there are too few white blood cells, particularly ones called neutrophils (neutropenia), the person is at increased risk of infection. Too few platelets means the person is at increased risk of excessive bleeding when injured, having nose-bleeds or bruising easily. If the counts get really low, your child may need a red cell transfusion or platelet transfusion. Only a small number of LCH patients require transfusions, but in some more severe cases, transfusions play an important part in supporting patients. Transfusions are also needed to support those patients who undergo intensive chemotherapy.

As the chemotherapy affects your child's immune system, he or she may need antibiotics to help fight infections. This means, if your child has a temperature of 38°C or more or becomes unwell (even with a normal temperature), you should immediately contact your medical team. Your child may need tests, antibiotics and admission to hospital for a few days. You may also be asked to avoid using paracetamol and ibuprofen since these drugs lower high temperatures and may mask an infection. In addition, if your child is in contact with someone who has chickenpox, shingles or measles, then you should let your hospital know as these are potentially dangerous infections for a child with a low (suppressed) immune system. Both your child and the rest of your family should ideally have the annual flu jab during the autumn/winter.

Chemotherapy can cause nausea and vomiting. Anti-sickness medicines, known as anti-emetic drugs, are given to control this side effect.

Hair loss can occur, but this does not always happen with the chemotherapy used to treat LCH. Young children often get used to hair loss fairly quickly but for parents it is far more traumatic, reminding them that their child is ill. Fortunately, all hair loss from chemotherapy is temporary and hair growth returns once treatment has stopped.

The chemotherapy normally used to treat LCH does not affect fertility, but you should discuss your child's particular treatment with your oncologist/haematologist for more information.

Chemotherapy can cause increased skin sensitivity to sunlight, therefore avoid intense sunlight and use sun block during treatment.

The two most commonly used chemotherapy drugs for LCH are *Vinblastine* and *6-Mercaptopurine*.

Vinblastine – is a chemotherapy drug that is injected into your child's blood stream through the central line. This drug is administered by a 'push' rather than a slow drip through. The process only takes about 10 minutes, but your child will be in hospital for longer since a number of checks need to be carried out before the drug is given. Once completed, your child will be able to go home.

In addition to the side-effects previously described, *Vinblastine* can sometimes, though rarely, cause tingling sensations in the hands and feet, hoarseness, constipation, muscle weakness and bone pain. It may also cause damage to the skin if it is injected directly into a vein (rather than through a central line) and then leaks into the skin.

6-Mercaptopurine – is a chemotherapy drug taken either as tablets or in a liquid form. It is usually taken at home.

In addition to the other side effects already mentioned, 6-mercaptopurine may affect the liver and this will be monitored by your doctor.

Sometimes other drugs are used to treat LCH, for instance when the above mentioned drugs are not successful or when the disease returns. Should this become necessary, your doctor will discuss the different options with you.

What Are Clinical Trials?

Clinical trials are an essential step in the development of treatment for various diseases and are necessary for improving the outcome for patients. Clinical trials are the only way to find out if a new treatment approach to a disease is better than the standard treatments currently used. Patients volunteering to take part in a trial are randomly assigned to either receiving the standard treatment or a different or experimental treatment. This is known as a randomised trial and is the most common way in which treatments are compared. This process (randomisation) ensures that the groups are similar in every way other than the fact that they receive different treatments. Any difference in outcome between the groups is then likely to be caused by the difference in the treatment and not by other differences between the groups. If an experimental treatment proves to be better than the standard treatment and does not cause unacceptable side-effects, it is recommended as the new standard treatment for future patients. This is how new treatments or approaches to treatment are introduced to patients.

The conduct of clinical trials is carefully regulated to ensure patients' well-being is protected. Patients will only be able to participate in a trial if there is an open trial at that time, if they meet the eligibility criteria, and if their treating hospital is taking part. Parents, and when appropriate children, must understand the implications of taking part in a clinical trial and give written consent (agreement) to allow them to be enrolled on a trial. If a trial is available to your child your doctor will explain it in more detail and you will be given the necessary information to allow you to decide whether to take part or not. While on a clinical trial your child will be very closely monitored.

