Georgia Childhood Cancer Report

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Georgia Department of Human Resources

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Dear Reader:

Although uncommon, childhood cancer deaths are among life's most depressing events. The "Georgia Childhood Cancer Report" provides a detailed picture of childhood cancer in Georgia, containing information on childhood cancer incidence, cancer deaths, risk factors and prevention. The report shows that every year more than 50 children die and that cancer is the third most common cause of death among Georgia children.

The "Georgia Childhood Cancer Report" represents a milestone for the Georgia Comprehensive Cancer Registry. It is the first time we have been able to report both childhood cancer incidence and mortality. The credit for the accomplishment goes to the many healthcare providers in Georgia who submit cancer data to the Georgia Comprehensive Cancer Registry. The Georgia Department of Human Resources, Division of Public Health, is dedicated to eliminating cancer as a major health problem. The data in this report will be used to guide statewide and local cancer control efforts; this will enable us to assure that every child with cancer in Georgia has access to the best treatment, improve the quality of information about childhood cancer and search for causes so that childhood cancer can be prevented.

Kathleen E. Toomey, M.D., M.P.H.
Director, Division of Public Health

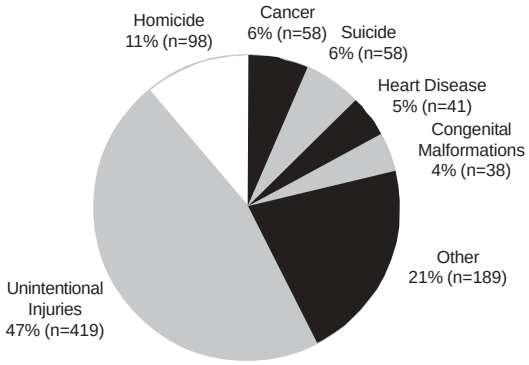
HIGHLIGHTS

- More than 50 children in Georgia die from cancer every year.
- Childhood cancer is the third most common cause of death among children 1-19 years of age.
- Leukemias, lymphomas, and cancers of the brain and central nervous system account for about half of all childhood cancers.
- The incidence of childhood cancers in Georgia has increased about 1.4% per year since 1975. The increase is presumed to be largely due to improved diagnostic methods and changes in disease coding practices.
- The death rate from childhood cancers in Georgia has decreased about 0.6% per year since 1979. The decrease is due to improved treatment.
- Because of advances in treatment, over 75% of children with cancer will survive 5 years or more.
- The cause or causes of almost all childhood cancers are unknown.

INTRODUCTION

Cancer is the third most common cause of death among children between 1 and 19 years of age, accounting for 6% of childhood mortality in Georgia in 1999 *(Figure 1)*. The most common forms of childhood cancer, leukemias, central nervous system (CNS) neoplasms, and lymphomas account for almost 50% of all childhood cancer diagnoses and over 60% of childhood cancer deaths *(Table 1)*.

Figure 1. Childhood Mortality by Cause, Ages 1 to 19 Years, Georgia, 1999 (n=901)



This report was written to assist health professionals, volunteers, and staff of cancer control organizations, community groups, and others who are working to reduce the burden of cancer throughout Georgia.

This report describes the burden of childhood cancer in Georgia and includes: 1) the number of cancer cases and incidence rates for children in selected areas of Georgia; and 2) the number of cancer deaths and mortality rates for Georgia children.

BASIC CANCER INFORMATION

What is Cancer?

Cancer is a group of diseases characterized by uncontrolled growth and spread of abnormal cells. If the growth is not controlled, it can result in death. Cancer is caused by both internal and external factors. Many cancers can be cured if detected and treated promptly.

Who Is at Risk of Developing Cancer?

Everyone, but children are less likely to develop cancer than adults. Since the risk of cancer increases as individuals age, most cases affect adults who are middle-aged or older. Nearly 72% of all cancers in Georgia are diagnosed at ages 55 and older and about 1% occurs among children 1 to 19 years of age. In the United States, males have a 1 in 2 lifetime risk of developing cancer, and for females the lifetime risk is 1 in 3. Lifetime risk refers to the probability that an individual, over the course of a lifetime, will develop cancer.

Table 1. Incidence vs Mortality for the Top Three Cancer Sites in Children Ages 0 to 19 Years in Georgia

	Incidence		Mortality	
	% of Cases	Cases*	% of Deaths	Deaths
Leukemia	22%	75	30%	16
CNS Cancer	15%	50	23%	12
Lymphoma	12%	40	8%	4

*Estimated cases per year in Georgia. Figures have been rounded to the nearest five.

CANCER IN CHILDREN

According to the American Cancer Society, about 12,400 children and adolescents in the United States were diagnosed with cancer in 2001. The types of cancers that occur in children differ from those most commonly seen in adults. Leukemias, brain and other nervous system tumors, and lymphomas are the most common cancers of children. Prostate, breast, lung, and colorectal cancers are the most common in adults. The stage of growth and development is another important difference between adults and children; the immaturity of children's organ systems often has important treatment implications.

Despite its rarity and the impressive strides in treatment and supportive care, cancer is still a leading cause of death in children between 1 and 19 years of age. Mortality rates have declined 50% since 1973. Because of significant advances in therapy, 75% of children diagnosed with cancer will survive 5 years or more, an increase of almost 40% since the early 1960's.

Many childhood cancers occur very early in life and many parents want to know why. The cause of most childhood cancers is not known. Radiation exposure contributes to a few types of childhood cancers. Some are the result of a familial predisposition. Unlike cancers of adults, childhood cancers are not significantly related to lifestyle-related risk factors of the patient such as tobacco or alcohol use, poor diet, or lack of physical activity.

