

Childhood Cancer for the Primary Care Physician



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KEYWORDS

- Childhood cancer • Acute leukemia • Lymphoma • CNS tumors • Bone tumors
- Sarcoma • Wilms tumors • Risk factors

KEY POINTS

- Childhood cancer is rare among childhood diseases; however, it is a leading cause of death in children.
- Childhood cancer is a challenging disease entity because of its rarity, and it often presents with symptoms that overlap with more common, benign diseases.
- Childhood cancer can present at any age or in any organ.
- Although childhood cancer is usually a random event, it can be predisposed by certain genetic, familial, or immunologic disorders. Physicians need to have a general knowledge about these disorders so that they have an increased level of suspicion for cancer in these patients.
- Childhood cancer can be a life-threatening medical emergency that requires immediate and swift action to prevent loss of life.

INTRODUCTION

Childhood cancer is rare among childhood diseases. Despite being a rare disease entity in children, approximately 10,000 to 15,000 new cases per year of cancer occur in the first 2 decades of life in the United States.¹ Before they reach adulthood, 1 child in every 600 develops some type of childhood cancer, which is a principal cause of death in children.¹⁻⁴ Despite being a leading cause of death, survival of childhood cancer is much higher than that of adult cancer and is improving over time.⁵ Making

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the diagnosis of childhood cancer can be challenging in the primary care setting. This difficulty may be caused by the vagueness of the presenting signs and symptoms of childhood cancer, the rarity of the cancer in childhood compared with adulthood, and the low index of suspicion that primary care providers have when dealing with such nonspecific symptoms.^{3,4,6} The mean time of delay in diagnosis was found in one large review of 23 studies of childhood cancer to range from as short as 2.5 weeks for neuroblastoma to almost 30 weeks for brain tumors. This article about common childhood cancers discusses the cancers commonly encountered by general practitioners, focusing on key points that can help physicians recognize cancer early in the diagnostic process.

ACUTE LEUKEMIA

Epidemiology

Acute leukemia in children is categorized into 2 major disease types: acute lymphoblastic leukemia (ALL) and acute myeloid leukemia (AML), based on the phenotype of the leukemia blast cells. ALL is the most common cancer in children in the United States, accounting for 26% of new cancer diagnoses in the birth to 14-year age group and 2670 new cases per year.⁷ ALL is more common in boys than in girls and in Hispanic and white children than in African American children. The peak incidence of ALL is among children aged 2 to 4 years. AML occurs in approximately 500 children in the United States per year, with the peak incidence in children less than 2 years old.⁷ In the United States, survival rates of pediatric leukemia have improved over the past few decades, and now most children treated for leukemia survive without disease recurrence.

Clinical Presentation

Signs and symptoms of acute leukemia in children include fever (61% of cases), bone pain, lymphadenopathy, hepatomegaly, splenomegaly, pallor, bruising, and petechiae.¹ Bone pain may manifest in a young child as a limp or refusal to walk.⁸ Constitutional symptoms, such as anorexia, weight loss, and fatigue, are also common. Complete blood count (CBC) findings may include leukocytosis or leukopenia (commonly neutropenia), anemia, and/or thrombocytopenia. Leukemia blast cells may be seen on the peripheral blood smear, but the absence of circulating blasts does not rule out acute leukemia. Other possible manifestations of acute leukemia include respiratory compromise secondary to a mediastinal mass; tumor lysis syndrome resulting in hyperuricemia, hyperkalemia, hyperphosphatemia, and acute kidney injury; opportunistic infections; disseminated intravascular coagulation (DIC); extramedullary infiltration of leukemia blasts into the skin or other soft tissues; and central nervous system (CNS) symptoms such as cranial nerve palsies and headaches.⁹

Differential Diagnosis

In the absence of blasts on the peripheral blood smear, the differential diagnosis should include infectious causes, autoimmune disease, and other malignancies. Lymphoma may present with features similar to acute leukemia. Neuroblastoma may also manifest with symptoms suggestive of leukemia, secondary to metastatic infiltration of the malignant cells into the bone marrow, liver, and bones.

Initial Diagnostic Work-up

Complete physical examination, with attention to vital sign stability, lymph nodes, liver and spleen size, skin, testes, and neurologic signs, is important at baseline. Initial

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