The Histiocyte Society has run several international trials in search of new and more effective treatments for LCH, including three large randomised trials (LCH I, LCH II and LCH III). The Histiocyte Society is a non-profit organisation whose members comprise an international group of over 200 doctors and scientists. It is committed to improving the lives of patients with histiocytosis by conducting clinical and laboratory research into causes and treatment. The international Histiocyte Society LCH study (LCHIV) was launched in 2012.

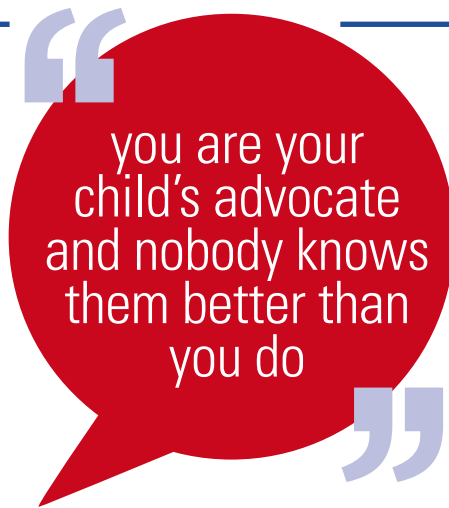
How do I cope with the diagnosis?

LCH can be difficult to diagnose and can be mistaken for other things. You may have been afraid that your child has a serious form of cancer and feel relieved that LCH is likely to be curable. You may feel angry that it wasn't picked up earlier. You will probably feel confused as you won't have heard of LCH before and worried and scared about what it means for your child. The consultant and the rest of the medical and nursing team are there to help you through this very difficult time. They will explain the disease and the treatment your child will need.

It is very likely at this stage that you will not take in much of the information you are given. You may be in shock and find it hard to function as normal. Don't worry if you have not asked all of the questions you meant to. You will have plenty of opportunities to do so later. Quite often, parents feel overwhelmed by the enormity of what has happened and cannot deal with anything else at this time.

As parents, being able to have even a few hours alone together to get used to the news and to talk about what has happened, before starting to tell everyone else, can be very helpful. If you are a single parent dealing with this news, it is helpful if you have a close friend or family member with whom you can share your worries and fears.

Telling friends and family can be emotionally difficult, and also takes a huge amount of time. It may help to tell one family member or friend and ask them to ring round others. Some parents also set up a webpage on one of the specialised websites such as Caringbridge or Carepages (see 'Other Links' section at the end). This means that the latest information can be quickly passed on to friends and family who desperately want to know what's happening, while not taking up all your time and energy having to repeat everything. It also gives people an easy way to let you know they care, through leaving messages in the guest books. Despite your best efforts, you will probably still find the telephone rings constantly. Don't feel bad about using the answerphone. Let people leave messages and call them back if and when you want to.



How do I get the information I want?

When you are worried about your child and meeting a doctor that you may not know very well, it is easy to forget what you meant to ask, and what you have been told. If you can, try to take notes when you attend the main meetings with the consultant. It might be better to take a friend or relative along with you, as they might be able to pay more attention to some of the discussion and take notes for you. It is often the case that you will remember different pieces of information and you may have different questions or concerns. Write down any questions beforehand, so that you don't forget them, and make sure you know whom to contact if you have any more questions, or urgent queries.

How do I listen to and talk to my child/their siblings about this?

A child of any age will notice a tense atmosphere and hushed conversations. It is very important that you talk to your children about what is happening. What you actually say to your child and their siblings will depend on their age. For younger children, it may be enough to say that they have some bad cells and the doctors are giving them some medicine to make the bad cells go away.

Older children and teenagers may want to know all that you know about their illness and may well have been with you when you were told the diagnosis. Being open and honest with them and talking together with the consultant about their questions will help you all to deal with the diagnosis. It may also be helpful for them to talk privately about their worries with members of the medical team, such as the doctor, social worker or psychologist. Some children may consider themselves mature enough to make their own decisions regarding their treatment and should therefore be involved.

Siblings may feel ignored and side-lined due to your child's illness. Their lives have changed a lot due to their sibling's illness and they may start to feel resentful about all the attention their sibling is receiving. They may also be extremely worried and upset to see what their sibling is going through and it can raise difficult questions in their minds. You will need to give them opportunities to ask these questions and bear in mind that they might find it easier to talk to another adult (close friend or family) as they may not want to upset you.

