

## Cancer in Children and Adolescents



### How common is cancer in children?

Although cancer in children is rare, it is the leading cause of death by disease past infancy among children in the United States. In 2017, it is estimated that 15,270 children and adolescents ages 0 to 19 years will be diagnosed with cancer and 1,790 will die of the disease in the United States.<sup>1</sup> Among children ages 0 to 14 years, it is estimated that 10,270 will be diagnosed with cancer and 1,190 will die of the disease in 2017.<sup>1</sup>

The most common types of cancer diagnosed in children ages 0 to 14 years in the United States are leukemias, followed by brain and other central nervous system tumors, lymphomas, soft tissue sarcomas (of which half are rhabdomyosarcoma), neuroblastoma, and kidney tumors.<sup>1</sup> The most common types of cancer diagnosed in 15- to 19-year-olds are lymphomas, followed by brain and other central nervous system tumors, leukemias, gonadal (testicular and ovarian) germ cell tumors, thyroid cancer, and melanoma.<sup>1</sup>

As of January 1, 2014 (the most recent date for which data exists), approximately 419,000 survivors of childhood

and adolescent cancer (diagnosed at ages 0 to 19 years) were alive in the United States.<sup>2</sup> The number of survivors will continue to increase, given that the incidence of childhood cancer has been rising slightly in recent decades and that survival rates overall are improving.

### What is the outlook for children with cancer?

The overall outlook for children with cancer has improved greatly over the last half-century. In 1975, just over 50% of children diagnosed with cancer before age 20 years survived at least 5 years.<sup>3</sup> In 2007–2013, 83% of children diagnosed with cancer before age 20 years survived at least 5 years.<sup>4</sup>

Although survival rates for most childhood cancers have improved in recent decades, the improvement has been especially dramatic for a few cancers, particularly acute lymphoblastic leukemia, which is the most common childhood cancer. Improved treatments introduced beginning in the 1960s and 1970s raised the 5-year survival rate for children diagnosed with acute lymphoblastic leukemia before age 20 years from less than 10% in the

1960s to about 88% in 2007–2013.<sup>4</sup> The 5-year survival rate for children diagnosed with non-Hodgkin lymphoma before age 20 years has also increased dramatically, from less than 50% in the late 1970s to about 89% in 2007–2013.<sup>4</sup>

A notable example of how treatment advances have improved the outlook for children with leukemia is reflected in recent data showing that during 1999–2014, brain cancer replaced leukemia as the leading cause of cancer death among 1- to 19-year-olds.<sup>5</sup>

By contrast, survival rates remain very low for some cancer types, for some age groups, and for some cancers within a site. For example, median survival for children with diffuse intrinsic pontine glioma (a type of brain tumor) is less than 1 year from diagnosis.<sup>6</sup> Among children with Wilms tumor (a type of kidney cancer), older children (those diagnosed between ages 10 and 16 years) have worse 5-year survival rates than younger children.<sup>7</sup> For soft tissue sarcomas, 5-year survival rates in 2007–2013 among children and adolescents ages 0 to 19 years ranged from 65% (rhabdomyosarcoma) to 95%



(chondrosarcoma),<sup>4</sup> but children with sarcomas who present with metastatic disease have much lower 5-year survival rates.

The cancer mortality rate—the number of deaths due to cancer per 100,000 people per year—among children ages 0 to 19 years declined by more than 50% from 1975–1977 to 2010–2014.<sup>8</sup> Specifically, the mortality rate was slightly more than 5 per 100,000 children in 1975 and about 2.1 per 100,000 children in 2010–2014. However, despite the overall decrease in mortality, approximately 1,800 children die of cancer each year in the United States, indicating that new advances and continued research to identify effective treatments are required to further reduce childhood cancer mortality.

### What are the possible causes of cancer in children?

The causes of most childhood cancers are not known. About 5% of all cancers in children are caused by an inherited genetic mutation (a mutation that can be passed from parents to their children). For example, 25% to 30% of cases of retinoblastoma, a cancer of the eye that develops mainly in children, are caused by an inherited mutation in a gene called RB1.<sup>9</sup> However, retinoblastoma accounts for only about 4% of all cancers in children ages 0 to 14 years. Inherited mutations associated with certain familial syndromes, such as Li-Fraumeni syndrome, Beckwith-Wiedemann syndrome, Fanconi anemia syndrome, Noonan syndrome, and von Hippel-Lindau syndrome, also increase the risk of childhood cancer.

