National Cancer Institute
at the National Institutes of Health

Langerhans Cell Histiocytosis Treatment (PDQ®) Patient Version

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General Information About Langerhans Cell Histiocytosis (LCH)

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<th>Key Points for This Section</th>
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<tr>
<td>- Langerhans cell histiocytosis is a disease that can damage tissue or cause lesions to form in one or more places in the body.</td>
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<tr>
<td>- Having a parent who was exposed to certain chemicals and family history may increase the risk of developing LCH.</td>
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<tr>
<td>- The cause of LCH is unknown.</td>
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<td>- The signs and symptoms of LCH depend on where it occurs in the body.</td>
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<td>- Skin</td>
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<td>- Lung</td>
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Langerhans cell histiocytosis (LCH) is a rare disease that occurs when the body makes too many Langerhans cells. A Langerhans cell is a type of white blood cell that helps the body fight infection. Langerhans cells (also called histiocytes) are normally found in the skin, lymph nodes, spleen, bone marrow, and lungs. In LCH, extra Langerhans cells spread through the blood and build up in certain parts of the body, where they can damage tissue or form tumors.

Scientists do not agree on whether LCH is a type of cancer or is a condition caused by a change in the immune system. LCH is often treated with anticancer drugs that may also be used to treat immune system conditions.

LCH may occur at any age, but is most common in young children. The treatment of LCH in children and adults is described in separate sections.

Check for U.S. clinical trials from NCI's list of cancer clinical trials that are now accepting patients with childhood Langerhans cell histiocytosis. For more specific results, refine the search by using other search features, such as the location of the trial, the type of treatment, or the name of the drug. General information about clinical trials is available from the NCI Web site.

Having a parent who was exposed to certain chemicals and family history may increase the risk of developing LCH.

Anything that increases your risk of getting a disease is called a risk factor. Having a risk factor does not mean that you will get the disease; not having risk factors doesn't mean that you will not get the disease. People who think they may be at risk should discuss this with their doctor. Risk factors for LCH may include the following:

- Having a parent who was exposed to certain chemicals such as benzene.
- Having infections as a newborn.
- Having a family history of thyroid disease.

The cause of LCH is unknown.

The signs and symptoms of LCH depend on where it occurs in the body.

These and other symptoms may be caused by LCH. Other conditions may cause the same symptoms. A doctor should be consulted if any of the following problems occur:

Skin

In infants, signs and symptoms of LCH may include:

- Flaking of the scalp that may look like “cradle cap”.
- Raised, brown or purple spots anywhere on the body.

In children and adults, signs and symptoms of LCH may include:

- Flaking of the scalp that may look like dandruff.
- Raised, red or brown, crusted rash in the groin area, abdomen, back or chest.
- Bumps or ulcers behind the ears, on the scalp, or in the groin area.

Mouth
Signs and symptoms of LCH may include:

- Swollen gums.
- Sores on the roof of the mouth, inside the cheeks, or on the tongue or lips.

**Bone**

Signs and symptoms of LCH may include:

- Swelling or a lump over a bone, such as the skull, hip, ribs, spine, or elbow.
- Pain where there is swelling or a lump over a bone.

**Lymph nodes and thymus**

Signs and symptoms of LCH may include:

- Swollen lymph nodes.
- Trouble breathing.

**Pituitary gland**

Signs and symptoms of LCH may include:

- Diabetes insipidus. This can cause a strong thirst and frequent urination.
- Slow growth.
- Late puberty.

**Thyroid**

Signs and symptoms of LCH may include:

- Swollen thyroid gland.
- Hypothyroidism. This can cause tiredness, lack of energy, being sensitive to cold, constipation, dry skin, thinning hair, memory problems, trouble concentrating, and depression. In infants, this can also cause a loss of appetite and choking on food. In children and teens, this can also cause behavior problems, weight gain, slowed growth, and late puberty.
- Trouble breathing.

**Central nervous system**

Signs and symptoms of LCH may include:

- Diabetes insipidus. This can cause a strong thirst and frequent urination.
- Trouble breathing.
- Loss of balance, uncoordinated body motions, and trouble walking.
- Trouble speaking.
- Changes in behavior.
- Memory problems.

**Liver and spleen**

Signs and symptoms of LCH may include:

- Swelling in the abdomen caused by a build up of extra fluid.
- Yellowing of the skin and whites of the eyes.
- Easy bruising or bleeding.

**Lung**
Signs and symptoms of LCH may include:

- Spontaneous pneumothorax. This can cause chest pain or tightness, trouble breathing, feeling tired, and the skin to turn a bluish color.
- Trouble breathing, especially in adults who smoke.

**Bone marrow**

Signs and symptoms of LCH may include:

- Easy bruising or bleeding.
- Fever.
- Frequent infections.

**Tests that examine the organs and body systems where LCH may occur are used to detect (find) and diagnose LCH.**

The following tests and procedures may be used to detect (find) and diagnose LCH or conditions caused by LCH:

- **Physical exam and history:** An exam of the body to check general signs of health, including checking for signs of disease, such as lumps or anything else that seems unusual. A history of the patient's health habits and past illnesses and treatments will also be taken.

- **Neurological exam:** A series of questions and tests to check the brain, spinal cord, and nerve function. The exam checks a person's mental status, coordination, and ability to walk normally, and how well the muscles, senses, and reflexes work. This may also be called a neuro exam or a neurologic exam.

- **Complete blood count (CBC) with differential:** A procedure in which a sample of blood is drawn and checked for the following:
  - The amount of hemoglobin (the protein that carries oxygen) in the red blood cells.
  - The portion of the blood sample made up of red blood cells.
  - The number and type of white blood cells.
  - The number of red blood cells and platelets.