There are many things you can do to help your family get through any difficult times during treatment. Some suggestions that have helped other families are:

- You and your partner spend time individually with your other children.
- Make sure that when your ill child receives presents from visitors, you even things up by giving something to your other children.
- Try and keep discipline consistent, even if it means relaxing things a little for everyone.

How do we deal with the treatment?

Parenting a sick child can be a very difficult experience. While it is good to keep things as normal as possible, worries about infections and the physical effects of the chemotherapy can make that impossible. How you change what you do depends on you and your family, on how your child is reacting and on advice from your doctors. For example, if your child loses their appetite, you may find that you are not so strict on table manners. Likewise, it may be fine for your child to attend nursery, while another family will prefer to keep them away from large groups of children.

COPING CONTINUED

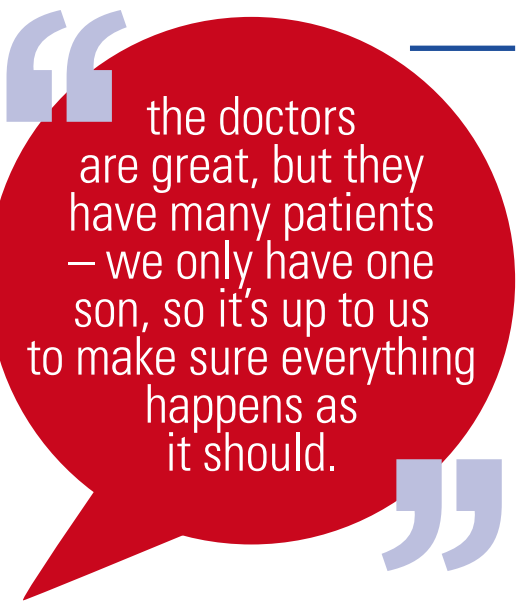
If your child is at school or nursery, you should talk to the staff as soon as you can. You could also write to them with the facts about your child's illness and ask that they pass on the information to other members of staff and parents of your child's classmates. This is particularly important if your child is immunosuppressed, as you will need to be told if anyone who is in contact with your child develops chicken pox, shingles or measles. Your child may be assigned an outreach / community nurse from the hospital who can meet with staff at your child's school or nursery to explain more about the disease and what they need to be aware of. This is important, as you will need to rely on the school or nursery to recognise when your child is very tired or ill.

Having a child with LCH can at times feel like a full-time job all by itself - just keeping track of medications, arranging and attending hospital appointments, getting test results, and generally making sure your child is getting the best treatment.

As well as the time spent dealing with hospital appointments and related tasks, the emotional trauma can make it hard to keep up with all the usual everyday jobs and work. Don't be shy to ask for help – friends and family are often grateful to be able to do something concrete, so ask them to make a meal, do the ironing, mow the lawn, pick up the prescriptions. There may also be organisations that can help, such as local charities that will offer support or services such as doing your shopping. CLIC Sargent provides social workers and community young adult workers to support patients and their families, so ask for a referral in your hospital or contact them yourselves.

There are variations in what your local health service may offer. Some areas offer children's community nurses (or paediatric home nursing teams) who will come to your house to take blood tests, change dressings etc, which will save you a hospital trip. Your local pharmacist may be able to supply some of the less specialised medications, on prescription from your GP, which can be more convenient than using the hospital pharmacy. There may also be a 'shared care' arrangement with a doctor in your local hospital so that you can go to a closer hospital for blood tests, chemotherapy or if your child gets an infection.

Your child's appetite may be affected by the chemotherapy and steroids and can change a lot depending on where they are in the treatment schedule. If you are worried about your child's nutrition, talk to your medical team who will be able to advise you, or refer you to a dietician.



During the period of treatment you will have ups and downs. Although the first shock of diagnosis will fade and you will feel that at least action is being taken to deal with the illness, worries about your child's progress and prognosis will come back from time to time, particularly if things don't go according to plan. It can be difficult to maintain the stability of your child's home environment and minimise the impact on your family. Help and support from close family and friends, online support networks or your community group can all help to keep your spirits up. Doing fun activities as a family can help distract you and give you all a break from the worry and stress.

Treatment can range from just the initial biopsy or surgery in some children, to 6 months or even 2 years chemotherapy in other cases. It is helpful when you are given some idea of what treatment is planned and how long it might carry on for, so that you can try and organise your lives to cope with this situation. Talk to your Consultant about this but keep in mind that it may need to change depending on your child's response to treatment.

