

# Care of the Pediatric Patient with a Brain Tumor

AANN Clinical Practice Guideline Series

This publication was made possible by an educational grant from the American Brain Tumor Association



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## Preface

In 1997, the American Association of Neuroscience Nurses (AANN) created a series of patient care guidelines, the *AANN Reference Series for Clinical Practice*, to meet its members' needs for educational tools. To better reflect the nature of the guidelines and the organization's commitment to developing each guideline based on current literature and evidence-based practice, the name of the series was changed in 2007 to the *AANN Clinical Practice Guideline Series*.

The goal of this guideline is to offer evidence-based recommendations on nursing activities that have the potential to maximize outcomes for pediatric patients with brain tumors. Not all recommendations concern activities independently performed by registered nurses (RNs), but nurses are responsible for implementing and monitoring the outcomes of these activities. The evidence presented here may help nurses make appropriate choices when caring for patients with brain tumors. Dependent on the scope of practice regulations, advanced practice nurses may have

independent or collaborative responsibilities for activity performance. This guideline may assist them in the treatment of patients with brain tumors as well. Resources and recommendations must describe the best practices that can enable RNs to provide optimal care for people with brain tumors. Accordingly, adherence to these guidelines is voluntary, and the ultimate determination regarding guideline application must be made by practitioners in light of each patient's individual circumstances. This reference is an essential resource for nurses providing care to pediatric patients with brain tumors. It is not intended to replace formal learning, but rather to augment the clinician's knowledge base and provide a readily accessible reference tool. The nursing profession and AANN are indebted to the volunteers who have devoted their time and expertise to this valuable resource, which was created for those who are committed to excellence in the care of pediatric patients with brain tumors.

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## I. Introduction

### A. Purpose

1. The purpose of the *Care of the Pediatric Patient with a Brain Tumor* clinical practice guideline (CPG) is to provide an up-to-date and thorough review and assessment of the literature; nursing care recommendations based on the evidence in the literature; and, in the absence of published scientific evidence, recommendations based on expert opinion. The intended audience is bedside nurses caring for pediatric patients with brain tumors.
2. This comprehensive, yet concise, review will serve as a reference for the care of children with brain tumors, from acute diagnosis to long-term survivorship.

### B. Guideline goal

1. The goal of this CPG is to establish current and consistent practices among nurses providing direct care to pediatric patients with brain tumors. The basic understanding of pediatric brain tumors, including the epidemiology, etiology, and most common types of brain tumors, is covered, as well as diagnostic testing, acute care, surgical management, treatments and long-term effects and the nursing assessment, and interventions related to those topics.
2. There is hope that recognition of the overall dearth of literature specific to nursing care regarding this topic will encourage bedside nurses to commit to evidence-based care and the development of evidence-based publications.

### C. Assessment of scientific evidence

1. A comprehensive review of literature published prior to July 2013 was performed using the PubMed/Medline, CINAHL, Cochrane, Google Scholar, and OVID databases. Key words (and combinations of them) included ICU management, acute seizures, resection, biopsy, pediatric, brain tumor, neuro-oncology, surgery, radiation, chemotherapy, end of life, palliative care, event free survival, craniospinal radiotherapy, central nervous system tumor, oncology, patient reported outcomes, cognitive training, childhood cancer, social, academic, neuropsychologic, off therapy, pediatric oncology nursing, attention problems, survivorship, late effects, nursing roles, and cancer. Preference was given to randomized clinical trials, especially those performed within the last 5 years; however, historically significant literature was referenced in the absence of more current and rigorous research.
2. Secondary references were discovered from primary references, but all efforts were made to retrieve secondary references and give appropriate credit to the original author and/or study.

3. Online references were used sparingly, and sources are included in the reference list.
4. Textbook references such as Adamson, Bagatell, Balis, and Blaney (2011); Blaney et al. (2011); and chemotherapy administration guidelines (Ettinger & Rale, 2011) from the American Pediatric Hematology Oncology Nursing Society and other publications appropriate to the topic were reviewed, used, and cited in the absence of evidence of clinical trials.
5. For the AANN Clinical Reference Series, data quality is classified as follows:
  - a. Class I: Randomized controlled trials without significant limitations or meta-analysis
  - b. Class II: Randomized controlled trials without important limitations (e.g., methodologic flaws or inconsistent results) and observational studies (e.g., cohort or case-control)
  - c. Class III: Qualitative study, case study, or series
  - d. Class IV: Evidence from expert committee reports and/or expert opinion of the guideline panel; standards of care and clinical protocols that have been identified.
6. The *Clinical Practice Guidelines* and recommendations for practice are established based upon the evaluation of the available evidence (AANN, 2005; Guyatt & Rennie, 2002; Melnyk, 2004).
  - a. Level 1 recommendations are supported by Class 1 evidence.
  - b. Level 2 recommendations are supported by Class II evidence.
  - c. Level 3 recommendations are supported by Class III and IV evidence.

## II. Epidemiology and Overview

### A. Epidemiology

Epidemiology of pediatric brain tumors in patients ages 0–19 years (Bondy et al., 2008; Dolecek, Propp, Stroup, & Kruchko, 2012; Rosemberg & Fujiwara, 2005)

1. Between 2005 and 2009, the overall incidence of brain tumors in children ages 0–19 years was 5.13 per 100,000 persons, accounting for 7% of all reported brain tumors and central nervous system (CNS) tumors.
2. The second most common malignancy among children and the most common solid tumor in children 0–19 years of age (2005–2009 data)
  - a. 3.3 tumors per 100,000 persons, age-adjusted to the 2000 U.S. standard population, are malignant.
  - b. 1.9 tumors per 100,000 persons, age-adjusted to the 2000 U.S. standard population, are nonmalignant.
3. Tumors may be located across the CNS (Dolecek et al., 2012). The cerebellum is the most common site, at 17%.

4. Age-related incidence of pediatric brain tumors (2005–2009 data)
  - a. Highest incidence among ages 15–19 years; 5.63 tumors per 100,000, age-adjusted to the 2000 U.S. standard population
  - b. Lowest incidence among ages 5–9 years; 4.70 per 100,000, age-adjusted to the 2000 U.S. standard population
5. Age-related incidence of specific tumor type varies by histopathology (2005–2009 data).
  - a. Ages 0–14 years
    - i. Pilocytic astrocytomas account for 18% of pediatric brain tumors.
    - ii. Embryonal tumors including medulloblastoma account for 15% of pediatric brain tumors.
    - iii. Malignant gliomas account for 14% of pediatric brain tumors.
  - b. Ages 15–19 years
    - i. Pituitary tumors account for 23% of pediatric brain tumors.
    - ii. Pilocytic astrocytomas account for 11% of pediatric brain tumors.
6. Gender characteristics (2005–2009)
  - a. The incidence of germ-cell tumors is higher in males than females (2:1).
  - b. The incidence of pituitary tumors is higher in females than males (3:1).
7. The overall incidence is 5.30 (per 100,000) among non-Hispanic whites and 4.55 (per 100,000) among blacks, age adjusted to the 2005–2009 U.S. standard population.

## B. Classification

1. Brain tumors are classified based upon their histopathology (Louis et al., 2007; Pfister, Hartmann, & Korshunov, 2009).
2. The World Health Organization (WHO) provides the established standard for clinicians regarding the definition of brain tumor based on the premise that each type of tumor results from the abnormal growth of a specific cell type.
  - a. Tumors are graded based on the abnormal appearance of the tumor cells and tumor tissue under a microscope.
  - b. The WHO grading system indicates how quickly a CNS tumor is likely to grow and spread (Louis et al., 2007).
    - i. Grade I: Slow-growing, nonmalignant, and associated with long-term survival
    - ii. Grade II
      - a) Relatively slow-growing, but sometimes recurs as a higher-grade tumor
      - b) Can be nonmalignant or malignant
    - iii. Grade III: Malignant and often recurs as a higher-grade tumor
    - iv. Grade IV: Reproduces rapidly; very aggressive malignant tumors

3. The classification of brain tumors can be further categorized into glial and nonglial tumors (Pfister et al., 2009).
  - a. Glial tumors
    - i. Originate from glial cells
    - ii. Astrocytomas, derived from astrocytes
    - iii. Ependymomas, derived from ependymal cells
    - iv. Oligodendrogliomas, derived from oligodendrocytes
  - b. Nonglial tumors
    - i. Embryonal tumors, such as medulloblastoma
    - ii. Craniopharyngiomas
    - iii. Pineal tumors
    - iv. Meningiomas
    - v. Germ-cell tumors

## C. Risk factors

1. Two factors are linked to an increased risk for primary CNS tumors in childhood.
  - a. A history of receiving significant doses of radiation to the CNS
  - b. Some genetic syndromes are associated with an increased risk for CNS tumors (Fleming & Chi, 2012).
    - i. Cowden syndrome (Hottinger & Khakoo, 2009; Lyons, Wilson, & Horton, 1993; Nelen et al., 1997)
      - a) An autosomal-dominant disorder affecting 1 in 250,000 children
      - b) Linked to germline mutations in the phosphatase and tensin homolog gene on chromosome 10q22-23
      - c) Associated with hamartomas, meningiomas, and dysplastic cerebellar gangliocytomas
    - ii. Gorlin syndrome
      - a) An autosomal-dominant disorder affecting 1 in 50,000 individuals (Hottinger & Khakoo, 2009)
      - b) Thirty percent of cases present when the first affected family member exhibits an alteration in the gene as a result of a mutation (Hottinger & Khakoo, 2009).
      - c) Characterized by the development of multiple jaw keratocysts; frequently begins in the second decade of life; and/or basal cell carcinomas, usually from the third decade onward (Farrell & Plotkin, 2007; Hottinger & Khakoo, 2009)
      - d) Macrocephaly found in approximately 60% of patients with Gorlin syndrome
      - e) Skeletal abnormalities (Farrell & Plotkin, 2007; Hottinger & Khakoo, 2009)

