Cancer Association of South Africa (CANSA)

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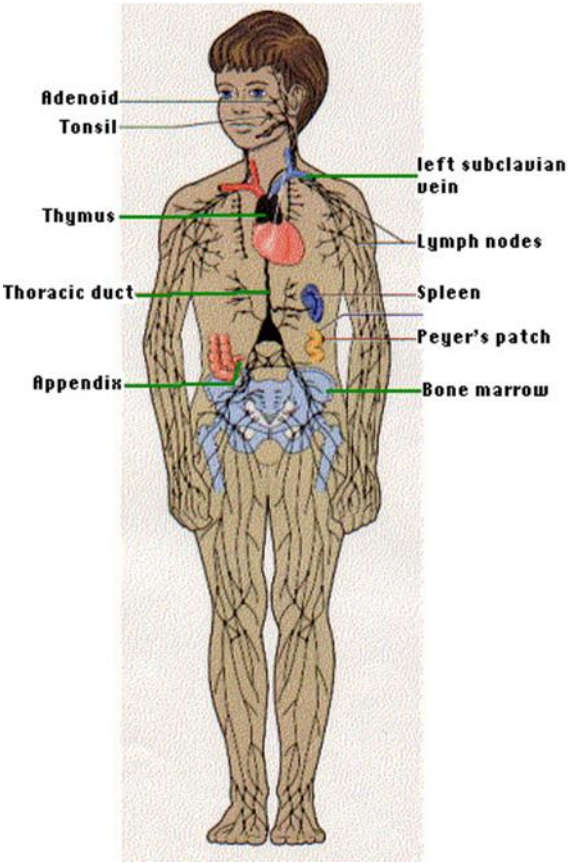
Fact Sheet on Childhood Hodgkin's Lymphoma

Introduction

The term 'lymphoma' refers to cancers that originate in the body's lymphatic tissues. Lymphatic tissues include the lymph nodes (also called lymph glands), thymus, spleen, tonsils, adenoids, and bone marrow, as well as the channels (called lymphatics or lymph vessels) that connect them. Although many types of cancer eventually spread to parts of the lymphatic system, lymphomas are distinct because they actually originate there.



[Picture Credit: Lymphatic System of a Child]

It is said that about 150 children younger than 19 years old are diagnosed with lymphoma each year in South Africa. Lymphomas are divided into three broad categories, depending on the appearance of their cancerous (malignant) cells. These are known as Hodgkin's lymphoma (HL), non-Hodgkin lymphoma (NHL), and Burkitt Lymphoma (BL). Together, they are one of the most common types of cancer in children.

Part of the body's immune system, the lymphatic system is a network of vessels and nodes that normally filters the fluid found within all tissues. Lymph nodes remove bacteria and other disease-causing organisms from the lymph fluid, and produce lymphocytes and antibodies needed to fight off infections caused by these organisms. An increase in the size of a lymph node (lymphadenopathy) indicates increased activity within the node, due to inflammation, infection, or cancer.

Malignancy (cancer) occurs when a cell's genetic code mutates, or changes, resulting in abnormal cells that grow rapidly and in uncontrolled fashion. Lymphomas are a group of cancers originating from

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lymphocytes, which are white blood cells whose normal function is to fight off infections within the body.
(KidsHealth; Lymphoma*Info*.Net).

Incidence of Lymphoma in Children
The incidence of the different types of lymphoma is provided under the description of each of the lymphomas.

Symptoms of Lymphoma in Children
Warning signs for lymphoma are similar in children and adolescents as well as in adults. Symptoms include:

- One or more enlarged lymph nodes in the neck, underarm, or groin, which are usually painless
- Chills
- Swelling of the lymph nodes, which may or may not be painless
- Abdominal swelling (lymphomas in the chest or abdomen can grow to a very large size before symptoms appear)
- Unexplained fever
- Night sweats
- Loss of appetite
- Unexplained weight loss
- Lack of energy
- Coughing
- Difficulty in breathing\
- Itchiness

If a child has a lymph node that becomes enlarged without explanation or remains enlarged for a prolonged period of time, a paediatrician should be consulted. He/she may prescribe a course of antibiotics to treat a possible infection before performing a more extensive evaluation (Memorial Sloan Kettering Cancer Center; Lymphoma Research Foundation).

