

Family Handout for Children with a Brain Tumor

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What do we know about childhood brain tumors?

- Brain tumors represent about 25% of all childhood cancer diagnoses.
- There are approximately 3200 cases of new primary brain tumors diagnosed in patients under 20 years of age each year in the United States.
- Brain tumors are more common in children under age 8 than in older children.
- There are many different types of brain tumors. Brain tumors are named according to the type of cells or the part of the brain in which they begin.
- About half (50%) of brain tumors in children are astrocytomas (including glioblastoma multiforme).
- About 23% of brain tumors in children are medulloblastomas or primitive neuroectodermal tumors.
- About 15% of brain tumors in children are brain stem gliomas.
- About 9% of brain tumors in children are ependymomas.
- About 3% of brain tumors in children are other types.

Only a few risk factors for brain tumors are known.

Genetic conditions

Children with some genetic syndromes are more likely to develop brain tumors than other children. The syndromes are neurofibromatosis, von Hippel-Lindau disease, Li-Fraumeni syndrome, ataxia telangiectasia, basal cell nevus syndrome, and hereditary non-polyposis colon cancer (Gorlin syndrome). Children with these genetic conditions are more at risk for brain tumors, but these account for only a small fraction of cases. These syndromes are usually recognized early in childhood, so it is most likely that you would know if your child has one of these conditions.

Prior radiation

Children who have received radiation therapy to the head as a part of treatment for an earlier cancer are at an increased risk for a new brain tumor.

Male/female

The patterns differ depending on the type of brain tumor. Boys and girls are equally likely to develop an astrocytoma. Boys are more likely to develop a medulloblastoma, ependymoma or germ cell tumor than girls.

Race and ethnicity

White children are more likely than African American children to develop a medulloblastoma or ependymoma. Other types of brain tumors affect white and African American children equally although children in Japan develop more germ cell tumors.

Symptoms of a Brain Tumor in Children

Typical symptoms of Central Nervous System (CNS) tumors are directly related to the location of the tumor, how fast it is growing, and any associated tissue swelling that occurs in conjunction with the tumor.

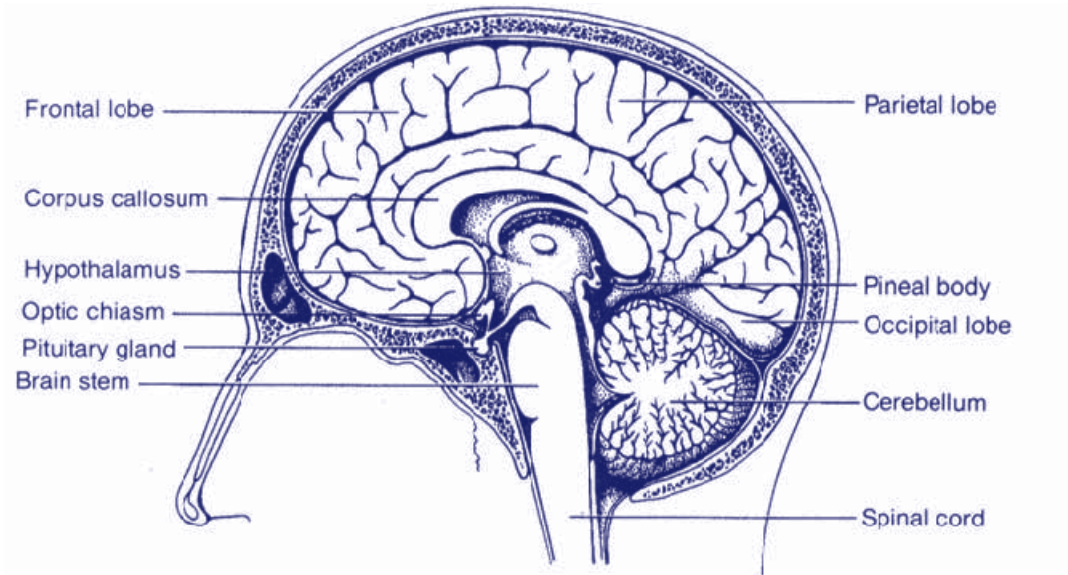
Typical symptoms of CNS tumors include:

- Headaches (frequent, recurrent), especially after waking up in the morning
- Vomiting, especially in the morning
- Eye movement problems and/or vision changes
- Unsteady gait or worsening balance
- Weakness of one side of the face
- Loss of previously acquired developmental milestones
- Weakness in the arms or legs
- Bulging fontanel or increased head size in infants
- Tilting of the head or neck to one side
- Seizures
- Difficulty with speech or swallowing or drooling
- Back or neck pain, often awakening the child at night