- f) Five percent of patients with Gorlin syndrome develop medulloblastoma with desmoplastic histology (Hottinger & Khakoo, 2009). Medulloblastoma in patients with Gorlin syndrome develops at a younger age than sporadic medulloblastoma (Farrell & Plotkin, 2007).
- g) Has a more favorable prognosis
- h) Patients are at risk for radiation therapy (RT)-induced basal cell carcinomas (Farrell & Plotkin, 2007; Hottinger & Khakoo, 2009).
- iii. Li-Fraumeni syndrome (LFS)
  - a) An autosomal-dominant disorder predisposing individuals to multiple cancers
  - b) Mutation-inactivating TP53 gene
  - c) Twelve percent of those with LFS develop brain tumors (Hottinger & Khakoo, 2009).
  - d) Sixteen years is the mean age of onset for development of a brain tumor associated with LFS (Farrell & Plotkin, 2007).
  - e) Increased incidence of astrocytic tumors, medulloblastoma, and supratentorial primitive neuroectodermal tumors (Farrell & Plotkin, 2007)
  - f) Individuals with LFS are at higher risk for radiation-induced secondary malignancies (Farrell & Plotkin, 2007).
- iv. Neurofibromatosis type 1 (NF1; Farrell & Plotkin, 2007; Hottinger & Khakoo, 2009)
  - a) An autosomal-dominant disorder characterized by the development of tumors along the nerve sheath
  - b) One in 4,000 births (Farrell & Plotkin, 2007; Hottinger & Khakoo, 2009)
  - c) The result of mutations in the NF1 gene along chromosome 17; responsible for making neurofibromin, a protein produced in many nerve cells responsible for tumor suppression (Hottinger & Khakoo, 2009)
  - d) Diagnosis of NF1 is usually evident by age 6 (Hottinger & Khakoo, 2009).
  - e) Eighty percent of children with NF1 develop cutaneous, subcutaneous, or deep neurofibromas by puberty; neurofibromas are peripheral nerve sheath tumors comprising Schwann cells, fibroblasts, and perineural cells (Farrell & Plotkin, 2007).
  - f) Optic pathway pilocytic astrocytoma is the most common CNS tumor associated with NF1; 6%–20% of patients with NF1 have an optic pathway pilocytic astrocytoma (Lewis, Gerson, Axelson, Riccardi, & Whitford, 1984). NF1-related tumors are often associated with better outcomes because they may remain static or even regress (Farrell & Plotkin, 2007).
- g) Nonoptic gliomas are most common in the brain stem and range from low to high grade (Farrell & Plotkin, 2007).
- v. Neurofibromatosis type 2 (NF2)
  - a) An autosomal-dominant disorder affecting 1 in 40,000 individuals; the result of a mutation in the NF2 gene located on chromosome 22q12 (Hottinger & Khakoo, 2009)
  - b) Average age of onset is between 17 and 21 years of age (Farrell & Plotkin, 2007).
  - c) Predisposes patients to peripheral and CNS tumors (Hottinger & Khakoo, 2009). The hallmark feature is the development of bilateral vestibular schwannomas (Farrell & Plotkin, 2007; Hottinger & Khakoo, 2009).
- vi. Tuberous sclerosis (TS; Datta, Hahn, & Sahin, 2008; Farrell & Plotkin, 2007; Hottinger & Khakoo, 2009)
  - a) An autosomal-dominant disorder affecting 1 in 6000–10,000 births (Datta et al., 2008; Farrell & Plotkin, 2007; Hottinger & Khakoo, 2009)
  - b) Two-thirds of cases occur sporadically because of mutation in the TSC1 or TSC2 genes (Datta et al., 2008).
  - c) Characterized by epilepsy; cognitive disability; neurobehavioral abnormalities; autism; and hamartomas of the heart, kidney, CNS, and skin (Farrell & Plotkin, 2007; Hottinger & Khakoo, 2009)
  - d) Associated abnormalities within the CNS (Farrell & Plotkin, 2007) include cortical tubers, subependymal nodules, and subependymal giant cell astrocytomas (develop most often in the Foramen of Monro, leading to obstructive hydrocephalus).
- vii. Turcot syndrome (Hottinger & Khakoo, 2009)
  - a) An autosomal-dominant disorder characterized by tumors of the CNS associated with familial polyposis of the colon
  - b) The result of mutations in the adenomatous polyposis coli gene,



**Table 1. Signs and Symptoms by Tumor Location**

Tumor Location	Presenting Signs and Symptoms
Cerebral Hemispheres	Headache, seizures, hemiparesis, hyperreflexia, clonus, sensory loss, speech disturbances, memory deficits, personality changes, visual disturbances
Posterior Fossa	Nausea and vomiting, headache, abnormal gait and coordination, papilledema, abnormal eye movements
Brain Stem	Cranial nerve deficits, gait and coordination disturbances, nystagmus, focal motor weakness, signs of increased intracranial pressure such as headache and papilledema
Spinal Cord	Radicular pain and/or weakness (symptoms correspond to level of lesion), loss of bowel/bladder control, gait abnormalities, scoliosis
Optic Pathway	Visual disturbances, proptosis, nystagmus
Hypothalamus/Thalamus	Endocrine disturbances including diabetes insipidus and growth failure, altered level of consciousness and memory, and academic problems

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associated with medulloblastoma or a mutation in the MLH1 or PMS2 genes associated with glioblastoma multiforme

viii. Von Hippel-Lindau disease

- a) An autosomal-dominant disorder occurring in 1 in 35,000 individuals (Farrell & Plotkin, 2007; Hottinger & Khakoo, 2009)
- b) Predisposes patients to hemangioblastomas of the brain, kidneys, adrenal glands, pancreas, spinal cord, and retina and renal cysts (Hottinger & Khakoo, 2009)
- c) Mutation in the VHL gene (Hottinger & Khakoo, 2009)
- d) CNS hemangioblastomas occur in 60%–80% of patients (Hottinger & Khakoo, 2009).
- e) CNS hemangioblastomas may cause life-threatening complications despite their benign nature and classic slow-growing course (Farrell & Plotkin, 2007).

**Nursing recommendations**

1. Nurses should be aware that children with hereditary syndromes may be at increased risk for brain tumors. Nurses caring for children with these genetic syndromes should observe for signs and symptoms that may suggest a CNS tumor (Level 3).
2. Nurses should have a basic understanding of genetic predisposition as it relates to pediatric brain tumors. This knowledge should be used to assess a child's risk for developing CNS tumors (Level 3).
3. Nurses should possess awareness of the resources available to optimize care coordination and use a multidisciplinary approach to manage symptoms and improve function and quality of life (Level 2).
4. Nurses should know how to identify risk

factors of hereditary syndromes that present unique and complex issues. Nurses will use this knowledge to guide families in future decision making and improve the care of long-term survivors (Level 2).

5. Nurses will assist families during genetic counseling by identifying personal risk and screening options, early detection methods, and risk-reduction strategies (Level 2).

**D. Signs and symptoms**

1. Arise as a result of four mechanisms (Armstrong, Cohen, Eriksen, & Hickey, 2004; Duffner, 2007; Gonzalez et al., 2009)
  - a. Invasion of the tumor into the brain parenchyma
  - b. Compression of brain tissue as a result of a tumor
  - c. Increased intracranial pressure (ICP)
  - d. Herniation
2. Symptoms may present as generalized or focal (Blaney et al., 2011; Dobrovoljac, Hengartner, Boltshauser, & Grotzer, 2002; Hayashi et al., 2010; Klitbo, Nielsen, Illum, Wehner, & Carlsen, 2011; Wilne et al., 2007).
  - a. The most common generalized symptoms are headaches accompanied by vomiting and unsteadiness.
  - b. Focal symptoms: Specific deficits are related to the location of the tumor and adjacent structures (**Table 1**).
  - c. Symptomatology in infants and young children differs because of the pliability of the cranium and open cranial fontanels (Blaney et al., 2011).
    - i. Irritability, anorexia, failure to thrive, developmental delays, or regression
    - ii. Tense and bulging anterior fontanel
    - iii. Sunsetting; loss of upward conjugate gaze
  - d. Diagnosis difficulties
    - i. Age and developmental level of the child may affect the ability to identify

**Table 2. Characteristic Clinical Behaviors Found in Molecular Subgroups of Medulloblastoma**

	<b>Wnt</b>	<b>Shh</b>	<b>Group 3</b>	<b>Group 4</b>
Age Group	Infant Adult	Infant Child Adult	Infant Child	Infant Child Adult
Gender M/F	1:1	1:1	2:1	2:1
Histology	Classic Rarely LCA	Desmoplastic/nodular Classic LCA	Classic LCA	Classic LCA
Prognosis	Very good	Infants good Others intermediate	Poor	Intermediate
Associated Genetic Syndrome	Turcot	Gorlin		

*LCA, Large-cell anaplastic*

Source. "Molecular subgroups of medulloblastoma: The current consensus," by M. D. Taylor, P. A. Northcott, A. Korshunov, M. Remke, Y. J. Cho, S. C. Clifford, . . . S. M. Pfister, 2012, *Acta Neuropathologica*, 123(4), 465–472.

symptoms and diagnose (Duffner, 2007; Reulecke, Erker, Fiedler, Niederstadt, & Kurlemann, 2008).

- ii. Children are not able to articulate symptomatology as clearly as adults (Wilne, Ferris, Nathwani, & Kennedy, 2006).
- iii. Children may require anesthesia for imaging, which may compromise access to timely diagnostic testing (Wilne et al., 2006).
- iv. Low parent educational level, social deprivation, and lack of familiarity with health care may lead to a delay in diagnosis (Wilne et al., 2010).
- v. When headache and vomiting occur without focal signs of infection for more than a few days, the diagnosis of a viral illness should be made with caution (Elgamal & Richards, 2006; Shemie, Jay, Rutka, & Armstrong, 1997).
- vi. Persistent lethargy should be regarded as a neurologic sign rather than a non-specific symptom (Elgamal & Richards, 2006; Shemie et al., 1997).

#### E. Tumor types

1. Medulloblastoma (small blue round-cell tumor)
  - a. Primary malignant embryonal tumor, classified as a Grade IV tumor (Louis et al., 2007)
  - b. Arises infratentorially in the cerebellum or fourth ventricle (Bartlett, Kortmann, & Saran, 2013)
  - c. The most common presenting symptoms are those that are associated with increased ICP.
  - d. Disease risk is based on a combination of several factors (Taylor et al., 2012).
    - i. Demographics: Age group and gender
    - ii. Clinical features
      - a) Histology
        - (1) Classic: 80% of all medulloblastomas
        - (2) Desmoplastic/nodular: 15% of all medulloblastomas

- (3) Large-cell anaplastic
  - b) Metastasis
- iii. Genetics
  - a) Gain of the 6q, 17q, or i(17q) chromosome
  - b) Deletion of the 6q chromosome
  - c) MYC or MYCN amplification.
    - (1) MYC and MYCN are regulator genes.
    - (2) If MYC and MYCN are overexpressed, poor cell regulation and proliferation can occur.
  - d) Gene expression: Amount of MYC or MYCN amplification
  - e) Molecular subgroups (**Table 2**)
    - (1) Most clinically relevant risk predictor
    - (2) Each molecular histological (or tissue) type is distinct in prognosis and response to therapy (Taylor et al., 2012).
    - (3) Clinical behaviors have been identified in each subgroup (Taylor et al., 2012).
      - (a) Wnt subgroup
        - (i) Primarily found in infants to early childhood (0–3 years of age)
        - (ii) Classic histology
        - (iii) Favorable prognosis
        - (iv) Can be associated with Turcot syndrome
      - (b). Shh subgroup
        - (i) Primarily found in infants to early childhood (0–3 years of age) and individuals older than 16 years
        - (ii) Desmoplastic/nodular histology
        - (iii) Favorable prognosis

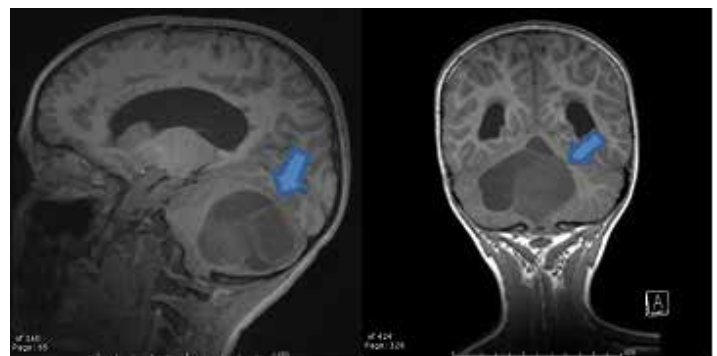
- (iv) Can be associated with Gorlin syndrome
- (c). Group 3
  - (i) Primarily found in children 3–16 years of age
  - (ii) More common in boys
  - (iii) Classic histology and a high incidence of large-cell histology
  - (iv) Frequently metastatic
  - (v) Poor prognosis
- (d). Group 4
  - (i) Primarily found in children 3–16 years of age
  - (ii) More common in boys
  - (iii) Contains classic and large-cell histology
  - (iv) Prognosis is similar to the Shh subgroup (favorable)

M0	No evidence of metastasis
M1	Tumor cells found in the cerebrospinal fluid (by lumbar peritoneal and cytology study)
M2	Tumor beyond primary site, but still in brain
M3	Tumor deposits (“seeds”) in spine area that are easily seen on magnetic resonance imaging
M4	Tumor spread to areas outside the central nervous system (outside both the brain and spine)

*Source.* “An operative staging system and a megavoltage radiotherapeutic technic for cerebellar medulloblastomas,” by C.H., Chang, E. M. Housepian, & C. Herbert Jr., 1969, *Radiology*, 93(6), 1351–1359.