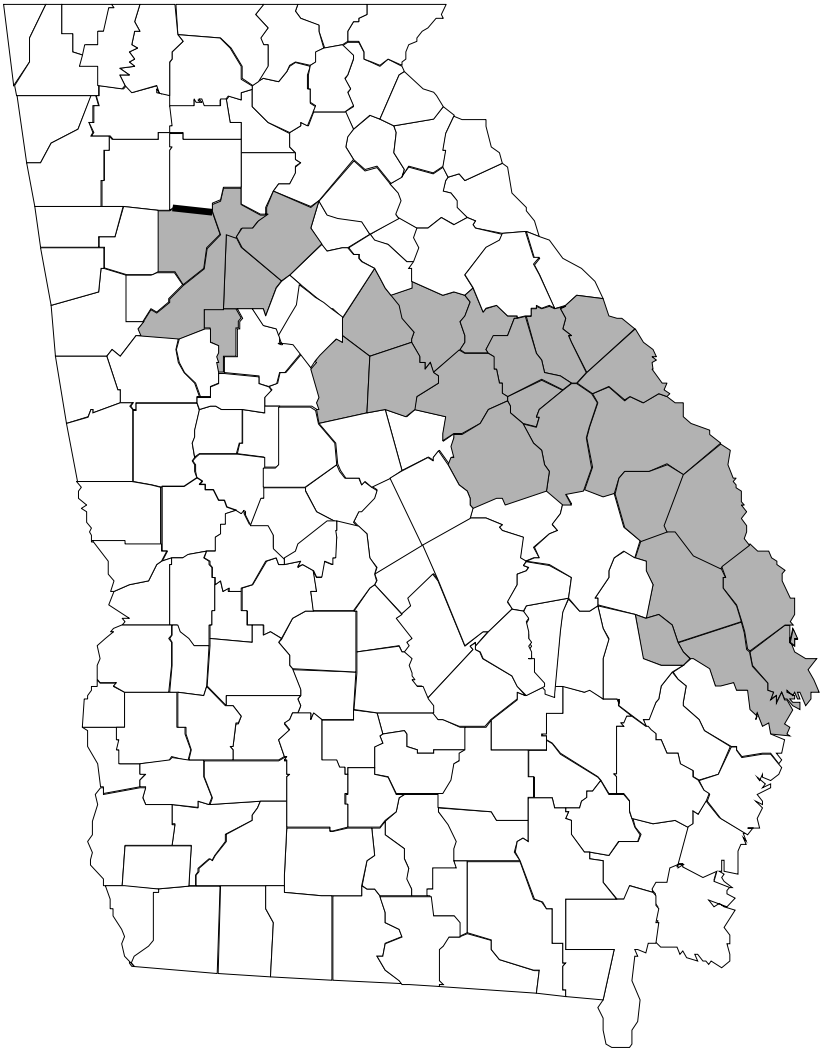
Childhood cancers can be treated with chemotherapy, surgery, radiation therapy, or by a combination of these therapies. Although there are exceptions, childhood cancers tend to respond well to chemotherapy because they grow fast. Most forms of chemotherapy specifically affect growing cells. Because treatment for some childhood cancers is more successful than for others, survival rates differ between cancer types.

	Children	Adults
Top 5 Cancers	Leukemias CNS Neoplasms Lymphomas Soft Tissue Sarcomas Sympathetic Nervous System Tumors	Prostate Female Breast Lung and Bronchus Colon and Rectum Lymphomas
Prevention	Since the cause of most childhood cancer is unknown, there is no known way to prevent it.	Nearly two-thirds of cancer deaths can be linked to modifiable risk factors such as tobacco use, diet, obesity, and lack of exercise. In addition, many skin cancers could be prevented by protection from the sun's rays.
Survival	Overall, 5-year relative survival rate for all cancers from 1992-1997 for children age 0 to 19 is 78%.	Overall, 5-year relative survival rate for all cancers from 1992-1997 for all ages is 62%.

COUNTIES INCLUDED IN THIS REPORT

For the purpose of this report, the counties with the most complete, long-term cancer incidence data were included in the incidence analysis. They include the five counties in Metro Atlanta and the ten rural counties served by the Surveillance, Epidemiology, and End Results (SEER) Program and the eleven counties originally part of the Savannah River Region Health Information System (SRRHIS) *(Figure 2)*. These counties represent 45% of the total population in Georgia. A description of the SEER and SRRHIS programs and a list of the included counties may be found in Appendix A of this report.

Figure 2. Georgia Counties Included in the Childhood Cancer Incidence Analysis



CHILDHOOD CANCER INCIDENCE

Trends in Childhood Cancer Incidence in Metro Atlanta, Georgia

Incidence rates for all childhood cancers combined increased steadily (estimated annual percent change = 1.4%) from 1975 to 1998 in Metro Atlanta (*Figure 3*). At least some of this increase is due to changes in diagnostic technology, reporting, and classification.

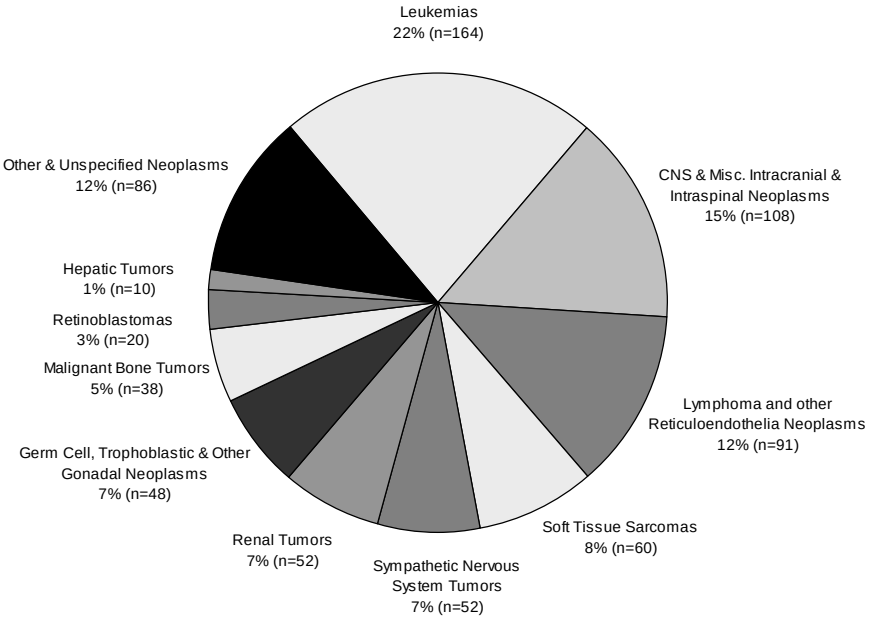
Figure 3. Childhood Cancer Incidence Rates*, Metro Atlanta, All Sites, Ages 0 to 19, 1975-1998

* Rate per 1,000,000, age-adjusted to the 1970 US standard population

Distribution of Childhood Cancer Incidence by Type in Selected Areas of Georgia

- Leukemia is the most common form of childhood cancer, accounting for 22% of all childhood cancer diagnoses between 1993 and 1997 in Georgia's SEER and SRRHIS counties (*Figure 4*).
- CNS tumors and lymphomas combined account for over one fourth of all childhood cancer diagnoses.