- **Blood chemistry studies:** A procedure in which a blood sample is checked to measure the amounts of certain substances released into the body by organs and tissues in the body. An unusual (higher or lower than normal) amount of a substance can be a sign of disease in the organ or tissue that makes it.

- **Liver function test:** A blood test to measure the blood levels of certain substances released by the liver. A high or low level of these substances can be a sign of disease in the liver.

- **Urinalysis:** A test to check the color of urine and its contents, such as sugar, protein, red blood cells, and white blood cells.

- **Water deprivation test:** A test to check how much urine is made and whether it becomes concentrated when little or no water is given. This test is used to diagnose diabetes insipidus, which may be caused by LCH.

- **Bone marrow aspiration and biopsy:** The removal of bone marrow, blood, and a small piece of bone by inserting a hollow needle into the hipbone or breastbone. A pathologist views the bone marrow, blood, and bone under a microscope to look for signs of LCH.
Bone marrow aspiration and biopsy. After a small area of skin is numbed, a Jamshidi needle (a long, hollow needle) is inserted into the patient's hip bone. Samples of blood, bone, and bone marrow are removed for examination under a microscope.

- **Bone scan**: A procedure to check if there are rapidly dividing cells in the bone. A very small amount of radioactive material is injected into a vein and travels through the bloodstream. The radioactive material collects in the bones and is detected by a scanner.

Bone scan. A small amount of radioactive material is injected into the patient's bloodstream and collects in abnormal cells in the bones. As the patient lies on a table that slides under the scanner, the radioactive material is detected and images are made on a computer screen or film.

- **X-ray**: An x-ray of the organs and bones inside the body. An x-ray is a type of energy beam that can go through the body and onto film, making a picture of areas inside the body.

- **CT scan (CAT scan)**: A procedure that makes a series of detailed pictures of areas inside the body, taken from different angles. The pictures are made by a computer linked to an x-ray machine. A dye may be injected into a vein or swallowed to help the organs or tissues show up more clearly. This procedure is also called computed tomography, computerized tomography, or computerized axial tomography.
Computed tomography (CT) scan of the abdomen. The patient lies on a table that slides through the CT machine, which takes x-ray pictures of the inside of the body.

- MRI (magnetic resonance imaging): A procedure that uses a magnet, radio waves, and a computer to make a series of detailed pictures of areas inside the body. A substance called gadolinium may be injected into a vein. The gadolinium collects around the LCH cells so that they show up brighter in the picture. This procedure is also called nuclear magnetic resonance imaging (NMRI).

Magnetic resonance imaging (MRI) of the abdomen. The patient lies on a table that slides into the MRI machine, which takes pictures of the inside of the body. The pad on the patient's abdomen helps make the pictures clearer.

- Somatostatin receptor scintigraphy: A type of radionuclide scan used to find certain tumors. A small amount of a radioactive drug similar to somatostatin is injected into a vein and travels through the bloodstream. The radioactive drug attaches to tumor cells that have receptors for somatostatin. A radiation-measuring device detects the radioactive drug and makes pictures showing where the tumor cells are in the body. Also called octreotide scan and SRS.

- PET scan (positron emission tomography scan): A procedure to find tumor cells in the body. A small amount of radioactive glucose (sugar) is injected into a vein. The PET scanner rotates around the body and makes a picture of where glucose is being used in the body. Tumor cells show up brighter in the picture because they are more active and take up more glucose than normal cells do.
PET (positron emission tomography) scan. The patient lies on a table that slides through the PET machine. The head rest and white strap help the patient lie still. A small amount of radioactive glucose (sugar) is injected into the patient's vein, and a scanner makes a picture of where the glucose is being used in the body. Cancer cells show up brighter in the picture because they take up more glucose than normal cells do.

- **Ultrasound exam**: A procedure in which high-energy sound waves (ultrasound) are bounced off internal tissues or organs and make echoes. The echoes form a picture of body tissues called a sonogram. The picture can be printed to be looked at later.

- **Endoscopy**: A procedure to look at organs and tissues inside the body to check for abnormal areas. An endoscope is inserted through an incision (cut) in the skin or opening in the body, such as the mouth. An endoscope is a thin, tube-like instrument with a light and a lens for viewing. It may also have a tool to remove tissue or lymph node samples, which are checked under a microscope for signs of disease.

- **Biopsy**: The removal of cells or tissues so they can be viewed under a microscope by a pathologist to check for Birbeck granules. Birbeck granules are found in Langerhans cells. To diagnose LCH, a biopsy of bone lesions, skin, lymph nodes, or the liver may be done.

**Certain factors affect prognosis (chance of recovery) and treatment options.**

LCH in organs such as the skin, bones, lymph nodes, or pituitary gland usually gets better with treatment and is called "low-risk". LCH in the spleen, liver, bone marrow, or lung is harder to treat and is called "high-risk".

The prognosis (chance of recovery) and treatment options depend on the following:

- Whether the disease is found in one or more places in the body.
- Whether the disease is found in the liver, spleen, lung, bone marrow, or certain bones in the skull.
- How quickly the disease responds to initial treatment.
- Whether the disease has just been diagnosed or has come back (recurred).

In infants up to one year of age, LCH may disappear without treatment.

