Brain and Spinal Cord Tumors in Children

What is cancer?

The body is made up of hundreds of millions of living cells. Normal body cells grow, divide, and die in an orderly fashion. During the early years of a person's life, normal cells divide faster to allow the person to grow. After the person becomes an adult, most cells divide only to replace worn-out or dying cells or to repair injuries.

Cancer begins when cells in a part of the body start to grow out of control. There are many kinds of cancer, but they all start because of out-of-control growth of abnormal cells.

Cancer cell growth is different from normal cell growth. Instead of dying, cancer cells continue to grow and form new, abnormal cells. Cancer cells can also invade (grow into) other tissues, something that normal cells cannot do. Growing out of control and invading other tissues are what makes a cell a cancer cell.

Cells become cancer cells because of damage to DNA. DNA is in every cell and directs all its actions. In a normal cell, when DNA gets damaged the cell either repairs the damage or the cell dies. In cancer cells, the damaged DNA is not repaired, but the cell doesn’t die like it should. Instead, this cell goes on making new cells that the body does not need. These new cells will all have the same damaged DNA as the first cell does.

People can inherit damaged DNA, but most DNA damage is caused by mistakes that happen while the normal cell is reproducing or by something in our environment. Sometimes the cause of the DNA damage is something obvious, like cigarette smoking. But often no clear cause is found.

In most cases the cancer cells form a tumor. Some cancers, like leukemia, rarely form tumors. Instead, these cancer cells involve the blood and blood-forming organs and circulate through other tissues where they grow.

Cancer cells often travel to other parts of the body, where they begin to grow and form new tumors that replace normal tissue. This process is called *metastasis*. It happens when the cancer cells get into the bloodstream or lymph vessels of our body.

No matter where a cancer may spread, it is always named for the place where it started. For example, breast cancer that has spread to the liver is still called breast cancer, not liver cancer. Likewise, prostate cancer that has spread to the bone is metastatic prostate cancer, not bone cancer.

Different types of cancer can behave very differently. For example, lung cancer and breast cancer are very different diseases. They grow at different rates and respond to different treatments. That is why people with cancer need treatment that is aimed at their particular kind of cancer.

Not all tumors are cancerous. Tumors that aren’t cancer are called *benign*. Benign tumors can cause problems – they can grow very large and press on healthy organs and tissues. But they cannot grow into (invade) other tissues. Because they can’t invade, they also can’t spread to other parts of the body (metastasize). These tumors are almost never life threatening.

What are the differences between cancers in adults and children?

The types of cancers that develop in children are different from the types that develop in adults. Although there are exceptions, childhood cancers tend to respond better to chemotherapy. Children's bodies also tend to tolerate chemotherapy better than adults' bodies do. But because chemotherapy can have some long-term side effects, children who survive their cancer need careful attention for the rest of their lives.

Since the 1960s, most children and adolescents with cancer have been treated at specialized centers designed for them. Being treated in these centers offers them the advantage of a team of specialists who know the differences between adult and childhood cancers, as well as the unique needs of children with cancers. This team usually includes pediatric oncologists, pathologists, surgeons, radiation oncologists, pediatric oncology nurses, and nurse practitioners.

These centers also have psychologists, social workers, child life specialists, nutritionists, rehabilitation and physical therapists, and educators who can support and educate the entire family.

Most children with cancer in the United States are treated at a center that is a member of the Children's Oncology Group (COG). All of these centers are associated with a university or children's hospital. As we have learned more about treating childhood cancer, it has become even more important that treatment be given by experienced experts.

What are brain and spinal cord tumors in children?

Brain tumors are masses of abnormal cells that have grown out of control. In most other parts of the body, it is very important to distinguish between benign (non-cancerous) tumors and malignant (cancerous) ones. Benign tumors in other parts of the body are almost never life- threatening. The main reason cancers are so dangerous is because they can spread throughout the body and interrupt the way normal organs function.

Most brain cancers can spread through the brain tissue, but they rarely spread to other parts of the body. Even so-called benign tumors can, as they grow, press on or invade normal brain tissues, causing damage that is often disabling and can sometimes cause death. This is why doctors usually speak of "brain tumors" rather than "brain cancers." The major differences between benign and malignant brain tumors are how readily they spread through the rest of the central nervous system and whether they can be removed and not come back. But both types can potentially be life threatening.

Brain and spinal cord tumors are different in adults and children. They often form in different places, develop from different cell types, and may have a different treatment and prognosis (outlook). **This document refers only to children's tumors. Brain and spinal cord tumors in adults are discussed in our separate document*, Brain and Spinal Cord Tumors in Adults*.**

The central nervous system

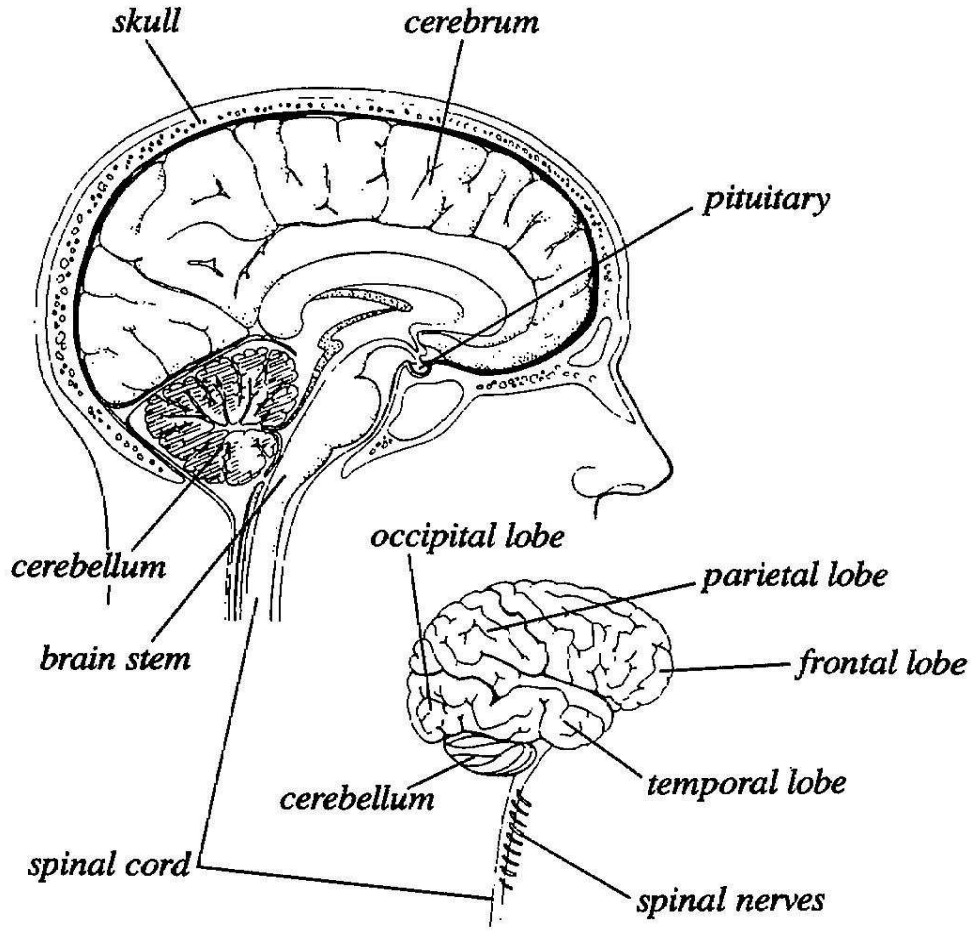
To understand brain and spinal cord tumors, it helps to know about the normal structure and function of the central nervous system (CNS), which is the medical name for the brain and spinal cord.

The brain is the center of thought, feeling, memory, speech, vision, hearing, movement, and much more. The spinal cord and special nerves in the head called cranial nerves carry messages between the brain and the rest of the body. These messages tell our muscles how to move, transmit information gathered by our senses, and help coordinate the actions of our internal organs.

The brain is protected by the skull. Likewise, the spinal cord is protected by the bones (vertebrae) of the spinal column.

The brain and spinal cord are surrounded and cushioned by a special liquid, called *cerebrospinal fluid (CSF).* Cerebrospinal fluid is made by the choroid plexus, which is located in spaces within the brain called ventricles. The ventricles, as well as the spaces around the brain and spinal cord, are filled with CSF.

Parts of the brain and spinal cord



The main areas of the brain include the cerebrum, cerebellum, and brain stem. Each area has a special purpose.

Cerebrum: The cerebrum is the large, outer part of the brain. It is made up of 2 hemispheres (halves) and controls reasoning, thought, emotion, and language. It is also responsible for planned muscle movements (throwing a ball, walking, chewing, etc.) and for taking in and interpreting sensory information such as vision, hearing, smell, touch, and pain.

The symptoms caused by a tumor in a cerebral hemisphere depend on where in the hemisphere the tumor develops. Common symptoms include:

- Seizures
- Trouble speaking
- A change of mood such as depression
- A change in personality

- Weakness or paralysis of part of the body
- Changes in vision, hearing, or sensation

Cerebellum: The cerebellum lies underneath the cerebrum at the back part of the brain. It helps coordinate movement. Tumors of the cerebellum can cause problems with coordination in walking, trouble with fine movements of arms and legs, problems with swallowing or synchronized eye movements, and changes in rhythm of speech.

Brain stem: The brain stem is the lower part of the brain that connects to the spinal cord. It has bundles of very long nerve fibers that carry signals controlling muscles and sensation or feeling between the cerebrum and the rest of the body. Special centers in the brain stem also help control breathing and the beating of the heart. In addition, most cranial nerves (which carry signals directly between the brain and the face, eyes, tongue, and mouth) start in the brain stem.

Tumors in this critical area of the brain may cause weakness, stiff muscles, or problems with sensation, hearing, facial movement, or swallowing. Double vision is a common early symptom of brain stem tumors, as are problems with coordination in walking. The brain stem is a small area that is so essential for life, it may not be possible to surgically remove tumors in this area.

Spinal cord: The spinal cord, like the brain stem, has bundles of very long nerve fibers that carry signals that control muscles, sensation or feeling, and bladder and bowel control. Spinal cord tumors may cause weakness, paralysis, or numbness. Because the spinal cord is such a narrow structure, tumors that develop there usually cause symptoms on both sides of the body (for example, weakness or numbness of both legs). This is different from tumors of the brain, which usually affect only one side of the body. Moreover, most tumors of the spinal cord develop below the neck, where the nerves to the arms have branched off the spinal cord, so that only lower body functions – bowel, bladder, or leg movement or sensation – are affected.

Cranial nerves: The cranial nerves are nerves that extend directly out of the base of the brain (as opposed to coming out of the spinal cord). The most common cranial nerve tumors in children are called *optic gliomas*, which are tumors of the optic nerve (the large nerve that runs between the brain and each eye) that cause vision problems. Tumors starting in other cranial nerves may cause trouble swallowing; hearing loss in one or both ears; or facial paralysis, numbness, or pain.

Peripheral nervous system: The peripheral nervous system consists of the parts of the nervous system other than the brain and spinal cord (which make up the central nervous system). Tumors that start in the nerves of the peripheral nervous system generally cause pain, weakness, and/or loss of sensation in the area served by that nerve.

Types of cells and body tissues in the brain and spinal cord

The brain and spinal cord contain different kinds of tissues and cells, which can develop into different types of tumors. These tumors can have different prognoses (outlooks) and may be treated differently.

Neurons (nerve cells): These are the most important cells within the brain. They send signals through their nerve fibers (axons). Axons in the brain tend to be short, while those in the spinal cord can be as long as several feet. Electric signals carried by neurons determine thought, memory, emotion, speech, muscle movement, and just about everything else that the brain and spinal cord do. Unlike many other types of cells that can grow and divide to repair damage from injury or disease, neurons stop dividing about a year after birth (with a few exceptions). Neurons do not usually form tumors, but they can be damaged by tumors that start nearby.

Glial cells: Glial cells are the supporting cells of the brain. Most brain and spinal cord tumors develop from glial cells. These tumors are sometimes referred to as a group called *gliomas*.

There are 3 types of glial cells – astrocytes, oligodendrocytes, and ependymal cells. A fourth cell type called microglia is part of the immune system and is not truly a glial cell.

- **Astrocytes** help support and nourish neurons. When the brain is injured, astrocytes form scar tissue that helps repair the damage. The main tumors arising from these cells are called *astrocytomas* or *glioblastomas*.