- f) Staging: Medulloblastomas are staged according to the Modified Chang Staging System, which is based on radiological findings and surgical assessment (**Table 3**; Chang, Housepian, & Herbert, 1969).
- g) Stratification is done by age and stage (Mueller & Chang, 2009; Zeltzer et al., 1999).
  - (1) Standard risk
    - (a) Patients older than 3 years of age
    - (b) Less than 1.5-cm residual disease after resection
    - (c) Magnetic resonance imaging (MRI) and cerebrospinal fluid (CSF) studies include M0 staging according to the Modified Chang Staging System.
  - (2) High risk
    - (a) Subtotal resection with more than 1.5 cm residual tumor on postoperative imaging

- and/or metastatic disease resulting in M1-M3 staging according to the Modified Chang Staging System
- (3) Atypical teratoid rhabdoid tumor
  - (a) A malignant embryonal tumor of the CNS, classified as a WHO Grade IV tumor, composed of rhabdoid cells; rhabdoid cells are high-grade malignant cells that are similar to rhabdoid cells found in the kidney, which are non-malignant (Buscariollo, Park, Roberts, & Yu, 2012)
  - (b) Represents 1%–2% of all pediatric brain tumors and up to 20% of cases among patients younger than 3 years of age (Buscariollo et al., 2012; Ginn & Gajjar, 2012)
  - (c) Can occur anywhere in the brain, but most commonly in the cerebellum and brain stem (Reddy, 2005)
  - (d) Symptoms reflect tumor location.
  - (e) Short history of progressive symptoms
  - (f) Usually occurs in children younger than 3 years of age, but can be seen in older children as well (Bikowska, Grajkowska, & Jozwiak, 2011; Buscariollo et al., 2012; Ginn & Gajjar, 2012)
  - (g) Multimodality treatment offers improved overall survival (Buscariollo et al., 2012; Tekautz et al., 2005).



**Figure 1.** A 6-year-old boy with a juvenile pilocytic astrocytoma. Note the cystic components of the mass as well as the significantly large size upon diagnosis. Photos courtesy of Boston Children’s Hospital. Used with permission.

- (h) Children older than 3 years of age tend to fare better than younger children because of the potential survival benefit derived from adjuvant RT (Chi et al., 2009; Tekautz et al., 2005).
- (i) Overall survival is 6–18 months (Buscariollo et al., 2012).

## 2. Gliomas

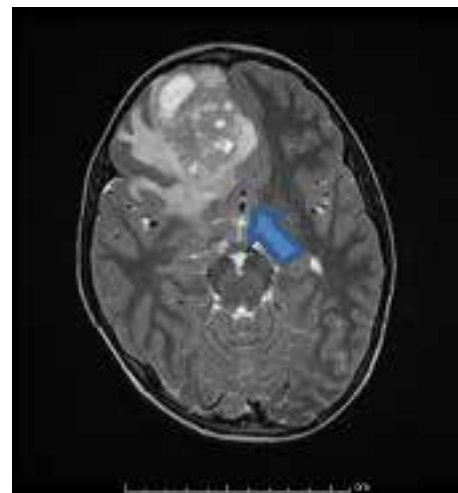
### a. Low-grade gliomas (LGG)

- i. Include Grade I and Grade II tumors according to the WHO criteria (Blaney et al., 2011)
- ii. The most common subtypes of LGG are
  - a) Pilocytic astrocytoma (WHO Grade I; **Figure 1**)
  - b) Diffuse fibrillary astrocytoma (WHO Grade II).
- iii. Rarer subtypes of Grade II astrocytomas
  - a) Pilomyxoid astrocytoma
  - b) Pleomorphic xanthroastrocytoma
- iv. LGG account for 30%–50% of CNS tumors in children and may occur in children of all ages (Blaney et al., 2011).
- v. LGG occur sporadically; however, they are also associated with familial cancer syndromes. Children with NF1 are at an increased risk for LGG, particularly optic pathway gliomas (Farrell & Plotkin, 2007).
- vi. Children who develop LGG associated with NF1 have better outcomes because the LGG remain static or even regress (Farrell & Plotkin, 2007)
- vii. LGG may occur anywhere in the CNS.
- viii. The cerebellum is the most prevalent location (Louis et al., 2007).
- iv. Gross total resection may be curative.
- x. In cases in which LGG cannot be safely resected, chemotherapy can be used to achieve disease stability (Gnekow, Kortmann, Pietsch, & Emser, 2004; Gurrangan et al., 2007; Mishra et al., 2010; Packer et al., 1997).

### b. High-grade gliomas (HGG) outside of the brain stem

- i. Supratentorial HGG tend to occur in older children (Fangusaro & Chi, 2009).
- ii. Identified risk factors
  - a) Previous RT exposure, LFS, Turcot syndrome, and NF1 (Fangusaro & Chi, 2009)
  - b) Fifty percent of HGG are located in the cerebral hemispheres.
- iii. Anaplastic astrocytoma; Grade III

- a) Defined as having cell atypia and having high mitotic features
- b) Invasion into the adjacent brain tissue and irregular margins are seen on MRI imaging (Cage, Mueller, Haas-Kogan, & Gupta, 2012).
- c) Represents 1%–2% of all pediatric brain tumors with equal distribution between boys and girls (Fangusaro & Chi, 2009)
- d) Median age of incidence is approximately 9 years.
- e) One-third occur in the supratentorium.
- f) Evolving symptoms present over a short time (weeks to months); symptoms vary according to location; general or focal (Fangusaro & Chi, 2009; Tamber & Rutka, 2003).
  - (1) Generalized symptoms often represent increased ICP.
  - (2) Focal symptoms reflect tumor location.
- g) Etiology is usually unclear, although there are several known predisposing risk factors (Cage et al., 2012).
  - (1) Previous exposure of cranial irradiation therapy
  - (2) LFS
  - (3) NFI
  - (4) Turcot syndrome
- h) Five-year progression-free survival is approximately 23% (Fangusaro & Chi, 2009; MacDonald, Aguilera, & Kramm, 2011).
  - (1) Degree of surgical resection is the most important prognostic indicator.



**Figure 2.** This 14-year-old boy with glioblastoma multiforme also has neurofibromatosis type 1. Photo courtesy of Boston Children's Hospital. Used with permission.

- (2) Age younger than 3 years is associated with a better prognosis.
  - (3) Multimodality therapy is used (Fangusaro & Chi, 2009; Sathornsumetee, Rich, & Reardon, 2007).
  - (4) Aggressive surgical resection is performed when clinically feasible.
  - (5) RT is the mainstay of treatment.
  - (6) The role of adjuvant chemotherapy is limited.
3. Glioblastoma (Grade IV; **Figure 2**)
    - a. Exhibits vascular proliferation and/or necrosis (Tamber & Rutka, 2003)
    - b. Locally invasive, with significant local mass effect because of rapid growth (Fangusaro & Chi, 2009; Tamber & Rutka, 2003)
    - c. Can be found anywhere in the CNS, but most commonly in the supratentorium (Fangusaro & Chi, 2009)
      - i. Thirty percent to 50% are located within the cerebral hemispheres.
    - d. Risk factors (Fangusaro & Chi, 2009)
      - i. Inherited defects such as LFS, NF1, and Turcot syndrome
      - ii. Previous exposure to radiation
    - e. Gross total or near-total resection is the most important prognostic indicator and should be attempted whenever feasible (Fangusaro & Chi, 2009; Sanders et al., 2007).
    - f. RT is the standard of care for children older than 3 years of age (Cage et al., 2012).
    - g. Five-year overall survival is approximately 10%–30% (Fangusaro & Chi, 2009).
  4. Diffuse intrinsic pontine glioma (**Figure 3**)
    - a. A highly infiltrative and aggressive lesion noted for cell atypia, high mitotic activity, tumor necrosis, and vascular proliferation
    - b. Derived from glial cells (i.e., astrocytes, oligodendrocytes, and ependymal cells)
      - c. Unknown etiology, although NF1 is a known risk factor (Fangusaro & Chi, 2009)
      - d. Involved in 10%–15% of all pediatric brain tumors and 75%–80% of pediatric brain stem tumors (Kieran & Marcus, 2013; Warren, 2012)
      - e. Most common in children ages 5–10 years
      - f. Diagnosis is based on radiographic findings and presentation (Fangusaro & Chi, 2009; Kieran & Marcus, 2013; Warren, 2012).
      - g. MRI demonstrates a large brain stem mass centered within and involving the majority of the pons.
        - i. Intratumoral hemorrhage and tumor necrosis are not uncommon.
        - ii. Encasement of the basilar artery is seen often, but is not inclusive of diagnosis.
      - h. Symptomatology is often acute in onset and short in duration (Fangusaro & Chi, 2009; Warren, 2012).
        - i. Cranial nerve deficits
        - ii. Long tract signs
          - a) Signs such as hyperreflexia, clonus, and/or increased muscle tone that indicate a lesion in the upper spinal cord or brain stem
          - b) Ataxia
      - i. Surgery is not recommended because of the eloquent region of the brain, in which the tumor is located.
      - j. RT offers a transient reduction in tumor.
      - k. Disease progression is often seen at 3–8 months following RT (Kieran & Marcus, 2013; Warren, 2012).
      - l. Adjuvant chemotherapy has not demonstrated significant improvement.
      - m. Corticosteroids are often used to improve symptoms associated with peritumoral edema (Kieran & Marcus, 2013).
  5. Other tumor types: Ependymoma (**Figure 4**)
    - a. Arise from ependymal cells lining the ventricles of the brain (Zacharoulis & Moreno, 2009)



**Figure 3.** This 9-year-old boy has diffuse intrinsic pontine glioma. Photo courtesy of Boston Children's Hospital. Used with permission.



**Figure 4.** This 3-year-old has ependymoma. Photo courtesy of Boston Children's Hospital. Used with permission.

- b. Approximately 60% of ependymomas are found in the fourth ventricle (Zacharoulis & Moreno, 2009).
  - c. Most common in children younger than 3 years of age (Reni, Gatta, Mazza, & Vecht, 2007)
  - d. Seeding of the tumor is found in approximately 15% of patients upon diagnosis (Zacharoulis & Moreno, 2009).
  - e. There is higher risk for ependymoma in patients with NF2 (Reni et al., 2007).
  - f. According to the WHO, there are four major subtypes of ependymoma, although grading does not predict evolution of the tumor or prognosis (Godfraind, 2009; Zacharoulis & Moreno, 2009).
    - i. Grade I: Myxopapillary
    - ii. Grade I: Subependymoma
    - iii. Grade II: Classic ependymoma, the most common (Zacharoulis & Moreno, 2009)
    - iv. Grade III: Anaplastic ependymoma (Godfraind, 2009)
  - g. Patients present with a short history of symptoms upon presentation; increased ICP because of the obstruction of CSF flow by the tumor in the fourth ventricle (Zacharoulis & Moreno, 2009).
  - h. Patients with supratentorial ependymomas present most commonly with lobar syndromes and headache.
  - i. Patients should have MRIs of the entire neuraxis to identify potential metastatic disease.
  - j. CSF analysis is important in staging the tumor and should be obtained within 2 weeks for cytology (Moreno et al., 2010).
  - k. Gross total resection offers a better prognosis (Godfraind et al., 2012; Lafay-Cousin et al., 2011; Moreno et al., 2010; Reni et al., 2007; Zacharoulis & Moreno, 2009).
  - l. There is a role for second-look surgery to obtain a higher resection.
  - m. RT is considered standard therapy in children with ependymoma (Godfraind et al., 2012; Zacharoulis & Moreno, 2009).
  - n. Response rates to chemotherapy in ependymoma are variable.
  - o. Chemotherapy continues to have a role in delaying postoperative RT in children younger than 3 years of age (Zacharoulis & Moreno, 2009).
6. Craniopharyngioma
- a. A nonglial intracranial tumor whose origins are derived from an embryogenic malformation of Rathke's pouch (Muller, 2010; Winkfield et al., 2011)
  - b. Found in the intrasellar or suprasellar region of the brain and may include the hypothalamus
  - c. Histologically considered benign, but often associated with a high incidence of morbidity attributable to the location and the degree of hypothalamic involvement (Harrington & Casella, 2012; Muller, 2010)
  - d. Among pediatric CNS tumors, 6%–10% are of this type
  - e. Symptoms reflect the tumor's location: headache, visual impairment, decrease in growth, polydipsia, and polyuria attributable to the compression of the pituitary, optic chiasm, and hypothalamus (Harrington & Casella, 2012; Muller, 2010; Winkfield et al., 2011)
  - f. A sharply bordered mass that includes a combination of solid, cystic, and calcified tumor components is a classic radiologic finding (Muller, 2010).
  - g. Complete surgical resection offers a better prognosis.
  - h. Sequelae and quality of life should be taken into account, considering the eloquent location within the brain.
  - i. Proton beam RT
7. Choroid plexus neoplasms
- a. Intraventricular tumors of neuroectodermal origin (Ogiwara, Dipatri, Alden, Bowman, & Tomita, 2012)
  - b. More commonly found in the lateral ventricles (Bettegowda et al., 2012)
  - c. Among all pediatric brain tumors, 2%–4% are of this type (Bettegowda et al., 2012; Lafay-Cousin et al., 2011).
  - d. Two variants of choroid plexus tumors
    - i. Choroid plexus papilloma; WHO Grade I; surgery can be curative (Bettegowda et al., 2012; Lafay-Cousin et al., 2011)
    - ii. Choroid plexus carcinoma; WHO Grade III
      - a) Aggressive, vascular tumor, poorly delineated (Gopal, Parker, Debski, & Parker, 2008)
      - b) Has the potential to metastasize; therefore, imaging of the entire neuraxis is necessary (Gopal et al., 2008)
      - c) May be a manifestation of LFS (Gopal et al., 2008)
      - d) The infiltrative nature of the tumor precludes the ability to obtain a complete resection.
      - e) The role of adjuvant chemotherapy remains unclear.
      - f) Presenting symptoms of increased ICP are related to overproduction, obstruction, and impaired absorption of CSF (Bettegowda et al., 2012).
      - g) Presentation of symptoms in children younger than 1 year of age:

increased head circumference, bulging fontanel, separated cranial sutures, and vomiting

**Nursing recommendations**

1. Nurses should apply their knowledge of tumor origins, functional anatomy, and physiology when completing a patient assessment (Level 3).
2. Nurses should correlate anatomical location of the brain tumor with clinical condition when monitoring for neurologic improvement or deterioration (Level 3).
3. Nurses should apply knowledge of tumor classification, site, tissue involved, and histology as they guide patients and families through the care continuum (Level 3).
4. Nurses should integrate basic comprehension of the molecular and histological diagnosis of pediatric brain tumors into patient and family education regarding treatment protocols specific to tumor subtype in preparation for treatment (Level 3).

**III. Diagnostic Techniques**

**A. Computed tomography (CT)**

1. A CT scan uses ionizing radiation to produce tomographic images of the brain.
  - a. The use of ionizing radiation places patients at increased risk for developing cancer later in life (King et al., 2009; Pearce et al., 2012; Smith-Bindman et al., 2009).
  - b. CT scan provides fewer details of varying soft tissues.
    - i. CT is not ideal for use in initial diagnosis or disease progression or to differentiate brain tumors from other intracranial abnormalities.
    - ii. Frequently used as screen for acute changes in neurological status
    - iii. Often used to detect presence of calcium
      - a) Calcium is found in oligodendroglioma, neurocysticercosis, and craniopharyngioma.
      - b) Calcium can also be seen on susceptibility-weighted MRI sequence (Haacke, Mittal, Wu, Neelavalli, & Cheng, 2009).
2. Contrast medium-induced nephrotoxicity following a contrast CT scan is rare in a healthy child; however, it can occur within 3 days after receiving iodine-based contrast medium (Mitchell, Jones, Tumlin, & Kline, 2011; Thomsen & Morcos, 2005).

**Nursing recommendations**

1. Nurses should advocate that CT scans only be performed when warranted to reduce exposure to radiation and decrease risk for radiation-induced cancer (Level 2).
2. Nurses should provide patient and family education regarding the signs and symptoms of

acute renal failure if a patient receives a contrast CT in the outpatient setting (Level 2).

3. A licensed independent provider caring for the patient should be notified by the nurse if the patient demonstrates signs and symptoms of acute renal failure (Level 2).

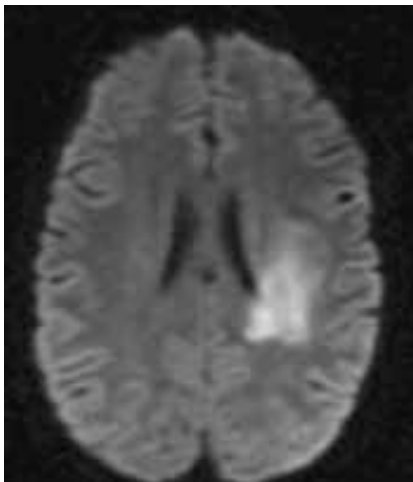
**B. Magnetic Resonance Imaging (MRI; Table 4)**

1. An MRI using gadolinium contrast medium is the most sensitive diagnostic test when evaluating a patient for a brain tumor because of its superior ability to provide contrast between the different soft tissues (Gooding, Brasch, Lallemand, Wesbey, & Brant-Zawadzki, 1984).
2. MRI technology uses a very large magnet and is the most cost-effective diagnostic study for children when a high suspicion for brain tumor exists (Medina, Kuntz, & Pomeroy, 2001).
3. Because of the strong magnetic field within the MRI machine, only MRI-compatible objects are allowed in the MRI suite.
  - a. Injury or death can occur if metal (ferrous-containing) objects are present.
  - b. There is a high potential for internal and external burns inflicted upon patients if they have metal objects inside or outside of their body (Shellock & Kanal, 1994).

<b>MRI</b>	<b>Best Used To</b>
Diffusion	Identify tumor grading and functional areas of the brain
Perfusion	Identify tumor grading and assess for progressive disease
Spectroscopy	Identify tumor type and grading
Angiography	Evaluate for obstruction or aneurysm
Functional	Map language and motor function
<b>Other</b>	
Positron emission tomography	Identify tumor grading, prognosis, evaluate response to therapy and tumor occurrence

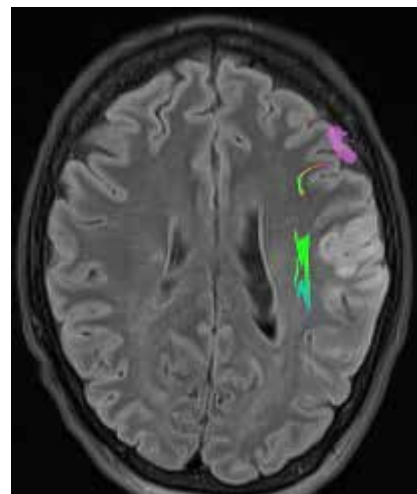
*Sources.* "Molecular imaging of pediatric brain tumors: Comparison of tumor metabolism using (1)(8)F-FDG-PET and MRSI," by S. J. Hipp, E. A. Steffen-Smith, N. Patronas, P. Herscovitch, J. M. Solomon, R. S. Bent . . . K. E. Warren, 2012, *Journal of Neuro-Oncology*, 109(3), 521–527. "Multimodal magnetic resonance imaging in the diagnosis and therapeutical follow-up of brain tumors," by A. Housni & S. Boujraf, 2013, *Neurosciences (Riyadh)*, 18(1), 3–10. "Clinical impact of MR spectroscopy when MR imaging is indeterminate for pediatric brain tumors," by J. F. Norfray, T. Tomita, S. E. Byrd, B. D. Ross, P. A. Berger, & R. S. Miller, 1999, *American Journal of Roentgenology*, 173(1), 119–125. doi:10.2214/ajr.173.1.10397111. "Toward functional neuronavigation: Implementation of functional magnetic resonance imaging data in a surgical guidance system for intraoperative identification of motor and language cortices. Technical note and illustrative case," by G. J. Rutten, N. Ramsey, H. J. Noordmans, P. Willems, P. van Rijen, J. W. Berkelbach van der Sprenkel . . . C. van Veelen, 2003, *Neurosurgical Focus*, 15(1), E6. "Incidence of vasculopathy in children with hypothalamic/chiasmatic gliomas treated with brachytherapy," by U. Tacke, D. Karger, J. Spreer, A. Berlis, G. Nikkhah, & R. Korinthenberg, 2011, *Child's Nervous System*, 27(6), 961–966. "Spectroscopic and perfusion magnetic resonance imaging predictors of progression in pediatric brain tumors," by A. A. Tzika, L. G. Astrakas, M. K. Zarifi, D. Zurakowski, T. Y. Poussaint, L. Goumnerova . . . P. M. Black, 2004, *Cancer*, 100(6), 1246–1256. "PET and brain tumor image fusion," by T. Z. Wong, T. G. Turkington, T. C. Hawk, & R. E. Coleman, 2004, *Cancer Journal*, 10(4), 234–242.

4. An MRI requires more time to complete than a CT scan and can be limited by the patient's movement.
  - a. Children who are prepared for procedures experience fewer negative side effects than children who do not receive any preparation (Kain, Mayes, & Caramico, 1996).
  - b. Children younger than 6–8 years of age or those who are developmentally delayed may require sedation or general anesthesia to complete the study (de Amorim e Silva, Mackenzie, Hallowell, Stewart, & Ditchfield, 2006).
5. Elevated patient anxiety during an MRI procedure may decrease quality of the images, increase sedation time and the number of tests performed, and increase risks for extended sedation and cost of the procedure (Hallowell, Stewart, de Amorim e Silva, & Ditchfield, 2008).
6. The settings of a programmable ventriculoperitoneal (VP) shunt may be inadvertently changed by the MRI magnet.
7. Nephrogenic systemic fibrosis has been reported in patients with renal insufficiency who are exposed to gadolinium contrast medium.
  - a. Usual elimination of gadolinium-containing products is about 2 hours; however, in patients with renal dysfunction, the elimination half-life is prolonged by 30–120 hours (Peak & Sheller, 2007).
8. Magnetic resonance (MR) diffusion
  - a. Diffusion-weighted imaging (DWI) uses the diffusion of water molecules in the specific section of the scan being studied to form quantitative values to calculate the apparent diffusion coefficient (ADC; **Figure 5**).
    - i. The ADC calculated by the DWI of the brain tumor allows for better tumor grading (Yamasaki et al., 2005).

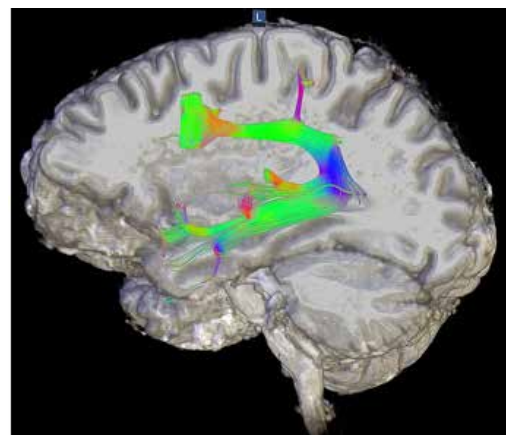


**Figure 5.** Diffusion-weighted imaging with area of restricted diffusion. Photo courtesy of Amar Gajjar, MD, St. Jude's Children's Research Hospital. Used with permission.

- ii. MR diffusion also encompasses diffusion tensor imaging and fiber tractography, which provide a three-dimensional (3-D) graphic representation of the tumor and important functional areas of the brain to better guide the surgeon in tumor resection and preservation of unaffected brain tissue (**Figure 6** and **Figure 7**).
- b. MR perfusion
    - i. Perfusion-weighted imaging (PWI) measures the cerebral hemodynamics at the microcirculation level, including cerebral blood volume (CBV), cerebral blood flow, and mean transit time.



