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Association of Pediatric Hematology/Oncology Nurses

A Handbook for Families

Langerhans Cell Histiocytosis

HEMATOLOGY SERIES

LANGERHANS CELL HISTIOCYTOSIS
A HANDBOOK FOR FAMILIES

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ABOUT THIS COVER

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WHAT IS LANGERHANS CELL HISTIOCYTOSIS?

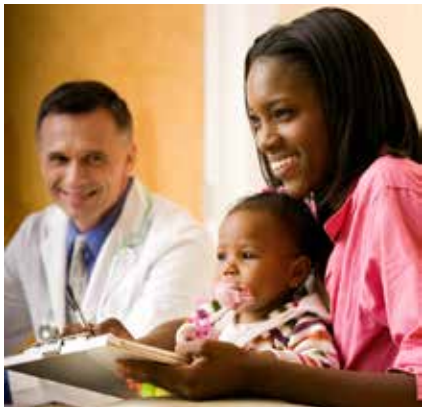
Langerhans cell histiocytosis is a rare disease that occurs when normal Langerhans cells lose their ability to fight infection and instead group together to destroy healthy tissue.

Langerhans cell histiocytosis has been known by many names including Histocytosis X malignant histiocytosis syndrome (now known as T cell lymphoma), eosinophilic granuloma Hand-Schüller-Christian disease, Letterer-Siwe disease, Hashimoto-Pritzker disease, self-healing histiocytosis, Langerhans cell granulomatosis, and Type II histiocytosis. To prevent confusion, researchers and healthcare professionals have agreed to use one name to refer to this disease: Langerhans cell histiocytosis (LCH).

WHAT ARE NORMAL LANGERHANS CELLS?

Normal Langerhans cells are part of our body's infection-fighting defense system. These Langerhans cells are usually found in body tissues such as the skin, lymph nodes, and spleen. These cells act like watch-dogs—constantly on the lookout for anything abnormal. If the body is invaded by germs (like bacteria or viruses), the Langerhans cells "sniff out" the invader and move through a series of orderly steps to alert other cells in the immune system to begin fighting the infection.

WHAT CAUSES LANGERHANS CELL HISTIOCYTOSIS?



LCH develops when normal Langerhans cells become overactive, lose control, and do not follow their usual orderly routine. The cells are stimulated to begin their defense work, but instead of going through their normal steps of gathering information and activating other cells in the immune system, these abnormal Langerhans cells cluster together like an unruly mob. This cluster of abnormal cells results in tumor-like lesions that begin to destroy normal tissues in the bones, skin, or other body systems. Scientists are not sure exactly what causes the cells' abnormal behavior, but research shows that infections or problems in the immune system may contribute.

WHO GETS LANGERHANS CELL HISTIOCYTOSIS?

Both children and adults can develop LCH. An estimated 1 in every 100,000 children younger than 15 years old will be diagnosed with LCH each year. In children younger than 2 years old, LCH usually affects multiple parts of the body and causes the child to become extremely ill. In older children, LCH is typically milder and may cause only bone or skin lesions.

IS LANGERHANS CELL HISTIOCYTOSIS INHERITED?

LCH experts do not think that LCH is inherited; however, there have been a few rare cases in which more than one family member has been diagnosed with LCH. No specific gene has been linked to the development of LCH.

IS LANGERHANS CELL HISTIOCYTOSIS A FORM OF CANCER?

LCH is a result of abnormal cell behavior in the immune system and behaves like inflammation that has gotten out of control. Although LCH shares some cancer-like characteristics, such as uncontrolled growth, it is not a cancer. However, chemotherapy is often used to treat LCH. Chemotherapy decreases immune system activity, including the over-activity of abnormal Langerhans cells.

WHAT ARE SOME SYMPTOMS OF LANGERHANS CELL HISTIOCYTOSIS?

Symptoms of LCH vary depending on which parts of the body are affected.

BONES

Bone lesions are the most common sites of LCH. LCH often develops in the skull, but bones in the legs, ribs, hips, jaw, spine and arms can also be affected as well. Symptoms can include swelling and pain in the affected area, headaches if there are lesions in the skull, and limping if there are lesions in the leg or hips. Fractures can occur for no apparent reason if tumors are present in weight-bearing bones, such as the legs or spine. Sometimes there are no symptoms at all, and the bone lesion is discovered accidentally when an X ray is taken for a different reason. Doctors often describe an X ray of a bone lesion as looking "punched-out" because the bone looks as though a cookie cutter punched out a hole in the bone.