Figure 4. Childhood Cancer Incidence, Ages 0 to 19, SEER and SRRHIS, 1993-1997 (n=729)



Childhood Cancer Incidence Rates in Selected Areas of Georgia

- Between 1993 and 1997, there were 729 cancer diagnoses among children aged 0 to 19 years living in Georgia's SEER and SRRHIS counties. The age-adjusted incidence rate for this time period was 154.2 per 1,000,000 *(Table 2)*, slightly higher than the U.S. rate of 145.6 per 1,000,000 *(Appendix C)*.
- The highest cancer incidence rate (34.2 per 1,000,000) among children aged 0 to 19 years was for leukemia, followed by CNS neoplasms with a rate of 23.5 per 1,000,000.
- Overall, males had a higher cancer incidence rate than females. This difference was most apparent in leukemias, lymphomas, and soft tissue sarcomas.
- Cancer incidence was highest in the youngest and oldest age categories *(Table 3)*.
- Children aged 0 to 4 years were most likely to be diagnosed with leukemias, neuroblastomas, or renal tumors. Children aged 15 to 19 years were more likely to be diagnosed with lymphomas, especially Hodgkin lymphoma.

Table 2. Age-Adjusted Childhood (Ages 0 to 19) Cancer Incidence Rates, Georgia SEER and SRRHIS, 1993-1997

	Total		Male		Female	
Site	**Cases**	**Rate***	**Cases**	**Rate***	**Cases**	**Rate***
All Types	729	154.2	410	169.6	319	138.2
Leukemias	164	34.2	106	43.3	58	24.6
- Lymphocytic	124	25.7	86	35.0	38	16.0
- Acute non-lymphocytic	35	7.3	18	—	17	—
Lymphoma and other reticuloendothelial neoplasms	91	20.4	59	25.7	32	14.9
- Hodgkin lymphoma	51	11.6	30	13.2	21	9.9
- Non-Hodgkin lymphoma	30	6.7	20	8.8	10	—
CNS & Misc. Intracranial & Intraspinal Neoplasms	108	23.5	59	25.3	49	21.6
- Astrocytoma	65	14.3	37	16.1	28	12.5
- Medulloblastoma	13	—	7	—	6	—
Sympathetic Nervous System Tumors	52	9.8	27	10.1	25	9.6
- Neuroblastoma & ganglioneuroblastoma	47	8.8	23	8.4	24	9.2
Retinoblastomas	20	3.6	7	—	13	—
Renal Tumors	52	9.9	23	8.5	29	11.4
- Wilms tumor	45	8.6	20	7.3	25	10.0
Hepatic Tumors	10	—	7	—	<5	—
Malignant Bone Tumors	38	8.7	25	11.1	13	—
- Osteosarcoma	22	5.0	18	—	<5	—
- Ewing's sarcoma	12	—	5	—	7	—
Soft Tissue Sarcomas	60	12.7	37	15.3	23	9.9
- Rhabdomyosarcoma & embryonal sarcoma	38	7.9	23	9.4	15	—
Germ Cell, Trophoblastic & Other Gonadal Neoplasms	48	10.2	27	10.9	21	9.4
Other Epithelial & Unspecified Neoplasms	86	19.1	33	14.1	53	24.5

* Average annual rate per 1,000,000, age-adjusted to the 1970 US standard population.

Table 3. Age-Specific Childhood Cancer Incidence Rates, Georgia SEER and SRRHIS, 1993-1997

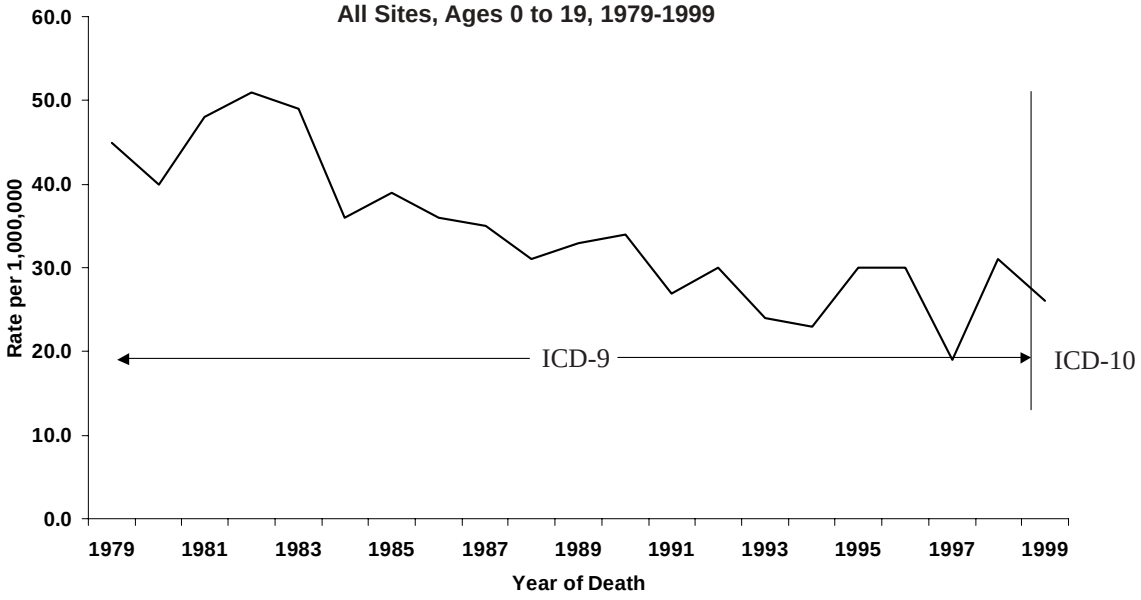
	Ages 0 to 4		Ages 5 to 9		Ages 10 to 14		Ages 15 to 19	
Site	**Cases**	**Rate***	**Cases**	**Rate***	**Cases**	**Rate***	**Cases**	**Rate***
All Types	235	187.9	134	114.1	139	123.8	221	198.8
Leukemias	63	50.4	46	39.2	32	28.5	23	20.7
Lymphoma and other reticuloendothelial neoplasms	6	—	13	—	22	19.6	50	45.0
CNS & Misc. Intracranial & Intraspinal Neoplasms	26	20.8	28	23.8	35	31.2	19	—
Sympathetic Nervous System Tumors	40	32.0	5	—	<5	—	<5	—
Retinoblastomas	18	—	<5	—	<5	—	<5	—
Renal Tumors	36	28.8	13	—	<5	—	<5	—
Hepatic Tumors	<5	—	<5	—	<5	—	<5	—
Malignant Bone Tumors	<5	—	<5	—	15	—	19	—
Soft Tissue Sarcomas	19	—	12	—	11	—	18	—
Germ Cell, Trophoblastic & Other Gonadal Neoplasms	13	—	7	—	<5	—	26	23.4
Other Epithelial & Unspecified Neoplasms	9	—	<5	—	17	—	58	52.2

* Average annual rate per 1,000,000.

Trends in Childhood Cancer Mortality in Georgia

Mortality rates for all childhood cancers combined decreased steadily (estimated annual percent change = -0.6%) from 1979 to 1999 in Georgia *(Figure 5)*. This decrease results from improvements in survival for most childhood cancers, especially leukemia and lymphoma. The availability of newer, more effective chemotherapy treatments is the principal cause of improved survival among childhood cancer patients.