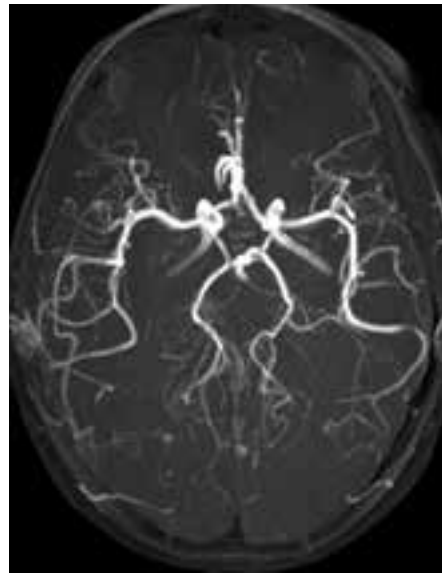
**Figure 6.** Axial fluid-attenuated inversion recovery image in a patient with a left inferior frontal tumor. Overlaid upon this image are functional MRI data showing expressive language (Broca's area, pink) and diffusion tensor imaging data showing the location of the white matter fibers connecting Wernicke's area to Broca's area, or the arcuate fasciculus. Photo courtesy of Asim Choudhri, MD, LeBonheur Children's Hospital. Used with permission.



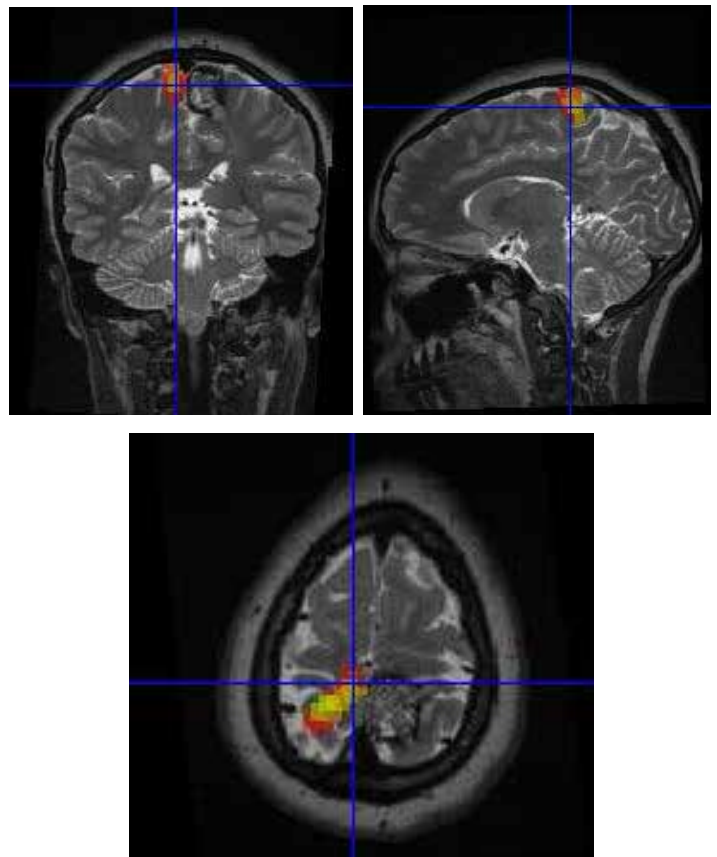
**Figure 7.** Sagittal three-dimensional (3-D) rendering of the brain with a cut-away showing the arcuate fasciculus, derived from diffusion tensor imaging data. The 3-D rendering shows the course of the fibers better than any single two-dimensional image can; however, detailed measurements are typically performed on a two-dimensional image. Photo courtesy of Asim Choudhri, MD, LeBonheur Children's Hospital. Used with permission.



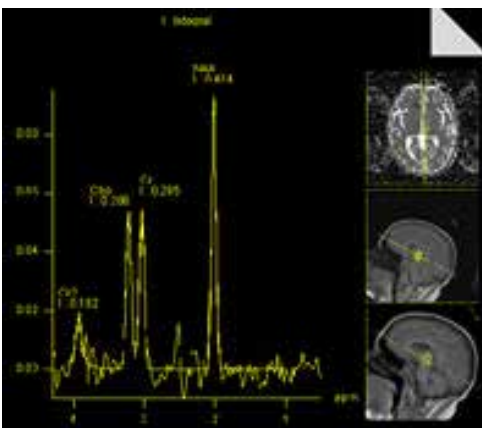
- ii. CBV is the most commonly evaluated parameter in patients with brain tumors (Covarrubias, Rosen, & Lev, 2004).
  - iii. This technique requires a bolus of intravenous (IV)-administered contrast medium followed by rapid sequencing of images (Lacerda & Law, 2009).
  - iv. The PWI is used to further guide tumor grading and identify progressive disease (Tzika et al., 2004).
- c. MR spectroscopy (**Figure 8**)
- i. MR spectroscopy uses the measurement of normal metabolites (N-acetylaspartate, choline, and creatine) and abnormal metabolites (lactate, lipids, and glutamine-glutamate complexes) found in the brain tumor and tissue.
  - ii. Provides complex neurochemical information to better identify the type and grade of a tumor (Norfray et al., 1999)
- d. MR angiography (**Figure 9**)
- i. This technique necessitates a bolus of IV-administered contrast medium.
  - ii. Images are acquired during the first pass of the contrast medium through the arteries.
  - iii. Evaluates for stenosis, obstruction, or an aneurysm that can be a result of external radiotherapy (Tacke et al., 2011)
9. Transverse MRI (**Figure 10**)
- a. Functional MRI (fMRI)
- i. fMRI is primarily used for the purpose of neurosurgical planning for resection of a brain tumor.
  - ii. Most commonly used to map language function
  - iii. Can also be used in motor mapping after performing cued movements (Rutten et al., 2003)
10. Positron emission tomography (PET; **Figure 11**)
- a. A PET scan is a noninvasive technique using radiotracers.



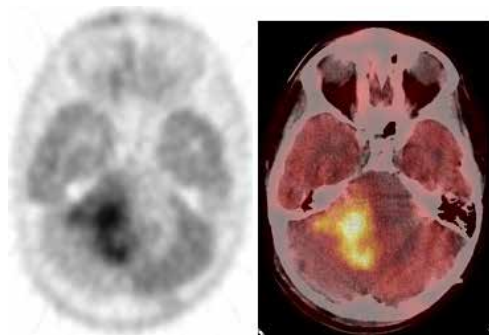
**Figure 9.** Magnetic resonance angiography. Photo courtesy of Amar Gajjar, MD, St. Jude's Children's Research Hospital. Used with permission.



**Figure 10.** Functional magnetic resonance imaging (fMRI) demonstrates location of motor movement of the leg. Photo courtesy of Jim Wheless, MD, LeBonheur Children's Hospital. Used with permission.



**Figure 8.** Magnetic resonance spectroscopy. Photo courtesy of Amar Gajjar, MD, St. Jude's Children's Research Hospital. Used with permission.



**Figure 11.** Transverse view of positron emission tomography indicating a brain tumor, demonstrating increased glucose metabolism. Images courtesy of Dr. Barry Shulkin, PhD, St. Jude Children's Research Hospital. Used with permission.

- b. Complements MRI imaging modalities by providing additional information that relates to the underlying cellular metabolism of a brain tumor
  - i. Evaluates a brain tumor for grading and prognosis and determines response to therapy and evaluation of tumor occurrence (Hipp et al., 2012; Wong, Turkington, Hawk, & Coleman, 2004)

#### Nursing recommendations

1. The patient should be assessed by the nurse prior to arrival into the MRI room to ensure that he or she is not wearing any metal and that there are no metal implants. This assessment will help to avoid potential harm or death (Level 3).
2. Nurses will ensure that a healthcare worker specially trained in MRI safety will perform an MRI screening assessment on the patient and any family member who will accompany the patient to the scanner prior to the exam and notify the ordering provider if the screening is positive for any metal devices (Level 2).
3. Nurses should educate patients regarding the large size of the MRI machine, the loud noise it makes, and the additional 10–15 minutes that some specialty MRIs necessitate. A child-life specialist should be consulted as needed to help provide age-appropriate education and distraction techniques (Level 2).
4. When the patient has a programmable VP shunt, the nurse will ensure that the setting has been verified, both prior to and after the MRI scan, and ensure that the shunt is reprogrammed after the MRI, if necessary (Level 3).
5. Before the imaging study begins, nurses should assess the patient for claustrophobia and their ability to lie still. The nurse should notify the provider if sedation or general anesthesia will be needed and administer sedation as prescribed (Level 2).
6. Serum blood urea nitrogen and creatinine levels should be drawn prior to a contrasted

MRI as ordered by the licensed independent practitioner, and abnormalities should be reported to the ordering provider (Level 2).

7. Nurses should ensure that a functional large-bore peripheral IV catheter is in place when a patient will be receiving IV contrast (Level 2).
8. Nurses should verify nothing-by-mouth (NPO) status as appropriate for the specific test being performed and notify and educate patients and families (Level 2).
9. Nurses will inform the patient that the person performing the scan may ask them to perform certain movements or lie still (Level 3).

#### C. Biopsy

1. A surgical procedure in which a sample of tissue is taken from the brain mass for closer examination
2. Currently the most accurate method for assessment and identification of brain tumors
3. A stereotactic biopsy uses a computer-based, 3-D image-guided system to direct the surgeon to the exact location of the tumor.
  - a. Stereotactic biopsy is advantageous over an open craniotomy because of its associated decrease in blood loss, faster recovery, and decreased risk for infection (Brady, Thornhill, & Colapinto, 1997; Prados, Berger, & Wilson, 1998).
  - b. Depending upon the stereotactic system used, a patient may require an MRI with specialized software and imaging sequences prior to the biopsy. Conversely, there may be availability of “real time” MRI in the operating room (OR) suite.

#### Nursing recommendation

1. Nurses should have a basic understanding of the surgical technique for biopsy so they can prepare/educate the patient and family on what to expect before and after surgery (Level 2).

### IV. Acute Care

#### A. Initial admission

Upon initial admission of a child with a newly diagnosed brain tumor, the nurse should perform

1. An initial head-to-toe assessment beginning with an airway, breathing, and circulation (ABC) assessment. If the patient is not maintaining a patent airway, spontaneous breathing, or circulation, assessment should cease, basic life support should begin immediately, and the cardiopulmonary resuscitation team and appropriate provider should be notified immediately.
2. A history and nursing assessment to allow for care planning and a baseline examination to be used as comparison during the postoperative period
3. A full neurological examination based on the child's age, including cranial nerve, motor, sensory, and cerebellar assessment

## B. Psychosocial assessment

A psychosocial assessment of the patient and family should be obtained. Assess for

1. Fear and anxiety
2. Patient or family understanding of surgery/treatment; request teach-back of the plan to confirm understanding.

## C. Involvement of a child-life specialist

A child-life specialist should be involved in the patient's care to ensure proper translation of the plan of care based on the child's developmental age.

## D. Continued close monitoring of neurologic status

1. Routine neurologic checks are completed preoperatively to obtain a current neurologic baseline.
2. For patients with tumors that cause hydrocephalus, neurological checks may be more frequent (Lacy, Saadati, & Yu, 2012).
  - a. The fontanel is a window into the brain for infants; palpation can help to detect increased ICP.
    - i. Bulging
    - ii. McEwen's sign: A dull cracked-pot sound suggests increased ICP.
  - b. Discuss any expected deficits the patient may have postoperatively with the neurosurgeon.

## E. Patient and family education and support

1. Trust in the medical staff during their child's diagnosis and hospitalization is an important aspect that promotes a positive hospital experience for families and helps parents cope (Jackson et al., 2007).

### Nursing recommendations

1. Nurses must obtain an assessment to become familiar with a patient's baseline neurologic status, which allows the ability for comparisons to detect any acute changes (Level 3).
2. If the patient is an infant, the nurse should include the fontanel as a part of the neurologic assessment for signs of increased ICP (Level 3).
3. Nurses should assess the family's and patient's level of understanding of the current diagnosis (Level 3).
4. Nurses should communicate openly and honestly using terms and methods appropriate to each family situation, taking into consideration education level, cultural background, and coping skills (Level 3).
5. Nurses should allow patients and family members to ask questions and provide answers as appropriate to the nurse's knowledge and role (Level 3).
6. Nurses should provide patients and families with writing materials so they may write down questions to prepare for their conversation with providers (Level 3).
7. Nurse should communicate next steps; for example, surgery, biopsy, pathology, and factors determining further treatment (Level 3).