**Stages of LCH**

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**There is no staging system for LCH.**

The extent or spread of disease is usually described as stages. There is no staging system for LCH. Before planning treatment, it is important to know whether there is single-site, multiple-site, or multisystem disease and whether "low-risk" organs (skin, bones, lymph nodes, pituitary gland) or "high-risk" organs (spleen, liver, bone
marrow, lung) are affected.

**LCH is described based on how many places it occurs and where it occurs in the body.**

LCH may be described as single-site, multiple-site, and multisystem disease.

- Single-site: Occurs in one organ or system in the body such as the skin, mouth, bone, lymph node, thymus, pituitary gland, or central nervous system.
- Multiple-site: Occurs in more than one part of the same organ or area of the body.
- Multisystem: Occurs in two or more organs or areas of the body.

**Recurrent LCH**

Recurrent Langerhans cell histiocytosis is disease that has recurred (come back) after it has been treated. The disease may come back in the same place or in other parts of the body.

**Treatment Option Overview for LCH**

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<td><strong>Six types of standard treatment are used:</strong></td>
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<td>- Watchful waiting</td>
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<td>- Surgery</td>
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<td>- Corticosteroid therapy</td>
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<td>- Chemotherapy</td>
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<td>- Photodynamic therapy</td>
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<td>- Radiation therapy</td>
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<td><strong>Patients can enter clinical trials before, during, or after starting their treatment.</strong></td>
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<tr>
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<tr>
<td><strong>Follow-up tests may be needed.</strong></td>
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**There are different types of treatment for patients with LCH.**

Different types of treatments are available for patients with LCH. Some treatments are standard (the currently used treatment), and some are being tested in clinical trials. A treatment clinical trial is a research study meant to help improve current treatments or obtain information on new treatments for patients. When clinical trials show that a new treatment is better than the standard treatment, the new treatment may become the standard treatment. Whenever possible, patients should be enrolled in a clinical trial in order to benefit from advances in LCH treatment and ongoing care.

Clinical trials are taking place in many parts of the country. Information about ongoing clinical trials is available from the NCI Web site. Choosing the most appropriate treatment is a decision that ideally involves the patient, family, and health care team.

**Children with LCH should have their treatment planned by a team of health care providers who are experts in treating this disease in children.**

Treatment will be overseen by a pediatric oncologist, a doctor who specializes in treating children with cancer.
The pediatric oncologist works with other pediatric health care providers who are experts in treating children with LCH and who specialize in certain areas of medicine. These may include the following specialists:

- Primary care physician.
- Pediatric surgeon.
- Pediatric hematologist.
- Radiation oncologist.
- Neurologist.
- Endocrinologist.
- Pediatric nurse specialist.
- Rehabilitation specialist.
- Psychologist.
- Social worker.
- Geneticist.

Some treatments for LCH cause side effects months or years after treatment has ended.

Some treatments cause side effects that continue or appear months or years after treatment has ended. These are called late effects. Late effects of treatment for LCH may include the following:

- Slow growth and development.
- Hearing loss.
- Bone, tooth, liver, and lung problems.
- Changes in mood, feeling, learning, thinking, or memory.
- Risk of cancer related to treatment.

Some late effects may be treated or controlled. It is important to talk with your child's doctors about the possible late effects caused by some treatments.

Six types of standard treatment are used:

LCH is usually treated with anticancer treatments. These treatments stop the LCH cells from growing and dividing.

**Watchful waiting**

Watchful waiting is closely monitoring a patient's condition without giving any treatment until symptoms appear or change.

**Surgery**

Surgery may be used to remove LCH lesions.

**Corticosteroid therapy**

Corticosteroids are steroids used to treat LCH lesions.

**Chemotherapy**

Chemotherapy is a treatment that uses drugs to stop the growth of cells, either by killing the cells or by stopping them from dividing. When chemotherapy is taken by mouth or injected into a vein or muscle, the drugs enter the bloodstream and can reach cells throughout the body (systemic chemotherapy). When chemotherapy is placed directly onto the skin or into the cerebrospinal fluid, an organ, or a body cavity such as the abdomen, the drugs mainly affect cells in those areas (regional chemotherapy).

Nitrogen mustard is a drug that is applied directly to the skin to treat small LCH lesions.

**Photodynamic therapy**
Photodynamic therapy is a treatment that uses a drug and a certain type of laser light to kill cells. A drug that is not active until it is exposed to light is injected into a vein. The drug collects more in LCH cells than in normal cells. For LCH, laser light is shined onto the skin and the drug becomes active and kills the LCH cells. Photodynamic therapy causes little damage to healthy tissue. Patients who have photodynamic therapy should not spend too much time in the sun.

In one type of photodynamic therapy, called psoralen and ultraviolet A therapy (PUVA), the patient receives a drug called psoralen and then ultraviolet radiation is directed to the skin.

**Radiation therapy**

Radiation therapy is a treatment that uses high-energy x-rays or other types of radiation to kill cells or keep them from growing. **External radiation therapy** uses a machine outside the body to send radiation toward the LCH lesion.

**New types of treatment are being tested in clinical trials.**

Information about clinical trials is available from the NCI Web site.

Patients may want to think about taking part in a clinical trial.

For some patients, taking part in a clinical trial may be the best treatment choice. Clinical trials are part of the medical research process. Clinical trials are done to find out if new treatments are safe and effective or better than the standard treatment.

Many of today's standard treatments for disease are based on earlier clinical trials. Patients who take part in a clinical trial may receive the standard treatment or be among the first to receive a new treatment.

Patients who take part in clinical trials also help improve the way diseases will be treated in the future. Even when clinical trials do not lead to effective new treatments, they often answer important questions and help move research forward.

Patients can enter clinical trials before, during, or after starting their treatment.