Figure 5. Childhood Cancer Mortality Rates*, Georgia, All Sites, Ages 0 to 19, 1979-1999

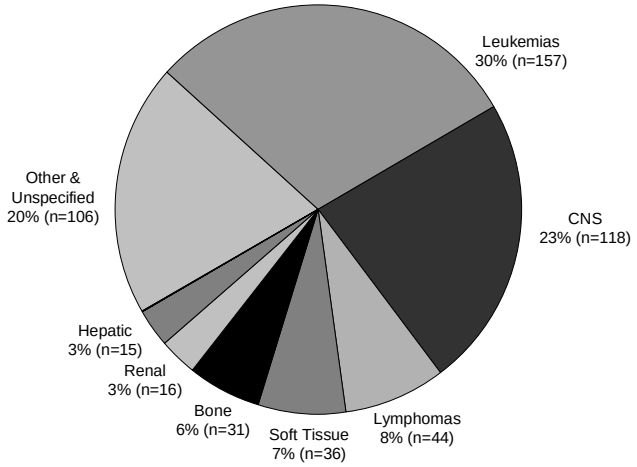


* Rate per 1,000,000, age-adjusted to the 1970 US standard population

Distribution of Childhood Cancer Mortality by Type in Georgia

- Leukemia is the most common cause of childhood cancer deaths, accounting for almost one third of all childhood cancer deaths in Georgia between 1990 and 1999 *(Figure 6)*.
- Central nervous system (CNS) neoplasms are second, responsible for about one fourth of all childhood cancer deaths.
- Lymphomas, soft tissue, bone, renal, and hepatic tumors account for another fourth of pediatric cancer deaths.

Figure 6. Childhood Cancer Mortality, Ages 0 to 19, Georgia, 1990-1999 (n=523)



Childhood Cancer Mortality Rates in Georgia

- Between 1990 and 1999, there were 523 cancer deaths among Georgia's children aged 0 to 19 years. The age-adjusted rate for this time period was 24.9 per 1,000,000 *(Table 4)*.
- The highest cancer mortality rate (7.5 per 1,000,000) among children aged 0 to 19 years was for leukemia, followed closely by CNS neoplasms with a rate of 5.6 per 1,000,000.
- Overall, males had a slightly higher cancer mortality rate than females. This difference was more apparent in the 15 to 19 year old age group.
- Cancer mortality increased with age, with the 15 to 19 year old age group having the highest number of cancer deaths among children for most cancer types *(Table 5)*.

Table 4. Age-Adjusted Childhood (Ages 0 to 19) Cancer Mortality Rates, Georgia, 1990-1999

	Total		Male		Female	
Site	**Deaths**	**Rate***	**Deaths**	**Rate***	**Deaths**	**Rate***
All Types	523	24.9	292	27.2	231	22.5
Leukemias	157	7.5	87	8.1	70	6.8
CNS	118	5.6	68	6.3	50	4.9
Lymphomas†	44	2.1	29	2.7	15	—
Soft Tissue, including heart	36	1.7	18	—	18	—
Renal Tumors	16	—	8	—	8	—
Bone Tumors	31	1.5	16	—	15	—
Hepatic Tumors	15	—	10	—	5	—
Other & Unspecified	106	5.0	56	5.3	50	4.8

* Average annual rate per 1,000,000, age-adjusted to the 1970 US standard population.
† Including Other Reticuloendothelial Neoplasms.

Table 5. Age-Specific Childhood Cancer Mortality Rates, Georgia, 1990-1999

	Ages 0 to 4		Ages 5 to 9		Ages 10 to 14		Ages 15 to 19	
Site	**Deaths**	**Rate***	**Deaths**	**Rate***	**Deaths**	**Rate***	**Deaths**	**Rate***
All Types	116	21.2	119	22.5	129	24.9	159	30.7
Leukemias	36	6.6	36	6.8	39	7.5	46	8.9
CNS	27	4.9	37	7.0	33	6.4	21	4.1
Lymphomas†	5	—	8	—	10	—	21	4.1
Soft Tissue, including heart	8	—	8	—	5	—	15	—
Renal Tumors	6	—	<5	—	<5	—	<5	—
Bone Tumors	<5	—	<5	—	11	—	19	—
Hepatic Tumors	9	—	<5	—	<5	—	5	—
Other & Unspecified	25	4.6	24	4.5	27	5.2	30	5.8

* Average annual rate per 1,000,000.
† Including Other Reticuloendothelial Neoplasms.

Childhood Leukemia

What is it?

Leukemia is a cancer of the white blood cells. This cancer starts in the bone marrow but can spread to the blood, lymph nodes, spleen, liver, central nervous system, or other organs.

In the United States, leukemia is the most common cancer in children and adolescents. It accounts for nearly one fourth of all cancers in children under age 19. Five-year survival rates have greatly increased over time, depending on type and other prognostic factors such as age, sex, race, and response to therapy.

Types of Childhood Leukemia

Leukemia is divided into two types: acute (rapidly growing) and chronic (slowly growing), with the majority of childhood leukemia being the acute form.

Acute lymphocytic leukemia (ALL) represents about 75% of all childhood leukemias. Five-year relative survival for children with ALL is about 80%.

Acute myelogenous leukemia (AML) represents most of the remaining leukemias in children. Five-year relative survival for children with AML is about 40%.

Risk Factors and Prevention

Most children with leukemia have no known risk factors and, at the present time, there is no way to prevent their leukemias from developing. However, doctors have found that this cancer is associated with a number of other conditions and risk factors.

Significant radiation exposure to the fetus within the first months of development may carry up to a 5-fold increased risk of developing ALL. Treatment of other cancers with radiation and chemotherapy and the use of immune-suppressing drugs are also causes of some leukemias.

Certain genetic diseases that cause children to be born with an abnormal or deficient immune system, such as Down's syndrome and Li-Fraumeni syndrome, increase the risk of developing leukemia.

Leukemia Incidence in Georgia Children

- Leukemia ranks first among cancer diagnoses for children ages 0 to 19 in Georgia SEER and SRRHIS counties, accounting for 22% of all childhood cancer morbidity.
- Between 1993 and 1997, there were 164 cases of leukemia in children. The age-adjusted rate was 34.2 per 1,000,000.
- Males were about twice as likely as females to be diagnosed with leukemia.
- The highest rates were among children under the age of five.

Leukemia Mortality in Georgia Children

- Leukemia ranks first among cancer deaths for children ages 0 to 19 in Georgia, accounting for 30% of all childhood cancer mortality.
- Between 1990 and 1999, there were 157 deaths from leukemia among children. The age-adjusted rate was 7.5 per 1,000,000.
- Males and females were equally affected.
- The highest rates were among children over the age of 15.