SKIN

LCH skin lesions typically appear as a rash that does not respond to usual treatments. The rash may show up in the diaper area, on the scalp, behind the ears, or on the trunk of the body. LCH skin rashes can be mistaken for cradle cap or diaper rash and often persist for months before the diagnosis is made.

MOUTH

LCH mouth lesions may appear as unexpected loose teeth, early tooth loss, or swollen gums.

EARS

LCH ear lesions often are accompanied by continuous ear drainage or a history of frequent ear infections.

GASTROINTESTINAL SYSTEM

LCH gastrointestinal lesions may cause long-lasting diarrhea and poor weight gain. In some cases, the liver and spleen may become enlarged.

LUNGS

LCH lung lesions may cause a chronic cough, chest pain, or poor weight gain.

CENTRAL NERVOUS SYSTEM

LCH central-nervous-system lesions may cause headaches, excessive thirst and urination, abnormal swelling around the eyes, bulging eyes, changes in behavior, or seizures.

WHAT TESTS AND PROCEDURES WILL MY CHILD NEED?

Your child's healthcare team will meet with you to discuss your child's symptoms and will order specific tests to make a diagnosis. Common tests and procedures your child might need include the following.

BLOOD TESTS

Blood tests may be done to rule out other diseases but are not diagnostic for LCH. They are performed during therapy to monitor both your child's response to treatment and any possible side effects of therapy. Blood tests check your child's blood cells, body salts, and chemistries. A complete blood count (CBC) is a type of blood test that is useful in detecting a drop in the number of red blood cells (cells that carry oxygen), white blood cells (cells that fight infection), or platelets (cells that help blood clot properly). Blood chemistries such as BUN (blood urea nitrogen) and creatinine monitor changes in kidney function. Other chemistries such as ALT (alanine aminotransferase), AST (aspartate aminotransferase), and bilirubin may be done to assess liver function.

X RAYS

Your child will need X rays to determine which bones are affected by LCH. A *bone survey* or *skeletal survey,* which X rays all the bones in your child's body from head to toe, is the most common X ray used to detect LCH. If LCH is suspected in a bone, the area is usually described as *lytic*, which means that some of the bone has been eaten away by the abnormal LCH cells. You might not be able to feel these areas with your hand, so the bone survey will show your child's healthcare team where these areas can be found. After your child's treatment is finished, a bone survey will be performed every few months as part of regular check-ups to make sure LCH has not returned.

CT SCAN

Computerized axial tomography (CT) scans are computer-assisted X rays that take detailed pictures of areas inside the body from different angles. Your child will have a CT scan of his or her chest if there is a possibility that LCH has developed in the lungs. If LCH is suspected in the skull, a CT scan of the head will be performed. CT scans are painless and quick, but your child must lie completely still during the scan. Some children may require sedation to help them lie still. Your child may be asked not to eat or drink for several hours before the scan. It may be necessary for your child to drink a liquid containing a flavorless dye to make the pictures clearer. Usually, however, a small amount of dye is injected into a vein during the CT scan. Generally, no side effects occur from either type of dye; however, allergic reactions are possible.

BIOPSY

To be absolutely certain that your child has LCH, a *biopsy*, or tissue sample, will need to be taken from the area that looks abnormal—usually a skin rash, a bone, the gums, or a lymph node. A pathologist, a specialized doctor who diagnoses tissue changes caused by disease, will look at the tissue sample under a microscope. The pathologist will also do special studies to look for LCH cells. These tests may take several days to finish. When the tests are completed, your child's healthcare team will talk to you about the results.

OTHER TESTS

Other tests may need to be performed depending on the location of the disease. These tests include an ultrasound of the abdomen, PET (positron emission tomography) scan, or bone marrow aspiration. Your child's doctor or nurse will explain these tests to you before they are performed.

HOW IS LANGERHANS CELL HISTIOCYTOSIS CLASSIFIED?

LCH is classified based on which parts or systems of the body are affected by the disease. Each of the following is considered a body system: bone, skin, muscles/tissues, pituitary, brain and central nervous system, blood and marrow, lungs, liver, spleen, lymph nodes, mouth, intestines, and the area around the eyes.