Some clinical trials only include patients who have not yet received treatment. Other trials test treatments for patients whose disease has not gotten better. There are also clinical trials that test new ways to stop a disease from recurring (coming back) or reduce the side effects of treatment.

Clinical trials are taking place in many parts of the country. See the Treatment Options for Childhood LCH and the Treatment Options for Adult LCH sections that follow for links to current treatment clinical trials. These have been retrieved from NCI's clinical trials database.

When treatment of LCH stops, new lesions may appear or old lesions may come back.

Many patients with LCH get better with treatment. However, when treatment stops, new lesions may appear or old lesions may come back. This is called reactivation and may occur within one year after stopping treatment. Patients with single-site disease are less likely to have a reactivation, while patients with multisystem disease are more likely to have a reactivation. More common sites of reactivation are bone, ears, or skin, and diabetes insipidus also may develop. Less common sites of reactivation include lymph nodes, bone marrow, spleen, liver, or lung. Some patients may have more than one reactivation over a number of years.

Follow-up tests may be needed.

Some of the tests that were done to diagnose the disease may be repeated. This is to see how well the treatment is working and if new lesions have appeared. These tests may include:

- Physical exam.
- Neurological exam.
- MRI.
- CT scan.
- PET scan.
Other tests that may be needed include:

- **Brain stem auditory evoked response (BAER) test**: A test that measures the brain's response to clicking sounds or certain tones.
- **Pulmonary function test (PFT)**: A test to see how well the lungs are working. It measures how much air the lungs can hold and how quickly air moves into and out of the lungs. It also measures how much oxygen is used and how much carbon dioxide is given off during breathing. This is also called lung function test.

Decisions about whether to continue, change, or stop treatment may be based on the results of these tests.

Some of the tests will continue to be done from time to time after treatment has ended. The results of these tests can show if your condition has changed or if the disease has recurred (come back). These tests are sometimes called follow-up tests or check-ups.

**Treatment Options for Childhood LCH**

**Treatment Options for Single- and Multiple-Site Lesions**

**Skin Lesions**

Treatment of childhood LCH single-site skin lesions may include the following:

- Watchful waiting.
- Steroids or nitrogen mustard applied to the skin.
- Chemotherapy, such as methotrexate or thalidomide, given by mouth.
- Photodynamic therapy with psoralen and ultraviolet A radiation (PUVA).

**Bone Lesions**

Treatment of childhood LCH single-site bone lesions may include surgery with or without steroid therapy.

Treatment of childhood LCH multiple-site bone lesions may include chemotherapy and steroid therapy.

Treatment of childhood LCH single-site bone lesions on the sides of the skull, behind the ears, or around the eyes may include:

- Chemotherapy and steroid therapy.
- Surgery.

Treatment of spine or hip bone lesions that have weakened the bone and may lead to a broken bone in childhood LCH may include:

- Radiation therapy.
- Surgery to strengthen the weakened bone by bracing or fusing the bones together.

**Central Nervous System Lesions**

Treatment of childhood LCH central nervous system lesions may include chemotherapy.

**Multiple-Site Lesions**

Treatment of childhood LCH multiple-site lesions (such as two or more single-site bone, skin, lymph node, or pituitary gland lesions) may include chemotherapy and steroid therapy. Treatment of childhood LCH that occurs in "low-risk" organs is about 6 months long.

Treatment of childhood LCH spleen, liver, bone marrow, or lung lesions (with or without skin, bone, lymph node, or pituitary gland lesions) may include chemotherapy and steroid therapy. Treatment of childhood LCH that occurs in "high-risk" organs is about 12 months long.

**Treatment Options for Multisystem Lesions**
Treatment of childhood LCH multisystem disease lesions may include:

- Chemotherapy with or without steroid therapy.
- A liver transplant for patients with severe liver damage.

**Treatment Options for Recurrent, Refractory, and Progressive Childhood LCH**

Recurrent LCH is disease that cannot be detected after treatment and then comes back. Treatment of recurrent childhood LCH may include chemotherapy with or without steroid therapy.

Refractory LCH is disease that does not get better with treatment. Treatment of refractory childhood LCH in "high-risk" organs or refractory LCH in multisystem "low-risk" organs may include high-dose chemotherapy with or without stem cell transplant.

Progressive LCH is disease that continues to grow during treatment. Treatment of progressive childhood LCH in patients with multisystem disease may include chemotherapy using anticancer drugs that have not been given to the patient before.

Check for U.S. clinical trials from NCI's list of cancer clinical trials that are now accepting patients with childhood Langerhans cell histiocytosis. For more specific results, refine the search by using other search features, such as the location of the trial, the type of treatment, or the name of the drug. General information about clinical trials is available from the NCI Web site.

**Treatment Options for Adult LCH**

Treatment of LCH in adults is usually the same as for children. (See the Treatment Options for Childhood LCH section above for more information).

Treatment may include:

- Combination chemotherapy with or without steroid therapy.
- Anti-inflammatory drugs combined with chemotherapy.
- A clinical trial of lung transplant for patients with severe lung damage.

For more information about LCH trials for adults, see the Histiocyte Society website.

Check for U.S. clinical trials from NCI's list of cancer clinical trials that are now accepting patients with adult Langerhans cell histiocytosis. For more specific results, refine the search by using other search features, such as the location of the trial, the type of treatment, or the name of the drug. General information about clinical trials is available from the NCI Web site.