What happens at the end of treatment?

Even though the treatment is stressful, many parents find the end of treatment difficult. Some parents have said they felt the disease had only been kept at bay while the drugs were being given. When treatment ends, it can feel as if the 'safety net' has been taken away, so you feel more worried about the return of symptoms. This uncertainty can be eased a little by the regular follow-up that your child will receive and by talking to your medical team about any concerns you have.

If the disease recurs, known as reactivation, most patients will again respond well to treatment. Your consultant will discuss the treatment options with you. Reactivation occurs in about one third of patients who had disease in more than one system/site but reactivation is rare in most other patients.

What resources are there for sibling or family support?

Families should be able to gain support from the group of professionals who work with their child's consultant. They are known as a multi-disciplinary team (or MDT) and usually include nurses, social workers, play therapists and psychologists (refer to page 27-28). Many centres also run patient, sibling and parent support groups.

Your GP will be kept informed by the hospital, but it is also useful to keep him or her up to date yourself. They can be a useful source of support for the whole family as he or she may be able to refer you for specialised support locally if needed.

Counselling can be useful for you and/or your children. Your GP may be able to refer you to a counsellor, or a child psychologist if required. 'Relate' provides counselling for couples, which can help to mitigate the strain that can be placed on your relationship with your partner.

There are a number of different charities, such as CLIC Sargent and Barretstown that offer holidays for families with children with serious diseases.

It is not always easy to find the help you need and you may need to ask a few people before you find the right support for you. As well as talking to your GP, try talking to the medical team at your hospital or shared care hospital, your community nurse, paediatric oncology outreach nurse specialist or CLIC Sargent social worker. There are a number of organisations listed in the 'Organisations and Links' section at the end of this booklet.

Where can I get financial support?

In the UK the NHS covers all medical treatment and tests free of charge. Prescriptions for medications are free for children. Having a sick child, however, inevitably puts additional financial pressure on families (travel, parking, loss of income etc).

Children and young people who receive chemotherapy can apply for Disability Living Allowance. CLIC Sargent has information on their website regarding benefits, parental leave and flexible working: www.clicsargent.org.uk. You can also contact the Disability Benefits Centre or the Citizens Advice Bureau for further information.

It is vital that you talk to your employer to explain what is going on and find out what they can provide in terms of flexibility.

A carer's allowance may also be available. This is means-tested, so depends on your income. For further information, talk to your outreach nurse or social worker.

CLIC Sargent offer grants and can apply to other charities to help with the additional financial costs, such as unpaid leave from work, after school care for siblings and travel. More information on these grants can be found on their website, or through your social worker.

How do I find other parents?

One of the real difficulties for parents of children with a rare disease, such as LCH, is being able to talk to other parents, who have been through, or are going through the same experience as you. Your local medical team may be able to put you in touch with other families who have been affected by LCH.

The Histiocytosis Research Trust (HRT) periodically arranges 'Road Shows' around the country where patients and parents can meet clinicians who have a specific interest in LCH. This is also an opportunity to meet other families affected by the disease. The HRT aims to provide the most current information on LCH and to support patients and their families. For more information visit www.histiocytosis.org.uk and www.hrtrust.org.uk

The Histiocytosis Association based in USA has an online forum, which provides a useful source of support and advice from other parents. For more information visit www.histio.org.uk

WHERE TO GET SUPPORT

Your multidisciplinary team:

Paediatric oncologist/haematologist – these are doctors who treat children with cancers and blood disorders.

Nurses - there will be a sister, ward manager or charge nurse in charge of the ward in the hospital. Staff nurses, student nurses and health care assistants work under their direction. There may also be a community nurse who will visit your child at home.

Surgeon - they carry out operations and can take a biopsy or put in a central line.

Radiologist - they undertake scans and may also do biopsies.

Shared care hospital – although your child's main care may be administered by a specialist hospital, it may be more convenient for you to go to your local hospital for more routine treatments such as regular administration of chemotherapy, blood tests and dealing with infections. This can be a very effective resource, saving long journeys to the specialist centre and also provide medical help close at hand in case of infections. This is referred to as a shared care hospital and your consultant will arrange this for you, if appropriate.