- **Oligodendrocytes** make myelin, a substance that surrounds and insulates the nerve cell axons of the brain and spinal cord. This helps neurons send electric signals through the axons. Tumors arising from these cells are called *oligodendrogliomas.*

- **Ependymal cells** line the ventricles (fluid-filled areas) within the central part of the brain and form part of the pathway through which CSF flows. Tumors arising from these cells are called *ependymomas.*

- **Microglia** are the immune (infection fighting) cells of the central nervous system.

Neuroectodermal cells: These are primitive cells that are probably the remains of embryonic cells. They are found throughout the brain. The most common tumors that come from these cells are called *medulloblastomas*, which arise in the cerebellum.

Meninges: These are layers of tissue that line the outer part of the brain and spinal cord. The meninges help form the spaces through which CSF travels. The most common tumors that start in these cells are called *meningiomas.*

Choroid plexus: The choroid plexus is the area of the brain within the ventricles that makes CSF, which nourishes and protects the brain. Tumors that start here include *choroid plexus papillomas* and *choroid plexus carcinomas*.

Pituitary gland and hypothalamus: The pituitary is a small gland at the base of the brain. The hypothalamus is the part of the brain to which the pituitary gland is connected. Both help regulate the activity of several other glands. For example, they control the amount of thyroid hormone made by the thyroid gland, the production and release of milk by the breasts, and the amount of male or female hormones made by the testicles or ovaries. They also make growth hormone, which stimulates body growth, and vasopressin, which regulates water balance by the kidneys.

The growth of tumors in or near the pituitary or hypothalamus, as well as surgery and/or radiation therapy in this area, can interfere with these functions. As a result, a child may have low levels of one or more hormones after treatment and may need to take hormones to correct any deficiencies.

Pineal gland: The pineal gland is not strictly part of the brain. It is, in fact, a small endocrine gland that sits between the cerebral hemispheres. Its main function is probably to make melatonin, a hormone that regulates sleep, in response to changes in light.

Blood-brain barrier: Unlike the case with most other organs, the small blood vessels (capillaries) in the brain and spinal cord create a very selective barrier between the blood and the tissues of the central nervous system. This normally keeps harmful toxins from getting into the brain. Unfortunately, it also keeps out most chemotherapy drugs that are used to kill cancer cells, which in some cases limits their usefulness.

Types of brain and spinal cord tumors in children

Tumors can form in almost any type of tissue or cell in the brain or spinal cord. Some tumors have a mixture of cell types. Tumors in different areas of the central nervous system may be treated differently and have different prognoses (outlooks).

Brain tumors in children are more likely to start in the lower parts of the brain, such as the cerebellum and brain stem, than they are in adults. However, they can start in the upper parts of the brain as well.

Unlike cancers that start in other parts of the body, tumors starting in the brain or spinal cord rarely spread (metastasize) to distant organs. They cause damage because they can grow and spread into nearby areas, where they can destroy normal tissue.

Gliomas

Gliomas are not a specific type of cancer. Glioma is a general term for a group of tumors that start in glial cells. A number of tumors can be considered gliomas, including glioblastoma (also known as glioblastoma multiforme), anaplastic astrocytoma, astrocytoma, oligodendroglioma, ependymoma, brain stem glioma, and optic glioma. Most brain and spinal cord tumors in children are gliomas.

Astrocytomas: Most tumors that develop in the brain itself start in cells called astrocytes, a kind of glial cell. These tumors are called astrocytomas. About half of all childhood brain tumors are astrocytomas. When these tumors occur in the brain stem, they are referred to as *brain stem gliomas*.

Most astrocytomas can spread widely throughout, and intermingle with, the normal brain tissue, which can make them very hard to remove by surgery. Sometimes they spread along the CSF pathways. It is very rare for them to spread outside of the brain or spinal cord.

Astrocytomas are classified as low grade, intermediate grade, or high grade, based on how the cells look under the microscope.

- Low-grade astrocytomas are the slowest growing and the most common type of astrocytoma in children.
- Intermediate-grade astrocytomas, or **anaplastic astrocytomas**, grow at a moderate rate.
- The highest-grade astrocytoma, known as **glioblastoma** (or glioblastoma multiforme), is the fastest growing.

There are some special types of astrocytomas that are low grade and tend to have a good prognosis.

- **Juvenile pilocytic astrocytomas** are slow growing and rarely infiltrate (grow into) nearby tissues. They most commonly occur in the cerebellum but can also occur in the optic nerve, hypothalamus, brain stem, or other areas.
- **Subependymal giant cell astrocytomas** occur in the ventricles. They are also slow growing and rarely infiltrate nearby tissues. These tumors are almost always linked with tuberous sclerosis (an inherited condition that may also cause epilepsy, mental retardation, and tumors of the skin and kidneys).
- **Optic gliomas** are low-grade astrocytomas of childhood that start in the optic nerve. They are often linked with an inherited condition called neurofibromatosis type 1. These tumors can sometimes be treated successfully by surgery. At other times they may require radiation therapy or chemotherapy. These tumors are rarely cause death but may cause vision loss and injury to nearby brain tissue.

Oligodendrogliomas: These tumors start in brain glial cells called oligodendrocytes. Like astrocytomas, most of these can spread or grow into nearby brain tissue and cannot be completely removed by surgery. Oligodendrogliomas rarely spread along the CSF pathways and even less frequently spread outside the brain or spinal cord.

Ependymomas: About 5% to 10% of brain tumors in children are ependymomas. These tumors arise from the ependymal cells that line the ventricles or central canal of the spinal cord. They can range from fairly low-grade (less aggressive) tumors to higher grade ones, which are called *anaplastic ependymomas*.

Ependymomas may spread along the CSF pathways but do not spread outside the brain or spinal cord. Ependymomas may block the flow of cerebrospinal fluid out of the ventricles, causing the ventricles to become very large – a condition called hydrocephalus.

Unlike astrocytomas and oligodendrogliomas, ependymomas usually do not spread into or infiltrate normal brain tissue. As a result, some (but not all) ependymomas can be removed and cured by surgery. But because they can spread along ependymal surfaces and CSF pathways, treating them can sometimes be difficult. Spinal cord ependymomas have the greatest chance of being cured, but treatment can cause side effects related to nerve damage.

Primitive neuroectodermal tumors (PNETs)

These tumors start in primitive (immature) cells of the central nervous system. About 1 out of 5 brain tumors in children are this type. They are more common in younger children than older ones, and are rare in adults. PNETs tend to grow fast and frequently spread throughout the CSF pathways. These tumors sometimes have different names depending on where they occur.

Medulloblastomas: PNETs that start in the cerebellum are called medulloblastomas. About 15% of childhood brain tumors are medulloblastomas. These tumors can often be treated effectively and tend to have a better prognosis than PNETs in other parts of the brain.

Pineoblastomas: PNETs are called pineoblastomas when they occur in the pineal gland. The outlook for pineoblastomas is not as favorable as that for medulloblastomas.

Craniopharyngiomas

These slow-growing tumors start above the pituitary gland but below the brain itself. They may compress the pituitary gland and the hypothalamus, causing hormonal problems. Most craniopharyngiomas are very close to the optic nerve, making them hard to remove completely without damaging the child's vision.

Mixed glial and neuronal tumors

Certain tumors that occur in children and young adults (and rarely in older adults) seem to have both glial and neuronal cell components. They tend to have a fairly good prognosis.

- **Pleomorphic xanthoastrocytoma** and **dysembryoplastic neuroepithelial tumors** appear malignant under the microscope, but these tumors tend to be fairly benign, and most are cured by surgery alone.
- **Gangliоglioma** is a type of tumor that has both mature neurons and glial cells. Most can be cured by surgery alone or surgery combined with radiation therapy.

Choroid plexus tumors

These rare tumors arise in the choroid plexus within the ventricles of the brain. Most are benign (choroid plexus papillomas) and cured by surgery. However, some are malignant (choroid plexus carcinomas).

Schwannomas (neurilemomas)

This type of tumor starts in Schwann cells that surround and insulate cranial nerves and other nerves. Schwannomas are usually benign tumors. They often form near the cerebellum in the cranial nerve responsible for hearing and balance, in which case they

are called *vestibular schwannomas* or *acoustic neuromas*. They may also arise from spinal nerves after the point where they have left the spinal cord. When this is the case, they can compress the spinal cord, causing weakness, sensory loss, and bowel and bladder problems.

These tumors are rare in children. When present in this age group, particularly if there is more than one, they might suggest an inherited tumor syndrome such as neurofibromatosis.

Other tumors that start in or near the brain

Meningiomas: These tumors arise from the meninges, the layers of tissue that surround the outer part of the brain and spinal cord. Meningiomas cause symptoms by pressing on the brain or spinal cord. They are much less common in children than in adults.

Meningiomas are almost always benign and are usually cured by surgery. Some, however, are located very close to vital structures in the brain and cannot be cured by surgery alone.

Meningiomas are often assigned a grade based on how the cells look under the microscope. Grade I tumors, which look most like normal cells, make up about 80% to 90% of meningiomas. Grade II (atypical) meningiomas look slightly more abnormal. Grade III (anaplastic) meningiomas, which look the most abnormal, make up only about 1% to 3% of meningiomas. Higher grade meningiomas are more likely to come back after treatment, and some grade III meningiomas can spread to other parts of the body.

Chordomas: These tumors start in the bone at the base of the skull or at the lower end of the spinal column. These tumors are not from the central nervous system, but they can cause injury to the nervous system by pressing on it. Chordomas may come back many times over a period of 10 to 20 years, causing more and more damage. They usually do not spread or metastasize to other organs.

Germ cell tumors: Germ cell tumors develop from germ cells that normally form eggs in women and sperm in men. During normal embryonic and fetal development, germ cells migrate to the ovaries or testicles and develop into eggs or sperm cells. Sometimes, however, a few germ cells may not migrate properly and end up in abnormal locations such as the brain. They may then develop into germ cell tumors similar to those that can form in the ovaries or testicles.

Germ cell tumors of the nervous system usually occur in children, most often in the pineal gland or above the pituitary gland. Germ cell tumors can sometimes be diagnosed without a biopsy by measuring certain chemicals in the cerebrospinal fluid or blood.

The most common germ cell tumor of the nervous system is the germinoma, which can be cured in almost all cases by radiation therapy and sometimes chemotherapy. Other tumors that start in germ cells, such as choriocarcinoma or yolk sac tumors are rarely cured by surgery. Both radiation therapy and chemotherapy are used in their treatment, but in some cases this may not control the tumor completely.

Neuroblastomas: These nerve cell tumors are the third most common cancer in children. Neuroblastomas rarely develop in the brain or spinal cord; most develop from nerve cells inside the abdomen or chest. This type of cancer is most commonly diagnosed during early infancy. For more information, see our separate document, *Neuroblastoma*.

Cancers that spread to the brain from other sites

Sometimes brain tumors are found not to have started in the brain but rather to have spread (metastasized) from some other part of the body. Tumors that start in other organs and then spread to the brain are called *metastatic brain tumors* (as opposed to primary brain tumors, which start in the brain). This is important because metastatic and primary brain tumors are usually treated differently. In children, metastatic tumors to the brain are much less common than primary brain tumors.

This document covers only primary brain tumors.

What are the key statistics about brain and spinal cord tumors in children?

Brain and spinal cord tumors are the second most common cancers in children (after leukemia). They account for about 21% of childhood cancers. Around 4,000 central nervous system tumors are diagnosed each year in children and teens. About one fourth of these are considered benign tumors. The incidence rate (number per 100,000 children) of these cancers has not changed much in recent years.

Boys are affected slightly more often than are girls.

About 3 out of 4 children with brain tumors (all types combined) survive *at least* 5 years after being diagnosed. The outlook can vary a great deal based on the type of tumor and where it is located. For more specific survival information on some particular tumor types, see "Survival rates for selected brain and spinal cord tumors."

What are the risk factors for brain and spinal cord tumors in children?

A risk factor is anything that affects a person's chance of getting a disease such as cancer. Different cancers have different risk factors. For example, smoking is a risk factor for several types of cancer in adults.

Lifestyle-related risk factors such as diet, body weight, physical activity, and tobacco use play a major role in many adult cancers. But these factors usually take many years to influence cancer risk, and they are not thought to play much of a role in childhood cancers, including brain tumors.

Very few risk factors for brain tumors have been found. There is no clear cause for most brain tumors.

Radiation exposure

The only well-established environmental risk factor for brain tumors is radiation exposure to the head, which most often comes from the treatment of other conditions. For example, before the risks of radiation were well known (more than 50 years ago), children with ringworm of the scalp (a fungal infection) often received low-dose radiation therapy. This was found to increase their risk of brain tumors later in life.