Genetic mutations that initiate cancer development can also arise during the development of a fetus in the womb. Evidence for this comes from studies of monozygotic (identical) twins in which both twins developed leukemia with an identical leukemia-initiating gene mutation.<sup>10</sup>

Children who have Down syndrome, a genetic condition caused by the presence of an extra copy of chromosome 21, are 10 to 20 times more likely to develop leukemia than children without Down syndrome.<sup>11</sup> However, only a very small proportion of childhood leukemia is linked to Down syndrome.

Most cancers in children, like those in adults, are thought to develop as a result of mutations in genes that lead to uncontrolled cell growth and eventually cancer. In adults, these gene mutations are often the result of exposure to environmental factors, such as cigarette smoke, asbestos, and ultraviolet radiation from the sun. However, environmental causes of childhood cancer have been difficult to identify, partly because cancer in children is rare, and partly because it is difficult to determine what children might have been exposed to early in their development.

Many studies have shown that exposure to ionizing radiation can damage DNA, which can lead to the development of childhood leukemia and possibly other cancers. For example, children and adolescents who were exposed to radiation from the World War II atomic bomb blasts had an elevated risk of leukemia,<sup>12</sup> and children and adults who were exposed to radiation from

accidents at nuclear power plants had an elevated risk for thyroid cancer.<sup>13</sup> Children whose mothers had x-rays during pregnancy (that is, children who were exposed before birth) and children who were exposed after birth to diagnostic medical radiation from computed tomography scans also have an increased risk of some cancers.<sup>14</sup>

Studies of other possible environmental risk factors, including parental exposure to cancer-causing chemicals, prenatal exposure to pesticides, childhood exposure to common infectious agents, and living near a nuclear power plant, have produced mixed results. Whereas some studies have found associations between these factors and risk of some cancers in children, other studies have found no such associations.<sup>15–17,18</sup> Higher risks of cancer have not been seen in children who have a parent who was diagnosed with and treated for a childhood cancer that was not caused by an inherited mutation.<sup>19</sup>

### Where do children with cancer get treated?

Children who have cancer are often treated at a children's cancer center, which is a hospital or a unit within a hospital that specializes in diagnosing and treating children and adolescents who have cancer. Most children's cancer centers treat patients up to 20 years of age. The health professionals at these centers have specific training and expertise to provide comprehensive care for children, adolescents, and their families.

Children's cancer centers also participate in clinical trials. The improvements in survival for children with

cancer that have occurred over the past half century have been achieved because of treatment advances that were studied and proven to be effective in clinical trials.

More than 90% of children and adolescents who are diagnosed with cancer each year in the United States are cared for at a children's cancer center that is affiliated with the NCI-supported Children's Oncology Group (COG). COG is the world's largest organization that performs clinical research to improve the care and treatment of children and adolescents with cancer. Each year, approximately 4,000 children who are diagnosed with cancer enroll in a COG-sponsored clinical trial.

### What should survivors of childhood cancer consider after they complete treatment?

Survivors of childhood cancer need follow-up care and enhanced medical surveillance for the rest of their lives because of the risk of complications that can occur many years after they complete treatment for their cancer. Health problems that develop months

or years after treatment has ended are known as late effects.

Long-term follow-up analysis of a cohort of survivors of childhood cancer treated between 1970 and 1986 has shown that cancer survivors remain at risk of complications and premature death as they age, with more than half of survivors having experienced a severe or disabling complication or even death by the time they reach age 50 years.<sup>21</sup> Children treated in more recent decades may have lower risks of late complication or mortality due to modifications in treatment regimens to reduce exposures to radiotherapy and chemotherapy, increased efforts to detect late effects as early as possible, and improvements in medical care for late effects of therapy.<sup>22</sup>

The specific late effects that a person who was treated for childhood cancer might experience depend on the type and location of his or her cancer, the type of treatment he or she received, and patient-related factors, such as age at diagnosis.

Children who were treated for bone cancer, brain tumors, and Hodgkin lymphoma, or who received radiation

to their chest, abdomen, or pelvis, have the highest risk of serious late effects from their cancer treatment, including second cancers, joint replacement, hearing loss, and congestive heart failure.<sup>23,24</sup>

It's important for childhood cancer survivors to have regular medical follow-up examinations so any health problems that occur can be identified and treated as soon as possible. The Children's Oncology Group (COG) has developed long-term follow-up guidelines for survivors of childhood, adolescent, and young adult cancers.

Many children's cancer centers have follow-up clinics where survivors of childhood cancer can go for follow-up until they reach their early 20s. Some cancer centers are now creating clinics dedicated to follow-up care for long-term survivors of pediatric and adolescent cancers.

*For full-length article and references, please see the online version of this article.*

**Source:** National Cancer Institute. ■

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