Paediatric Oncology Outreach Nurse Specialists (POONS) – these are specialist nurses who can provide support for you as you care for your child at home. They can visit your child's school with you to explain your child's condition and treatment to the teaching staff. They may also act as a liaison point between you and your hospital.

Psychologist/Counsellor – they may be able to help children who have difficulty with behaviour or learning during their treatment. They also can support your family.

CLIC Sargent social workers – they are funded by the charity CLIC Sargent to support children and young people with cancer and their families. They provide this support to LCH patients and their families. Social workers can help with practical, emotional and financial problems families may face. This help includes help with obtaining the non-means tested Disability Living Allowance and a disabled person's parking badge if appropriate.

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Medical Illustration Leicester

WHERE TO GET SUPPORT CONTINUED

Play specialists – they use play to help children cope with the experience of being ill. They can help provide toys, jigsaws, DVDs and arts and craft activities during stays in hospital.

Hospital school teachers – they can provide educational support for your child during extended stays in hospital.

Community nurses – they are available in many areas and may be able to visit you at home, usually to take blood tests and monitor your child. This can be very useful as it can help minimise trips to the hospital.

Dieticians - can provide advice on what foods your child should eat to ensure they are getting all the nutrients they need.

GP (General Practitioner)/ Family doctor - as LCH is very rare, your own GP is unlikely to know much about LCH, but your hospital doctors will write regularly to your GP to keep him or her up to date with your child's progress. They will continue to provide health advice and support for you as a family.

ORGANISATIONS AND LINKS



“ there are several organisations worldwide dedicated to improving our understanding of LCH... ”

ORGANISATIONS AND LINKS



The Histiocytosis Research Trust

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.

Phone: +44 7850 740241
www.hrtrust.org



Artemis Association

The Artemis Association is a group of parents, patients, doctors and friends based in Greece. Its objectives include supporting patients and their families, encouraging and supporting research and keeping up to date with the latest treatments.

Phone: +30 210 45 20 453
www.histoartemis.gr



Histiocytosis Association (HA)

The HA is based in the United States and is dedicated to raising awareness about histiocytic disorders, providing educational and emotional support, and funding research leading to better treatments and a cure.

Phone: +1 856 589 6606
www.histio.org



Histiocyte Society

The Histiocyte Society is a group of more than 200 physicians and scientists from around the world committed to improving the lives of patients with histiocytic disorders by conducting clinical and laboratory research into the causes and treatment of these diseases.

Phone: +1 856 589 6606
www.histiocytesociety.org



Nikolas Symposium

The Nikolas Symposium is an annual scientific conference hosted in Greece and funded by the Kontoyannis family in honor of Nikolas Kontoyannis who had severe multi-system LCH as a child and lives with the late effects of the disease. The symposium brings together scientists, pathologists and clinicians in search of a rational cure for the Histiocytic disorders.

Phone: +30 210 45 20 453
wwwwww.niksym.org



LCH Belgium

Een onafhankelijke patiëntenvereniging voor kinderen en volwassenen met de ziekte Langerhanscelhistiocytose

wwwwww.lch.be



Histio Net

This is a reference network for Langerhans Cell Histiocytosis and associated syndromes. Several partners – medical experts in the field of care for Langerhans Cell Histiocytosis (LCH), patients and support groups from the European Union (EU) and from outside the EU – cooperates to share and disseminate knowledge and experience. Their objective is to participate in improving care of patients with LCH and other rare diseases belonging to the same 'family' of diseases.

www.eurohistio.net

OTHER USEFUL LINKS



CCLG (Children's Cancer and Leukaemia Group)

CCLG is the UK professional body for those working in the area of childhood cancer. LCH is usually treated by this group of clinicians and the CCLG hosts a Histiocytosis Interest Group which has specific expertise in LCH. The CCLG produces a variety of leaflets (e.g. 'A Guide to Clinical Trials – For Parents and Young People', 'How to help brothers and sisters'), all of which are available online and in hard copy. The CCLG website has numerous links to other sites that may be useful.

Phone: +44 116 249 4460 (Main Office)
www.cclg.org.uk

WE ARE MACMILLAN. CANCER SUPPORT

Macmillan

Macmillan provides practical, medical and financial support for families and promotes and campaigns for better cancer care. Macmillan also helps patients and their families with LCH and has a range of booklets and leaflets available (including information on LCH and chemotherapy).