## F. Emergency intervention

Emergent treatment may include an external ventricular drain (EVD) in the presence of hydrocephalus (Santos de Oliveira, Barros Juca, Valera, & Machado, 2008; Walker, Stone, Jacobson, Phillips, & Silberstein, 2012).

1. Emergent reduction of ICP may be required before tumor resection may be performed.
2. Commonly needed for posterior fossa tumors because of the proximity to the fourth ventricle and CSF pathways
3. Following tumor excision, the obstructive component of the hydrocephalus is relieved in most patients.
4. An EVD may be used in conjunction with corticosteroid therapy and early surgery.
5. An EVD is kept in place postoperatively to determine the need for a long-term shunt. Shunt determination is made by either raising the EVD or clamping it to evaluate patients' continued or ceased hydrocephalus.

### Nursing recommendations

1. Nurses should monitor patients for signs and symptoms of increased ICP (Level 3).
2. Nurses should manage the EVD to monitor ICP, maintain patency, and to drain CSF as prescribed (Level 3).

## G. Medications used during the acute phase

1. Corticosteroids for treatment of vasogenic edema: Vasogenic edema is caused by the disruption of the blood-brain barrier (BBB) by the tumor, resulting in increased capillary permeability. Plasma proteins and water-soluble substances move into the brain surrounding the tumor, resulting in edema (Batchelor & Byrne, 2006).
2. Corticosteroids are thought to decrease edema by decreasing the permeability of tumor capillaries, improving the integrity of the BBB (Jarden, Dhawan, Moeller, Strother, & Rottenberg, 1989; Sinha, Bastin, Wardlaw, Armitage, & Whittle, 2004).
3. In the absence of randomized controlled trials, clinical experience has created practice guidelines based on evidence supporting the use of corticosteroids in the management of metastatic brain tumors in adult patients, but not primary brain tumors and not in pediatric patients (Hempfen, Weiss, & Hess, 2002; Ryken et al., 2010; Sturdza et al., 2008; Vecht, Hovestadt, Verbiest, van Vliet, & van Putten, 1994).
  - a. Dexamethasone (Decadron) is the drug of choice for the treatment of vasogenic edema in symptomatic patients.
  - b. Dosing is customized for every patient to minimize side effects and maximize edema reduction.

- c. Should be tapered slowly to the lowest dose that maximizes neurologic function to prevent hypothalamic-pituitary-adrenal axis insufficiency (Zollner et al., 2012)
- d. Gastrointestinal (GI)-protective medications (H2 receptor antagonists such as ranitidine [Zantac] or proton pump inhibitors such as omeprazole [Prilosec]) are recommended for all patients taking corticosteroids to reduce the potential for ulceration or gastric bleeding.

## H. Seizures

1. Children with cancer have a higher incidence of seizures than adults with cancer (DiMario & Packer, 1990; Drane & Meador, 2002; Khan et al., 2005).
2. Conversely, in children with new-onset seizures, only 1%–3% have brain tumors (Sjors, Blennow, & Lantz, 1993; Williams, Abbott, & Manson, 1992).
3. Predominantly partial-onset with secondary generalization (Fattal-Valevski, Nissan, Kramer, & Constantini, 2013)
  - a. Attributable to areas of focal injury
  - b. May secondarily generalize if not treated or treated inadequately
4. Seizures may be an initial or presenting symptom, or may occur later in the disease or treatment (Packer et al., 2003).
5. Dependent upon histological type (based on adult data)
  - a. Less aggressive, more epileptogenic (Moots et al., 1995)
    - i. Dysembryoblastic neuroepithelial tumors (DNET): 100% of patients with DNET have seizures (Herman, 2002; Morrell & deToledo-Morrell, 1999).
    - ii. Gangliogliomas: 80%–90% of patients with gangliogliomas have seizures (Herman, 2002; Villemure & de Tribolet, 1996).
    - iii. Low-grade astrocytoma: 75% of patients with low-grade astrocytomas have seizures (Cascino, 1990; Villemure & de Tribolet, 1996).
    - iv. Glioblastoma multiforme: 29%–49% of patients with glioblastoma multiforme have seizures (Cascino, 1990; Herman, 2002).
  - b. Cortical brain tumors are more epileptogenic, especially frontal, temporal, and parietal lobes.
6. Current recommendations for antiepileptic drug (AED) use
  - a. Prophylactic use of AEDs is not recommended (Stevens, 2006).
  - b. Withdrawal of an AED within 1 week of surgery for patients who were started on an AED preoperatively or perioperatively but have not had a seizure is recommended (Stevens, 2006).
  - c. Surgeons should communicate the potential for electrolyte disturbances to all team members (Hardesty, Sanborn, Parker, & Storm, 2011).
    - i. Diabetes insipidus (DI), syndrome of inappropriate antidiuretic hormone (SIADH), and cerebral salt wasting (CSW) increase the risk for seizures attributable to rapid shifts in serum sodium. These conditions may be prevented with close monitoring and treatment.

Body system/symptom	Symptom and management
Gastrointestinal	Ulcer, gastritis; bleeding and/or perforation can occur without gastric-protective medication.
Immunosuppression	Poor wound healing; assess for thrush
Weight gain	Can occur because of increased appetite and fluid retention; encourage a healthy diet with reduced salt intake
Insomnia	When possible, schedule outside of nighttime sleep schedule.
Behavioral changes	Patients may experience mood swings and irritability.
Skin	Fragility, acne, bruising (attention to skin care is warranted)
Electrolyte/metabolic disturbances	Hyperglycemia, glycosuria, hypokalemia, hypocalcemia, hyperlipidemia. Assess these measures and provide dietary guidance and supplements as appropriate.
<i>Sources:</i> "Complications of brain tumors and their treatment," by J. Lacy, H. Saadati, & J. B. Yu, 2012, <i>Hematology/Oncology Clinics of North America</i> , 26(4), 779–796. <i>Physicians' Desk Reference</i> (67th ed., text rev.) by Physicians' Desk Reference Staff, 2012, Montvale, NJ: PDR Network.	

4. Further research is needed to determine dosage, accepted side effects, and length of treatment.
5. Common steroid side effects (**Table 5**)

### Nursing recommendations

1. Nurses should use their knowledge of the potential side effects of steroid use in pediatric patients with brain tumors and provide ongoing assessment of possible symptoms, applying that knowledge to assess for complications associated with their use (Level 3).
2. Nurses should communicate any signs or symptoms that may indicate a need for steroid dose change with the healthcare team to assist in determining treatment duration and/or symptom management (Level 3).
3. Nurses should educate patients and/or family members regarding known side effects and their symptoms and assist them in symptom management (Level 3).
4. Nurses should teach patients and/or families that abrupt discontinuation of corticosteroids must be avoided, and that a taper schedule must be followed as prescribed (Level 2).

- d. An AED may be weaned within 3 months of surgery in children who experience preoperative seizures.
- e. Patients at high risk for seizure recurrence include those with (Das et al., 2012)
  - i. Tumor recurrence
  - ii. Tumor location in the temporal lobe
  - iii. Incomplete resection
  - iv. Structural lesions (compared with patients with idiopathic seizures; Berg et al., 2004).
- f. Children who go on to experience epilepsy may be weaned after a 2-year seizure-free period.
- g. If an AED is indicated, nonenzyme-inducing AEDs such as levetiracetam (Keppra), lamotrigine (Lamictal), and gabapentin (Neurontin) are preferred (Maschio, Dinapoli, Mingoia, et al., 2011; Maschio, Dinapoli, Sperati, et al., 2011; Vecht & van Breemen, 2006).
  - i. Enzyme-inducing AEDs such as carbamazepine (Tegretol), phenytoin (Dilantin), and phenobarbital increase drug-drug interactions. Enzyme-inducing AED use may be prohibited by research protocols (especially Phase I and II studies) because of potential drug-drug interactions (Broniscer et al., 2013; Pollack et al., 2011).
- h. Valproic acid (Depakene, Depakote) may have antitumoral effects, but thrombocytopenia and platelet dysfunction may occur, increasing the chance of intratumoral bleeding (Su et al., 2011; Weller et al., 2011).
  - i. Valproic acid is highly teratogenic, making it a significantly undesirable choice for females who have started their menses.
- i. An ideal AED should have minimal side effects, no drug-drug interaction, and be available in an IV formulation. At this time, levetiracetam (Keppra) is a common choice because it is the only AED that has these attributes (Usery, Michael, Sills, & Finch, 2010).
  - i. Levetiracetam use does not necessitate the drawing of levels to determine efficacy.
  - ii. Can cause behavior changes such as irritability, mood swings, and irrational behavior (Kern et al., 2012)
- j. Because AEDs have not been studied directly with chemotherapeutic agents, serum drug levels of AEDs are indicated (Singh, Rees, & Sander, 2007).
- k. Phenytoin and/or fosphenytoin (Dilantin) is also commonly used, especially for acute, prolonged seizures; however, levels need to be monitored for efficacy if it continues to be used for maintenance dosing.
  - i. Available in oral or IV formulations
    - a) The IV formulation of phenytoin lacks solubility, which may result in precipitation if administered with other medications or if the IV line is not flushed well.
    - b) The IV formulation of phenytoin may cause local tissue death if it leaks through the vein and should only be used when a central line is available.
    - c) Has the potential to cause cardiovascular toxicities such as hypotension, bradycardia, ventricular tachycardia, and ventricular fibrillation if infused at a high rate (Osborn, 2004)
  - ii. Levels must be drawn at appropriate times in relation to the time of administration, per provider orders (Kern et al., 2012).
  - iii. Adverse effects
    - a) May occur more frequently in patients with cancer (Drane & Meador, 2002)
    - b) Higher likelihood of Stevens-Johnson syndrome and toxic epidermal necrolysis, especially during the first 4–8 weeks of dose escalation (Delattre, Safai, & Posner, 1988)
    - c) Associated with the coadministration of phenytoin and carbamazepine during RT (Delattre et al., 1988; Hoang-Xuan, Delattre, & Poisson, 1990; Mamon, Wen, Burns, & Loeffler, 1999)

#### Nursing recommendations

1. Nurses should be aware of the potential risk and individual risk factors for seizures in pediatric patients with brain tumors and provide appropriate seizure care (Level 2).
2. Nurses should monitor for the side effects and drug interactions of AEDs used in children with newly diagnosed brain tumors, particularly in patients receiving multiple classes of medications (Level 1).
3. Nurses should educate patients and families about compliance with AED therapy, drug interactions, and potential side effects (Level 1).

## V. Surgery

### A. Primary goal

Surgery is a primary treatment for pediatric brain tumors. The goal of surgery is to remove as much tumor as possible, to improve survival and allow for other treatment modalities, and to obtain a tissue diagnosis (Lassen et al., 2012).

1. Types of procedures
  - a. Gross total resection in children with low-grade gliomas resulted in 5-year

progression-free survival exceeding 90%.

Children with less extensive tumor resection had disease progression (Wisoff et al., 2011).

- b. Advances in image-guided tumor localization, treatment planning, functional brain mapping, intraoperative imaging, and neurophysiological monitoring have improved the safety of deep-seated tumor resections (Pollack, 2011).
  - c. Stereotactic tumor resection of thalamic juvenile pilocytic astrocytomas with diffusion tensor imaging minimized neurological morbidity and improved ability to attain complete resection (Moshel, Elliott, Monoky, & Wisoff, 2009; **Figures 12a** and **Figure 12b**).
  - d. Brain stem gliomas can usually be diagnosed with MRI, diminishing the role of biopsy (Albright et al., 1993).
2. A neuroendoscopic biopsy is associated with acceptable risk for obtaining tissue from periventricular and intraventricular tumors (Song, Kong, & Shin, 2010).
  3. “Second look” or staged surgical resection often is used to increase extent of tumor removal while limiting morbidity related to surgery (Shiminski-Maher, 2013).