Treatment is then determined according to the following groups:

- Single-system disease—involves only one body system, usually bone, skin, or lymph nodes. Single-system disease can be a single lesion or can involve several sites within the same body system (several bone lesions, for instance).
- Multi-system, high-risk disease—involves more than one body system, affecting the function of one or more of the following organs: bone marrow, liver, spleen, or lung.
- Multi-system, low-risk disease—involves more than one body system, but does not include any of the high-risk sites.

HOW IS LANGERHANS CELL HISTIOCYTOSIS TREATED?

CHEMOTHERAPY

The most common chemotherapy medicines used to treat LCH are prednisone, which is given by mouth, and vinblastine, which is given in the vein. Some other agents that may be used include Etoposide, Methotrexate, or Cyclophosphamide, which are also given in the vein.

RADIATION THERAPY

Radiation therapy is rarely used.

CURETTAGE

If there is just one spot of LCH on a bone, the doctor may be able to remove all of the disease when he or she does the biopsy. In this case, no other treatment is necessary.

WILL NEEDLES BE USED TO ADMINISTER CHEMOTHERAPY?

If LCH is to be treated with chemotherapy, a surgeon often will insert a small plastic tube called a *venous access device* (VAD) or *line* ("port") into a large blood vessel, usually under the collar bone. The VAD can make it easier for your child to have blood drawn for blood tests and medications, chemotherapy, blood products, and nutritional support given when needed. You and your child's healthcare team will decide whether or not your child needs a VAD. The device is usually left in place for the entire treatment and removed after completion of therapy.

HOW LONG WILL MY CHILD'S THERAPY LAST?

If chemotherapy is recommended, the treatment usually lasts 12 months.

HOW SUCCESSFUL IS THE TREATMENT?

Most children with LCH respond well to treatment. Long-term survival for single-system disease is nearly 100%. Survival for multi-system disease is more varied. Many children with multi-system disease may experience periods of remission and reactivation before the disease is cured.

WHAT IF THE DISEASE RECURS?

If LCH recurs, it is not the same as a cancer relapse. Some children with LCH may go through several remissions and reactivations before the disease finally goes away for good. If the disease recurs, your child's doctor may recommend a different group of chemotherapy medicines or other types of medications that are known to have an effect on the immune system.

WHAT NEW TREATMENTS EXIST?

In addition to the standard chemotherapy, a variety of other promising agents, including cladribine, thalidomide, and biotherapy medications, as well as stem-cell transplants are being investigated to treat LCH. The effectiveness of these treatments is still being studied.

WHAT IS DIABETES INSIPIDUS AND WHY IS IT ASSOCIATED WITH LANGERHANS CELL HISTIOCYTOSIS?

Diabetes insipidus (DI) is a hormonal imbalance that is caused by dysfunction of the pituitary gland in the brain. DI occurs in more than 50% of children diagnosed with LCH. Children who are most likely to develop DI are those with bone lesions in the front of the head, including the face. Symptoms may not be present at the time of diagnosis but may show up 6–12 months later. Symptoms of DI include excessive thirst, an obsession with wanting to drink fluids all the time (day and night), and increased urination. There is no cure for DI, but symptoms can be controlled with medication. If your child wants to drink more frequently than usual—and needs to get up several times during the night to drink—and is going to the bathroom frequently during the day and night or has started wetting the bed, tell your child's healthcare team about these symptoms. It is important to know that DI does not go away when the LCH is cured; however your child may see improvement in symptoms.

HOW CAN I WORK WITH THE HEALTHCARE TEAM?

Your child's care requires a team approach. As a parent, you are a very important part of the team. Other members of the team may include doctors, nurse practitioners, nurses, social workers, child life specialists, therapists, and others. Because you know your child better than anyone else, the healthcare team will need your help to give your child the very best, comprehensive treatment.

It is important to communicate openly with your child's healthcare team. Be sure to ask your child's doctor or nurse questions whenever there is anything that you are unsure about. It helps to write down your questions when you think of them.

Here are some examples of questions to ask:

- What is Langerhans cell histiocytosis?
- Is it cancer?
- What parts of my child's body are involved?
- What group (or stage) is my child's disease?
- What treatment choices are available?
- What treatment do you recommend, and why?
- What risks or side effects does the recommended treatment have?
- What should I do to prepare for the treatment?
- What is my child's chance for survival?
- What are the chances that the disease will come back?
- What long-term effects might occur as a result of treatment?