**Get More Information From NCI**

**Call 1-800-4-CANCER**

For more information, U.S. residents may call the National Cancer Institute's (NCI's) Cancer Information Service toll-free at 1-800-4-CANCER (1-800-422-6237) Monday through Friday from 8:00 a.m. to 8:00 p.m., Eastern Time. A trained Cancer Information Specialist is available to answer your questions.

**Chat online**

The NCI's LiveHelp® online chat service provides Internet users with the ability to chat online with an Information Specialist. The service is available from 8:00 a.m. to 11:00 p.m. Eastern time, Monday through Friday. Information Specialists can help Internet users find information on NCI Web sites and answer questions about cancer.

**Write to us**
For more information from the NCI, please write to this address:

NCI Public Inquiries Office
Suite 3036A
6116 Executive Boulevard, MSC8322
Bethesda, MD 20892-8322

Search the NCI Web site

The NCI Web site provides online access to information on cancer, clinical trials, and other Web sites and organizations that offer support and resources for cancer patients and their families. For a quick search, use the search box in the upper right corner of each Web page. The results for a wide range of search terms will include a list of "Best Bets," editorially chosen Web pages that are most closely related to the search term entered.

There are also many other places to get materials and information about cancer treatment and services. Hospitals in your area may have information about local and regional agencies that have information on finances, getting to and from treatment, receiving care at home, and dealing with problems related to cancer treatment.

Find Publications

The NCI has booklets and other materials for patients, health professionals, and the public. These publications discuss types of cancer, methods of cancer treatment, coping with cancer, and clinical trials. Some publications provide information on tests for cancer, cancer causes and prevention, cancer statistics, and NCI research activities. NCI materials on these and other topics may be ordered online or printed directly from the NCI Publications Locator. These materials can also be ordered by telephone from the Cancer Information Service toll-free at 1-800-4-CANCER (1-800-422-6237).

Changes to This Summary (08/13/2010)

The PDQ cancer information summaries are reviewed regularly and updated as new information becomes available. This section describes the latest changes made to this summary as of the date above.

Changes were made to this summary to match those made in the health professional version.

About PDQ

PDQ is a comprehensive cancer database available on NCI's Web site.

PDQ is the National Cancer Institute's comprehensive cancer information database. Most of the information contained in PDQ is available online at NCI's Web site. PDQ is provided as a service of the NCI. The NCI is part of the National Institutes of Health, the federal government's focal point for biomedical research.

PDQ contains cancer information summaries.

The PDQ database contains summaries of the latest published information on cancer prevention, detection, genetics, treatment, supportive care, and complementary and alternative medicine. Most summaries are available in two versions. The health professional versions provide detailed information written in technical language. The patient versions are written in easy-to-understand, nontechnical language. Both versions provide current and accurate cancer information.

The PDQ cancer information summaries are developed by cancer experts and reviewed regularly.

Editorial Boards made up of experts in oncology and related specialties are responsible for writing and maintaining the cancer information summaries. The summaries are reviewed regularly and changes are made as new information becomes available. The date on each summary ("Date Last Modified") indicates the time of the most recent change.

PDQ also contains information on clinical trials.

A clinical trial is a study to answer a scientific question, such as whether one treatment is better than another. Trials are based on past studies and what has been learned in the laboratory. Each trial answers certain scientific
questions in order to find new and better ways to help cancer patients. During treatment clinical trials, information is collected about the effects of a new treatment and how well it works. If a clinical trial shows that a new treatment is better than one currently being used, the new treatment may become "standard." In the United States, about two-thirds of children with cancer are treated in a clinical trial at some point in their illness.

Listings of clinical trials are included in PDQ and are available online at NCI's Web site. Descriptions of the trials are available in health professional and patient versions. For additional help in locating a childhood cancer clinical trial, call the Cancer Information Service at 1-800-4-CANCER (1-800-422-6237).

The PDQ database contains listings of groups specializing in clinical trials.

The Children's Oncology Group (COG) is the major group that organizes clinical trials for childhood cancers in the United States. Information about contacting COG is available on the NCI Web site or from the Cancer Information Service at 1-800-4-CANCER (1-800-422-6237).

Glossary Terms

**abdomen** (AB-doh-men)

The area of the body that contains the pancreas, stomach, intestines, liver, gallbladder, and other organs.

**abnormal** (ab-NOR-mul)

Not normal. An abnormal lesion or growth may be cancer, premalignant (likely to become cancer), or benign (not cancer).

**appetite** (A-peh-tite)

A desire to satisfy a physical or mental need, such as for food, sex, or adventure.

**benzene** (BEN-zeen)

A chemical that is used widely by the chemical industry, and is also found in tobacco smoke, vehicle emissions, and gasoline fumes. Exposure to benzene may increase the risk of developing leukemia.

**biopsy** (BY-op-see)

The removal of cells or tissues for examination by a pathologist. The pathologist may study the tissue under a microscope or perform other tests on the cells or tissue. There are many different types of biopsy procedures. The most common types include: (1) incisional biopsy, in which only a sample of tissue is removed; (2) excisional biopsy, in which an entire lump or suspicious area is removed; and (3) needle biopsy, in which a sample of tissue or fluid is removed with a needle. When a wide needle is used, the procedure is called a core biopsy. When a thin needle is used, the procedure is called a fine-needle aspiration biopsy.

**blood** (blud)

A tissue with red blood cells, white blood cells, platelets, and other substances suspended in fluid called plasma. Blood takes oxygen and nutrients to the tissues, and carries away wastes.

**blood chemistry study** (blud KEH-mih-stree STUH-dee)

A procedure in which a sample of blood is examined to measure the amounts of certain substances made in the body. An abnormal amount of a substance can be a sign of disease in the organ or tissue that produces it.