Today, most radiation-induced brain tumors are caused by radiation to the head given for the treatment of other cancers. This is most common in children who received radiation to the brain as part of their treatment for leukemia. These brain tumors usually develop around 10 to 15 years after the radiation.

These tumors are still fairly rare, but because of the increased risk (as well as the other side effects), radiation therapy to the head is only given after careful consideration of benefits and risks. For most patients with cancer involving the brain or other areas of the head, the benefits of radiation therapy far outweigh the small risk of developing a brain tumor years later.

Inherited and genetic conditions

In rare cases (less than 1 in 10 brain tumors), children may have inherited abnormal copies of genes from a parent that put them at increased risk for certain types of brain tumors. In other cases, the abnormal genes are not inherited but occur as a result of changes (mutations) in the gene before birth.

People with inherited tumor syndromes often have many tumors that start when they are young. Some of the more well-known disorders are listed below.

Neurofibromatosis type 1 (von Recklinghausen disease): This is the most common syndrome linked to brain or spinal cord tumors. It is usually inherited from a parent, but it can also start in some children whose parents don't have it. Children with this syndrome may have optic gliomas or other gliomas of the brain or spinal cord, or neurofibromas (benign tumors of peripheral nerves). Changes in the NF1 gene cause this disorder.

Neurofibromatosis type 2: Less common than von Recklinghausen disease, this condition can also either be inherited or may start in children without a family history. It is associated with cranial or spinal nerve schwannomas, especially acoustic neuromas (usually on both sides of the head). It is also linked to an increased risk of meningiomas, as well as spinal cord gliomas or ependymomas. Changes in the NF2 gene are responsible for neurofibromatosis type 2.

Tuberous sclerosis: This condition may be associated with subependymal giant cell astrocytomas, as well as with other benign tumors of the brain, skin, heart, or kidneys. It is caused by changes in either the TSC1 or the TSC2 gene.

Von Hippel-Lindau disease: Children with this disease have an inherited tendency to develop hemangioblastomas (blood vessel tumors) of the cerebellum, spinal cord, or retina, as well as kidney cancer. It is caused by changes in the VHL gene.

Li-Fraumeni syndrome: People with this syndrome have an increased risk of gliomas, as well as breast cancer, soft tissue sarcomas, leukemia, and adrenal gland cancers. It is caused by changes in the p53 gene.

Other inherited conditions, including Gorlin syndrome, Turcot syndrome, Cowden syndrome, and hereditary retinoblastoma are also linked with increased risks of certain types of brain and spinal cord tumors. Some families may have genetic disorders that are not well recognized or that may even be unique to a particular family.

Do we know what causes brain and spinal cord tumors in children?

The cause of most central nervous system tumors is not fully understood. But researchers have found some of the chemical changes that occur in normal brain cells that may lead them to form brain tumors.

Normal human cells grow and function based mainly on the information contained in each cell's chromosomes. Chromosomes are long molecules of DNA in each cell. Brain tumors, like other tumors, are usually caused by changes (mutations) in a person's DNA. DNA is the chemical in each of our cells that makes up our genes – the instructions for how our cells function. We usually look like our parents because they are the source of our DNA. But DNA affects more than how we look.

Some genes control when our cells grow, divide, and die. Certain genes that speed up cell division are called *oncogene*s. Others that slow down cell division, or cause cells to die at the right time, are called *tumor suppressor genes*. Cancers can be caused by DNA mutations that turn on oncogenes or turn off tumor suppressor genes. These gene changes can be inherited from a parent (as is sometimes the case with childhood cancers) or may happen during a person's lifetime as cells in the body divide to form 2 new cells.

In recent years, researchers have found the gene mutations that cause some rare inherited syndromes (like neurofibromatosis, tuberous sclerosis, Li-Fraumeni, and von Hippel-Lindau) and increase the risk of developing some central nervous system tumors. For example, the Li-Fraumeni syndrome is caused by mutation of the p53 tumor suppressor gene. Normally, this gene prevents cells with damaged DNA from growing. Changes in this gene increase the risk of developing brain tumors (particularly astrocytomas), as well as some other cancers.

In most cases, it is not known why people without inherited syndromes develop changes in their central nervous system cells. Most risk factors for cancer somehow damage genes. For example, cigarette smoke is a risk factor for lung cancer and several other cancers because it contains chemicals that can damage genes. The brain is relatively protected from cigarette smoke and other cancer-causing chemicals that we all breathe or eat, so these factors are not likely to play a major role in these cancers.

Normal cells usually require several different gene changes before they become cancerous. There are many kinds of brain tumors, each of which may have different sets

of gene changes. A number of gene or chromosome changes have been found in different brain tumor types, but there are probably many others that have not yet been discovered.

Researchers now understand some of the gene changes that may occur in different types of brain tumors, but it's still not clear what might cause these changes. Some gene changes may be inherited, but most brain and spinal cord tumors in children are not the result of known inherited syndromes. Other gene changes may just be a random event that sometimes happens inside a cell, without having an external cause. Other than radiation, there are no known lifestyle-related or environmental causes of childhood brain tumors, so it is important to remember that there is nothing these children or their parents could have done to prevent these cancers.

Can brain and spinal cord tumors in children be prevented?

The risk of many adult cancers can be reduced with certain lifestyle changes (such as maintaining a healthy weight or quitting smoking), but at this time there are no known ways to prevent most cancers in children.

Other than radiation exposure, there are no known lifestyle-related or environmental causes of brain and spinal cord tumors in children, so at this time there is no way to protect against most of these cancers.

For most children with cancer involving the brain or other areas of the head, the benefits of radiation therapy far outweigh the small risk of developing a brain tumor years later. Still, when it is needed, doctors try to limit the dose of radiation as much as possible.

Can brain and spinal cord tumors in children be found early?

At this time there are no widely recommended blood tests or other screening exams for children to detect brain tumors before they start to cause symptoms. These tumors usually come to light as a result of signs or symptoms the child is having. In most cases, survival of patients with brain tumors depends on the type of tumor and its location, not how early it is detected. But as with any disease, earlier detection and treatment is likely to be helpful.

For children diagnosed with certain inherited syndromes that put them at higher risk for brain tumors, such as neurofibromatosis or tuberous sclerosis, doctors may recommend frequent physical exams and other tests. In some cases these tests may find tumors when they are still small. Not all tumors related to these symptoms may need to be treated right away, but finding them early may help doctors monitor them so that they can be treated quickly if they begin to grow or cause problems.

How are brain and spinal cord tumors in children diagnosed?

Brain and spinal cord tumors are usually found because of signs or symptoms a child is having. If a tumor is suspected, tests will be needed to confirm the diagnosis.

Symptoms of brain and spinal cord tumors

Symptoms from brain and spinal cord tumors may occur gradually and become worse over time, or they may happen suddenly.

General symptoms

Tumors within any part of the brain may cause pressure to rise within the skull. This can be caused by growth of the tumor, swelling in the brain, or blockage of the flow of cerebrospinal fluid. Increased pressure can lead to general symptoms such as:

- Headache
- Nausea
- Vomiting
- Crossed eyes or blurred vision
- Balance problems
- Behavior changes
- Seizures
- Drowsiness or even coma

A doctor can often tell if intracranial pressure is increased by looking in the child's eyes for swelling of the optic nerve (known as papilledema). In very young children who can't complain of symptoms, a parent may notice an increase in head size, with or without bulging of the soft spots of the skull (fontanelles). This happens because the several bones that make up the skull haven't grown together yet, and increased pressure from a tumor can push them apart.

Headache can be a major symptom of brain tumors. However, not all brain tumors cause headaches, and most headaches are not caused by tumors.

In some children, seizures are the first symptom of a brain tumor. Most seizures in children are not caused by brain tumors, but if your child has a seizure, your child's doctor may refer you to a neurologist (a doctor who specializes in brain and nervous system problems) to make sure it wasn't caused by a brain tumor.

In the school-aged child, other general symptoms of tumors can include poor school performance, fatigue, and personality changes.

In the first few years of life, other symptoms of tumors can include irritability, loss of appetite, developmental delay, and a drop in intellectual and physical abilities.

Symptoms of tumors in different parts of the central nervous system

Tumors in different parts of the central nervous system can cause different symptoms. But these symptoms can be caused by any disease in that particular location in the brain – they do not always mean a brain tumor is present.

Brain and spinal cord tumors often cause problems with the specific functions of the region they develop in. For example:

- Tumors in parts of the cerebrum (the large, outer part of the brain) that control movement or sensation may cause weakness or numbness in a part of the body.
- Tumors in or near the parts of the cerebrum responsible for language may cause problems with speech or even understanding words.
- Tumors in the front part of the cerebrum can sometimes affect thinking and personality.
- Tumors in an area of the brain called the basal ganglia typically cause abnormal movements and abnormal body positions.
- If the tumor is in the cerebellum, where coordination is controlled, the child may have trouble walking or with other normal functions, even eating.
- Tumors in the back part of the cerebrum, or around the pituitary gland, the optic nerve, or certain other cranial nerves may cause vision problems.
- Tumors in or near other cranial nerves may lead to loss of hearing, balance problems, weakness of some facial muscles, or trouble swallowing.
- Spinal cord tumors may cause numbness and/or weakness in both legs, as well as bladder or bowel problems.

Having one or more of the symptoms above does not mean that your child definitely has a brain or spinal cord tumor. All of these symptoms may have other causes. Still, if your child's symptoms suggest that a brain or spinal cord tumor may be present, make an appointment with a doctor so that the cause can be evaluated and treated, if needed.

Medical history and physical exam

If your child has symptoms that suggest a central nervous system tumor may be present, the doctor will take a complete medical history and do a neurologic exam to evaluate your child's brain and spinal cord function, if possible. Your child's doctor may do this

special type of exam. It tests reflexes, sensation, muscle strength, eye and mouth movement, coordination, alertness, and other functions.

If the results are abnormal, your child's doctor may refer you to a neurologist (a doctor specializing in nervous system diseases) or a neurosurgeon (a surgeon specializing in operations to treat nervous system diseases) to do a more detailed exam or for other tests.

Imaging tests

If your child's doctor thinks a brain problem may be present, he or she will probably order one or more imaging tests. These tests use x-rays, strong magnets, or radioactive substances to create pictures of internal organs such as the brain and spinal cord. The pictures may be looked at by doctors specializing in this field (neurosurgeons, neurologists, and neuroradiologists) as well as by your child's doctor.

Magnetic resonance imaging (MRI) or computed tomography (CT) scans are used most often for brain diseases. These scans will show a brain tumor, if one is present, in almost all cases, and can often tell the doctors exactly where the tumor is in the brain.

Magnetic resonance imaging (MRI) scan

MRI scans are very good for looking at the brain and spinal cord and are considered the best way to look for tumors in these areas. The images they provide are usually more detailed than those from CT scans (described below). But they do not image the bones of the skull as well as CT scans and therefore may not see the effects of tumors on the skull.

MRI scans provide detailed images of soft tissues in the body. They use radio waves and strong magnets instead of x-rays, so they do not expose the child to radiation. The energy from the radio waves is absorbed and then released in a pattern formed by the type of body tissue and by certain diseases. A computer translates the pattern into a very detailed image of parts of the body. A contrast material called *gadolinium* may be injected into a vein before the scan to better see details.

MRI scans can take a long time to complete – often up to an hour. Your child may have to lie inside a narrow tube, which is confining and can be distressing, so sedation is sometimes needed. Open MRI machines may be another option, though they may provide less detailed images. The MRI machine makes loud buzzing and clicking noises that your child may find disturbing. Some places provide headphones or earplugs to help block this noise out.

Magnetic resonance angiography (MRA): In some cases, a special form of MRI known as magnetic resonance angiography may be done to look at the structure of the blood vessels in the brain. This can be very useful before surgery to help the surgeon plan an operation.

Magnetic resonance spectroscopy: This test (also known as MR spectroscopy or MRS) is like an MRI, except that the radio wave interactions with different atoms within the tissues are measured. MRS highlights some features of brain tumors that may not be seen

clearly with MRI. It generally produces graph-like results called spectra (but crude images can also be created). This may help narrow the possible type of tumor, but in most cases a biopsy of the tumor is still needed to be sure. MRS can also be used after treatment to help determine if an abnormal area is remaining tumor or if it is more likely to be scar tissue.

Magnetic resonance perfusion: For this test, also known as perfusion MRI, a contrast dye is injected quickly into a vein. A special type of MR image is then obtained to look at the amount of blood going through different parts of the brain and tumor. Tumors need a bigger blood supply than normal areas of the brain. The faster a tumor is growing, the more blood it needs.