Less common symptoms include:

- Changes in eating or thirst
- Growth problems
- Dizziness
- Lethargy, irritability, or other behavior changes
- Deterioration in school performance
- Loss of sensation in the arms or legs
- Loss of consciousness, without history of injury
- Changes in, or loss of control of, bowel or bladder
- Hearing loss, without evidence of infection

Anatomy and Function of the Brain



Cross-Section of Brain

Frontal lobe

- movement
- reasoning
- behavior
- memory
- personality
- planning
- decision making
- judgment
- initiative
- inhibition
- mood

Parietal lobe

- telling right from left
- calculations
- sensations
- reading
- writing

Occipital lobe

- vision

Cerebellum

- balance
- coordination
- fine muscle control

Temporal lobe

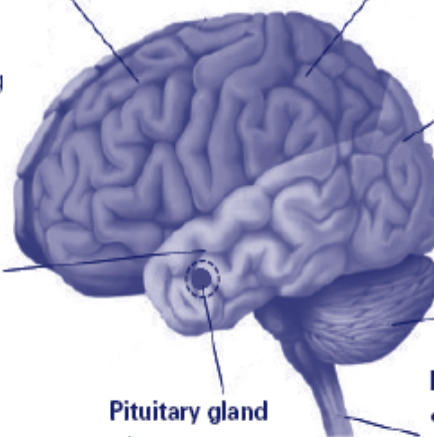
- language comprehension
- behavior
- memory
- hearing
- emotions

Pituitary gland

- hormones
- growth
- fertility

Brain stem

- breathing
- blood pressure
- heartbeat
- swallowing



How Brain Tumors are Diagnosed and Treated

Standard methods used to diagnose these tumors include:

- Computerized Tomography scans (CT Scans)
- Magnetic Resonance Imaging (MRI) scans

Additional procedures often performed during the diagnostic work-up of CNS tumors may include any or all of the following:

- Positron Emission Tomography (PET) scan, similar to CT and MRI, but assesses the metabolic activity of the tumor.
- SPECT scan, similar to an MRI but measures the uptake of certain substances in the brain; is also helpful in differentiating normal brain, tumor and scar tissue following surgery.
- Electroencephalogram (EEG), used to evaluate seizure activity.

Once the location of the tumor is precisely defined, in most cases, surgery must be performed to obtain a portion of the tumor for the pathologist to examine carefully under the microscope. This is necessary to make the correct diagnosis and to plan subsequent treatment. The surgeon will attempt to remove as much of the tumor as possible, without damaging any normal adjacent brain tissue.

After the diagnosis is established, a variety of other tests may be necessary to determine whether the tumor has spread to any other organs in the body, and to evaluate the function of other body systems before starting any subsequent therapy. Such tests may include:

- Lumbar puncture (spinal tap) to evaluate spinal fluid for tumor cells
- Audiogram (hearing test)
- Blood tests of liver and kidney function, salt balance.

Certain important facts are needed to help determine the prognosis and treatment for each type of brain tumor:

- The specific type of tumor, as determined by the pathologist(s)
- The specific area of the brain involved
- The ability to surgically remove the tumor partially or completely
- The growth rate of the tumor
- The age of the child at the time of diagnosis

Questions You Should Ask Your Doctor

Is my tumor benign or malignant?

What is the name and grade of my tumor?

What are the treatment options?

What are the side effects of each treatment option?

Are there any clinical trials for which I am eligible?

Surgical Interventions

Craniotomy

Craniotomy is the surgical opening of the skull (cranium). During this procedure, the patient is usually under general anesthesia. The surgeon makes an incision through a section of the scalp. Then, using sophisticated surgical tools, a part of the skull is removed and the covering of the brain (dura) is opened to reach the tumor. As much of the tumor as possible is removed. The dura is sutured and the part of the skull is put back into place. Then the scalp is closed with staples or stitches. These are usually removed after about seven to 10 days.

Biopsy

A biopsy is a surgical procedure in which a small sample of tissue is taken from the tumor and examined under a microscope. The purpose of a biopsy is to diagnose a tumor; to find out its type and grade.

Stereotactic or Needle Biopsy

A stereotactic or needle biopsy is used to take tissue samples from tumors in hard-to-reach areas of the brain. The patient is usually under general anesthesia during this procedure. A special frame called a static head frame holds the patient's head in place. A newer method, called "frameless" or computer-assisted image-guided brain needle biopsy, does not use a static head frame. The neurosurgeon makes a small hole in the skull. A narrow, hollow needle is inserted through the hole to extract tumor tissue. After a needle biopsy, the patient is monitored for several hours in the recovery room.