Central Nervous System Cancer in Children

What is it?

The central nervous system (CNS) includes the brain and spinal cord. CNS tumors and associated neoplasms are the second largest category of cancer in children, constituting about 20% of all childhood cancers under age 15. The highest incidence rates of CNS cancer occur among infants and children through age seven. Survival for children with CNS cancer is poorest among infants but improves with increasing age. Five-year relative survival rates have improved over time to 65%.

Types of Brain and Spinal Cord Cancer

Tumors of different parts of the CNS disrupt different functions and cause different symptoms. Also, tumors in different areas of the CNS may be treated differently and have a different prognosis. Most CNS cancers do not spread outside of the brain or spinal cord.

Astrocytomas account for about half of all childhood brain cancers. Many astrocytomas cannot be cured because they spread widely throughout the normal brain tissue. These astrocytomas are called infiltrating astrocytomas.

Oligodendrogliomas spread or infiltrate in a manner similar to astrocytomas and, in most cases, cannot be completely removed by surgery. A small number of these tumors, however, are associated with long-term survivals of 30 or 40 years.

Ependymomas usually do not spread or infiltrate into normal brain tissue. As a result, some, but not all, can be removed and cured by surgery.

Medulloblastomas represent approximately 25% of childhood brain cancers. They are fast-growing tumors but can be treated, and up to 50% of cases are cured by radiation therapy, sometimes with added chemotherapy. These tumors are common in children, but rare in adults.

Risk Factors and Prevention

The majority of CNS cancers are not associated with any known risk factors. As a result, most CNS cancers cannot be prevented

The only established environmental risk factor for brain tumors is ionizing radiation, usually as a result of treatment for other cancers.

Occasionally, cases of CNS cancers occur in families. Recently, researchers have found that some rare inherited syndromes (like tuberous sclerosis, neurofibromatosis, and Li-Fraumeni) are associated with an increased risk of developing some CNS cancers.

CNS Cancer Incidence in Georgia Children

- CNS cancer ranks second among cancer diagnoses for children ages 0 to 19 in Georgia SEER and SRRHIS counties, accounting for 15% of all childhood cancer morbidity.
- Between 1993 and 1997, there were 108 cases of CNS cancer in children. The age-adjusted rate was 23.5 per 1,000,000.
- Males and females were equally affected.
- The highest rates were among children between the ages of 10 and 14.

CNS Cancer Mortality in Georgia Children

- CNS cancer ranks second among cancer deaths for children ages 0 to 19 in Georgia, accounting for 23% of all childhood cancer mortality.
- Between 1990 and 1999, there were 118 deaths from CNS cancer among children. The age-adjusted rate was 5.6 per 1,000,000.
- Males and females were equally affected.
- The highest rates were among children between the ages of 5 and 9.

Lymphoma in Children

What is it?

Lymphoma is a cancer that starts in lymphoid tissue and is divided into two main types: Hodgkin lymphoma and non-Hodgkin lymphoma (NHL). Non-Hodgkin lymphoma differs significantly from Hodgkin lymphoma in behavior, pathology, spread, and responsiveness to treatment.

Hodgkin lymphoma is rare in children. The five-year relative survival rate is about 82%.

NHL represents about 4% of cancers in children. NHL occurs from infancy through adolescence, with a peak between the ages of 7 and 11 years. NHL is about three times more common in males than in females, and is about twice as common in white children as in black children. The five-year relative survival rate for children with early stage NHL is above 90%. The relative survival for children with more advanced stages is about 75%-85%.

Types of NHL Among Children

Lymphoblastic lymphoma (LBL) accounts for about 30% of NHL in children. LBL tends to spread very quickly to the bone marrow, other lymph nodes, the surface of the brain, and the membranes that surround the lungs and heart.

Small noncleaved lymphoma (SNCL) accounts for about 40% of childhood NHL. SNCL is one of the most rapidly growing cancers. It may spread to many organs including the surface of the brain or inside the brain.

Large cell non-Hodgkin lymphoma (LCL) represents about 30% of all NHL in children. Unlike lymphoblastic lymphoma and small noncleaved non-Hodgkin lymphoma, spread to bone marrow and the brain is not common.

Risk Factors and Prevention

Scientists have found few risk factors that are associated with Hodgkin lymphoma. Since most cases are not associated with any known risk factors, it is impossible to prevent.

Epstein-Barr virus (EBV) infection is associated with a slightly increased rate of Hodgkin lymphoma, but its role is unclear. Immunodeficiency has also been associated with a slightly increased risk.

The exact cause of NHL is not known. Scientists have found that this cancer is associated with a number of other conditions. However, most children with NHL do not have any known risk factors.

It has been suggested that an altered immune state provides the necessary susceptibility for NHL. Organ transplant, HIV infection, and congenital immunodeficiency syndromes have all been linked to an increased risk for developing NHL. The combination of immune deficiencies and EBV infection can also cause NHL to develop.

Lymphoma Incidence in Georgia Children

- Lymphoma ranks third among cancer diagnoses for children ages 0 to 19 in Georgia SEER and SRRHIS counties, accounting for 12% of all childhood cancer morbidity.
- Between 1993 and 1997, there were 91 cases of lymphoma in children. The age-adjusted rate was 20.4 per 1,000,000.
- Males were about twice as likely as females to be diagnosed with lymphoma.
- The highest rates were among children over the age of 15.

Lymphoma Mortality in Georgia Children

- Lymphoma ranks third among cancer deaths for children ages 0 to 19 in Georgia, accounting for 8% of all childhood cancer mortality.
- Between 1990 and 1999, there were 44 deaths from lymphoma among children. The age-adjusted rate was 2.1 per 1,000,000.
- Males were about twice as likely as females to die of lymphoma.
- The highest rates were among children over the age of 15.

Soft Tissue Sarcoma

What is it?

Rhabdomyosarcoma is a cancer of the skeletal muscles of the body. It is the most common type of soft tissue sarcoma in children, accounting for 3.4% of childhood cancers. Over 85% of rhabdomyosarcomas are diagnosed in infants, children, and teenagers.

The exact prognosis for each child with rhabdomyosarcoma depends on many factors, including the location of the tumor, type of tumor, and whether it has metastasized. Overall, two-thirds of children will survive with intensive treatment.

Types of Childhood Rhabdomyosarcoma

Embryonal rhabdomyosarcoma is the most common type in children. It tends to occur in the head and neck area, bladder, vagina, and in or around the prostate and testes and usually affects infants and young children.

Alveolar rhabdomyosarcoma occurs more often in large muscles of the trunk, arms, and legs, and typically affects older children and teenagers. This type is called alveolar because the malignant cells form little hollow spaces, or alveoli, in the affected tissue.