Perfusion MRI can give doctors an idea of how quickly a tumor is growing or help show them the best place to take a biopsy. It can also be used after treatment to help determine if an abnormal area is remaining tumor or if it is more likely to be scar tissue.

Computed tomography (CT) scan

The CT scan is an x-ray test that can produce detailed cross-sectional images of your child's brain and spinal cord. Instead of taking one picture, like a regular x-ray, a CT scanner takes many pictures as it rotates around your child while he or she lies on a table. A computer then combines these pictures into images of slices of the body. Unlike a regular x-ray, a CT scan creates detailed images of the soft tissues in the body.

CT scans formerly were used quite often to find brain and spinal cord tumors, but they have been largely replaced by MRI scans, which provide slightly more detailed images. Still, there are instances where CT scans may have advantages over MRI scans:

- CT scans take much less time than MRIs, which can be particularly helpful for children who have trouble staying still.
- CT scans provide greater detail of the bone structures near the tumor than MRIs do.
- CT angiography (CTA), which is described below, can provide better details of the blood vessels in and around a tumor than MRA in selected cases.

Before the scan, your child may get an injection of a contrast dye through an IV (intravenous) line. This helps better outline any tumors that are present. The contrast contains iodine and may cause some flushing (a feeling of warmth, especially in the face). Some people are allergic to the dye and get hives. Rarely, more serious reactions like trouble breathing or low blood pressure can occur. Be sure to tell the doctor if your child has any allergies or has ever had a reaction to any contrast material used for an imaging test.

CT scans take longer than regular x-rays (but not as long as MRI scans). Your child will need to lie still on a table while they are being done. During the test, the table moves in and out of the scanner, a ring-shaped machine that completely surrounds the table. Some people feel a bit confined by the ring they have to lie in while the pictures are being

taken. In some cases, your child may need to be sedated before the test to keep them as still as possible and help make sure the pictures come out well.

Spiral CT (also known as helical CT) is now available in many medical centers. This type of CT scan uses a faster machine. The scanner part of the machine rotates around the body continuously, allowing doctors to collect the images much more quickly than with a standard CT. This lowers the chance of blurred images from body movements. It also lowers the dose of radiation received during the test. The slices it images are also thinner, which gives more detailed pictures.

CT angiography (CTA): For this test, your child is injected with a contrast material through an IV line while he or she is in the CT scanner. The scan creates detailed images of the blood vessels in the brain, which can help doctors plan surgery. CT angiography can provide better details of the blood vessels in and around a tumor than MR angiography in selected cases.

Positron emission tomography (PET) scan

For a PET scan, a radioactive substance (usually a type of sugar related to glucose, known as FDG) is injected into the blood. The amount of radioactivity used is very low. Because cancer cells in the body are growing quickly, they absorb larger amounts of the sugar than most other cells. A special camera can then create a picture of areas of radioactivity in the body. The picture is not finely detailed like a CT or MRI scan, but it can provide helpful information about whether abnormal areas seen on other tests (such as MRIs) are likely to be cancerous or not.

This test is also useful after treatment, as it can help tell whether the tumor cells have been killed. Abnormal areas may still show up on an MRI scan. PET scans can help determine if the abnormal area is the remaining part of an active tumor or if it is more likely to just be scar tissue.

Angiogram

For this test, a special dye is injected into blood vessels near the tumor and the area then is viewed with x-rays. This helps doctors look at a tumor's blood supply. This test is not done much any more, as it has largely been replaced by magnetic resonance angiography (MRA) or computerized tomographic angiography (CTA) in recent years.

The brain tumor team

If a brain tumor is diagnosed or strongly suspected, your child should be seen at a children's cancer center by a team of specialists. A neurosurgeon usually heads the team that evaluates and treats your child and does the surgery, if it is needed. Other doctors on the team may include:

- Pediatric neurologist: a doctor who treats brain and nervous system diseases in children

- Radiation oncologist: a doctor who uses radiation to treat cancer
- Pediatric oncologist: a doctor who uses chemotherapy and other medicines to treat children's cancers
- Endocrinologist: a doctor who treats diseases in glands that secrete hormones

Before treatment, the team's social worker will help you, as a parent, understand the tests that will need to be done. The social worker will also counsel you about the problems you and your child may have during and after surgery, and may be able to help you find housing and financial aid if needed.

Other team members, such as a psychologist and specialists in rehabilitation, may also see your child before treatment begins. For example, if the tumor is slow growing and your child's condition is stable, he or she may be seen by a psychologist before treatment to assess any damage the tumor may have caused. Most of the work of these specialists takes place after treatment.

Biopsies to obtain tumor or other samples

A biopsy removes a sample of tumor to see whether cancer cells are present. Different kinds of biopsies may be used to diagnose a brain tumor or to help determine how far it may have spread.

Brain or spinal cord tumor biopsy

Imaging tests such as MRI and CT scans may show that a brain or spinal cord tumor is present (or very likely). But usually a definite diagnosis of the type of tumor can be made only by removing a sample of the tumor, which is called a biopsy. A neuropathologist (a doctor who uses lab tests to diagnose diseases of the nervous system) then looks at the biopsy sample under a microscope.

In some cases, such as for many brain stem gliomas, it may not be possible to biopsy the tumor safely, so the diagnosis may be made based only on how the tumor looks on imaging tests.

Biopsies may be done in different ways.

Stereotactic needle biopsy: This type of biopsy may be used in cases where the risks of surgery might be too high (such as with some tumors deep within the brain) but a sample is still needed to make a diagnosis.

Depending on the situation, the biopsy may be done with the child awake or under general anesthesia (asleep). For this procedure, the neurosurgeon injects a local anesthetic into areas of skin over the skull to numb them. (The skull and brain itself do not feel pain.) A rigid frame may then be fixed onto the child's head. This helps make sure the surgeon will target the tumor precisely. A small incision is made in the scalp and a small hole is drilled in the skull. An MRI or CT scan is used along with the frame to help the neurosurgeon guide a hollow needle into the tumor to remove a small piece of tissue.

Another approach is to attach markers to the scalp, obtain an MRI or CT scan, and then use an image-guidance system to direct the needle into the tumor.

The biopsy samples are then looked at under a microscope by a neuropathologist. The doctor can usually tell exactly what type of tumor it is. This helps determine the best course of treatment and the prognosis (outlook).

Craniotomy: If the tumor appears to be treatable with surgery based on the imaging tests, the neurosurgeon may not do a needle biopsy. Instead, he or she may do an operation called a *craniotomy* (described in the "Surgery" section) to remove all or most of the tumor. (Removing most of the tumor is known as *debulking*.)

Small samples of the tumor are looked at right away by the neuropathologist while the child is still in the operating room, to get a preliminary diagnosis. This can help guide treatment, including whether further surgery should be done at that time. A final diagnosis is arrived at a few days later in most cases.

Lumbar puncture (spinal tap)

This test is used to look for cancer cells in the cerebrospinal fluid (CSF), which is the liquid that bathes the brain and spinal cord. For this test, the doctor first numbs an area in the lower part of the back over the spine. The doctor may also recommend that the child be given something to make them sleep so the lumbar puncture can be done without difficulty or causing harm. A small, hollow needle is then placed between the bones of the spine to withdraw some of the fluid.

The fluid is looked at under a microscope for cancer cells. Tests can be also done on the CSF to check for certain substances released by some germ cell tumors.

Bone marrow aspiration and biopsy

Because some tumors (especially medulloblastomas) can spread beyond the nervous system, in some instances the doctor may recommend looking at cells in your child's bone marrow to see if the tumor has spread there.

The bone marrow aspiration and biopsy are often done at the same time. The samples are usually taken from the back of the pelvic (hip) bone, but in some cases they may be taken from the sternum (breastbone) or other bones.

In bone marrow *aspiration*, the skin over the hip and the surface of the bone are cleaned and numbed with local anesthetic. In most cases, the child is also given other medicines to reduce pain or even be asleep during the procedure. A thin, hollow needle is then inserted into the bone, and a syringe is used to suck out (aspirate) a small amount of liquid bone marrow.

A bone marrow *biopsy* is usually done just after the aspiration. A small piece of bone and marrow is removed with a slightly larger needle that is twisted as it is pushed down into the bone. Once the biopsy is done, pressure is applied to the site to help stop any bleeding

The specimens are then looked at under a microscope for tumor cells.

Blood and urine tests

These lab tests are rarely used to diagnose brain and spinal cord tumors, but if your child has been sick for some time they may be done to check how well the liver, kidneys, and some other organs are working. This is especially important before any planned surgery. If your child is getting chemotherapy, blood tests will be done routinely to check blood counts and to see if the treatment is affecting other parts of the body.

How are brain and spinal cord tumors in children staged?

Staging is the use of exams and imaging tests to determine how widespread a cancer is. For most cancers, the stage (extent) of the cancer is one of the most important factors in selecting treatment options and in determining the outlook (prognosis).

But tumors of the central nervous system (CNS) differ in some important ways from cancers in other parts of the body. The most deadly aspect of other cancers is their ability to spread throughout the body. Tumors starting in the brain or spinal cord can spread to other parts of the CNS, but they almost never spread to other organs. The most dangerous aspect of these tumors is that they can interfere with essential functions of the brain.

Because most tumors in the brain or spinal cord do not usually spread, they are not formally staged. Some of the most important factors that determine your child's prognosis include:

- The type of tumor (such as astrocytoma, ependymoma, etc.)
- The grade of the tumor (how quickly the tumor is likely to grow, based on how the cells look under a microscope)
- The location of the tumor
- How much of the tumor can be removed by surgery (if it can be done)
- Your child's age
- Your child's functional level (whether the tumor has started to interfere with normal brain functions)
- Whether or not the tumor has spread through the cerebrospinal fluid (CSF) to other parts of the brain and/or spinal cord
- Whether or not tumor cells have spread beyond the central nervous system

Medulloblastoma risk groups

More formal staging systems have been proposed for some childhood brain tumors. For example, many clinical trials for treating medulloblastoma use a system that places children into either standard-risk or high-risk groups. Children are placed in the high-risk group if they are younger than 3, have a lot of tumor that can't be removed during surgery, and/or have cancer cells in the CSF or spread to other parts of the brain or elsewhere. Doctors are still refining this system to make it as accurate as possible.

How are brain and spinal cord tumors in children treated?

This information represents the views of the doctors and nurses serving on the American Cancer Society's Cancer Information Database Editorial Board. These views are based on their interpretation of studies published in medical journals, as well as their own professional experience.

The treatment information in this document is not official policy of the Society and is not intended as medical advice to replace the expertise and judgment of your cancer care team. It is intended to help you and your family make informed decisions, together with your doctor.

Your doctor may have reasons for suggesting a treatment plan different from these general treatment options. Don't hesitate to ask him or her questions about your treatment options.

The next few sections describe the various types of treatments used for brain tumors in children. This is followed by a description of the most common treatment approaches for these tumors based on the type of tumor.

General comments about treatment

Children and teens with brain and spinal cord tumors and their families have special needs. These needs can be met best by cancer centers for children and teens, working closely with the child's primary care doctor. Treatment in these centers takes advantage of teams of specialists who know the differences between cancers in adults and those in children and teens, as well as the unique needs of younger people with cancer.

For childhood brain and spinal cord tumors, this team is often led by a pediatric neurosurgeon, a doctor who uses surgery to treat brain and nervous system tumors in children. Other doctors on the team may include:

- Pediatric neurologist: a doctor who treats brain and nervous system diseases in children
- Radiation oncologist: a doctor who uses radiation to treat cancer
- Pediatric oncologist: a doctor who uses chemotherapy and other medicines to treat children's cancers
- Endocrinologist: a doctor who treats diseases in glands that secrete hormones

Many other specialists may be involved in your child's care as well, including nurse practitioners, nurses, psychologists, social workers, rehabilitation specialists, and other health professionals.

The main treatments for children with central nervous system tumors are:

- Surgery
- Radiation therapy
- Chemotherapy

In many cases children will get some combination of these. Treatment is based on the type of tumor and other factors. Doctors plan each child's treatment individually to give them the best chance of cure. The possible long-term side effects of treatment are also considered.

Surgery

Surgery to remove the tumor

In most cases, the first step in brain tumor treatment is an operation called a craniotomy to remove as much of the tumor as possible. Surgery alone or combined with radiation therapy may cure many tumors, including some low-grade astrocytomas, pleomorphic xanthoastrocytomas, dysembryoplastic neuroepithelial tumors, ependymomas, craniopharyngiomas, gangliogliomas, and meningiomas.