Ask Macmillan: +44 808 808 00 00
www.macmillan.org.uk



CLIC Sargent

CLIC Sargent is a UK cancer charity for children and young people (up to the age of 25 years), and their families. They provide clinical, practical, financial and emotional support and aims to help the whole family deal with the impact of the disease.

Free Child Cancer Helpline:
+44 800 197 0068 9am-5pm (UK)
www.clicsargent.org.uk

OTHER USEFUL LINKS

www.carepages.com

Carepages websites are free patient blogs that connect family and friends during a health challenge.

www.caringbridge.org

Caringbridge provides free websites that connect family and friends during a serious health event. You can share health updates, and receive messages of support.

www.citizensadvice.org.uk

Citizens Advice Bureau can advise on employment issues.

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GLOSSARY

Absolute Neutrophil Count (ANC) – this refers to the number of a particular type of white blood cell (neutrophil) per volume of blood. Neutrophils are important for fighting infections

Acute – disease or symptoms of rapid onset or short duration

Alanine transaminase (ALT) – this is a blood test used in the diagnosis and study of acute liver disease

Alkaline phosphatase (ALP) – this is an enzyme found throughout the body. High blood levels can indicate liver disease or bone problems

Allergy – an extreme sensitivity to a substance, which may need to be avoided

Alopecia – hair loss

Anaemia – occurs when the number of red blood cells per volume of blood is reduced

Anaesthetic – this is used to sedate or temporarily cause sleep so that surgery can be carried out painlessly

Analgesic – medication used to reduce pain

Antibiotic – medications that destroy or stop the growth of bacteria and so helps treat bacterial infection

Anti-emetic – medicines that controls nausea and vomiting

Anti-fungal – medications that destroy or stop the growth of fungi and so helps to treat fungal infection

Anti-pyretic – medications that reduce fever (pyrexia) by lowering the body temperature, for example, paracetamol

Aspartate transaminase (AST) – this is a blood test used in the diagnosis and study of acute liver disease

Biopsy – the removal of a small piece of tissue from an organ or part of the body for microscopic examination

Blood Count (BC) – the numbers of different blood cells in a volume of blood. A full blood count (FBC) measures the amount of haemoglobin, red cells and white cells as well as platelets per volume of blood

Blood group – this is how a person's blood may be classified and the most common types are A, B, AB and O

Blood transfusion – this involves giving blood previously donated by one person to another person

Bone marrow – this spongy material in the centre of the large bones produces all the different blood cells

Catheter – a thin flexible tube used to pass fluid into the body or to drain fluid out

Cell – the living units from which animals and plants are built (for example, blood cells, brain cells). They are so tiny that thousands of cells could sit on a pin head

Central line – a tube inserted under the skin into a large vein to make the giving of medication and the taking of blood samples easier

Central Nervous System (CNS) – the brain and the spinal cord

Chemotherapy (Chemo) – chemical substances (medication) used to treat cancer as well as LCH

Chronic – describing a disease of long duration, often of gradual onset. The term does not imply anything about the severity of a disease

GLOSSARY CONTINUED

Computerised Tomography (CT) scan – takes a number of x-ray pictures of the body from different angles and uses a computer to convert them into cross sectional x-ray pictures or 'slices' of the body. A higher radiation dose is received by the patient than with some conventional X-ray techniques, but the diagnostic information obtained is far greater

Culture – when infection is suspected, samples of blood, urine, throat secretions etc. are taken and tested to try to identify the type of infection and the most appropriate treatment required

Cytotoxic – a drug that damages or destroys cells

Diagnosis – the exact name and type of the illness

Electrolytes – a general term for the many minerals necessary to provide the proper environment for the cells of the body. These minerals include calcium, potassium, sodium, and chloride

Emesis – vomiting

Erythrocyte sedimentation rate (ESR) – the distance red blood cells travel in one hour in a sample of blood as they settle to the bottom of a test tube. The sedimentation rate is increased in inflammation, infection, cancer, rheumatic diseases, and diseases of the blood and bone marrow.