Causes and Risk Factors of Lymphoma in Children
Although the causes of lymphoma remain unknown, the following may increase the risk of childhood or adolescent lymphomas:

- Family history (though no hereditary pattern has been firmly established)
- Presence or history of an autoimmune disease
- Receipt of an organ transplant
- Exposure to chemicals such as pesticides, fertilizers or solvents
- Infection with viruses such as Epstein-Barr, human T-lymphotropic virus type 1, HIV, hepatitis C, or certain bacteria such as *Helicobacter pylori*

The cause of lymphoma is not known, but there is a genetic component. Incidence rates are higher for those who have a family member diagnosed with lymphoma, especially a sibling. While environmental

and lifestyle factors are known to play a role in the development of cancer among adults, these factors have less of an impact on the development of childhood cancer.

Hodgkin's Lymphoma

Cancer cells of these patients are usually abnormal B-cells referred to as a Reed-Sternberg (R-S) cells. Although less commonly observed, R-S cells may also develop from T-cells. R-S cells most often develop in lymph nodes located in the upper body regions and spread to neighbouring lymph nodes via lymphatic vessels. There are two distinct types of Hodgkin's lymphoma: *classical* and *non-classical* Hodgkin's lymphoma.

[Picture Credit: Hodgkin's Lymphoma]

- Classical Hodgkin's Lymphoma: This lymphoma features R-S cells with a classical appearance. It may be diagnosed as Nodular Sclerosis Hodgkin Disease, Mixed Cellularity Hodgkin's Disease, Lymphocyte-Rich Hodgkin Disease or Lymphocyte-Depleted Hodgkin Disease.
- Non-classical Hodgkin's Lymphoma: This lymphoma features larger cancer cells that are variants of R-S cells and is most often found in the nodes of the upper body, arms and neck.

Classic Hodgkin lymphoma is divided into four subtypes, based on how the cancer cells look under a microscope:

- Nodular-sclerosing Hodgkin lymphoma occurs most often in older children and adolescents. It is common to have a chest mass at diagnosis.
- Mixed cellularity Hodgkin lymphoma most often occurs in children younger than 10 years of age. It is linked to a history of Epstein-Barr Virus (EBV) infection and often occurs in the lymph nodes of the neck.
- Lymphocyte-rich classic Hodgkin lymphoma is rare in children. When a sample of lymph node tissue is looked at under a microscope, there are Reed-Sternberg cells and many normal lymphocytes and other blood cells.
- Lymphocyte-depleted Hodgkin lymphoma is rare in children and occurs most often in adults or adults with the Human Immunodeficiency Virus (HIV). When a sample of lymph node tissue is looked at under a microscope, there are many large, oddly shaped cancer cells and few normal lymphocytes and other blood cells.

Incidence of Childhood Hodgkin's Lymphoma

According to the National Cancer Registry (2017) the following number of Hodgkin's Lymphoma cases were histologically diagnosed in South Africa during 2017:

Group Boys: 0 to 19 Years 2017	Actual No of Cases
All boys	56
Asian boys	0
Black boys	39
Coloured boys	7
White boys	11

Group Girls: 0 to 19 Years 2017	Actual No of Cases
All girls	32
Asian girls	4
Black girls	20
Coloured girls	2
White girls	6

The frequency of histologically diagnosed cases of Hodgkin Lymphoma in South Africa for 2017 was as follows (National Cancer Registry, 2017):

Group Boys: 0 to 19 Years 2017	0 – 4 Years	5 – 9 Years	10 – 14 Years	15 – 19 Years
All boys	7	14	16	19
Asian boys	0	0	0	0
Black boys	5	10	10	14
Coloured boys	0	2	3	2
White boys	2	2	3	4

Group Girls: 0 to 19 Years 2017	0 – 4 Years	5– 9 Years	10 – 14 Years	15 – 19 Years
All girls	4	3	8	17
Asian girls	1	0	0	3
Black girls	3	2	6	9
Coloured girls	0	0	1	1
White girls	0	1	1	4

N.B. In the event that the totals in any of the above tables do not tally, this may be the result of uncertainties as to the age, race or sex of the individual. The totals for 'all boys' and 'all girls', however, always reflect the correct totals.

Diagnosis of Hodgkin's Lymphoma (HL)

The following tests and procedures may be used:

Physical examination and history - an examination 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.

CT Scan (CAT scan) - a procedure that makes a series of detailed pictures of areas inside the body, such as the neck, chest, abdomen, or pelvis, taken from different angles. The pictures are made by a computer linked to an X-ray machine A dye may be infected 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.