Children with infiltrating tumors (tumors that grow into surrounding areas), such as anaplastic astrocytomas or glioblastomas, cannot be cured by surgery alone. But surgery can reduce the amount of tumor that needs to be treated by radiation or chemotherapy, which might improve the results of these treatments.

Surgery may also improve some of the symptoms caused by brain tumors, particularly those caused by increased pressure within the skull. These can include headaches, nausea, vomiting, and blurred vision. Surgery may also make seizures easier to control with medicines.

Surgery may not be a good option in some cases, such as if the tumor is too deep within the brain, or if it is in a part of the brain that can't be removed, such as the brain stem. If this is the case, other treatments may be used instead.

Craniotomy: A craniotomy is a surgical opening made in the skull. This is the most common surgery to remove a brain tumor. For this operation, the child may either be put under general anesthesia (in a deep sleep) or may remain awake (with the surgical area numbed) if brain function needs to be assessed during the operation.

The neurosurgeon makes an incision in the scalp and then removes a piece of bone from the skull to expose the area of brain over the tumor. Many devices can help the surgeon see the tumor and surrounding brain tissue. The surgeon often operates while looking at

the brain through a microscope. In some cases, imaging tests such as MRI, CT, or ultrasound can be used to help locate tumors buried deep in the brain.

Once the tumor is located, the surgeon removes as much as is safely possible. This can be done in several ways depending on how hard or soft the tumor is, and whether it contains many or just a few blood vessels. One way is to cut it out with a scalpel or special scissors. In other cases, a probe attached to an ultrasonic generator may be placed into the tumor to break it up and liquefy it. A small vacuum device is then used to suck it out. The surgeon is very careful to avoid damaging normal brain tissue as much as possible. After removing the tumor, the surgeon replaces the bone and closes the incision.

After the operation, the child may have a drain coming out of the incision that allows excess cerebrospinal fluid (CSF) to leave the skull. This drain is usually removed after a few days. Recovery time in the hospital is usually 4 to 6 days, but this may vary according to the size and location of the tumor and whether other treatments are given.

Surgery to place a shunt or ventricular access catheter

If the tumor blocks the CSF flow it can cause increased pressure inside the skull. This can cause symptoms such as headaches, nausea, vomiting, and blurred vision, and may even permanently damage the brain.

To drain away excess CSF and lower the pressure, the neurosurgeon may put in a silicone tube called a *shunt* (sometimes referred to as a *ventriculoperitoneal* or *VP shunt*). One end of the shunt is placed in a ventricle of the brain (an area filled with CSF) and the other end is placed in the abdomen or, less often, the heart. The tube runs under the skin of the neck and chest. The flow of CSF is controlled by a valve placed along the tubing. Shunts may be temporary or permanent. They may be placed before or after the surgery to remove the tumor.

Surgery may also be used to insert a ventricular access catheter, such as an Ommaya reservoir, to help deliver chemotherapy directly to the brain or spinal cord. A small incision is made in the scalp, and a small hole is drilled in the skull. A flexible tube is then threaded through the hole until the open end of the tube is in a ventricle, where it reaches the CSF. The other end, which has a dome-shaped reservoir, remains just under the scalp. After the operation, doctors and nurses can use a thin needle to give chemotherapy drugs through the reservoir or to remove CSF from the ventricle for testing.

Possible risks and side effects of surgery

Surgery on the brain or spinal cord is a serious operation, and surgeons are very careful to try to limit any problems either during or after surgery. Complications during surgery such as bleeding or reactions to anesthesia are rare, but they can happen.

Swelling in the brain is a major concern after surgery. Drugs called corticosteroids are typically given for several days after surgery to help lessen this risk.

One of the biggest concerns when removing brain tumors is the possible loss of brain function afterward, which is why doctors are very careful to remove only as much tissue as is necessary.

For more extensive information on surgery as a treatment for cancer, see our separate document, *Surgery*.

Radiation therapy

Radiation therapy uses high-energy x-rays or subatomic particles to kill cancer cells. This type of treatment is given by a doctor called a *radiation oncologist*. Radiation therapy may be used in different situations:

- After surgery to try to kill any remaining cancer cells
- As part of the main treatment if surgery is not a good option
- To help prevent or relieve symptoms, especially for some spinal cord tumors

Children younger than 3 years, however, are usually not given radiation because of potential long-term side effects with brain development. Instead, their treatment relies mainly on surgery and chemotherapy. Radiation treatment can also cause some problems in older children. Radiation oncologists try very hard to deliver high doses of radiation to the tumor while limiting the radiation to normal surrounding brain areas as much as possible.

Types of radiation therapy

In most cases, the radiation is focused precisely on the tumor from a source outside the body. This is called *external beam radiation therapy (EBRT).*

External beam radiation therapy is usually given daily (Monday thru Friday) over many weeks. Before your child's treatments start, the radiation team will take careful measurements to determine the correct angles for aiming the radiation beams and the proper dose of radiation. Each treatment is much like getting an x-ray, but the dose of radiation is much higher. It is not painful. For each session, your child lies on a special table while a machine delivers the radiation from a precise angle. Some younger children may need to be sedated to make sure they don't move during the treatment. Each session lasts about 15 to 30 minutes, but most of the time is spent making sure the radiation is aimed correctly. The actual treatment time each day is much shorter.

Because high doses of radiation therapy can damage normal brain tissue, doctors try to deliver high doses of radiation to the tumor with the lowest possible dose to normal surrounding brain areas. Several newer techniques help doctors focus the radiation more precisely:

Three-dimensional conformal radiation therapy (3D-CRT): 3D-CRT uses the results of imaging tests such as MRI and special computers to precisely map the location of the tumor. Several radiation beams are then shaped and aimed at the tumor from different

directions. Each beam alone is fairly weak, which makes it less likely to damage normal tissues, but the beams converge at the tumor to give a higher dose of radiation there. Your child may be fitted with a plastic mold resembling a body cast to keep him or her in the same position so that the radiation can be aimed more accurately.

Intensity modulated radiation therapy (IMRT): IMRT is an advanced form of 3D therapy. In addition to shaping the beams and aiming them at the tumor from several angles, the intensity (strength) of the beams can be adjusted to limit the dose reaching the most sensitive normal tissues. This may allow the doctor to deliver a higher dose to the tumor. Many major hospitals and cancer centers now use IMRT.

Conformal proton beam radiation therapy: Proton beam therapy is related to 3D-CRT and uses a similar approach. But instead of using x-rays, it focuses proton beams on the cancer. Protons are positive parts of atoms. Unlike x-rays, which release energy both before and after they hit their target, protons cause little damage to tissues they pass through and then release their energy after traveling a certain distance. Doctors can use this property to deliver more radiation to the tumor and do less damage to nearby normal tissues. The machines needed to make protons are expensive, and there are only a handful of them being used in the United States at this time.

Stereotactic radiosurgery/stereotactic radiotherapy: This type of radiation treatment delivers a large, precise radiation dose to the tumor area in a single session (radiosurgery) or in a few sessions (radiotherapy). (There is no actual surgery involved in this treatment.) It may be useful for some tumors in parts of the brain or spinal cord that can't be treated with surgery or when a child's health does not permit surgery.

First, a head frame is attached to the skull to help precisely aim the radiation beams. Once the exact location of the tumor is known from CT or MRI scans, radiation from a machine can be focused at the tumor from many different angles. This can be done in 2 ways.

In one approach, radiation beams are focused at the tumor from hundreds of different angles for a short period of time. An example of such a machine is the Gamma Knife.

Another approach uses a movable linear accelerator (a machine that creates radiation) that is controlled by a computer. Instead of delivering many beams at once, this machine moves around the head to deliver radiation to the tumor from different angles. Several machines with names such as X-Knife, CyberKnife, and Clinac are used in this way for stereotactic radiosurgery.

Stereotactic radiosurgery typically delivers the whole radiation dose in a single session, though it may be repeated if needed. Sometimes doctors give the radiation in several treatments to deliver the same or a slightly higher dose. This is called *fractionated radiosurgery* or *stereotactic radiotherapy*.

Brachytherapy (interstitial radiotherapy): Unlike the external radiation approaches above, brachytherapy involves inserting radioactive material directly into or near the tumor. The radiation given off travels a very short distance, so it affects only the tumor. This technique is most often used along with external radiation. It provides a high dose of

radiation at the tumor site, while the external radiation treats nearby areas with a lower dose.

Whole brain and spinal cord radiation therapy (craniospinal radiation): If tests such as an MRI scan or lumbar puncture find the tumor has spread along the spinal cord covering, the meninges, or into the surrounding fluid, then radiation may be given to the whole brain and spinal cord. Some tumors such as ependymomas and medulloblastomas are more likely to spread this way, and therefore may require craniospinal radiation.

Effectiveness of radiation therapy

Up to one half of all medulloblastomas and virtually all germinomas are cured by radiation therapy. Unfortunately, radiation does not cure most other brain tumors. If there is a small amount of the tumor remaining after surgery, astrocytomas, oligodendrogliomas, and ependymomas can sometimes be controlled by radiation therapy, but usually are not cured.

Possible effects of radiation therapy

Radiation is more harmful to tumor cells than it is to normal cells. Still, normal brain tissue is also damaged by radiation, especially in younger children.

During the course of radiation therapy, some children may become irritable and tired. Nausea, vomiting, and headaches are possible but are uncommon. Spinal radiation can cause nausea and vomiting more often than brain radiation. Sometimes dexamethasone (Decadron), a cortisone-like drug, can help relieve these symptoms.

Some weeks after radiation therapy, children may become drowsy or have other nervous system symptoms. This is called the *radiation somnolence syndrome* or *early-delayed radiation effect*. It usually passes after a few weeks.

Children may lose some brain function if large areas of the brain get radiation. Problems can include memory loss, personality changes, and trouble learning at school. These may get better over time, but some effects may be long-lasting. Other symptoms could include seizures and slowed growth. There may also be other symptoms depending on the area of the brain treated and how much radiation was given. The risk of these effects must be balanced against the risks of not using radiation and having less control of the tumor. If problems are seen after treatment, it is often hard to determine whether they were caused by damage from the tumor itself, from surgery or radiation therapy, or from some combination of these. Doctors are constantly testing lower doses or different ways of giving radiation to see if they can be as effective without causing as many problems.

Normal brain cells grow quickly in the first several years of life, making them very sensitive to radiation. Because of this, radiation therapy is often not used or is postponed in children younger than 3 years old to avoid damage that might affect brain development. This needs to be balanced with the risk of tumor regrowth, as early radiation therapy may be lifesaving in some cases. It is important that you talk with your child's doctor about the risks and benefits of treatment.

Rarely (in less than 5% of patients) after radiation therapy, a large mass of dead (necrotic) tissue forms at the site of the tumor. This occurs months to many years after radiation is given and is called *radiation necrosis*. In most cases the dead tissue includes both cancerous and non-cancerous cells. Occasionally, surgery is needed to remove the necrotic tissue.

Radiation can damage genes in normal cells. As a result, there is a small risk of developing a second cancer in the area that got the radiation – for example, a meningioma of the coverings of the brain, or less likely a bone cancer in the skull. If this does occur, it is usually many years after the radiation is given. This small risk should not keep children who need radiation from getting treatment. It's important to continue close follow-up with your child's doctor so that if problems do come up they can be found and treated as early as possible.

For more information on radiation therapy, see our separate document *Understanding Radiation Therapy: A Guide for Patients and Families*.

Chemotherapy

Chemotherapy uses anti-cancer drugs that are usually given into a vein (IV) or taken by mouth. These drugs enter the bloodstream and reach almost all areas of the body. However, many chemotherapy drugs are not able to enter the brain and reach tumor cells.

For some brain tumors, the drugs may be given directly into the cerebrospinal fluid (CSF) in the brain or into the spinal canal below the spinal cord. To help with this, a thin tube, known as a ventricular access catheter, may be inserted through a small hole in the skull and into a ventricle during a minor operation (see the "Surgery" section).

In general, chemotherapy is used for higher grade tumors. Some types of brain tumors, such as medulloblastoma, tend to respond well to chemotherapy.

Chemotherapy may be given before or after surgery and radiation therapy. It may be used instead of radiation therapy in children 3 years and younger.

Some of the chemotherapy drugs used to treat children with brain tumors include:

- Carboplatin
- Carmustine (BCNU)
- Cisplatin
- Cyclophosphamide
- Etoposide
- Lomustine (CCNU)
- Methotrexate
- Temozolomide

- Thiotepa
- Vincristine

These drugs may be used alone or in various combinations, depending on the type of brain tumor. Chemotherapy is given in cycles. Each cycle generally lasts about 3 to 4 weeks and is followed by a rest period to give the body time to recover.