Febrile neutropenia – having a raised body temperature/fever while having low levels of white blood cells which fight infection (neutrophils) in the circulating blood

Gastro intestinal – relating to the stomach and the intestines

Gastrostomy tube – a tube inserted through the wall of the abdomen directly into the stomach. This is a surgical procedure and is done under general anaesthesia. It can be used to give drugs, liquids and liquid food. This is also called a PEG tube.

Glomerular Filtration Rate (GFR) – a test to see how well the kidneys are working

Haematology – the study of blood and blood diseases

Haematuria – blood in the urine

Haemoglobin – the substance within red blood cells that carries oxygen to the tissues of the body. The concentration of haemoglobin in blood is used as a measure of how many red cells there are per volume of blood

Haemorrhage – bleeding

Histopathology – the microscopic study of cells in disease

Hydration – this reflects how much body water a person has. Dehydration means too little, well-hydrated means normal and over hydrated means too much

Hypotension – low blood pressure

Immune system – the body's defence against infection, disease and foreign substances

Immunoglobulin – antibodies (type of protein) that are produced by certain white blood cells and play an essential role in the body's immune system. It is sometimes necessary to give immunoglobulin (antibodies) collected from donors to patients to support their immune system (e.g. IVIG, ZIG)

Immuno-suppression – reduced function of the immune system leading to an increased risk of infection

GLOSSARY CONTINUED

Infusion – the introduction of a fluid into a vein (into the blood stream)

Intravenous (I.V.) – the administration of a drug or fluid directly into a vein (into the blood stream)

Kidney – organ that removes waste substances from the body and also maintains the body's mineral and water balance

Lesion – a change in tissue structure due to injury or disease. Ulcers, tumours, abscesses etc. may all be referred to as lesions

Leucocytes – white blood cells

Leucocytosis – a higher than normal concentration of white blood cells in the circulating blood

Leucopenia – a lower than normal concentration of white blood cells in the circulating blood

Liver – this part of the body takes part in many complex functions necessary for life, including digestion, production of certain blood proteins and elimination of many of the body's waste products

Lymph nodes – these are more commonly known as 'glands' and are located in many places throughout the body. Lymph nodes filter the lymphatic fluid and store special cells that play an important role in defending the body against infections

Lymphocytes – white blood cells responsible for the production of antibodies and for the direct destruction of invading organisms

Nasogastric tube (NG tube) – a thin tube that is inserted through the nose, down the throat and into the stomach. It can be used to give drugs, liquids and liquid food.

Nasojejunal tube (NJ tube) – a thin tube that is inserted through the nose, down the throat, through the stomach and into the small bowel. It can be used to give drugs, liquids and liquid food.

Nausea – feeling sick

Neurology – branch of medical science dealing with the nervous system

Neutrophils – white blood cells that fight acute infection

Oncology – the study, diagnosis and treatment of tumours, especially cancers

Paediatric – the branch of medicine that specialises in the study and treatment of children

Palliation – to relieve a symptom (like pain) but not necessarily a cure

Pancytopenia – decrease of red cells, white cells and platelets in the blood

PET scan – Positron Emission Tomography Scan – uses short-lived radioactive substances linked to a substance commonly used by the body (eg sugar) to produce three-dimensional colour pictures of where in the body this substance is being used

Plasma – the liquid portion of blood. It's a fluid which contains water and other components in which red cells, white cells and platelets are suspended

Platelets – the tiny cells in circulating blood which aid blood clotting

Prognosis – an estimate of the outcome of a disease based on the patient's current condition and accumulated medical knowledge about that disease and its best treatment

Pyrexia – fever or abnormally high body temperature

Reactivation – return of symptoms and signs of disease after a period of improvement (often used in the context of LCH)

Radiotherapy – the use of high-energy radiation to kill cells and shrink lesions

Red blood cell – this is the cell in the blood that carries oxygen and contains the pigment haemoglobin. It is produced in the bone marrow

Regression – a decrease in the size of a lesion or in the extent of disease in the body

Relapse – the return of signs and symptoms of disease after a period of improvement (often used in the context of cancer)

Remission – a period of well-being with no signs of disease. There is no longer any evidence of the disease using the available investigations

Renal – relating to kidneys

Respiratory – related to the process of breathing

Sepsis – infection

Septicaemia - bacteria within the bloodstream causing someone to be very unwell

Shingles – a painful or itchy rash in a particular area of the body caused by the same virus that causes chickenpox. People can only get shingles if they previously had chickenpox. Someone who has not had chickenpox can catch chickenpox from a person with shingles if they were in direct touching contact with the rash

Sinuses – hollow spaces within the bones of the face

Spleen – an organ near the stomach that forms part of the immune system. It is made up mainly of white blood cells.