**bone marrow** (bone MAYR-oh)
The soft, sponge-like tissue in the center of most bones. It produces white blood cells, red blood cells, and platelets.

**bone marrow aspiration and biopsy** (bone MAYR-oh AS-pih-RAY-shun ... BY-op-see)

A procedure in which a small sample of bone marrow (soft, sponge-like tissue in the center of most bones) and bone is removed. A small area of skin and the surface of the bone underneath are numbed with an anesthetic. Then a special wide needle is pushed into the bone. A sample of liquid bone marrow is removed with a syringe attached to the needle. The syringe is then removed and the needle is rotated to remove a sample of the bone and the bone marrow. Both the bone marrow and bone samples are sent to a laboratory to be looked at under a microscope.

**bone scan** (bone skan)

A technique to create images of bones on a computer screen or on film. A small amount of radioactive material is injected into a blood vessel and travels through the bloodstream; it collects in the bones and is detected by a scanner.

**breastbone** (brest...)

The long flat bone that forms the center front of the chest wall. The breastbone is attached to the collarbone and the first seven ribs. Also called sternum.

**cancer** (KAN-ser)

A term for diseases in which abnormal cells divide without control and can invade nearby tissues. Cancer cells can also spread to other parts of the body through the blood and lymph systems. There are several main types of cancer. Carcinoma is a cancer that begins in the skin or in tissues that line or cover internal organs. Sarcoma is a cancer that begins in bone, cartilage, fat, muscle, blood vessels, or other connective or supportive tissue. Leukemia is a cancer that starts in blood-forming tissue such as the bone marrow, and causes large numbers of abnormal blood cells to be produced and enter the blood. Lymphoma and multiple myeloma are cancers that begin in the cells of the immune system. Central nervous system cancers are cancers that begin in the tissues of the brain and spinal cord. Also called malignancy.

**cell** (sel)

The individual unit that makes up the tissues of the body. All living things are made up of one or more cells.

**complete blood count** (kum-PLEET blud kownt)

A test to check the number of red blood cells, white blood cells, and platelets in a sample of blood. Also called blood cell count and CBC.

**condition** (kun-DIH-shun)

In medicine, a health problem with certain characteristics or symptoms.

**constipation** (KAHN-stih-PAY-shun)

A condition in which stool becomes hard, dry, and difficult to pass, and bowel movements don’t happen very often. Other symptoms may include painful bowel movements, and feeling bloated, uncomfortable, and sluggish.

**CT scan** (… skan)

A series of detailed pictures of areas inside the body taken from different angles. The pictures are created by a computer linked to an x-ray machine. Also called CAT scan, computed tomography scan, computerized axial tomography scan, and computerized tomography.

**depression** (dee-PREH-shun)
A mental condition marked by ongoing feelings of sadness, despair, loss of energy, and difficulty dealing with normal daily life. Other symptoms of depression include feelings of worthlessness and hopelessness, loss of pleasure in activities, changes in eating or sleeping habits, and thoughts of death or suicide. Depression can affect anyone, and can be successfully treated. Depression affects 15-25% of cancer patients.

device (deh-VISE)

An object that has a specific use. In medicine, wheelchairs, pumps, and artificial limbs are examples of devices.

diagnosis (DY-ug-NOH-sis)

The process of identifying a disease, such as cancer, from its signs and symptoms.

drug

Any substance, other than food, that is used to prevent, diagnose, treat or relieve symptoms of a disease or abnormal condition. Also refers to a substance that alters mood or body function, or that can be habit-forming or addictive, especially a narcotic.

doscopy (EN-doh-SKOPE)

A thin, tube-like instrument used to look at tissues inside the body. An endoscope has a light and a lens for viewing and may have a tool to remove tissue.

doscopy (en-DOS-koh-pee)

A procedure that uses an endoscope to examine the inside of the body. An endoscope is a thin, tube-like instrument with a light and a lens for viewing. It may also have a tool to remove tissue to be checked under a microscope for signs of disease.

family history (FAM-ih-lee HIH-stuh-ree)

A record of the relationships among family members along with their medical histories. This includes current and past illnesses. A family history may show a pattern of certain diseases in a family. Also called family medical history.

fever (FEE-ver)

An increase in body temperature above normal (98.6 degrees F), usually caused by disease.

fluid (FLOO-id)

A substance that flows smoothly and takes the shape of its container. Liquids and gases are fluids.

gadolinium (GA-duh-LIH-nee-um)

A metal element that is used in magnetic resonance imaging (MRI) and other imaging methods. It is a contrast agent, which helps show abnormal tissue in the body during imaging with a special machine.

glucose (GLOO-kose)

A type of sugar; the chief source of energy for living organisms.

groin (groyn)

The area where the thigh meets the abdomen.

gums
The tissue of the upper and lower jaws that surrounds the base of the teeth. Also called gingiva.

**hemoglobin (HEE-moh-GLOH-bin)**

The substance inside red blood cells that binds to oxygen in the lungs and carries it to the tissues.

**hypothyroidism (HY-poh-THY-roy-dih-zum)**

Too little thyroid hormone. Symptoms include weight gain, constipation, dry skin, and sensitivity to the cold. Also called underactive thyroid.

**immune system (ih-MYOON SIS-tem)**

The complex group of organs and cells that defends the body against infections and other diseases.

**incision (in-SIH-zhun)**

A cut made in the body to perform surgery.