Pleomorphic rhabdomyosarcoma occurs mainly in adults 30 to 50 years of age, practically never occurring in children.

Risk Factors and Prevention

Rhabdomyosarcoma is unlike most adult cancers in that there are no known environmental or lifestyle risk factors associated with it. However, there is evidence of an increased risk for rhabdomyosarcoma from genetic disorders including Li-Fraumeni syndrome, Beckwith-Wiedemann syndrome, and neurofibromatosis.

Because no avoidable or modifiable risk factors have yet been found, there is no known way to prevent rhabdomyosarcoma.

Soft Tissue Sarcoma Incidence in Georgia Children

- Soft tissue sarcoma ranks fourth among cancer diagnoses for children ages 0 to 19 in Georgia SEER and SRRHIS counties, accounting for 8% of all childhood cancer morbidity.
- Between 1993 and 1997, there were 60 cases of soft tissue sarcoma in children. The age-adjusted rate was 12.7 per 1,000,000.
- Males were about one and a half times as likely as females to be diagnosed with soft tissue sarcoma.
- All ages were equally affected.

Soft Tissue (Including Heart) Cancer Mortality in Georgia Children

- Soft tissue cancer ranks fourth among cancer deaths for children ages 0 to 19 in Georgia, accounting for 7% of all childhood cancer mortality.
- Between 1990 and 1999, there were 36 deaths from soft tissue cancer among children. The age-adjusted rate was 1.7 per 1,000,000.
- Males and females were equally affected.
- The highest rates were among children over the age of 15.

Neuroblastoma

What is it?

Neuroblastoma is a form of cancer that occurs in infants and children, and very rarely, adults. The cells of this cancer usually resemble very primitive, developing embryonic or fetal nerve cells called neuroblasts. About one-third of neuroblastomas start in the adrenal glands, another third begin in the sympathetic nervous system ganglia of the abdomen, and most others start in the sympathetic ganglia of the chest or neck or the parasympathetic ganglia in the pelvis.

Overall, about 7 to 10% of all childhood cancers are neuroblastomas. In infants, neuroblastoma accounts for 50% of cancers. Nearly 90% of cases are diagnosed by age six. When detected early, neuroblastomas can be treated effectively. However, in as many as 70% of cases, the disease is not diagnosed until it has already metastasized. Overall, about 40% of children with neuroblastomas can be cured. Sometimes the tumor regresses with little or no treatment or matures into a benign tumor.

Ganglioneuromas and Ganglioneuroblastomas

Ganglioneuromas are benign tumors composed of mature ganglion and nerve sheaths that do not continue to grow. They are usually removed by surgery and carefully examined to be certain they do not have areas of malignancy.

Ganglioneuroblastomas are malignant tumors that contain immature neuroblasts that can grow and spread abnormally, as well as areas of tissue that have matured and are similar to ganglioneuroma.

Risk Factors and Prevention

Neuroblastomas have not been associated with any environmental or lifestyle risk factors. There is evidence suggesting that neuroblastoma may be inherited. Familial cases differ from sporadic cases in age of onset and patterns of spread.

Since there are no avoidable risk factors for neuroblastoma, there is no known way to prevent the disease. If there is a family history of neuroblastoma, genetic counseling may be considered.

Sympathetic Nervous System Cancer Incidence in Georgia Children

- Sympathetic nervous system cancer ranks fifth among cancer diagnoses for children ages 0 to 19 in Georgia SEER and SRRHIS counties, accounting for 7% of all childhood cancer morbidity.
- Between 1993 and 1997, there were 52 cases of sympathetic nervous system cancer in children. The age-adjusted rate was 9.8 per 1,000,000.
- Males and females were equally affected.
- The highest rates were among children under the age of five.

Sympathetic Nervous System Cancer Mortality in Georgia Children

- Due to ICD coding limitations, we were unable to produce mortality rates for sympathetic nervous system cancer.

Wilms Tumor

What is it?

Wilms tumor (also called nephroblastoma) is the most common type of kidney cancer affecting children. About 95% of kidney cancers in children are Wilms tumors. About six percent of all cancers in children are Wilms tumors, with most cases occurring in the first five years of life. The overall five-year relative survival rate for children with Wilms tumor is about 92%.

Types of Wilms Tumor

Wilms tumors are classified into two major types depending on how they look under the microscope. These two categories differ in prognosis and treatment.

Wilms tumors of unfavorable histology contain anaplasia (the presence of large, irregular nuclei in the tumor's cells). The prognosis tends to be worse than that of Wilms tumors without anaplasia. Also, tumors with diffuse anaplasia have a worse prognosis than tumors with focal anaplasia.

Wilms tumors of favorable histology do not contain anaplasia. Patients with these tumors usually have a much better outlook for cure. About 95% of Wilms tumors have favorable histology.

Risk Factors and Prevention

Scientific research has not shown a connection between Wilms tumor and environmental risk factors either before or after a child's birth.

Some genetic and hereditary risk factors have been associated with Wilms tumor. Such risk factors include a family history of Wilms tumor, mental retardation (WAGR) syndrome, Beckwith-Wiedemann syndrome, and Denys-Drash syndrome.

While there is a clear connection between Wilms tumors, certain birth defect syndromes, and genetic deletions or mutations, most children with this type of cancer do not have any known birth defects or inherited gene changes. Not enough is known about the causes of Wilms tumor to say how it can be prevented.

Renal Cancer Incidence in Georgia Children

- Renal cancer ranks sixth among cancer diagnoses for children ages 0 to 19 in Georgia SEER and SRRHIS counties, accounting for 7% of all childhood cancer morbidity.
- Between 1993 and 1997, there were 52 cases of renal cancer in children. The age-adjusted rate was 9.9 per 1,000,000.
- Males and females were equally affected.
- The highest rates were among children under the age of five.

Renal Cancer Mortality in Georgia Children

- Renal cancer ranks sixth among cancer deaths for children ages 0 to 19 in Georgia, accounting for 3% of all childhood cancer mortality.
- Between 1990 and 1999, there were 16 deaths from renal cancer among children.
- Males and females were equally affected.
- The highest rates were among children under the age of five.

Bone Cancer in Children

What is it?

Osteosarcoma is the most common type of childhood bone cancer. It represents about 2.4% of all childhood cancers. It is almost twice as common in males as in females, and adolescents are the most commonly affected age group. About 80% of childhood osteosarcomas develop at the ends of the long bones that form the knee. However, osteosarcoma can develop in any bone of the body. Five-year relative survival rates vary from 20 to 85% depending on tumor grade, metastasis, and response to treatment.

