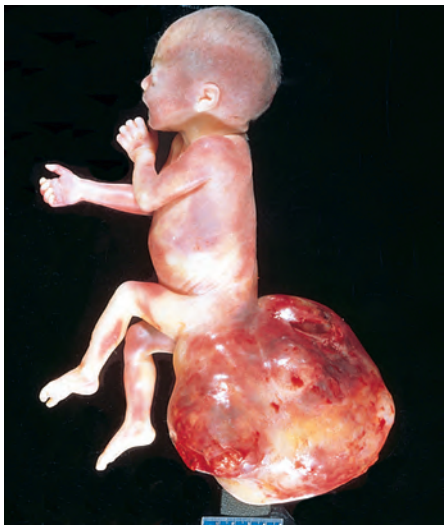

Figure 10-23 Sacrococcygeal teratoma. Note the size of the lesion compared with that of the stillbirth.

Approximately 75% of these tumors are mature teratomas, and about 12% are unequivocally malignant and lethal. The remainder is immature teratomas; their malignant potential correlates with the amount of immature tissue, usually immature neuroepithelial elements, that are present. Most of the benign teratomas are encountered in younger infants (<4 months), whereas children with malignant lesions tend to be somewhat older. Other sites for teratomas in childhood include the testis (Chapter 21), ovaries (Chapter 22), and various midline locations, such as the mediastinum, retroperitoneum, and head and neck.

Malignant Tumors

Cancers of infancy and childhood differ biologically and histologically from their counterparts occurring later in life. The main differences, some of which have already been alluded to, include the following:

- Incidence and type of tumor
- Relatively frequent demonstration of a close relationship between abnormal development (teratogenesis) and tumor induction (oncogenesis)
- Prevalence of underlying familial or genetic aberrations spontaneously
- Tendency of fetal and neonatal malignancies to regress or differentiate spontaneously
- Improved survival or cure of many childhood tumors, so that more attention is now being devoted to minimizing the adverse delayed effects of chemotherapy and radiation therapy in survivors, including the development of second malignancies

Incidence and Types. The most frequent childhood cancers arise in the hematopoietic system, nervous tissue (including the central and sympathetic nervous system, adrenal medulla, and retina), soft tissues, bone, and kidney. This is in sharp contrast to adults, in whom the skin, lung, breast, prostate, and colon are the most common sites of tumors.

Neoplasms that exhibit sharp peaks in incidence in children younger than age 10 years include (1) leukemia (principally acute lymphoblastic leukemia), (2) neuroblastoma, (3) Wilms tumor, (4) hepatoblastoma, (5) retinoblastoma, (6) rhabdomyosarcoma, (7) teratoma, (8) Ewing sarcoma, and posterior fossa neoplasms—principally (9) juvenile astrocytoma, (10) medulloblastoma, and (11) ependymoma. Other forms of cancer are also common in childhood but do not have the same striking early peak. The approximate age distribution of these cancers is indicated in Table 10-7. Within this large array, leukemia alone accounts for more deaths in children younger than age 15 years than all of the other tumors combined.

Histologically, many of the malignant non-hematopoietic pediatric neoplasms are unique. In general, they tend to have a more primitive *(embryonal)* undifferentiated appearance, are often characterized by sheets of cells with small, round nuclei, and frequently show features of organogenesis specific to the site of tumor origin. Because of this latter characteristic, these tumors are frequently designated by the suffix *-blastoma*, for example, nephroblastoma (Wilms tumor), hepatoblastoma, and neuroblastoma. Because of their primitive histologic appearance, many childhood tumors have been collectively referred to as *small round blue cell tumors.* The differential diagnosis of such tumors includes neuroblastoma, Wilms tumor, lymphoma (Chapter 13), rhabdomyosarcoma (Chapter 27), Ewing sarcoma/primitive neuroectodermal tumor (Chapter 26), medulloblastoma (Chapter 28), and retinoblastoma (Chapter 29). If the anatomic site of origin is known, diagnosis is usually possible on histologic grounds alone. Occasionally, a combination of chromosome analysis, immunoperoxidase stains, or electron microscopy is required. Two of these tumors are particularly illustrative and are discussed here: the neuroblastic tumors, specifically neuroblastoma, and Wilms tumor. The remaining tumors are discussed in their respective organ-specific chapters.

Neuroblastic Tumors

The term neuroblastic tumor includes tumors of the sympathetic ganglia and adrenal medulla that are derived from primordial neural crest cells populating these sites. As a family, neuroblastic tumors demonstrate certain characteristic features including *spontaneous or therapy-induced*

Table 10-7 Common Malignant Neoplasms of Infancy and Childhood

0 to 4 Years	5 to 9 Years	10 to 14 Years
Leukemia	Leukemia	
Retinoblastoma	Retinoblastoma	
Neuroblastoma	Neuroblastoma	
Wilms tumor		
Hepatoblastoma	Hepatocellular carcinoma	Hepatocellular carcinoma
Soft tissue sarcoma (especially rhabdomyosarcoma)	Soft tissue sarcoma	Soft-tissue sarcoma
Teratomas		
Central nervous system tumors	Central nervous system tumors	Osteogenic sarcoma
	Ewing sarcoma	Thyroid carcinoma
	Lymphoma	Hodgkin disease