**infection (in-FEK-shun)**

Invasion and multiplication of germs in the body. Infections can occur in any part of the body and can spread throughout the body. The germs may be bacteria, viruses, yeast, or fungi. They can cause a fever and other problems, depending on where the infection occurs. When the body’s natural defense system is strong, it can often fight the germs and prevent infection. Some cancer treatments can weaken the natural defense system.

**injection (in-JEK-shun)**

Use of a syringe and needle to push fluids or drugs into the body; often called a "shot."

**Langerhans cell histiocytosis (LANG-er-HANZ sel HIS-tee-oh-sy-TOH-sis)**

A group of rare disorders in which too many Langerhans cells (a type of white blood cell) grow in certain tissues and organs including the bones, skin, and lungs, and damage them. Langerhans cell histiocytosis may also affect the pituitary gland (which makes hormones that control other glands and many body functions, especially growth). Langerhans cell histiocytosis is most common in children and young adults. Also called LCH.

**LCH**

A group of rare disorders in which too many Langerhans cells (a type of white blood cell) grow in certain tissues and organs including the bones, skin, and lungs, and damage them. LCH may also affect the pituitary gland (which makes hormones that control other glands and many body functions, especially growth). LCH is most common in children and young adults. Also called Langerhans cell histiocytosis.

**lens (lenz)**

A clear disk that focuses light, as in a camera or microscope. In the eye, the lens is a clear, curved structure at the front of the eye behind the pupil. It focuses light rays that enter the eye through the pupil, making an image on the retina (light-sensitive layers of nerve tissue at the back of the eye).

**lesion (LEE-zhun)**

An area of abnormal tissue. A lesion may be benign (not cancer) or malignant (cancer).

**liver (LIH-ver)**

A large organ located in the upper abdomen. The liver cleanses the blood and aids in digestion by secreting bile.
liver function test (LIH-ver FUNK-shun ...)

A blood test to measure the blood levels of certain substances released by the liver. A high or low level of certain substances can be a sign of liver disease.

lung

One of a pair of organs in the chest that supplies the body with oxygen, and removes carbon dioxide from the body.

lymph node (limf node)

A rounded mass of lymphatic tissue that is surrounded by a capsule of connective tissue. Lymph nodes filter lymph (lymphatic fluid), and they store lymphocytes (white blood cells). They are located along lymphatic vessels. Also called lymph gland.

microscope (MY-kroh-SKOPE)

An instrument that is used to look at cells and other small objects that cannot be seen with the eye alone.

MRI

A procedure in which radio waves and a powerful magnet linked to a computer are used to create detailed pictures of areas inside the body. These pictures can show the difference between normal and diseased tissue. MRI makes better images of organs and soft tissue than other scanning techniques, such as computed tomography (CT) or x-ray. MRI is especially useful for imaging the brain, the spine, the soft tissue of joints, and the inside of bones. Also called magnetic resonance imaging, NMRI, and nuclear magnetic resonance imaging.

nerve (nerv)

A bundle of fibers that receives and sends messages between the body and the brain. The messages are sent by chemical and electrical changes in the cells that make up the nerves.

neurological exam (NOOR-oh-LAH-jih-kul eg-ZAM)

A series of questions and tests to check brain, spinal cord, and nerve function. The exam checks a person’s mental status, coordination, ability to walk, and how well the muscles, sensory systems, and deep tendon reflexes work.

organ (OR-gun)

A part of the body that performs a specific function. For example, the heart is an organ.

oxygen (OK-sih-jen)

A colorless, odorless gas. It is needed for animal and plant life. Oxygen that is breathed in enters the blood from the lungs and travels to the tissues.

pathologist (puh-THAH-loh-jist)

A doctor who identifies diseases by studying cells and tissues under a microscope.

PDQ

PDQ is an online database developed and maintained by the National Cancer Institute. Designed to make the most current, credible, and accurate cancer information available to health professionals and the public, PDQ contains peer-reviewed summaries on cancer treatment, screening, prevention, genetics, complementary and alternative medicine, and supportive care; a registry of cancer clinical trials from around the world; and directories of physicians, professionals who provide genetics services, and organizations that provide cancer care. Most of this information, and more specific information about PDQ, can be found on the NCI’s Web site at http://www.cancer.gov
PET scan

A procedure in which a small amount of radioactive glucose (sugar) is injected into a vein, and a scanner is used to make detailed, computerized pictures of areas inside the body where the glucose is used. Because cancer cells often use more glucose than normal cells, the pictures can be used to find cancer cells in the body. Also called positron emission tomography scan.

physical examination (FIH-zih-kul eg-ZA-mih-NAY-shun)

An exam of the body to check for general signs of disease.

pituitary gland (pih-TOO-ih-TAYR-ee...)

The main endocrine gland. It produces hormones that control other glands and many body functions, especially growth.

platelet (PLA TE-let)

A tiny piece of a cell found in the blood that breaks off from a large cell found in the bone marrow. Platelets help wounds heal and prevent bleeding by forming blood clots. Also called thrombocyte.

prognosis (prog-NO-sis)

The likely outcome or course of a disease; the chance of recovery or recurrence.

protein (PROH-teen)

A molecule made up of amino acids that are needed for the body to function properly. Proteins are the basis of body structures such as skin and hair and of substances such as enzymes, cytokines, and antibodies.

puberty (PYOO-ber-tee)

The time of life when a child experiences physical and hormonal changes that mark a transition into adulthood. The child develops secondary sexual characteristics and becomes able to have children. Secondary sexual characteristics include growth of pubic, armpit, and leg hair; breast enlargement; and increased hip width in girls. In boys, they include growth of pubic, face, chest and armpit hair; voice changes; penis and testicle growth, and increased shoulder width.

radiation (RAY-dee-AY-shun)

Energy released in the form of particle or electromagnetic waves. Common sources of radiation include radon gas, cosmic rays from outer space, medical x-rays, and energy given off by a radioisotope (unstable form of a chemical element that releases radiation as it breaks down and becomes more stable).

radioactive (RAY-dee-oh-AK-tiv)

Giving off radiation.

radioactive drug (RAY-dee-oh-AK-tiv...)

