

Childhood Hodgkin Lymphoma

What is Hodgkin Lymphoma (HL)?

Hodgkin Lymphoma is a cancer arising within cells of the lymph glands (which includes the Spleen). There are many kinds of lymphoma, but broadly speaking these are divided into Hodgkin Lymphoma (HL) which spreads through the lymphatic system slowly, and Non Hodgkin Lymphomas (NHL) which *in children* tends to grow more quickly and spread through the blood stream. With modern treatment programs using multiagent chemotherapy, with or without radiation therapy, over 80% of children with HL will be cured. Cure rates vary depending on the risk factors and your doctor will give you a better estimate after taking those all into account.

What are the signs of childhood HL?

These are related to invasion of lymph nodes or lymph tissue but can also be due to substances released by the cancer:

- Painless lumps in the neck, collarbone area, armpits or groin
- Mass inside the chest
- Feeling tired or looking pale
- Weight loss, night sweats or fevers
- Itching

What causes HL?

Nobody knows for sure, there is nothing in the diet that caused this. It is typically not inherited so other children in the family should not be at risk. It has been linked to viral infections such as “mono” (EBV), but it is not infectious so it cannot be passed to other children or playmates.

How is HL diagnosed?

- A careful physical exam and history may show some or all of the signs above.
- Blood tests such as complete blood count (CBC), blood chemistry and viral tests support but do not confirm the diagnosis.
- Chest x-ray may show if the lymphoma has formed a mass in the chest.
- CT scan: is a special kind of X-ray that shows more clearly enlargement of lymph nodes, liver, spleen and chest masses in detail.
- Lymph node biopsy is the key test in which an affected lymph node is removed (biopsy) by a surgeon and sent to a pathologist who reviews the lymph tissue under a microscope looking for cancerous cells (Reed-Sternberg cells), and looking at markers on the surface of the cancer cells. This allows an accurate diagnosis and sub-type
- PET-CT scan is a special kind of scan that can show where the lymphoma has spread in the body (and later can also show whether it is responding to treatment).
- Bone marrow aspiration and biopsy, removal of liquid bone marrow and a tiny piece of bone by inserting a needle into the hipbone while the child is sedated can check for spread of HL, it may not be needed if a PET-CT scan has already been done.

What is risk grouping in HL?

Risk factors tell us how likely the HL will come back without adequate treatment (relapse) and are used to plan chemotherapy, which includes information at diagnosis:

- Ann Arbor Stage (disease spread, I being localized, and IV being widespread)
- Presence of fever, weight loss, and night sweats (B symptoms), absence labeled as A
- Bulky disease.

Based on these findings your child will be allotted a stage e.g. stage IIB involves more than one lymph node group still relatively localized, but with weight loss and night sweats. The risk categories vary between US and European protocols, nevertheless all localized asymptomatic disease (IA, IIA) is low risk, all patients with widespread symptomatic disease (IIIB, IVB) are high risk, and in-between stages are intermediate risk. In the figure below low risk is blue, intermediate risk orange, and high risk red in color.

Study Group	Risk	IA	IB	IIA	IIB	IIIA	IIIB	IVA	IVB
COG	AHOD0431 – Low	Blue		Blue					
	AHOD0031 – Intermediate	E		E					
	AHOD0831 – High	X		X					
EuroNet-PHL-C1*	TG1 – Low	Blue		Blue					
	TG2 – Intermediate	E	E	E					
	TG3 – High	RF	RF	RF			E	E	
EuroNet-PHL-C2	TL1 – Low	Blue		Blue					
	TL2 – Intermediate	E	E	E					
	TL3 – High	RF	RF	RF			E	E	
Pediatric Hodgkin Consortium	HOD99/HOD08 – Low	Blue		< 3 ns					
	HOD05 – Intermediate	E		E					
	HOD99/HLHR13 – High	mX		mX					

We also look at how at a CT or PET-CT scan done after 1-2 cycles of chemotherapy to see how the tumor has shrunk or responded to treatment, which helps us adjust treatment.

Finally looking under the microscope, the pathologist can subtype HL into 1) Nodular Lymphocyte-Predominant and 2) Classical, which can affect the treatment you receive.

What treatment will my child receive?

Multiple treatment protocols are used for HL: a common protocol is ABVD consisting of the following four cancer fighting (chemotherapy) drugs given into a vein on days 1 and 15 of each month: Doxorubicin (Adriamycin™), Bleomycin, Vinblastine, Dacarbazine.

Chemotherapy drugs are given as a combination to increase the chance of killing all the cancer cells, and injected into a vein to reach cancer cells throughout the body. Typically 4-6 cycles are administered in an outpatient or day care unit. However if your child develops fever or low blood counts admission to hospital may be needed. If your child has bulky disease or other risk factors your physician may decide to also administer low dose radiation over 2 weeks. A very small number of children may rarely need a bone marrow transplant (BMT) if treatment fails.

All people who receive chemotherapy are at risk of side effects, which usually get better when the medication is stopped. Common side effects include nausea, vomiting, hair loss, low blood counts and fatigue (tiredness). Drugs are given to prevent nausea and vomiting. Hair loss is usually temporary but rarely it may be permanent. Low blood counts may require



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transfusion or increase the risk of infection. Chemotherapy side effects for each medication are listed on the sheets your doctor will give you. It is important to remember that the treatment given is essential for cure and most patients will have very few of the side effects listed.

What dietary and other precautions should I take?

Chemotherapy suppresses your child's immune system so infections are a concern. We prohibit street/outside food and recommend home food be thoroughly cooked. No special foods will help with cure but we ban vitamin or nutritional supplements that might interfere with chemotherapy. Visits to movie theaters, malls, weddings, or other crowded places and plane, bus or railway travel are all discouraged. You should contact your doctor immediately if in spite of these precautions your child develops a fever >100.4 F, diarrhea, cough, vomiting, abdominal pain, headaches or looks unwell.

What are long term outcomes?

HL is highly curable, and patients lead long happy lives after finishing treatment. Because a few patients may have long-term side effects or "late effects" from the chemotherapy and radiation given, it is important to follow up with your doctor after finishing treatment.