Possible side effects of chemotherapy

Chemotherapy drugs attack cells that are dividing quickly, which is why they often work against cancer cells. But other cells in the body, such as those in the bone marrow, the lining of the mouth and intestines, and the hair follicles, also divide quickly. These cells are also likely to be affected by chemotherapy, which can lead to side effects.

The side effects of chemotherapy depend on the type of drugs, the amount taken, and how long they are taken. Possible side effects can include:

- Hair loss
- Mouth sores
- Loss of appetite
- Nausea and vomiting
- Increased chance of infections (from low white blood cell counts)
- Easy bruising or bleeding (from low blood platelet counts)
- Fatigue (from low red blood cell counts)

These side effects are usually short-term and go away once treatment is finished. Your child's doctor and treating team will watch closely for any side effects that develop. There are often ways to lessen these side effects. For example, drugs can be given to help prevent or reduce nausea and vomiting.

Along with the risks above, some chemotherapy drugs may have specific side effects (although these are fairly uncommon). For example, cisplatin and carboplatin can also cause kidney damage and hearing loss. Your doctor will check your child's kidney function and hearing periodically if he or she is given these drugs.

Be sure to ask your doctor or nurse about medicines to help reduce side effects, and let him or her know if your child has side effects so they can be managed effectively. In some cases, the doses of the chemotherapy drugs may need to be reduced or treatment may need to be delayed or stopped to prevent the effects from getting worse.

For more information on chemotherapy, see our separate document, *Understanding Chemotherapy: A Guide for Patients and Families*.

Other drug treatments

Other drugs may be used to help control symptoms or other effects caused by tumors, but they don't treat the tumors directly.

Corticosteroids: Cortisone-like drugs such as dexamethasone (Decadron) are often given to reduce the swelling that may occur around brain tumors. This may help relieve headaches and other symptoms.

Anti-seizure drugs (anti-epileptics): Drugs may also be given to lower the chance of seizures, which may happen in people with brain tumors. Many anti-seizure drugs are available. One of the more commonly used drugs is called phenytoin (Dilantin).

Hormones: If the pituitary gland has been damaged by the spread of the tumor or by treatments (surgery or radiation therapy), your child may need to take pituitary hormones to replace those missing.

Clinical trials

You may have had to make a lot of decisions since you've been told your child has cancer. One of the most important decisions you will make is deciding which treatment is best. You may have heard about clinical trials being done for this type of cancer. Or maybe someone on your health care team has mentioned a clinical trial to you.

Clinical trials are carefully controlled research studies that are done with patients who volunteer for them. These studies are done to get a closer look at promising new treatments or procedures.

If you would like your child to take part in a clinical trial, you should start by asking your doctor if your clinic or hospital conducts clinical trials. You can also call our clinical trials matching service for a list of clinical trials that meet your medical needs. You can reach this service at 1-800-303-5691 or on our Web site at www.cancer.org/clinicaltrials. You can also get a list of current clinical trials by calling the National Cancer Institute Cancer Information Service toll free at 1-800-4-CANCER (1-800-422-6237) or by visiting the NCI clinical trials Web site at www.cancer.gov.

Your child will have to meet certain requirements to take part in any clinical trial. If your infant or young child does qualify for a clinical trial, you will have to decide whether or not to enter (enroll) the child into it. Older children, who can understand more, usually must also agree to take part in the clinical trial before the parents' consent is accepted.

Clinical trials are one way to get state-of-the-art cancer care for your child. They are the only way for doctors to learn better methods to treat cancer. Still, they are not right for every child.

You can get a lot more information on clinical trials in our document called *Clinical Trials: What You Need to Know*. You can read it on our Web site or call our toll-free number (1-800-227-2345) and have it sent to you.

Complementary and alternative therapies

When your child has cancer you are likely to hear about ways to treat his or her cancer or relieve symptoms that your doctor hasn't mentioned. Everyone from friends and family to Internet groups and Web sites offer ideas for what might help. These methods can include vitamins, herbs, and special diets, or other methods such as acupuncture or massage, to name a few.

What exactly are complementary and alternative therapies?

Not everyone uses these terms the same way, and they are used to refer to many different methods, so it can be confusing. We use *complementary* to refer to treatments that are used *along with* your regular medical care. *Alternative* treatments are used *instead of* a doctor's medical treatment.

Complementary methods: Most complementary treatment methods are not offered as cures for cancer. Mainly, they are used to help the person with cancer feel better. Some methods that are used along with regular treatment are: art therapy or play therapy to reduce stress; acupuncture to help relieve pain; or peppermint tea to relieve nausea. Some complementary methods are known to help, while others have not been tested. Some have been proven not be helpful, and a few have even been found harmful.

Alternative treatments: Alternative treatments may be offered as cancer cures. These treatments have not been proven safe and effective in clinical trials. Some of these methods may pose danger, or have life-threatening side effects. But the biggest danger in most cases is that your child may lose the chance to be helped by standard medical treatment. Delays or interruptions in medical treatments may give the cancer more time to grow and make it less likely that treatment will help.

Finding out more

It is easy to see why parents who have children with cancer think about alternative methods. You want to do all you can to help fight the cancer, and the idea of a treatment with few or no side effects sounds great. Sometimes medical treatments like chemotherapy can be hard to take, or they may no longer be working. But the truth is that most of these alternative methods have not been tested and proven to work in treating cancer.

As you consider your child's options, here are 3 important steps you can take:

- Look for "red flags" that suggest fraud. Does the method promise to cure all or most cancers? Are you told not to use regular medical treatments? Is the treatment a "secret" that requires you to take your child to certain providers or to another country?
- Talk to your child's doctor or nurse about any method you are thinking about.

- Contact us at 1-800-227-2345 to learn more about complementary and alternative methods in general and to find out about the specific methods you are looking at.

The choice is yours

You always have a say in how your child is treated. If you want to use a non-standard treatment, learn all you can about the method and talk to your child's doctor about it. With good information and the support of your child's health care team, you may be able to safely use the methods that can help your child while avoiding those that could be harmful.

Treatment of specific types of brain and spinal cord tumors

The treatment options for brain and spinal cord tumors depend on several factors, including the type of tumor and how far it has grown or spread.

Non-infiltrating astrocytomas (Juvenile pilocytic astrocytomas, subependymal giant cell astrocytomas)

Many doctors consider these benign tumors because they tend to grow very slowly and do not grow into (infiltrate) nearby tissues. Juvenile pilocytic astrocytomas occur most often in the cerebellum in young children, while subependymal giant cell astrocytomas grow in the ventricles and are almost always seen in children with tuberous sclerosis.

In most cases, these astrocytomas can be cured by surgery alone. Radiation therapy may be given if the tumor is not completely removed, although many doctors will wait until there are signs the tumor is growing back before considering it. Even then, another operation to remove the remaining tumor may be the first option. The outlook is not as good if the astrocytoma occurs in a place that does not allow it to be removed surgically, such as the hypothalamus or brain stem.

For subependymal giant cell astrocytomas that can't be removed completely by surgery, treatment with a newer drug called everolimus (Afinitor) may shrink the tumor or slow its growth for some time. This drug is taken daily as a pill. Common side effects can include mouth sores, fever, respiratory infections, and lowered blood cell counts.

Low-grade astrocytomas

The initial treatment for these tumors is surgery when possible. Because these tumors often grow into nearby normal brain tissue, they may be hard to cure with just surgery. Usually the surgeon will try to remove as much of the tumor as safely possible. If the surgeon can remove it all this may be curative.

Radiation therapy may be given after surgery, especially if a lot of tumor remains. Otherwise, it may be postponed until the tumor starts to regrow. (Sometimes, a second surgery may be tried before giving radiation.) Radiation may also be used as the main treatment if surgery is not a good option because of where the tumor is located.

For children younger than 3, if the tumor cannot be completely removed or if it grows back, chemotherapy may be used until they are older. They may then be treated with radiation.

Intermediate- and high-grade astrocytomas (Anaplastic astrocytomas, glioblastomas)

Although surgery is often the first treatment, these infiltrating astrocytomas are not curable by surgery. After as much of the tumor as possible is removed, radiation therapy is given, often followed by chemotherapy. For children younger than 3, radiation may be postponed until they are older. Surgery may be repeated in some cases if the tumor comes back after the initial treatment. Because these tumors are hard to cure with current treatments, clinical trials of promising new treatments may be a good option.

Oligodendrogliomas

If possible, surgery is the first option for these infiltrating tumors. Although they are usually not curable by surgery, it can relieve symptoms and prolong survival. Many of these tumors grow slowly, and surgery may be repeated if it grows back in the same spot. Chemotherapy and/or radiation therapy may be given after surgery.

If surgery is not an option, chemotherapy, with or without radiation therapy, may be helpful. These tumors may respond to chemotherapy better than other brain tumors if certain chromosome changes are in the tumor cells. You can ask your child's doctor about testing for these changes.

Ependymomas and anaplastic ependymomas

These tumors do not infiltrate normal brain tissue as extensively as astrocytomas and may sometimes be cured by surgery if the entire tumor can be removed. If some of the tumor is left behind, a second operation may be done in some cases (often after a short course of chemotherapy). Radiation therapy is recommended after surgery in most patients to try to prevent recurrence, even if it appears that all of the tumor has been removed.

The use of chemotherapy after surgery is still being tested in clinical trials. It may be recommended, but its benefit is still uncertain. It may be more helpful for anaplastic ependymomas. Very young children may be given chemotherapy to avoid or delay the use of radiation.

Sometimes the tumor cells can spread into the cerebrospinal fluid (CSF). The doctor may test the CSF for cancer cells by doing a lumbar puncture (spinal tap). Radiation may be extended to include the entire brain and spinal cord if tumor cells are found in the CSF or growing on the surface the nervous system.

Optic gliomas

These tumors invade the nerves leading to the eye. They are often hard to operate on because these nerves are very sensitive and may be harmed by surgery. Depending on where the tumor is, removing it could lead to loss of vision in one or both eyes. This is why surgery has to be considered carefully. Sometimes surgery might not be needed, because these tumors can grow very slowly.

If treatment is needed and the tumor can be completely removed without the loss of vision or other serious problems, surgery is preferred. If the patient cannot be cured with surgery, radiation therapy to this area is the preferred treatment even though it can also affect a child's vision. Often, chemotherapy may be given to younger children instead of radiation.

Brain stem gliomas

Most of these tumors are astrocytomas, although a small number are ependymomas or other tumors. These tumors usually have a characteristic appearance on MRI, so the diagnosis can often be made without surgery or a biopsy.

A small number of brain stem gliomas occur as a small tumor with very distinct edges (called a focal brain stem glioma). These tumors can often be treated successfully with surgery. In cases where surgery can't be done or doesn't completely remove the tumor, radiation therapy may be effective in slowing its growth.

Most brain stem gliomas grow throughout the brain stem, which is vital to life and can't be removed. Surgery in these cases would most likely do more harm than good, so it is usually not attempted. These brain stem gliomas are usually treated with radiation therapy. Chemotherapy is sometimes added, although it's not clear how helpful it might be. These tumors are very hard to control, and they tend to have a poor prognosis. Certain children, namely those with neurofibromatosis type I, may have a better outlook.

Primitive neuroectodermal tumors (including medulloblastoma and pineoblastoma)

Primitive neuroectodermal tumors (PNETs) are all treated in similar ways, but medulloblastomas tend to have a better outlook than other types of PNETs.

Medulloblastomas: These tumors start in the cerebellum. They tend to grow quickly and are among those most likely to spread outside of the brain (usually to the bones or the bone marrow). But they also tend to respond well to treatment. Children in the high-risk group are usually given more intensive treatment than children in the average-risk group (see "How are brain and spinal cord tumors in children staged?").

Medulloblastomas are treated with surgery followed by radiation therapy to the area where they started. High doses of radiation are aimed at the area of the tumor. Because these tumors tend to spread to the cerebrospinal fluid (CSF), lower doses of radiation may be given to the whole brain and the spinal cord (craniospinal radiation), as well.

Chemotherapy is usually given after radiation therapy, and may allow doctors to use lower doses of radiation in some cases. But if the tumor has spread through the CSF, standard doses of radiation will be needed.

For children younger than 3, doctors try to use as little radiation as possible. Chemotherapy is typically the first treatment given after surgery. Depending on how the tumor responds, the chemotherapy may or may not be followed by radiation therapy.