Shunt

A shunt, or catheter, is a device consisting of a thin tube and a valve that controls the flow of fluid. A shunt is used to divert CSF from the brain into the abdominal cavity, where it is absorbed into the bloodstream. During a cerebral shunt procedure, the surgeon drills a small hole in the skull. One end of the shunt is inserted into a ventricle; the other end is tunneled under the skin to the abdomen. Then, excess CSF drains into the abdominal cavity.

Some children may not have surgery because it is too dangerous and/or there has not been a benefit shown for such patients. Examples:

- Children with tumor that has spread to other areas of the brain or spinal cord
- Children with tumors in the brain stem (the upper part of the back of the neck)
- Children with tumors in the pituitary, pineal or thalamic areas (in the deep center of the brain)

Recovery from the Surgery

Most children recover very well from surgery, and many gradually regain neurological function lost because of the tumor. Occasionally, a child will have additional brain damage caused by unexpected bleeding or pressure on the brain during surgery, but these children frequently recover as well.

Posterior Fossa Syndrome

A child with a tumor removed from the posterior fossa in the lower back of the brain, may have post-operative mutism (inability to talk) caused by distortion of the cerebellum. This can be quite frightening for families but it is important to know that most of these children will regain their speech over time.

Chemotherapy

Chemotherapy is the treatment of disease by means of chemicals (drugs) that have a toxic effect on tumor cells as they divide. Chemotherapy is given in cycles, which consist of “on” and “off” phases— days of treatment followed by periods of time between treatments. Chemotherapy is usually taken orally or by injection. It may be given alone or in combination with other treatments. It is also used to delay or replace radiation treatment in young children. The brain has a defense mechanism called the *blood brain barrier*, which keeps out harmful substances such as bacteria and chemicals. The blood brain barrier can also prevent some chemotherapy drugs from entering the brain. There are some cases where chemotherapy treatment is not used. Certain types of tumors do not respond to chemotherapy.

Side Effects of Chemotherapy

Chemotherapy drugs target rapidly dividing cells, including normal ones. When normal cells are damaged, it can cause side effects. But normal cells can repair the damage or be replaced by other healthy cells, which is why side effects are usually temporary. Factors influencing side effects include:

- The specific chemotherapy drug
- The dose of the drug
- The health of the patient

Please refer to “Treatment Side Effect” Section of the Oncology Family Handbook for more details.

In general, the chemotherapy used to treat people with brain tumors is well tolerated. Newer *antiemetics* (anti-nausea drugs) have greatly reduced the nausea that chemotherapy patients may have experienced in the past. Oncologists or oncology nurses will work with patients to minimize or prevent anticipated side effects.

Your doctors and nurses will provide you with the information on the side effects of the chemotherapy agents you or your child will be receiving.

Despite monitoring the effects of chemotherapy very closely, some long-term effects can occur, sometimes years after therapy is completed. Therefore, it is important that every patient be followed throughout his or her life by a physician who is aware of the late effects of treatment.

Tips for Parents

- Review each drug with your doctor including all potential side effects.
- Discuss what can be done to prevent or treat side effects.
- Understand the tests that will be done to monitor side effects.
- *Remember that you are the expert on your child. Notify the healthcare team of any changes you notice, or concerns you may have.*

High Dose Chemotherapy with Stem Cell Support

In order to overcome the resistance of cancer cells and achieve more tumor kill, high dose chemotherapy may need to be used in an attempt to improve patient survival. The high dose chemotherapy may permanently destroy the normal stem cells in the bone marrow. Blood stem cells are the cells from which new blood cells develop such as red blood cells, white blood cells, and platelets.

Blood stem cells can be collected and stored following routine chemotherapy. After the high dose chemotherapy, the patient's own blood stem cells will be thawed and given back to the patient as "rescue" to help the recovery. This approach has been used in an attempt to improve patient survival and to delay, reduce, and possibly eliminate radiation therapy in certain situations.

Radiation Therapy

Radiation therapy (RT) uses high energy x-rays or other types of ionizing radiation to stop cancer cells from dividing. Radiation therapy may be used when surgery is not advised, or for tumors that cannot be completely resected. It may be used after surgery to destroy residual tumor cells and prevent or delay tumor recurrence.