Stomatitis – sore mouth

Temperature spike – when body temperature suddenly rises

Thrombocytopenia – less than the normal number of platelets in the blood

Total Parenteral Nutrition (TPN) – a method of delivering nutrition or other substances directly into a vein (IV), bypassing the digestive system

Toxicity – unpleasant or dangerous side-effects of treatment

Urinary tract – the organs and structures involved with the production and removal of urine (i.e. kidneys, ureters, bladder, urethra)

Varicella – Chickenpox – an infection caused by the Varicella Zoster virus (VZV). The same virus causes shingles

Viruses – a group of very small organisms that can produce disease. Viral infections include the common cold, chickenpox, measles, mumps and cold sores.

White blood cells – cells in the blood that are important in fighting infection (neutrophils, lymphocytes, etc)

Zoster ImmunoGlobulin (ZIG) – antibodies against Chickenpox (obtained from donors who previously had chickenpox). This can be given to individuals with lowered immunity who have not had chickenpox before to protect them if they came into contact with someone who has chickenpox.

ABOUT THE AUTHORS

Keri Taylor

Keri's son, Liam, was diagnosed with multi-system Langerhans Cell Histiocytosis at the age of 23 months after suffering nappy rash and cradle cap for a year. He was referred to Great Ormond Street Hospital where the diagnosis of LCH of the skin and liver was made.

Dr Johann Visser

Dr Johann Visser is a consultant paediatric oncologist in the East Midlands Children's and Young Persons' Integrated Cancer Service. He is based at the Leicester Children's Hospital and has a special interest in histiocytosis, sarcoma and clinical trials. He is a member of the Histiocyte Society and the UK Children's Cancer and Leukaemia Group. He is a trustee of the Histiocytosis Research Trust.

Dr Harriet Holme

Dr Harriet Holme is a paediatric oncology registrar/Academic Clinical Fellow in Haematology with special interest in paediatric oncology and histiocyte disorders. Harriet is a member of Histiocyte Society and UK Children's Cancer and Leukaemia Group.

Dr Peppy Brock

Paediatric oncologist Dr Penelope Brock (Peppy) is an expert on paediatric LCH and based at Great Ormond Street Hospital. She is a member of the International Society of Paediatric Oncology and the UK Children's Cancer and Leukaemia Group and leads internationally on a number of clinical trial groups.

Dr Vasanta Nanduri

Dr Vasanta Nanduri is a paediatric consultant with special interests in oncology and endocrinology based at Great Ormond Street and Watford General Hospital. She is an expert in histiocytic disorders. Vasanta is a member of the Histiocyte Society and has served on the Executive Board of the society for a number of years. She is also a member of the UK Children's Cancer and Leukaemia Group and is a trustee of the Histiocytosis Research Trust.

DR JON PRITCHARD 1942–2007



In 1981, Dr Pritchard met the Kontoyannis family after their son Nikolas was diagnosed with multi-system LCH. He suggested to Nikolas' father Paul that the best way to seek a cure for LCH would be to organize a meeting where scientists and clinicians could get together to discuss the disease. The first Nikolas Symposium was held in 1989 in Cambridge and has been organized annually ever since. Following on from this first meeting, Dr Pritchard suggested that a charity should be set up to raise money to fund research into the disease and The Histiocytosis Research Trust was born in 1991.

There are now nearly 300 people registered with the Trust, with a strong nucleus raising money. Dr Pritchard is remembered as being an outstanding individual who contributed and worked tirelessly for the causes he strongly believed in. He was very energetic, endearing, intelligent, cheerful and reassuring. He led the way and supported both The Trust and The Nikolas Symposium in their efforts to search for a cure for LCH. He provided unrelenting support to all his patients and their families and will never be forgotten by the many he helped. He is remembered for bringing a smile to every ill child as well providing strength and hope.

This Langerhans Cell Histiocytosis parent's guide is dedicated to his memory.



**The Histiocytosis
Research Trust**
together we'll find a cure