Pet Scan (positron emission tomography scan) – a procedure to find malignant tumour 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. Malignant tumour cells show up brighter in the picture because they are more active and take up more glucose than normal cells do. Sometimes a PET scan and a CT scan are done at the same time. If there is any cancer, this increases the chance that it will be found.

Chest X-ray - an x-ray of the organs and bones inside the chest. 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.

Complete Blood Count (CBC) - a procedure in which a sample of blood is drawn and checked for the following:

- The number of red blood cells, white blood cells, and platelets.
- The amount of haemoglobin (the protein that carries oxygen) in the red blood cells.
- The portion of the blood sample made up of red blood cells.

Blood Chemistry Studies - a procedure in which a blood sample is checked to measure the amounts of certain substances released into the blood 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.

Sedimentation Rate - a procedure in which a sample of blood is drawn and checked for the rate at which the red blood cells settle to the bottom of the test tube.

Lymph node biopsy - the removal of all or part of a lymph node. The lymph node may be removed during a thoracoscopy, mediastinoscopy, or laparoscopy. One of the following types of biopsies may be done:

- Excisional biopsy - the removal of an entire lymph node.
- Incisional biopsy - the removal of part of a lymph node.
- Core biopsy - the removal of tissue from a lymph node using a wide needle.
- Fine-needle aspiration (FNA) biopsy - the removal of tissue from a lymph node using a thin needle.

A pathologist views the tissue under a microscope to look for cancer cells, especially Reed-Sternberg cells. Reed-Sternberg cells are common in classical Hodgkin lymphoma.

The following test may be done on tissue that was removed:

- Immunophenotyping - a laboratory test used to identify cells, based on the types of antigens or markers on the surface of the cell. This test is used to diagnose the specific type of lymphoma by comparing the cancer cells to normal cells of the immune system.

Treatment of Childhood Lymphoma

Treatment of childhood lymphoma is largely determined by staging. Staging is a way to categorise or classify patients according to how extensive the disease is at the time of diagnosis.

The types of standard treatment used include:

- Chemotherapy
- Radiation therapy
- Targeted therapy
- Immunotherapy
- Surgery
- High-dose chemotherapy with stem cell transplant

New types of treatment are being tested in clinical trials

Chemotherapy (the use of highly potent medical drugs to kill cancer cells) is the primary form of treatment for all types of lymphoma.

In certain cases, radiation therapy (the use of high-energy rays to shrink tumours and keep cancer cells from growing), may also be used.
Short-term and long-term side effects - Intensive lymphoma chemotherapy affects the bone marrow, causing anaemia and bleeding problems, and increasing the risk for serious infections. Chemotherapy and radiation treatments have many other side effects — some short-term (such as hair loss, changes in skin colour, increased infection risk, and nausea and vomiting) and some long-term (such heart and kidney damage, reproductive problems, thyroid problems, or the development of another cancer later in life) — that parents should discuss with their doctor.

Relapses - Although most kids do recover from lymphoma, some with severe disease will have a relapse (reoccurrence of the cancer). For these children, bone marrow transplants and stem-cell transplants are often among the newest treatment options.

During a bone marrow/stem cell transplant, intensive chemotherapy with or without radiation therapy is given to kill residual cancerous cells. Then, healthy bone marrow/stem cells are introduced into the body in the hopes that it will begin producing white blood cells that will help the child fight infections.

New Treatments - Promising new treatments being developed for childhood lymphomas include several different types of immunotherapy, specifically the use of antibodies to deliver chemotherapy medicines or radioactive chemicals directly to lymphoma cells. This direct targeting of lymphoma cells may avoid the toxic side effects that occur when today's chemotherapy and radiation treatments damage normal, noncancerous body tissues.

About Clinical Trials
Clinical trials are research studies that involve people. They are conducted under controlled conditions. Only about 10% of all drugs started in human clinical trials become an approved drug.

Clinical trials include:

- Trials to test effectiveness of new treatments
- Trials to test new ways of using current treatments
- Tests new interventions that may lower the risk of developing certain types of cancers

- Tests to find new ways of screening for cancer

The South African National Clinical Trials Register provides the public with updated information on clinical trials on human participants being conducted in South Africa. The Register provides information on the purpose of the clinical trial; who can participate, where the trial is located, and contact details.

For additional information, please visit: www.sanctr.gov.za/

Medical Disclaimer

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Hodgkin's Lymphoma

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