Pineoblastomas and other PNETs: These tumors also tend to grow quickly, and they are generally harder to treat than medulloblastomas. Surgery is the main treatment for these tumors, but they are usually hard to remove completely. Still, surgery can relieve symptoms and may help other treatments to be more effective. Children 3 or older are given radiation therapy after surgery. Because these tumors tend to spread to cerebrospinal fluid (CSF), radiation therapy is often given to the whole brain and the spinal cord (craniospinal radiation).

Chemotherapy may be given with radiation therapy so that a lower dose of radiation can be used. But if the tumor has spread to the CSF, standard doses of radiation will be required. Chemotherapy is also used to treat tumor recurrence.

For children younger than 3 years, doctors try to use as little radiation as possible. Chemotherapy is typically the first treatment given after surgery. Some studies have achieved very good results using chemotherapy in young children. Depending on how the tumor responds, the chemotherapy may or may not be followed by radiation therapy.

There are some reports that giving high-dose chemotherapy followed by an autologous stem cell transplant may be an effective treatment for children with medulloblastomas or other PNETs. Several clinical trials are now studying this. For more information on stem cell transplants, see our separate document, *Bone Marrow and Peripheral Blood Stem Cell Transplant*.

Meningiomas

Surgery is the main treatment for these tumors. Children are usually cured if the surgery completely removes the tumor. Some tumors, particularly those at the base of the brain, cannot be removed completely, and some are malignant and come back in spite of apparent complete removal. Radiation therapy may control the growth of meningiomas that cannot be removed completely or those that come back after surgery. Chemotherapy may be tried if surgery and radiation aren't effective, but it is not helpful in many cases.

Schwannomas (including acoustic neuromas)

These slow-growing tumors are usually benign and are effectively cured by surgical removal. In some centers, small vestibular schwannomas (also known as acoustic neuromas) are treated by stereotactic radiosurgery (see the section, "Radiation therapy"). For the rare malignant schwannoma, radiation therapy is often given after surgery.

Spinal cord tumors

These tumors are usually treated similarly to those of the same type in the brain. Astrocytomas of the spinal cord usually cannot be completely removed. They may be treated with surgery to remove as much tumor as possible, followed by radiation therapy, or with radiation therapy alone. Chemotherapy may be used in younger children or if the tumor appears to be aggressive.

Meningiomas near the spinal cord are often cured by surgical removal. Some ependymomas can be cured by surgery as well. If an ependymoma cannot be completely removed, radiation therapy will follow surgery.

Choroid plexus tumors

Benign choroid plexus papillomas are usually cured just with surgery. Choroid plexus carcinomas are malignant tumors that are only sometimes cured by surgery. After surgery, these carcinomas are usually treated with radiation and/or chemotherapy.

Craniopharyngiomas

Craniopharyngiomas can be treated by surgically removing most of the tumor (debulking) followed by radiation in most cases. Partial surgical removal followed by very focused radiation therapy may cause fewer severe side effects than complete removal.

Germ cell tumors

The most common germ cell tumor, germinoma, is usually cured by radiation therapy alone. Chemotherapy may be added if the tumor is very large or if radiation doesn't destroy it completely. In very young children, chemotherapy may be used instead of radiation therapy. If other types of germ cell tumors are present, either mixed or not mixed with germinoma, the outlook is usually not as good.

Other types of germ cell tumors (such as teratomas and yolk sac tumors) are usually treated with both radiation therapy and chemotherapy. Sometimes these tumors spread to the cerebrospinal fluid (CSF), and radiation therapy to the spinal cord and brain is needed.

Survival rates for selected brain and spinal cord tumors

Survival rates are often used by doctors as a standard way of discussing a person's prognosis (outlook). Some parents may want to know the survival statistics for children in similar situations, while others may not find the numbers helpful, or may even not want to know them. Whether or not you want to read about the survival statistics below for brain and spinal cord tumors is up to you.

The 5-year survival rate refers to the percentage of children who live *at least* 5 years after their cancer is diagnosed. Of course, many children live much longer than 5 years (and some are cured).

In order to get 5-year survival rates, doctors have to look at children who were treated at least 5 years ago. Improvements in treatment since then may result in a more favorable outlook for children now being diagnosed with brain tumors.

Survival rates are often based on previous outcomes of large numbers of children who had the disease, but they cannot predict what will happen in any particular child's case. Knowing the type of a child's brain tumor is important in estimating their outlook. But many other factors may also affect a child's outlook, such as the location and extent of the tumor and how well it responds to treatment. Even when taking these other factors into account, survival rates are at best rough estimates. Your child's doctor can tell you if the numbers below may apply, as he or she is familiar with the aspects of your child's particular situation.

The numbers below come from the Central Brain Tumor Registry of the United States (CBTRUS) and are based on children aged 19 or younger who were treated between 1995 and 2006. There are some important points to note about these numbers:

- These numbers are for some of the more common types of tumors. Numbers are not readily available for all types of tumors that occur in children, often because they are rare or are hard to classify.
- In some cases, the numbers include a wide range of different types of tumors that may have different outlooks. For example, the numbers for PNETs include medulloblastomas, which tend to have a better outlook than pineoblastomas or PNETs in other parts of the brain.

Type of Tumor	**5-Year Survival Rate**
Juvenile pilocytic astrocytoma/subependymal giant cell astrocytoma	About 95%
Low-grade astrocytoma	About 85%
Anaplastic astrocytoma	About 30%
Glioblastoma	About 20%
Oligodendroglioma	About 95%
Ependymoma/anaplastic ependymoma	About 70%
PNETs (includes medulloblastoma and pineoblastoma)	About 60%

More treatment information

For more details on treatment options – including some that may not be addressed in this document – the National Cancer Institute (NCI) and CureSearch are good sources of information.

The NCI provides treatment guidelines via its telephone information center (1-800-4-CANCER) and its Web site (www.cancer.gov). Detailed guidelines intended for use by cancer care professionals are also available on www.cancer.gov.

CureSearch is a combined effort of the National Childhood Cancer Foundation and the Children's Oncology Group (COG). CureSearch can be contacted via telephone at 1-800-458-6223 or on the Web at www.curesearch.org.

What should you ask your doctor about your child's brain or spinal cord tumor?

It is important for you to have frank, open discussions with your child's cancer care team. They want to answer all of your questions, no matter how minor you might think they are. Here are some questions to consider:

- What kind of tumor does my child have?
- Where is the tumor located, and how far has it spread?
- Are there other tests that need to be done before we can decide on treatment?
- How much experience do you have treating this type of tumor?
- What treatment options do we have? What do you recommend? Why?
- What are the possible risks and side effects of treatment?
- How might treatment affect my child's ability to learn, grow, and develop?
- Will treatment affect my child's future ability to have children?
- What should we do to be ready for treatment?
- How long will treatment last? What will it involve? Where will it be done?
- How will treatment affect our daily activities?
- Based on what you've learned about my child's tumor, what is the expected prognosis (outlook)?
- What will we do if the treatment doesn't work or if the cancer recurs?
- What type of follow-up will my child need after treatment?

- Are there nearby support groups or other families who have been through this that we could talk to?

Along with these sample questions, be sure to write down any others you might want to ask. For instance, you might want information about recovery times so you can plan your work and your child's school and activity schedule. Or you may want to ask about second opinions concerning the diagnosis and treatment options, or about clinical trials for which your child may qualify.

What happens after treatment for brain and spinal cord tumors in children?

After treatment, the main concerns for most families are the immediate and long-term effects of the tumor and its treatment, and concerns about the tumor still being present or coming back.

It is certainly normal to want to put the tumor and its treatment behind you, and to get back to a life that doesn't center around cancer. But it's important to realize that follow-up care is a central part of this process that offers your child the best chance for recovery and long-term survival.

Looking for tumor progression or recurrence

In some cases, even with slow growing tumors, some of the tumor may still be present after treatment. Even with childhood tumors that are treated successfully, it is important to remember that some may come back even many years later. (Your child's doctor should be able to give you an idea of how likely this might be in his or her case.)

Imaging tests (CT or MRI scans) and physical exams will be done after treatment to help determine how successful it was. Whether there is evidence of the tumor still being present or not, your child's cancer care team will probably want to follow your child closely, especially in the first few months and years after treatment to make sure there is no progression or recurrence. Depending on the type and location of the tumor and the extent of the treatment, the team will decide which tests should be done and how often.

During this time, it is important to report any new symptoms to your child's doctor right away, so that the cause can be determined and treated, if need be. Your child's doctor can give you an idea of what to look for. Should further treatment be needed at some point, the doctor will go over potential treatment options with you.

Recovering from the effects of the tumor and its treatment

The possible effects of the tumor and its treatment on physical and mental function can range from very mild to fairly severe. Fortunately, a child's brain is often better able to adjust to changes than an adult's.

Once your child has recovered from treatment, doctors will try to determine the extent of any damage to the brain or other areas. In a very young child this may take time. It is likely that physical exams and imaging tests (CT or MRI scans) will be done after treatment to determine the extent and location of any changes in the brain.

Several types of doctors and other health professionals may be involved in assessing any damage and helping your child recover.

A neurologist (a doctor who specializes in treating the nervous system) may assess your child's physical coordination and muscle strength. If there is muscle weakness or paralysis, your child will be seen by physical and/or occupational therapists and perhaps a physiatrist (a doctor who specializes in rehabilitation) while in the hospital and/or as an outpatient for physical therapy.

If the speech center of the brain is damaged, a speech therapist will help your child improve talking and communicating.

If needed, an ophthalmologist (a doctor who specializes in eye problems) will check your child's vision and an audiologist can check your child's hearing. If problems with vision or hearing are found, your child may require some type of special education.

After surgery, your child may also be seen by a psychiatrist or psychologist to determine the extent of any damage caused by the tumor and surgery. If your child gets radiation therapy and/or chemotherapy, this process may be repeated again after treatment is finished. The doctor will document your child's development in areas such as general intelligence, speech and hearing, memory, and learning skills.

If your child is in school, a teacher working with the hospital (called the school liaison) may become involved. Before going (back) to school, the liaison may help pave the way for your child by talking with the teachers, explaining your child's health issues, and discussing any special education techniques that may be required. In some cases, medicines may be helpful as well. (For more information, see our separate document, *Children Diagnosed With Cancer: Returning to School.*)

If the tumor was in or near the base of the brain (such as a craniopharyngioma) or radiation therapy was given to this area, pituitary hormone production may be affected. Symptoms of pituitary problems can include fatigue, listlessness, poor appetite, cold intolerance, and constipation, which may point to low levels of cortisol and/or thyroid hormone. Other problems can include delayed growth and/or sexual maturation. Sometimes these symptoms may appear even before treatment, as a result of the tumor.

If there is reason to think the pituitary may have been affected, your child may be seen by an endocrinologist (a specialist in hormone disorders). Hormone treatments may be prescribed to restore normal hormone levels. For example, growth hormone can be given to help restore normal growth.

Long-term effects

A major concern of both parents and doctors is the potential for long-term effects from treatment. Some of these, such as learning problems or delayed growth and development, were mentioned above. Others may include effects on the reproductive system or an increased risk of other cancers later in life. While doctors do everything they can to minimize the chance of these complications, in some cases they may be an unavoidable part of ensuring the tumor is treated properly.

To help increase awareness of late effects and improve follow-up care of childhood cancer survivors throughout their lives, the Children's Oncology Group (COG) has developed long-term follow-up guidelines for survivors of childhood cancers. These guidelines, written for doctors and other health care professionals, describe in detail the suggested long-term follow-up care based on the treatments the child has received.

It is very important to discuss possible long-term complications with your child's health care team, and to make sure there is a plan in place to watch for these problems and treat them, if needed. To learn more, ask your child's doctors about the COG survivor guidelines, and see our document, *Childhood Cancer: Late Effects of Cancer Treatment*.

Keeping good medical records

As much as you might want to put the experience behind you once treatment is completed, it is also very important to keep good records of your child's medical care during this time. Eventually, your child will grow up, be on his or her own, and have new doctors. It is important that he or she be able to give the new doctors the details of the cancer diagnosis and treatment. Gathering the details soon after treatment may be easier than trying to get them at some point in the future. There are certain pieces of information that your child's doctors should have, even after the child has become an adult. These include:

- A copy of the pathology report from any biopsies or surgeries.
- If surgery was done, a copy of the operative report(s).
- If there were hospitalizations, a copy of the discharge summaries (forms that doctors prepare when patients leave the hospital).
- A list of the final doses of each chemotherapy drug or other drug your child received.
- If radiation therapy was given, a final summary of the dose and field.

What's new in research and treatment for brain tumors in children?

There is always research going on in the area of brain tumors. Scientists are looking for causes and ways to prevent brain tumors, and doctors are working to improve treatments.