#### Nursing recommendations

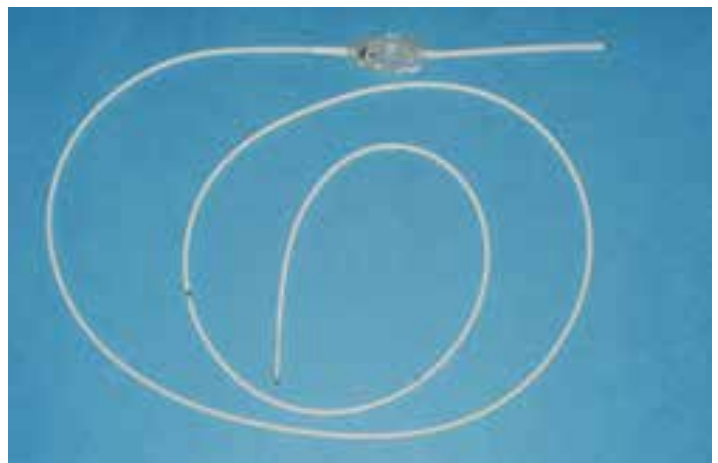
1. Because gross total resection of low-grade gliomas, if achieved with acceptable functional outcomes, offers the best chance for progression-free survival, nurses should emphasize preoperatively that technology (e.g., image guidance, functional brain mapping, intraoperative imaging,



**Figure 12a.** The figure shows a neurosurgeon making a burr hole through the skull. **Figure 12b** shows the long needle inserted into the brain to obtain the biopsy. The patient's head has a frame attached that allows a specific trajectory into the tumor. Photos courtesy of Cathy Cartwright. © 2013. Used with permission.

neurophysiological monitoring) has improved the safety of tumor resection, and gross total resection improves survival (Level 1).

2. When performing preoperative education with parents, nurses will use the knowledge that the most important prognosticator for outcome in children with most types of malignant brain tumors is the extent of the tumor removal. This education will provide important information when discussing preoperative preparation and surgical outcomes with the parents and patient (Level 3).
  3. Nurses should know that brain stem gliomas can usually be identified noninvasively with the use of MRI and plan the patient's care accordingly (Level 3).
  4. Nurses should be aware of the need for staged surgical resection because patients and families will need preparation and support if they are to undergo more than one surgical procedure (Level 3).
- #### B. Shunts (Figure 13)
1. Of 131 children with malignant astrocytomas, 19 had preoperative or intraoperative ventriculostomy.
    - a. Twenty-two percent of deep tumors necessitated ventriculostomy compared with only 6% of superficial hemisphere neoplasms (Wisoff et al., 1998).
    - b. One-half of the deep tumors and 8% of the hemispheric astrocytomas ultimately necessitated a permanent CSF shunt.
  2. Of 1,250 cases of primary pediatric brain tumors in patients younger than 18 years of age, 56.7% of patients presented with hydrocephalus—51% at tumor diagnosis and 5.1% after tumor diagnosis (Wong, Liang, Chen, & Chang, 2011).
    - a. Ninety-eight percent of hydrocephalus was obstructive.
    - b. Of the children with hydrocephalus, 54.4%



**Figure 13.** Ventriculoperitoneal shunt. Photo courtesy of Cathy Cartwright. © 2013. Used with permission.

had a VP shunt, 10.9% had an endoscopic third ventriculostomy (ETV) shunt, 4.8% had a subdural-to-peritoneal shunt, 0.7% had a septostomy, 0.6% had a lumbar peritoneal shunt, and 0.1% had a ventriculoatrial shunt.

- c. Fifty-two patients with craniopharyngioma with a single cyst had an Ommaya reservoir placed on the day of surgery and the cyst aspirated. Over 7 years' follow-up, 38 (73%) did not develop recollection of the cyst and showed significant clinical improvement (Moussa, Kerasha, & Mahmoud, 2013).

#### **Nursing recommendations**

1. During parent/patient education, nurses will mention in their discussion that fewer than 25% of children with malignant tumors require postoperative shunting based on information obtained in the late 1990s (Level 1).
2. Nurses will teach parents/patients that during the past 2 decades, the VP shunt has been used more cautiously for obstructive hydrocephalus. Instead, more radical tumor resections and more frequent use of ETV are performed to reduce shunting (Level 2).
3. Nurses should be aware of signs and symptoms of hydrocephalus when assessing a child with a newly diagnosed brain tumor and closely monitor these children for early signs of neurological deterioration (Level 3).
4. Nurses should know that children with programmable valves must have their settings rechecked after MRI because the MRI magnet may alter the settings. Nurses will ensure that the need for shunt valve setting verification is communicated to all team members, and especially to the person responsible for performing the shunt reprogramming (Level 2).
5. Nurses will explain to parents that an Ommaya reservoir may be used to aspirate any recurrence of the cystic portion of the craniopharyngioma (Level 3).

#### **C. Postoperative complications**

1. Surgery for pediatric brain tumors is becoming safer as mortality rates decrease.
  - a. The most common complications are CSF leak leading to postoperative meningitis, wound infections, hematomas, and neurological deficits (Lassen et al., 2012).
  - b. In one study, a better neurological status was observed postoperatively in 23% of patients (27 of 117) and worse status in 7.6% (9 of 117). Infections were the most frequent complication (10.3%), followed by CSF leaks (< 10%) and hemorrhage (6%; Houdemont et al., 2011).

#### **Nursing recommendations**

1. Nurses should reassure patients/parents that in regard to surgery for brain tumor

resection, benefits generally outweigh risks. They should reinforce that surgery allows for diagnosis, symptom improvement, and reduction of tumor burden so other treatments such as radiation and chemotherapy are more effective (Level 3).

2. Nurses should carefully observe for signs of infection (fever, redness, discharge, swelling at incision site, stiff neck, photophobia) and hemorrhage (decreasing hemoglobin, increased heart rate, decreased blood pressure) and increased ICP (decrease in level of consciousness, bradycardia, hypertension; Level 3).
  3. Nurses should report CSF leaks to the patient's medical team immediately (Level 3).
- b. Cerebellar mutism syndrome (CMS)
    - i. In one study, 9.2% of children with posterior fossa tumors developed akinetic mutism (Houdemont et al., 2011).
    - ii. Patients who developed CMS after surgery have significantly lower performance in processing speed, attention, working memory, executive process, cognitive efficiency, reading, spelling, and mathematics (Palmer et al., 2010).

#### **Nursing recommendation**

Careful follow-up for patients with CMS includes neuropsychological evaluation and critical support for patients and their families, and nurses should collaborate with the multidisciplinary team to provide these services (Level 3).

- c. Electrolyte disturbance (Madden, Dobyns, Handler, & Foreman, 2010).
  - i. In this study, 30% of patients with suprasellar, thalamic, or hypothalamic tumors had hyponatremia, and 8% had hypernatremia.
  - ii. Thirty-eight percent had disordered serum sodium levels from the perioperative through postoperative periods.
  - iii. No abnormal sodium levels occurred postoperatively in patients with posterior fossa tumors.

#### **Nursing recommendations**

1. Nurses should collaborate and plan with neurosurgery, endocrine, and intensive care providers to monitor electrolytes after surgery in patients with brain tumor who undergo craniotomy, particularly patients who have suprasellar (i.e., germinoma, craniopharyngioma), thalamic, or hypothalamic tumors (Level 2).
2. Exogenous vasopressin/antidiuretic hormone, desmopressin acetate (DDAVP)

may be ordered for patients with DI, and fluid restriction may be required for patients with SIADH. Nurses will monitor intake and output, daily weights, and laboratory results such as urine and serum sodium and osmolality as ordered by the licensed healthcare provider (Level 3).

3. Nurses will administer DDAVP as ordered and monitor the outcomes of each treatment (Level 2).
- d. Dysphagia outcome: Eleven children ages 3 years 6 months to 13 years 5 months were assessed preoperatively with a bedside dysphagia screening tool before and after resection of a posterior fossa tumor (Morgan, Sell, Ryan, Raynsford, & Hayward, 2008).
  - i. No child had dysphagia before surgery, but 73% (8/11) had some dysphagia at 1–2 weeks postsurgery.
  - ii. At 2 months postsurgery, 75% could manage a full oral diet.

#### **Nursing recommendation**

Nurses should be aware that children who undergo posterior fossa tumor resection are at risk for aspiration and should be considered for evaluation by speech pathology to detect any swallowing impairment before attempting oral feedings. Nurses should collaborate with the provider to obtain a swallow evaluation as needed (Level 3).

- e. Lower cranial nerve dysfunction: Forty-one children and adolescents underwent radical resection for intrinsic medullary tumor (Jallo et al., 2005).
  - i. Fifteen previously underwent surgery.
  - ii. Twenty-two (54%) did not require a tracheostomy and/or gastrostomy tube postoperatively, but 19 (46%) had lower cranial nerve dysfunction requiring placement of a tracheostomy and/or gastrostomy tube.
  - iii. Some required ventilator support.
  - iv. All 19 patients had preoperative evidence of cranial nerve dysfunction such as choking, pneumonia, and voice changes.
  - v. On follow-up, 13 (68%) had complete recovery of lower cranial nerve function, 2 (11%) had significant improvement, and 4 (21%) had no lower cranial nerve function.

#### **Nursing recommendation**

Nurses should know that children with intrinsic intramedullary tumors have lower cranial nerve dysfunction. With this knowledge, nurses should participate in discussions with parents about the possibility of

postoperative tracheostomy and gastrostomy tube placement, but should also reassure them that most patients recover their lower cranial nerve function (Level 3).

#### **D. Rehabilitation**

1. Significant improvements have occurred in areas of self-care, locomotion, and mobility from rehabilitation admission, discharge, and follow-up. Communication and social cognition also showed improvement in follow-up (Philip, Ayyangar, Vanderbilt, & Gaebler-Spira, 1994).
2. Children and adolescents with CNS tumors (n = 190) were screened for speech-language and hearing disorders, and 81% were affected by some type of speech-language and/or hearing disorder (Goncalves, Radzinsky, da Silva, Chiari, & Consonni, 2008).

#### **Nursing recommendations**

1. Nurses should support and encourage appropriate rehabilitation referral services to improve function after brain tumor resection (Level 3).
2. Nurses should collaborate with the provider to obtain a speech-language hearing screen/evaluation as needed (Level 3).

#### **E. Preparation for surgery**

The separation of child and parents in the preoperative setting can add to the anxiety of both the child and the parents. In one prospective randomized control study of 103 participants, parental presence during induction of anesthesia in addition to 0.5 mg/kg of oral midazolam (Versed) had no additive effects in terms of reducing a child's anxiety, but parents who accompanied their children to the OR were less anxious and more satisfied (Kain et al., 2000).

#### **Nursing recommendation**

1. Allowing parents to accompany their child into the OR for induction of anesthesia in addition to premedicating the child with midazolam can decrease the parents' anxiety and increase their satisfaction (Level 1).
2. Nurses should advocate for parents to accompany their child into the OR and prepare the parent for what they may observe (Level 1).

#### **F. Preoperative shaving (Tang, Yeh, & Sgouros, 2001)**

1. Ninety pediatric patients 7–16.8 years of age were involved in a prospective nonrandomized study of 100 consecutive neurosurgical procedures. These patients were divided into hair-shaved and no-hair-shaved groups.
2. Wound dehiscence (n = 4) occurred in two patients in the hair-shaved group and two in the no-hair-shaved group.
3. There were two shunt infections in the hair-shaved group and one in the no-hair-shaved group.



### **Nursing recommendations**

1. When performing preoperative teaching, the nurse should explain that shaving the head is not necessary for wound healing and is done by physician preference (Level 3).
2. If the child is anxious about loss of hair due to preoperative shaving, the nurse should encourage the child and family to talk with the neurosurgeon about their concerns before surgery (Level 3).
3. The OR nurse may consider saving a small portion of a child's shaved hair to give to the parents, particularly if the child is an infant because this may be his or her first haircut (Level 3).

