

Non-Hodgkin Lymphoma in Children

Description/Etiology

Non-Hodgkin lymphoma (NHL) is cancer of the lymphatic system characterized by uncontrolled proliferation of lymphocytes (i.e., white blood cells [WBCs] that comprise much of the body's immune system). Congenital or acquired genetic abnormalities transform B or T lymphocytes, or a combination of B and T lymphocytes, to the malignant cells characteristic of NHL that invade and compromise the function of various organs. NHL is less predictable than Hodgkin disease and more likely to involve non-lymph node areas. The three most common types of NHL in children (i.e., denoting a pediatric population from birth to 18 years of age) are Burkitt lymphoma (involving B cells), lymphoblastic lymphoma (involving T cells), and large-cell lymphoma (involving B cells or, less commonly, T cells).

The clinical features of NHL in children vary according to the primary sites of involvement and the extent of disease. Although laboratory tests and imaging studies are typically performed to evaluate the extent of disease, laboratory examination of cerebrospinal fluid and/or biopsied tissue is necessary to confirm the diagnosis (for details, see *Laboratory Studies That May Be Performed*, below).

Treatment depends on the type, grade, and stage of disease, and a single or combination treatment regimen may be appropriate. Treatment options are chemotherapy, monoclonal antibody-based treatments, bone marrow or peripheral stem cell transplantation, radiation therapy, and, in cases of cutaneous T-cell lymphoma, photochemotherapy (i.e., exposure to sunlight or ultraviolet light). Prevention of complications (e.g., infection, adverse effects of treatment, multiple organ failure), emotional support, patient education, and family involvement play a significant role in treatment. Because children with NHL are more likely than adults to present with high-grade (i.e., characterized by acute onset and rapid progression), disseminated disease with extranodal involvement, prompt diagnosis and treatment are essential. NHL usually responds well to treatment, and the long-term prognosis is excellent for most types of NHL in children.

ICD-10

C85.7, C85.9

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Facts and Figures

Lymphomas, including both NHL and Hodgkin lymphoma, are the third most common childhood cancers after acute leukemias and brain cancer (for more information on Hodgkin lymphoma, see *Quick Lesson About ... Hodgkin's Lymphoma in Childhood/Adolescence*). NHL accounts for an estimated 7% of all malignancies in children. In the United States, the age-adjusted incidence of NHL is 1.1 per 100,000 individuals from birth to 19 years of age. The incidence of NHL is almost 2 times higher among males than females, and NHL is twice as common in Whites compared with Blacks. NHL in children is highly curable. With current therapies, cure rates are 70–90%.

Risk Factors

Most children with NHL have no known risk factors. Children who are at highest risk for NHL are those with congenital or acquired immunodeficiency syndromes, DNA repair syndromes (ie. Nijmegen breakage syndrome), a previous neoplasm, or Epstein-Barrvirus (EBV) infection, and those who have received immunosuppressive therapy for organ or bone marrow transplants. In sub-Saharan Africa, development of Burkitt lymphoma is associated with exposure to both malaria and EBV.

Signs and Symptoms/Clinical Presentation

The clinical features of NHL in children vary according to the primary sites of involvement and the extent of disease. Children usually present with extranodal masses in the abdomen, head and neck, or mediastinum. Signs and symptoms of abdominal involvement include ascites, nausea, vomiting, abdominal pain, and bowel or bladder obstruction. Primary involvement of the mediastinum can result in respiratory symptoms and swelling of the face, neck, and shoulder. CNS involvement may cause increased intracranial pressure, which causes nausea, vomiting, headache, vision changes, and neurologic impairment.

Assessment

› Patient History

- Obtain detailed information regarding the onset and duration of signs and symptoms

› Laboratory Tests That May Be Ordered

- CBC can identify anemia (indicated by a low Hgb), infection (indicated by elevated WBC), and decreased immune function (indicated by low platelet count and/or WBC) resulting from chemotherapy and radiation therapy
- Serum chemistry panel can identify complications of NHL (e.g., hyperuricemia, elevated lactic dehydrogenase [LDH], electrolyte abnormalities, acidosis), if present
- Cytologic examination of cerebrospinal fluid or histologic examination of bone marrow aspirate, tumor, or lymph node tissue will identify cancer cell type if present
 - Cytologic examination of cerebrospinal fluid can identify metabolic abnormalities, if present, which would affect NHL treatment

› Other Diagnostic Tests/Studies

- X-ray, MRI and CT scan will identify abnormal regions that require biopsy for histologic analysis, if present
- PET scan and bone scan will identify areas of NHL involvement throughout the body, if present