Ewing's family of tumors (EFT) accounts for about 5% of childhood bone tumors. EFT can occur at any age but is most common in early teenage years. About 64% of all cases occur in adolescents between 10 and 19 years. It occurs more often among males, and most patients are white. Unlike osteosarcoma, EFT usually occurs in the middle of the long bones of the legs or arms. It can also occur in the pelvic bones or ribs. EFT can occur inside or outside of the bone and spread to nearby tissues or throughout the body. Overall, five-year survival is about 66%.

Types of Bone Cancer in Children

Based on their appearance under a microscope, osteosarcomas can be classified as low, intermediate, or high grade. The grade largely determines prognosis and treatment strategy.

EFT is described by origin, size, and metastasis. Prognosis depends on these three factors, whether the tumor can be removed by surgery, and response to therapy.

Risk Factors and Prevention

Most patients with osteosarcoma do not have any known risk factors and the causes of their cancers remain unknown at this time. The risk of osteosarcoma is highest during the teenage "growth spurt," suggesting a link between rapid bone growth and tumor formation. Patients who were treated with radiation for another cancer, especially at a young age, are at increased risk of developing osteosarcoma. Certain benign bone diseases, such as Paget disease of the bone and multiple hereditary osteochondromas, increase the risk of developing osteosarcoma. Also, at an increased risk are children with Li-Fraumeni syndrome or retinoblastoma.

Age, sex, and race are the only risk factors found to be related to the risk of developing EFT. There has been no link between EFT and environmental, lifestyle, or genetic risk factors. There is currently no way to prevent EFT in children.

Bone Cancer Incidence in Georgia Children

- Bone cancer ranks eighth among cancer diagnoses for children ages 0 to 19 in Georgia SEER and SRRHIS counties, accounting for 5% of all childhood cancer morbidity.
- Between 1993 and 1997, there were 38 cases of bone cancer in children. The age-adjusted rate was 8.7 per 1,000,000.
- Males were about twice as likely as females to be diagnosed with bone cancer.
- The highest rates were among children over the age of ten.

Bone Cancer Mortality in Georgia Children

- Bone cancer ranks fifth among cancer deaths for children ages 0 to 19 in Georgia, accounting for 6% of all childhood cancer mortality.
- Between 1990 and 1999, there were 31 deaths from bone cancer among children. The age-adjusted rate was 1.5 per 1,000,000.
- Males and females were equally affected.
- The highest rates were among children over the age of ten.

CONCLUSIONS

Childhood cancers are uncommon but they remain an important public health issue. The information in this report summarizes current incidence and mortality rates for childhood cancers in Georgia. It also provides short summaries about what is known about the more common types of childhood cancers. The information will be useful to many individuals concerned about cancer such as public health workers, health care providers, volunteer workers and groups, and families and friends of children with cancer. The report also calls attention to needed actions.

To reduce the incidence and mortality from childhood cancers, several steps are being taken:

Search for causes so that childhood cancer can be prevented. The cause or causes of most childhood cancers are unknown, making preventive actions impossible. The search for causes is difficult because the cancers are rare. Efforts are underway to pool information from states with high quality cancer registries for the purposes of research. Georgia will participate in these group research projects.

Assure that every child with cancer in Georgia has access to the best treatment. In recent decades, the treatment of childhood cancers has improved markedly. Presently more than 75% of children with cancer survive 5 years or more. National estimates suggest that about 20% of children with cancer may not receive the most appropriate treatment.

Improve the quality of information about childhood cancer in Georgia. Presently, accurate and reliable information about childhood cancer in Georgia is available for only the 26 counties included in this report. The quality of information reported by hospitals and other cancer care providers to the Georgia Comprehensive Cancer Registry varies across the state but is improving. Efforts to achieve accurate and reliable information from the entire state are in progress.

Thus, by participating in the search for causes, by assuring access to appropriate treatment, and by improving the quality of information contained in the Georgia Comprehensive Cancer Registry, Georgians will be participating in efforts to reduce the burden of childhood cancers, a group of uncommon but devastating illnesses.

METHODOLOGY

Data Sources

The number of deaths for the state of Georgia for 1990-1999 was obtained from the Georgia Department of Human Resources, Division of Public Health, Vital Records Branch. The population counts used to calculate the 1990-1999 death rates were obtained from the United States (U.S.) Census Bureau. The number of deaths and death rates used in the trend analysis for the state of Georgia 1979-1998 were obtained from CDC Wonder, a web-based system that provides a single point of access to a wide variety of Centers for Disease Control and Prevention (CDC) reports, guidelines, and numeric public health data.

The number of cases for Georgia SEER and SRRHIS counties for 1993-1997 were obtained from the Georgia Department of Human Resources, Division of Public Health, Georgia Comprehensive Cancer Registry. The population counts used to calculate the 1993-1997 incidence rates were obtained from the U.S. Census Bureau. The number of cases and incidence rates used in the trend analysis for SEER Metro Atlanta 1975-1998 were obtained from SEER*Stat version 4.0, a statistical system for the analysis of SEER databases.

Estimation of Mortality and Incidence Rates

Incidence counts and rates for 1993-1997 were generated using SAS, a computer based statistical analysis system. Rates for International Classification of Childhood Cancers (ICCC) groupings by sex and age were estimated using data for the SEER Metro Atlanta, SEER Rural Georgia, and SRRHIS counties. Age-adjusted rates based on five-year age groupings were standardized to the 1970 U.S. standard population.

Incidence trends for 1975-1998 were generated using SEER*Stat version 4.0. Rates were estimated using data from the Metro Atlanta SEER Registry. Rates were age-adjusted to the 1970 U.S. standard population.

Mortality counts and rates for 1990-1999 were generated using SAS. Rates for selected groupings by sex and age were estimated using data for the entire state of Georgia. Age-adjusted rates based on five-year age groupings were standardized to the 1970 U.S. standard population.

Mortality trends for 1979-1998 were generated using CDC Wonder. Rates were age-adjusted to the 1970 US standard population.

Rates were not calculated where the count (cases or deaths) was less than twenty.

The ICD codes used for disease categories may be found in Appendix C.

REFERENCES

1. U.S. Bureau of the Census, CO-99-12 Population Estimates for Counties by Age, Race, Sex and Hispanic Origin, Annual Time Series: July 1, 1990 to July 1, 1999
2. American Cancer Society. Cancer Facts and Figures 2000.
3. American Cancer Society. http://www.cancer.org.
4. DeVita VT, Hellman S, Rosenberg SA, Cancer Principles and Practice of Oncology, Volume 2, 5th Edition, Philadelphia, PA: Lippincott-Raven Publishers; 1997.