A drug that contains a radioactive substance and is used to diagnose or treat disease, including cancer. Also called radiopharmaceutical.

radionuclide scanning (RAY-dee-oh-NOO-klide SKAN-ing)

A procedure that produces pictures (scans) of structures inside the body, including areas where there are cancer cells. Radionuclide scanning is used to diagnose, stage, and monitor disease. A small amount of a radioactive chemical (radionuclide) is injected into a vein or swallowed. Different radionuclides travel through the blood to different organs. A machine with a special
camera moves over the person lying on a table and detects the type of radiation given off by the radionuclides. A computer forms an image of the areas where the radionuclide builds up. These areas may contain cancer cells. Also called scintigraphy.

**receptor** (reh-SEP-ter)

A molecule inside or on the surface of a cell that binds to a specific substance and causes a specific physiologic effect in the cell.

**recover** (ree-KUH-ver)

To become well and healthy again.

**recur**

To come back or to return.

**recurrent cancer** (ree-KER-ent KAN-ser)

Cancer that has recurred (come back), usually after a period of time during which the cancer could not be detected. The cancer may come back to the same place as the original (primary) tumor or to another place in the body. Also called recurrence.

**red blood cell**

A cell that carries oxygen to all parts of the body. Also called erythrocyte and RBC.

**risk factor** (... FAK-ter)

Something that increases the chance of developing a disease. Some examples of risk factors for cancer are age, a family history of certain cancers, use of tobacco products, being exposed to radiation or certain chemicals, infection with certain viruses or bacteria, and certain genetic changes.

**scanner** (SKA-ner)

In medicine, an instrument that takes pictures of the inside of the body.

**scientist** (SY-en-tist)

A person who has studied science, especially one who is active in a particular field of investigation.

**somatostatin receptor scintigraphy** (SOH-muh-toh-STA-tin reh-SEP-ter sin-THI-gruh-fee)

A type of radionuclide scan used to find carcinoid and other types of tumors. Radioactive octreotide, a drug similar to somatostatin, is injected into a vein and travels through the bloodstream. The radioactive octreotide attaches to tumor cells that have receptors for somatostatin. A radiation-measuring device detects the radioactive octreotide, and makes pictures showing where the tumor cells are in the body. Also called octreotide scan and SRS.

**spinal cord** (SPY-nul kord)

A column of nerve tissue that runs from the base of the skull down the back. It is surrounded by three protective membranes, and is enclosed within the vertebrae (back bones). The spinal cord and the brain make up the central nervous system, and spinal cord nerves carry most messages between the brain and the rest of the body.

**spine** (spine)

The bones, muscles, tendons, and other tissues that reach from the base of the skull to the tailbone. The spine encloses the spinal cord and the fluid surrounding the spinal cord. Also called backbone, spinal column, and vertebral column.
spleen (spleen)

An organ that is part of the lymphatic system. The spleen makes lymphocytes, filters the blood, stores blood cells, and destroys old blood cells. It is located on the left side of the abdomen near the stomach.

symptom (SIMP-tum)

An indication that a person has a condition or disease. Some examples of symptoms are headache, fever, fatigue, nausea, vomiting, and pain.

thyroid (THY-royd)

A gland located beneath the larynx (voice box) that makes thyroid hormone and calcitonin. The thyroid helps regulate growth and metabolism. Also called thyroid gland.

thyroid gland (THY-royd...)

A gland located beneath the larynx (voice box) that makes thyroid hormone and calcitonin. The thyroid gland helps regulate growth and metabolism. Also called thyroid.

tissue (TISH-oo)

A group or layer of cells that work together to perform a specific function.

tumor (TOO-mer)

An abnormal mass of tissue that results when cells divide more than they should or do not die when they should. Tumors may be benign (not cancer), or malignant (cancer). Also called neoplasm.

ulceration (UL-ser-RAY-shun)

The formation of a break on the skin or on the surface of an organ. An ulcer forms when the surface cells die and are cast off. Ulcers may be associated with cancer and other diseases.

ultrasound (UL-truh-SOWND)

A procedure in which high-energy sound waves are bounced off internal tissues or organs and make echoes. The echo patterns are shown on the screen of an ultrasound machine, forming a picture of body tissues called a sonogram. Also called ultrasonography.

urinalysis (YOOR-in-AL-ih-siss)

A test that determines the content of the urine.

urine (YOOR-in)

Fluid containing water and waste products. Urine is made by the kidneys, stored in the bladder, and leaves the body through the urethra.

vein (vayn)

A blood vessel that carries blood to the heart from tissues and organs in the body.

white blood cell (hwite blud sel)

A type of immune cell. Most white blood cells are made in the bone marrow and are found in the blood and lymph tissue. White blood cells help the body fight infections and other diseases. Granulocytes, monocytes, and lymphocytes are white blood cells. Also called leukocyte and WBC.

x-ray (EX-ray)
A type of high-energy radiation. In low doses, x-rays are used to diagnose diseases by making pictures of the inside of the body. In high doses, x-rays are used to treat cancer.

**Table of Links**

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