Treatment Goals

› Promote Optimal Physiologic Function and Reduce Risk of Complications

- Monitor vital signs, all physiologic systems, and laboratory/other diagnostic study results; report abnormalities and treat, as ordered
- Frequently assess for pain; administer analgesic, as ordered; reassure patient and family that pain can be controlled
 - Notify the treating clinician of unresolved pain, and request referral to a pain management clinician if warranted
- Follow facility pre- and post-treatment protocols if patient becomes a candidate for surgery (e.g., for bone marrow transplantation), chemotherapy, or radiation therapy; reinforce pre- and post-treatment education and verify completion of informed consent documents by a parent or legal guardian
- Administer or monitor the administration of systemic chemotherapy (e.g., cyclophosphamide-based combination chemotherapy), as ordered
- Assess for myelosuppression, a common side effect of chemotherapy
 - Monitor laboratory values and vital signs, with attention to absolute neutrophil count and body temperature
 - Administer transfusion of blood products, erythropoietin, growth factors, aminocaproic acid, and anti-infective agents, as ordered, to restore hematologic cells and prevent infection
 - Encourage adequate rest and periods of exercise, as tolerated, to manage fatigue
- Assess for tumor lysis syndrome (TLS), a side effect of chemotherapy
 - Monitor for signs and symptoms of TLS such as restlessness, irritability, diarrhea, decrease in urination, or leg cramps
 - Monitor lab values and vital signs, with attention to potassium, uric acid, phosphate, and calcium
 - Administer TLS treatment as prescribed (e.g. allopurinol, Imbruvica, or venclaxa)
 - Encourage fluids and closely monitor intake and output
- Assess for and treat gastrointestinal side effects of chemotherapy and radiation therapy (e.g., nausea, vomiting, diarrhea)
 - Administer antiemetic and antidiarrheal medications, as ordered, and encourage adequate fluid intake to prevent dehydration
 - Encourage and/or provide nonpharmacologic therapy (e.g., distraction, gentle massage) for management of related discomfort
- Provide good hygiene and assess skin integrity in area of radiation therapy; apply topical agents, as ordered
- Provide a high-energy, high-protein diet, as ordered, with consideration to food preferences; request referral to a dietitian, if appropriate, for patient assessment and patient/parent education

- Monitor closely for signs and symptoms of infection, drug toxicity, or organ failure and administer prescribed treatment, as ordered
- › **Promote Emotional Well-Being, Educate, and Encourage Active Participation in Decisions for the Therapeutic Plan**
 - Assess anxiety level and coping ability of patient and family; provide emotional support and promote a positive self-image for patients who have experienced a dramatic change in lifestyle due to NHL-related functional limitations
 - Educate and encourage discussion regarding NHL, diagnostic procedures, risks and benefits of treatment (including implications regarding fertility) and pain management options, changes in body image and function, what to expect during recovery from treatment and individualized prognosis; provide age-appropriate written information to reinforce verbal education
 - Encourage patient and family involvement in treatment planning and communication with the treating clinician, dietitian, social worker, and/or specialty clinicians
 - For children who are self-conscious about hair loss from chemotherapy, request referral to an oncology-related source for wigs and provide encouragement, understanding, and creativity in coping with having a disturbed body image
 - Request referral, if appropriate, to a
 - mental health clinician, facility chaplain, or the patient’s clergy person for counseling on strategies for coping with NHL
 - social worker for identification of local resources for in-home care, transportation, financial planning, and support groups

Food for Thought

- › In a 2016 study aimed at determining the relationship between adolescent health and occurrence of NHL, researchers found that overweight and obese adolescents as well as those with a taller stature were at an increased risk for NHL (Leiba et al., 2016)

Red Flags

- › Chemotherapy has the potential to produce multiple severe side effects and adverse reactions; knowledge of all prescribed agents must precede administration of the agents and providing care for a patient who is receiving chemotherapy

What Do I Need to Tell the Patient/Patient’s Family?

- › Advise the patient to seek immediate medical attention for new or worsening signs and symptoms (e.g., extranodal mass, abdominal pain, nausea) and for complications or side effects of treatment (e.g., vomiting, skin breakdown, diarrhea)
- › Provide detailed information about what to expect during recovery, how to recognize signs and symptoms of relapse, how to manage pain, how to recognize activity tolerance limits, and what continued outpatient therapy is required
- › Educate that immunocompromised patients who are receiving radiation therapy or chemotherapy should not eat raw fruits, vegetables, or meats (e.g., sushi); all foods must be thoroughly cooked
- › Encourage children (as age-appropriate) and parents to join a support group for contact with others who face similar health challenges

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