APPENDIX A

The Georgia Comprehensive Cancer Registry

The Georgia Comprehensive Cancer Registry (GCCR) was established to be a tool for Georgia health professionals at the state and local level to better understand the state's cancer burden. The central cancer registry collects statewide data on new cases of cancer and these data are used to estimate cancer incidence rates within Georgia, monitor cancer trends, evaluate possible clusters of cancer, respond to inquiries about cancer from the public, and conduct research. Data from the central cancer registry also assist state and local agencies in focusing their early detection and risk reduction programs. The state's goal is to produce cancer incidence and mortality reports regularly for use by the public and health professionals.

Surveillance, Epidemiology, and End Results Program

In order to accurately understand the burden of cancer in Georgia, it is important to have at least 90% of the newly diagnosed cancer cases reported to GCCR every year. For 1995, the first year of mandatory reporting by all health care providers, reporting is estimated to be 94% complete; for 1996 and 1997 it is 78% complete; for 1998 it is 84% complete; for 1999 it is 81% complete; and for 2000 it is 56% complete. The primary reason for incomplete data is that there are still hospitals, laboratories, and outpatient treatment and diagnostic facilities that are not reporting their cancer data to the central cancer registry. The State's goal is to have all health care providers reporting data that are timely, complete, and accurate to the GCCR.

The Surveillance, Epidemiology, and End Results (SEER) Program of the National Cancer Institute is the most authoritative source of information on cancer incidence and survival in the United States. Data collection for SEER began on January 1, 1973. In 1975, five counties in metropolitan Atlanta were added, and in 1978, ten predominantly black rural counties in Georgia were added.

Geographic areas were selected for inclusion in the SEER program based on their ability to operate and maintain a high quality population-based cancer reporting system and for their epidemiologically significant population subgroups. The population covered by SEER is comparable to the general U.S. population with regard to measures of poverty and education. The SEER population tends to be somewhat more urban and has a higher proportion of foreign-born persons than the general U.S. population.

The SEER program currently collects and publishes cancer incidence and survival data from 11 population-based cancer registries and 3 supplemental registries covering approximately 14 percent of the U.S. population. The SEER Program is the only comprehensive source of population-based information in the United States that includes stage of cancer at the time of diagnosis and survival rates within each stage.

The SEER Program is considered to be the standard for quality among cancer registries around the world.

APPENDIX A(CONTINUED)

Savannah River Region Health Information System

The Savannah River Region Health Information System (SRRHIS) was a joint program of the Medical University of South Carolina and Emory University. Spurred by public concern about the risk for cancer from living near or downstream from the Savannah River Site (a nuclear materials production site), the two universities obtained support for a population-based registry, to be used to evaluate risk and to inform the community in a timely and understandable fashion. Cancer data collection began as of January 1, 1991 and continued through the end of 1995. The Georgia Comprehensive Cancer Registry continued data collection efforts through the end of 1997.

To this date, an elevated risk for cancer among residents of the twelve counties has not been found.

Counties Included in the Childhood Cancer Incidence Analysis

SEER Metro Atlanta	Clayton, Cobb, DeKalb, Fulton, and Gwinnett
SEER Rural Georgia	Glascock, Greene, Hancock, Jasper, Jefferson, Morgan, Putnam, Taliaferro, Warren, and Washington
SRRHIS	Bryan, Bulloch, Burke, Chatham, Columbia, Effingham, Evans, Jefferson, Jenkins, McDuffie, Richmond, and Screven

APPENDIX B

Age-Adjusted Childhood (Ages 0 to 19) Cancer Incidence Rates

United States vs Georgia SEER and SRRHIS, 1993-1997

	United States		Georgia	
Site	Cases	Rate	Cases	Rate
All Types	27422	145.6	729	154.2
Leukemias	7357	38.0	164	34.2
- Lymphocytic	5692	29.2	124	25.7
- Acute non-lymphocytic	1267	6.7	35	7.3
Lymphoma and other reticuloendothelial neoplasms	3924	21.9	91	20.4
- Hodgkin disease	2040	11.6	51	11.6
- Non-Hodgkin lymphoma	1282	7.1	30	6.7
CNS & Misc. Intracranial & Intraspinal Neoplasms	4817	25.6	108	23.5
- Astrocytoma	2396	12.9	65	14.3
- Medulloblastoma	1133	5.9	13	—
Sympathetic Nervous System Tumors	1444	6.9	52	9.8
- Neuroblastoma & ganglioneuroblastoma	1382	6.5	47	8.8
Retinoblastomas	604	2.8	20	3.6
Renal Tumors	1217	5.9	52	9.9
- Wilms tumor	1133	5.5	45	8.6
Hepatic Tumors	291	1.4	10	—
Malignant Bone Tumors	1464	8.3	38	8.7
- Osteosarcoma	803	4.6	22	5.0
- Ewing's sarcoma	488	2.8	12	—
Soft Tissue Sarcomas	1942	10.5	60	12.7
- Rhabdomyosarcoma & embryonal sarcoma	859	4.5	38	7.9
Germ Cell, Trophoblastic & Other Gonadal Neoplasms	1931	10.6	48	10.2
Other Epithelial & Unspecified Neoplasms	2431	13.7	86	19.1

* Average annual rate per 1,000,000, age-adjusted to the 1970 US standard population.

APPENDIX C

ICD-9 Codes for Childhood Cancer Mortality (1990-1998)*

All Sites	140-208
Leukemias	202.4, 203.1, 204-208
CNS	191-192
Lymphomas & Other Reticuloendothelial Neoplasms	200.0-202.3, 202.5-203.0, 203.2-203.8
Soft Tissue, including heart	171, 164.1
Renal Tumors	189.0, 189.1
Bone Tumors	170
Hepatic Tumors	155.0-155.2
Other & Unspecified	179, 185, 193, 140.0-154.8, 156.0-164.0 164.2-165.9, 172.0-188.9, 189.2-190.9, 194.0-199.1

ICD-10 Codes for Childhood Cancer Mortality (1999)*

All Sites	C00-C97
Leukemias	C90.1, C91-C95
CNS	C70-C72
Lymphomas & Other Reticuloendothelial Neoplasms	C81.0-C90.0, C90.2, C96
Soft Tissue, including heart	C38.0, C45.2, C46.1, C47, C49
Renal Tumors	C64-C65
Bone Tumors	C40-C41
Hepatic Tumors	C22
Other & Unspecified	C00-C21, C23-C37, C38.1-C39.9, C43.0-C45.1, C45.7-C46.0, C46.2-C46.9, C48, C50-C63, C66-C69, C73-C80, C97

ICD-O-2 Codes for Childhood Cancer Incidence

The International Classification of Childhood Cancer (ICCC) groupings were used.
They can be found in NAACCR's Cancer in North America 1993-1997, Vol 1: Incidence, Appendix B.

* The ICD10-ICD9 comparability ratio for malignant neoplasms is 1.0068. No adjustments were made to the data to take this into account.

Georgia Childhood Cancer Report 2002