Often the use of radiation therapy to a large area of brain or spine is avoided in children below the age of three because it may damage the developing brain. Ionizing radiation damages the basic building material in cells (DNA). Normal healthy cells can repair the damage better than tumor cells. Over time, irradiated tumor cells die. Thus, the results of radiation therapy may not be apparent until several months after treatment.

- Most patients with brain tumors will be treated with radiation therapy over a period of approximately 6 weeks following recovery from surgery.
- Use of radiation therapy to a large area of brain or spine is avoided in children below the age of three because it may damage the developing brain.
- Very young children and those with low grade tumors may have radiation therapy delayed or significantly reduced, or omitted if chemotherapy alone is judged to be effective.
- Radiation is given on weekdays and only takes a few minutes, provided that the child can lie still with the head supported by a custom-made "frame". Young children and those who have difficulty lying still usually require anesthesia. These children cannot eat or drink in the morning prior to the anesthesia.

Side Effects of Radiation Treatment

- Localized hair loss.
- Non-painful temporary redness/darkening of the overlying skin.
- Nausea and vomiting and fatigue occurs occasionally in older children or adolescents.
- Some children may experience increased sleepiness and fatigue during radiation treatment and for 1-2 months after the completion of radiation.
- Sometimes high dose radiation to brain stem tumors may cause dry or sore throat.

The late effects of radiation may include:

- Decreased thyroid function treatable with thyroid medication.
- Decreased growth which may be treated with growth hormone injections.
- Hormone deficits which may require treatment with medication.
- Intellectual deficits that are worse in those children who have high-dose, whole brain radiation at a young age.

Special Types of Radiation Therapy

Conformal Radiation

Conformal radiation is a type of conventional radiation treatment. A linear accelerator sends a high dose of radiation that has been modified by computer to match or conform to the shape of the tumor. The objective is to apply a uniform level of radiation to the tumor while reducing the amount of radiation that reaches other parts of the brain.

Intensity Modulated Radiation Therapy (IMRT)

Intensity Modulated Radiation Therapy (IMRT), is a type of conformal radiation therapy. IMRT is designed to restrict the treatment beam to the tumor, regardless of its shape. The intensity of radiation is made to vary across the

beam, so that a higher intensity reaches the thickest parts of the tumor. Multiple beams are used simultaneously and meet at the target site.

Gamma Knife/Radiosurgery

The Gamma Knife machine contains 201 sources of radioactive cobalt. It focuses a high dose of radiation to a small target area. The Gamma Knife is most effective for small tumors that are round or oval shaped. Gamma Knife can be used as a primary form of treatment or as a secondary treatment after surgery.

Proton Beam Radiotherapy

Proton beam radiotherapy is also called heavy particle radiation therapy. This method uses beams of charged protons (atomic particles) produced by a machine called a cyclotron. The cyclotron sends a single beam that is designed by a computer to match the shape of the tumor. Proton beam radiotherapy is used to treat tumors at the base of the skull and tumors of unusual shapes.

Neuroscience Conference

At CHOC Children's we provide an interdisciplinary team approach. All newly diagnosed brain tumor cases and many follow up cases are presented at the Neuroscience Conferences which are attended by Neurosurgeons, the Oncology Team, Neuro-radiologists, pathologist, radiation oncologists and the neuropsychologist. The diagnosis and management of patient and tumor type are discussed by the interdisciplinary team at these regular meetings.

Clinical Research

Everything now known about the cure of children with cancer has been learned from research. CHOC Children's collaborates with the Children's Oncology Group (COG) and Pharmaceutical Companies to provide cutting edge Clinical Trials for patients with various types of cancer.

COG has been the primary innovator in new treatments for children with cancer. COG conducts over 150 concurrent studies covering all the principal cancers of infants, children and adolescents. Over 40,000 patients worldwide are being treated according to COG research protocols. These clinical trials explore the safety, optimal dosage and response rate of new promising cancer drugs; compare the best available treatment to one or more experimental treatments, which are carefully developed with the goal of yielding even better results. By treating children in clinical trials, COG members have increased the cure rate for children with cancer.

CHOC Children's Research has many clinical trials open to find out the prevalence, the cause and innovative treatment for brain tumors.

Your doctor will provide you with the information on Clinical Trials relevant to your child's diagnosis. You can find more information about clinical trials in the *Clinical Trials* section of this handbook.

Neuropsychological Testing

A neuropsychological evaluation is a comprehensive assessment of thinking and behavior using standardized tests and procedures. Areas assessed may include: intelligence, academic achievement, language, attention, learning and memory, visual-perceptual skills, motor abilities, the ability to plan, organize and solve problems (executive functions), mood regulation and personality traits.