Use this space to write down some other questions you might have:

ARE MY FEELINGS NORMAL AND WHAT CAN I DO ABOUT THEM?

Hearing that your child has a serious illness can be shocking and overwhelming. At first you may not believe it, or you may hope that the diagnosis is wrong. These are normal feelings to have after the diagnosis is made.

Many family members feel that they are responsible somehow for the child's disease, or they feel guilty that they were not able to detect it sooner. It is important to remember there is no way you could have predicted that your child would get this disease or that you could have prevented it from happening. This disease was not caused by anything you or your child did. The important thing is that you are here now and are ready to begin the treatment to make your child better.

In addition to shock and guilt, you and your family may feel angry and sad. Even the youngest members of your family may be affected by this illness. Feelings of anger, sadness, guilt, and shock are all normal. Each member of your family will express these feelings in different ways and at different times. It can be difficult to feel so many strong emotions at once. Talking honestly with each other about feelings, reactions, and questions will help everyone in the family.

It may seem difficult to talk to friends, family, or even the healthcare team, but sharing your feelings can help you cope with the situation. Remember that your child will benefit if family members continue to show that they care by being supportive and communicating openly. If your friends and family give you information that seems to conflict with what your child's healthcare team has told you, be sure to talk to your child's healthcare team to clear up any confusion.

HOW CAN I HELP MY CHILD?

As a parent, you will often notice changes in your child during treatment that can make you feel even more helpless. It is important to always remember that your child is still the same person on the inside, despite any changes on the outside. Hair loss and other changes in body appearance are temporary and often bother adults much more than they bother your child or your child's siblings and friends. All of your feelings about what your child is going through during treatment must be balanced by remembering that the treatment provides an opportunity to cure the disease and to have your child go on to live a normal and meaningful life.

It is important to reinforce to your child that nothing he or she did caused this disease. Tell your child that your angry or sad feelings are directed at the disease and not at him or her. This will help maintain honesty and closeness in your relationship. Like you, your child will need someone with whom he or she can share feelings. Don't hesitate to ask your child to express his or her feelings. Explain, in age-appropriate terms, what is happening and why. Do not avoid using direct terms and explanations with your child. Children tolerate treatment better if they understand it and are allowed to be active decision makers whenever possible. The same is true for parents.

As you begin to learn the new, special needs of your child, it is important to remember that he or she is still a normal child and is still growing and developing. All children—both sick and well—need love, attention, discipline, limits, and the opportunity to learn new skills and try new activities that are age appropriate.

IS MY CHILD'S DIET IMPORTANT?

Yes. We know from research that well-nourished children tolerate therapy better and have fewer treatment delays due to illness. It may be difficult for your child to resume normal eating habits during therapy, so you will need to be flexible and creative. Often, several small meals during the day are tolerated better than three large meals. Children are usually more interested in eating foods that they have helped prepare. It is also important to include your child in the social activity of family meals even if he or she is not eating a full meal. Remember, nobody wins food fights—it is best not to force your child to eat.

Make sure that foods high in protein and carbohydrates are readily available. Multivitamins, herbs, supplements, and all other medicines (both over the counter and those prescribed by other doctors) should be approved by your healthcare team before you give them to your child, because they may cause an interaction with your child's chemotherapy treatment or change the effectiveness of the treatment.

A dietician trained in children's calorie and energy needs can offer you guidance and food suggestions. The healthcare team can also intervene if your child develops a nutritional problem.

CAN MY CHILD ATTEND SCHOOL?

Your child's ability to attend school will depend on the intensity of the therapy and his or her response to treatment. Some children tolerate therapy better than others. Your child may not be able to attend school for extended periods of time because of the treatment schedule or hospitalizations. However, it is important for your child to keep up with his or her schoolwork. Talk to the staff at your child's school about arranging for a tutor or homebound teacher until he or she is able to return to school. You should also discuss school attendance with your child's healthcare team. Many hospitals have programs that help children attend school while they are hospitalized.

School is important because it helps children maintain social contact with other children of the same age. Maintaining time with friends will be an important part of your child's recovery and will ease the adjustment when he or she returns to school. It is important for your child to return to school as soon as he or she is medically able to do so.

ONLINE RESOURCES

Histiocytosis Association of America

www.histio.org

SUGGESTED READING

IMPORTANT PHONE NUMBERS

NOTES

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