Imaging and surgery techniques

Recent advances have made surgery for brain tumors much safer and more successful. Such techniques include:

- Functional magnetic resonance imaging (fMRI), which can identify the site of important areas of the brain and how close they are to the tumor.
- Image-guided surgery, which allows for safer and more extensive removal of the tumor.

Radiation therapy

Several newer types of radiation therapy now allow doctors to deliver radiation more precisely to the tumor, which helps spare normal brain tissue from getting too much radiation. Newer techniques such as stereotactic radiosurgery, 3-dimensional conformal radiation therapy (3D-CRT), intensity modulated radiation therapy (IMRT), and proton beam therapy are described in the section "Radiation therapy."

The brain is very sensitive to radiation, which can lead to side effects if normal brain tissue receives a large dose, especially if the child is very young. Clinical trials have shown that in some situations, using chemotherapy can let doctors use lower doses of radiation therapy without lowering the chance that treatment will be effective. Doctors are now trying to determine if even lower doses of radiation can be used and still yield the same results.

Chemotherapy

New approaches may help make chemotherapy more useful against brain and spinal cord tumors.

Adjuvant chemotherapy

In some children and infants with brain tumors, chemotherapy is given right after surgery to either delay radiation therapy (particularly in infants) or to decrease the radiation dose needed to treat the tumor. This is known as adjuvant chemotherapy. Some studies are looking at whether giving prolonged chemotherapy may help avoid the need for radiation therapy at all in certain cases.

High-dose chemotherapy and stem cell transplant

One of the main factors that limits the doses of chemotherapy that can be given safely is its effects on the bone marrow, where new blood cells are normally made. A stem cell transplant allows higher doses of chemotherapy to be given than would normally be possible. First, blood stem cells are removed from either the child's blood or the bone marrow. The child is then treated with very high doses of chemotherapy. The blood stem

cells are then infused back into the body, where they settle in the bone marrow and start making new blood cells.

Although some children have had very good responses to this very intensive treatment, it is not yet known if it is effective enough to become standard. For now, most doctors consider this treatment experimental for brain tumors. Clinical trials are under way to determine how useful it is.

Modification of chemotherapy drugs

Many chemotherapy drugs are limited in their effectiveness because the normal tightly controlled openings in the brain capillaries, sometimes referred to as the *blood-brain barrier*, prevents them from getting from the bloodstream to the brain tumor. Researchers are now trying to modify some of these drugs by coating them with a tiny layer of fat (liposomal encapsulation) or attaching them to molecules that normally cross the blood-brain barrier, to help them work better. This is an area of active research and clinical trials.

Getting chemotherapy directly to tumors

Some new approaches may allow doctors to get chemotherapy directly to tumors.

In adults, doctors sometimes place special wafers containing chemotherapy directly at the site of a brain tumor during surgery. The wafers dissolve and give off doses of chemotherapy over several weeks, keeping a high concentration of the drug at that spot while sparing the rest of the body from the possible side effects. Studies are now under way to see if this will work in children as well.

In another method of getting chemotherapy to the tumor, a small tube is placed into a ventricle in the brain through a small hole in the skull during surgery. The tubing extends through the scalp and is connected to an infusion pump, through which drugs can be given. This is referred to as convection enhanced delivery. This may be done for hours or days and may be repeated more than once, depending on the drug used. This is an investigational method at this time, and studies continue.

Other new treatment strategies

Researchers are also testing some newer approaches to treatment that may help doctors target tumors more precisely. In theory, this should allow for more effective treatments that cause fewer side effects. These treatments are all being studied now.

Growth factor inhibitors

Tumor cells are often very sensitive to proteins called growth factors, which cause them to grow and divide. Newer drugs target these growth factors, which may slow the growth of tumor cells or even make them die. One example of such a targeted drug is everolimus (Afinitor), which can be used to shrink or slow the growth of subependymal giant cell

astrocytomas that can't be removed with surgery. Several other targeted drugs are already being used to treat other types of cancer, and some are being studied to see if they will work for brain tumors as well.

Angiogenesis inhibitors

Tumors have to create new blood vessels (a process called angiogenesis) to keep their cells nourished. New drugs that attack these blood vessels are used to help treat some cancers. One such drug, known as bevacizumab (Avastin), is used to treat glioblastomas in adults, although it's not clear if it will help children.

Hypoxic cell sensitizers

Some drugs increase the oxygen content in the tumor, which makes tumor cells more likely to be killed by radiation therapy if they are given before treatment. Studies are now looking to see if this affects the outcome of treatment.

Immunotherapy

The goal of immunotherapy is to make the body's immune system fight the brain tumor more effectively. Vaccines against brain cancer cells are being tested in clinical trials. Immune modulators such as interferon are also being studied.

Therapeutic viruses

Researchers have done a great deal of lab work with viruses that reproduce only within brain tumor cells and then cause those cells to die, while leaving normal cells alone. Research using these viruses in humans with brain tumors is still in the earliest stages.

Although the treatment approaches mentioned above are promising, they are mainly still in the experimental stage at this time, and are generally only available through clinical trials.

Additional resources

More information from your American Cancer Society

The following related information may also be helpful to you. These materials may be ordered from our toll-free number, 1-800-227-2345.

After Diagnosis: A Guide for Patients and Families (also available in Spanish)

Childhood Cancer: Late Effects of Cancer Treatment

Children Diagnosed With Cancer: Dealing with Diagnosis (also available in Spanish)

Children Diagnosed With Cancer: Financial and Insurance Issues

Children Diagnosed With Cancer: Returning to School

Children Diagnosed With Cancer: Understanding the Health Care System (also available in Spanish)

Clinical Trials: What You Need to Know

Family and Medical Leave Act (FMLA)

Fertility and Cancer: What Are My Options?

Nutrition for Children with Cancer (also available in Spanish)

Pediatric Cancer Centers (also available in Spanish)

Surgery (also available in Spanish)

Understanding Chemotherapy: A Guide for Patients and Families (also available in Spanish)

Understanding Radiation Therapy: A Guide for Patients and Families (also available in Spanish)

What Happened to You, Happened to Me (children's booklet)

When Your Brother or Sister Has Cancer (children's booklet)

When Your Child's Treatment Ends: A Guide for Families (booklet)

Books

The following books are available from the American Cancer Society. Call us at 1-800-227-2345 to ask about costs or to place your order.

Because... Someone I Love Has Cancer (kids' activity book)

Caregiving: Step-By-Step Resource for Caring for the Person with Cancer at Home

Jacob Has Cancer: His Friends Want to Help (coloring book for a child with a friend who has cancer)

Let My Colors Out (picture book for young children)

National organizations and Web sites*

In addition to the American Cancer Society, others sources of patient information and support include:

American Brain Tumor Association
Toll-free number: 1-800-886-2282
Web site: www.abta.org

American Childhood Cancer Organization (formerly Candlelighters)
Toll-free number: 1-800-366-2223
Web site: www.candlelighters.org

Cancer Kids
Web site: www.cancerkids.com

Children's Brain Tumor Foundation
Toll-free number: 1-866-228-HOPE (1-866-228-4673)
Web site: www.cbtf.org

CureSearch (National Childhood Cancer Foundation and Children's Oncology Group)
Toll-free number: 1-800-458-6223
Web site: www.curesearch.org

National Cancer Institute
Toll-free number: 1-800-4-CANCER (1-800-422-6237)
Web site: www.cancer.gov

National Children's Cancer Society, Inc.
Toll-free number: 1-800-5-FAMILY (1-800-532-6459)
Web site: www.children-cancer.org

Pediatric Brain Tumor Foundation
Toll-free number: 1-800-253-6530
Web site: www.pbtfus.org

Starlight Children's Foundation
Toll-free number: 1-800-315-2580
Web site: www.starlight.org

**Inclusion on this list does not imply endorsement by the American Cancer Society.*

Other publications*

For adults

100 Questions & Answers About Your Child's Cancer, by William L. Carroll and Jessica Reisman. Jones and Bartlett Publishers, 2004.

Cancer & Self-Help: Bridging the Troubled Waters of Childhood Illness, by Mark A. Chester and Barbara K. Chesney. University of Wisconsin Press, 1995.

Care for Children and Adolescents with Cancer: Questions and Answers. National Cancer Institute. Available at: www.cancer.gov/cancertopics/factsheet/NCI/children-adolescents or call 1-800-332-8615.

Childhood Brain and Spinal Cord Tumors: A Guide for Families, Friends, and Caregivers, by Tania Shiminski-Maher, Patsy McGuire Cullen, and Maria Sansalone. O'Reilly and Associates, 2001.

Childhood Cancer: a Parent's Guide to Solid Tumor Cancers, by Honna Janes-Hodder and Nancy Keene.O'Reilly and Associates, 1999.

Childhood Cancer: A Handbook from St Jude Children's Research Hospital, by Grant Steen and Joseph Mirro (editors). Perseus Publishing, 2000.

Childhood Cancer Survivors: A Practical Guide to Your Future, by Nancy Keene, Wendy Hobbie, and Kathy Ruccione. O'Reilly and Associates, 2000.

Children with Cancer: A Comprehensive Reference Guide for Parents (2nd Ed.), by Jeanne Munn Bracken and Pruden Pruden. Oxford University Press, 2005.

Educating the Child With Cancer: A Guide for Parents and Teachers, edited by Nancy Keene. Candlelighters Childhood Cancer Foundation, 2003.

Living with Childhood Cancer: A Practical Guide to Help Families Cope, by Leigh A. Woznick and Carol D. Goodheart. American Psychological Association, 2002.

Surviving Childhood Cancer: A Guide for Families, by Margo Joan Fromer. Published by New Harbinger Publications, 1998.

When Bad Things Happen to Good People, by Harold Kushner.G.K. Hall, 1982.

When Someone You Love Is Being Treated for Cancer. National Cancer Institute. Available at: www.cancer.gov/cancertopics/when-someone-you-love-is-treated, or call 1-800-332-8615.

Young People with Cancer: A Handbook for Parents. National Cancer Institute, 2003. Available at: www.cancer.gov/cancertopics/youngpeople, or call 1-800-332-8615.

Your Child in the Hospital: A Practical Guide for Parents (2nd Ed.), by Nancy Keene. Published by N. O'Reilly & Associates. 1999. (Also available in Spanish.)

Books for teens and children

Although these books are intended for children, younger kids are helped more when an adult reads with and helps the child reflect about what different parts of the book mean to the child.

The Amazing Hannah, Look at Everything I Can Do! by Amy Klett. Published by Candlelighters Childhood Cancer Foundation, 2002. For ages 1 to 6. (Also available in Spanish.)

Chemo, Craziness and Comfort: My Book about Childhood Cancer, by Nancy Keene. Published by Candlelighters Childhood Cancer Foundation, 2002. Can be ordered from www.candlelighters.org. For ages 6 to 12.

Childhood Cancer Survivors: A Practical Guide to Your Future (2nd Edition), by Kathy Ruccione, Nancy Keene, and Wendy Hobbie. Patient Centered Guides, 2006. For older teens.

Going to the Hospital, by Fred Rogers. Paperstar Book, 1997. For ages 4 to 8.

Life Isn't Always a Day at the Beach: A Book for All Children Whose Lives Are Affected by Cancer, by Pam Ganz. High-Five Publishing, 1996. Workbook for ages 6 to 10.

Little Tree: A Story for Children with Serious Medical Problems, by Joyce C. Mills. Published by Magination Press, 2003. For ages 4 to 8.

Living Well with My Serious Illness, by Marge Heegaard. Fairview Press, 2003. For ages 8 to 12.

Me and My Marrow, by Karen Crowe. Fujsawa Healthcare, 1999.

My Book for Kids with Cansur [sic], by Jason Gaes. Published by Viking Penguin, 1998. For ages 4 to 8.

Oncology, Stupology…I Want to Go Home! by Marilyn K. Hershey. Published by Butterfly Press, 1999. For ages 8 to 12. (Also available in Spanish.)

What About Me? When Brothers and Sisters Get Sick, by Allan Peterkin and Frances Middendorf. Magination Press, 1992. For brothers and sisters (ages 4 to 8) of a child with cancer.

When Someone Has a Very Serious Illness: Children Can Learn to Cope with Loss and Change, by Marge Heegaard. Published by Woodland Press, 1991. For ages 6 to 12.

Why, Charlie Brown, Why? A Story About What Happens When a Friend Is Very Ill, by Charles M. Schultz. Ballantine Publishing Group, 1990. For ages 6 to 12.

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No matter who you are, we can help. Contact us anytime, day or night, for information and support. Call us at **1-800-227-2345** or visit www.cancer.org.

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