Neuropsychological evaluations may include some of the same tests as a school psychological evaluation but differ from school evaluations in assessing a broader range of skills and interpreting findings in relation to brain processes. Neuropsychologists also have more extensive training and experience working with children diagnosed with complex medical and neurological disorders.

Neuropsychological evaluations are often the best manner to identify neurocognitive deficits that may have resulted from your child's medical condition and treatment. A brief baseline neurocognitive screening may be performed if your child is at higher risk of experiencing neurocognitive deficits. A more comprehensive neuropsychological evaluation at the completion of treatment will be vital in helping prepare for the transition back to school and determining appropriate educational placement the need for any additional services.

Regular serial neuropsychological evaluations will be important in tracking future neurocognitive development and assisting with educational needs.

About Follow-Up Care

At CHOC Children's we provide a long term interdisciplinary follow up program. After treatment, patients need monitoring or follow up with a member of their medical team who knows the original diagnosis and treatment history. This person can determine how often and for how long follow-up care will be necessary.

Important questions to ask your interdisciplinary team:

1. How often should I see you or other members of my health care team for follow-up visits?

2. What follow-up tests should be done and how often?

3. What symptoms should I be concerned about?

Useful Web sites: www.curesearch.org, www.brainumor.org, www.cancer.gov

Adapted from The Essential Guide to Brain Tumors, National Brain Tumor Foundation 2007 and Curesearch.org

Information supplemented by the following members of the Neuro-Oncology Team: Violet Shen MD, Marcos DiPinto PhD, Van Huynh MD, Jody Pathare MSN,FNP, Julene Schenk RN,BSN,CPON and Tina Templeman, RN. Many thanks for the work of all team members.

Neuro-Oncology Team Members

Neurosurgeon A surgeon specializing in the diagnosis, treatment and surgical management of disorders and diseases of the brain, spine and nervous system.

Neuro-oncologist An oncologist specializing in the diagnosis and treatment of cancers affecting the brain, spinal cord.

Neurologist A doctor specializing in the diagnosis and treatment of disorders and diseases affecting the brain, spinal cord and nervous system.

Neuro-radiologist A doctor that specializes on evaluating brain and imagining such as: MRI and CT scans.

Oncology Fellow A doctor who has completed pediatric residency training is now receiving specialty training in pediatric Hematology-Oncology

Radiation Oncologist A doctor specializing in the radiation therapy treating certain cancers.

Endocrinologist A doctor that specializes in caring for patients with hormone hormone imbalances and replacing normal hormones that the brain may not be releasing properly, such as diabetes insipidus and thyroid dysfunction.

Neuropsychologist A neuropsychologist is a licensed psychologist who has specialized training and experience studying how the brain functions. A neuropsychologist does testing and research to attempt to explain the relationship between brain activity and behavior and to understand how functions within the brain influence thinking, learning and emotions. He or she studies the impact that brain damage has on a person's functioning and abilities. A neuropsychologist can help brain tumor patients and survivors evaluate changes resulting from their brain tumor or treatment, and can help develop a plan for rehabilitation.

Case Coordinator A person who provides coordination of care for complex neuro-oncology patients. Responsible for coordination of quality healthcare services to meet each individual patient's specific healthcare needs. Sets up family conferences and assists in discharge planning, making arrangements for medical needs that patient may require at home. Serves as a community and insurance liaison.

Child Life Specialist A person who has special training in child development and how children react to illness and being in the hospital. A Child Life Specialist helps children to cope with cancer and its treatment.

Clinical Research Nurse A person who is familiar with Clinical Research protocols, assists in screening, enrolling and following patients for the clinical research studies, and submitting diagnostic, treatment and follow-up data to the Research Data Center.

Nurse Practitioner/ Clinical Nurse Specialist A nurse who has completed a master's program and specialty training in caring for children with cancer. He/She may coordinate the medical and nursing care of your child.

Nutritionist A person who is trained to evaluate your child's nutritional needs and weight. The nutritionist also helps to provide teaching and support about eating and drinking when your child goes home.

Oncology Nurse A person who is trained to provide daily nursing care and teaching to children and their families in the hospital or clinic.

Oncology Pharmacist A pharmacist who is specially trained to prepare the medicines and nutritional support that your child will need that is specific to Oncology patients.

Rehabilitation Services including physical therapy, occupation therapy and speech therapy are provided to patients with impaired neurological function to give patients the best chance for recovery.

Social Workers A person who is trained to help you and your child cope with illness and hospitalization through counseling, support groups, financial assistance, and resource referral.