### **G. Pain**

In a study of 52 pediatric patients undergoing craniotomy, postoperative pain, analgesic regimens used, and factors associated with significant pain were assessed. The duration of procedure was the only factor associated with parenteral morphine use for more than 24 hours, and older age was the only factor associated with having an episode of pain scoring higher than 3 on a scale of 0 to 10, with 0 being no pain and 10 being maximum pain (Teo, Palmer, & Davidson, 2011).

### **Nursing recommendations**

1. Nurses should carefully assess postoperative pain in patients who undergo craniotomy and consistently use age-appropriate pain scales (Level 3).
2. Nurses should recognize that older children or those who undergo lengthy procedures may need more medication to control their pain and administer analgesics accordingly (Level 3).

### **H. Venous thromboembolism (VTE) prevention**

1. The rarity of VTE events in children and the lack of controlled, trial-based evidence have led to few indications for VTE prophylaxis in the pediatric population.
  - a. Much of what is known about VTE prophylaxis is based on findings from adult patient studies and extrapolated for pediatric use (Van Wicklin, 2012).
  - b. Anticoagulant prophylaxis may be advisable for children at high risk based on underlying conditions and type and length of surgery.

### **Nursing recommendations**

1. Nurses should assess risk factors of VTE in children with brain tumors, including immobility, prolonged bed rest, infection, obesity, and neurological surgery and take appropriate precautions (Level 3).
2. Nurses should implement prophylaxis such as early ambulation, adequate hydration, graduated compression stockings, and intermittent pneumatic compression devices (Level 3).
3. Nurses should discuss anticoagulant prophylaxis with the provider for children at high risk for VTE (Level 3).

## **VI. Treatment**

### **A. Risk-adapted treatment strategy**

1. RT is delivered to children with brain tumors in accordance with a risk-adapted treatment strategy (Pollack, 2011).
  - a. Risk-adapted treatment strategy is targeted treatment modulated to an individual patient's predefined risk.
  - b. A patient with low-grade tumor may be given lower-dose radiation therapy in a more targeted area, while a patient with a high-grade tumor may be given a larger dose in a larger area.
2. Age consideration
  - a. The management of malignant brain tumors in children younger than 3–5 years of age has historically incorporated somewhat different strategies than used for comparable tumors in older children because of the particular sensitivity of the young brain to the toxic sequelae of RT (Pollack, 2011).
  - b. Justifiable concerns about the toxicity of radiation in young children may contribute to the higher mortality seen in infants with brain tumors than in older children (Bishop, McDonald, Chang, & Esiashvili, 2012).

### **Nursing recommendations**

1. Nurses should recognize that children younger than 3 years of age with brain tumors have overall worse prognostic outcomes. Nurses should apply this knowledge to educate parents that intensive chemotherapy/biotherapy are often given to avoid or delay RT to the developing brain (Level 3).
2. Although certain malignant diagnoses (e.g., ependymoma) necessitate RT in the younger-than-3 age group, nurses should anticipate early signs of learning delay and assist parents in referral for neurocognitive evaluation, audiology, and neuro-ophthalmology follow-up (Level 3).
2. Types of radiation
  - a. Radiation for pediatric brain tumors is usually delivered either to the primary tumor site (involved field) or to the entire cranio-spinal axis (for tumors with an underlying tendency to metastasize throughout the CNS; Robertson, 2006) via external beam RT (Chalabi & Patel, 2009).
  - b. External beam radiotherapy can be directed from a linear accelerator (i.e., with photons) in one (i.e., radiosurgery) or multiple (i.e., intensity-modulated RT) directions.
  - c. RT can also be delivered via a cyclotron (proton therapy) that deposits the majority of energy at a designated depth with less "drop off" or exit dose than conventional radiation (Chalabi & Patel, 2009; Swift, 2002).

**Table 6. Potential Acute Effects of Radiation Therapy**

System Affected	Side Effect	Care Strategies
Integumentary	Skin erythema, hyperpigmentation, ulceration and desquamation, alopecia	Vaseline/petroleum jelly, lotion (only after radiation therapy each day), cut hair short, hats, wigs
Nervous	Hypothalamic dysfunction, pituitary dysfunction, headache, somnolence/fatigue	Endocrine referral, adherence to prescribed medications, rest
Ophthalmologic	Dry eyes, blurry vision	Eye drops
Cardiac	Pericarditis	Monitor for signs and symptoms of chest pain or discomfort
Pulmonary	Radiation pneumonitis	Monitor for signs and symptoms of chest pain or shortness of breath; try relaxation techniques for improved ventilation
Gastrointestinal	Mucositis, esophagitis, nausea, vomiting, diarrhea, abdominal cramping, anorexia, malnutrition	Try new foods of all varieties, mouth rinse after meals (glutamine, lidocaine, chlorhexidine gluconate), lozenges, adherence to prescribed medications

The data in column 3 are summarized from "Symptom monitoring and dependent care during cancer treatment in children: Pilot study," by Williams, P. D., Schmideskamp, J., Ridder, E. L., & Williams, A. R., 2006, *Cancer Nursing*, 29(3), 188–197.

- d. Brachytherapy involves a radioactive source that is placed inside or next to the area requiring treatment. It is still an emerging form of therapy for pediatric solid tumors and is not often used (Chalabi & Patel, 2009).
- e. Radiosensitizers are used to increase tumor sensitivity to radiation or decrease radiation resistance.
  - i. Pretreatment with chemotherapeutic agents such as carboplatin (Paraplatin) or biologic response-modifiers such as cetuximab (Erbix) can enhance the effect of irradiation on tumor cell killing (Robertson, 2006).
  - ii. These are also used to maximize the benefit of lower doses of radiation (Swift, 2002).

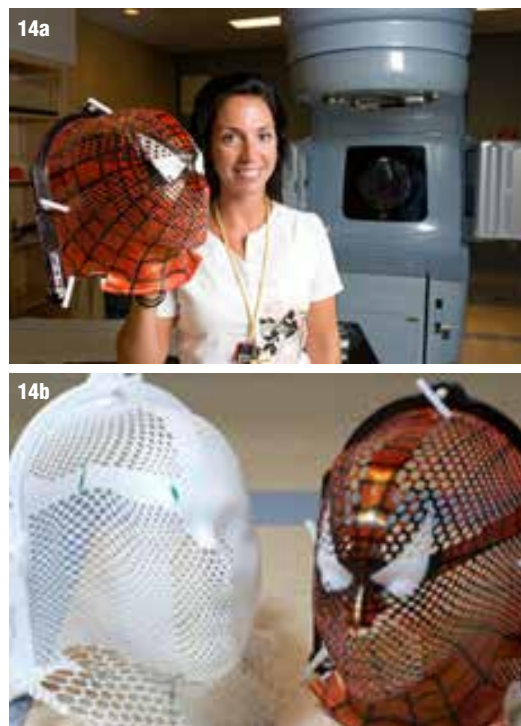
#### Nursing recommendations

1. Nurses should be able to distinguish between the various methods of therapy that deliver radiation to the brain and the ways in which these methods may affect normal tissue surrounding the treatment area. They should use this knowledge to develop appropriate nursing care interventions (Level 3).
2. Nurses should teach patients/families to anticipate side effects dependent upon the type of radiation being administered (Table 6; Level 3).
3. Potential RT complications (Table 6)
  - a. Immobilization is required for delivery of external beam radiotherapy (Klosky et al., 2004). Anesthesia is often necessary for young children who are unable to lie motionless. Examples of sedation include general anesthesia, IV (conscious) sedation, oral sedation, or any combination of these.
  - b. Successful alternatives to sedation include the use of interactive intervention and cognitive behavioral strategies that can alleviate anxiety and distress, improve physiologic findings (including heart rate), and decrease risks for complications associated with anesthesia.

- c. Immobilization masks are painted to resemble superhero masks in an attempt to alleviate anxiety and provide a means of imaginative play during RT (Figure 14a and Figure 14b).

#### Nursing recommendations

1. Nurses should articulate their role in providing continuity, alleviating anxiety, and managing potential side effects associated with RT (Level 3).
2. Nurses should advocate for child-life consultation during the simulation period, which



**Figure 14a and Figure 14b.** Examples of painted immobilization masks used during external beam radiotherapy to reduce anxiety and provide imaginative play during radiation therapy. From "London program helps mask their fears" by J. Sher. *Woodstock Sentinel Review*. Retrieved from [www.woodstocksentinelreview.com/2012/09/05/london-program-helps-mask-their-fears](http://www.woodstocksentinelreview.com/2012/09/05/london-program-helps-mask-their-fears). © Derek Ruttan / QMI Agency. Used with permission.

will serve as a practice session for therapy as well as during RT (Level 3).

3. Nurses should provide daily continuity with interactive interventions and play therapy when child-life personnel are not available (Level 3).
4. Nursing care
  - a. Young children who require sedation will also require strict adherence to NPO status each night before RT (Chalabi & Patel, 2009); NPO status can be complicated by nausea and vomiting-associated dehydration.
  - b. IV hydration may be necessary before or after RT (McFadyen, Pelly, & Orr, 2011).
  - c. Antiemetics should be administered aggressively during RT, especially when delivering doses to the craniospinal axis, which targets the abdomen as well (Chalabi & Patel, 2009).
  - d. Maintaining normal or even elevated hematocrit levels by way of packed red blood cell transfusion in patients undergoing irradiation is important. Ensuring normal tissue oxygenation is of particular importance when using combined modality regimens that include myelosuppressive chemotherapy that could result in significant anemia during RT (Finlay, Uteg, & Giese, 1987).

#### **Nursing recommendations**

1. Patient and family education should include NPO instructions. Nurses should instruct the child and family that the child will not be able to eat or drink prior to sedation with RT (Level 3).
  2. Nurses should maintain levels of competency in administering antiemetics, IV fluids, and blood products during the period surrounding RT to provide safe and effective care to their patients (Level 3).
  3. Nurses should encourage the use of IV antiemetics for children receiving cranial and craniospinal radiation (Level 3).
  4. Nurses should monitor for signs of low hemoglobin and hematocrit levels such as headache, fatigue, and pallor and administer packed red blood cells in patients per complete blood count results and as prescribed (Level 3).
5. Patient-family education topics (McFadyen et al., 2011)
    - a. If the child develops symptoms of upper respiratory infection or intractable vomiting during the course of RT, the risks and benefits of proceeding with anesthesia and RT should be weighed in collaboration with the radiation oncology team and parents.
    - b. Weight gain from concurrent use of corticosteroids may affect the fit of the immobilization

mask, which may necessitate a new simulation and creation of a new mask to maintain an accurate treatment path.

- c. Parents can apply local anesthetic cream or a patch to the port site before arrival to the radiation facility to minimize pain associated with accessing the central venous access port (Chalabi & Patel, 2009; Klosky et al., 2004).

#### **Nursing recommendations**

1. Nurses should help parents to prepare their children for RT and anticipate potential stressors to reduce the child's overall distress (Level 3).
2. Nurses should help parents advocate for anesthetic creams for needle sticks/port access (Level 3).
3. Nurse should maintain communication with the child's oncologist regarding corticosteroid taper in relation to the fit of the immobilization mask (Level 3).

#### **B. Chemotherapy**

1. Since the introduction of chemotherapy more than 50 years ago, the prognosis for childhood cancer has improved dramatically (Adamson et al., 2011).
2. Anticancer drugs are most effective when administered in an adjuvant setting in conjunction with local control with surgery or radiation.
  - a. Types of chemotherapy (Nixon & Rae, 2011)
    - i. IV chemotherapy is often administered through a central venous catheter (e.g., peripherally inserted central catheter, implanted port, external tunneled catheter) because some chemotherapy medications are vesicants and cannot be administered through a peripheral IV catheter because of risk for tissue damage. Blood return must be confirmed before infusion of parenteral chemotherapy.
    - ii. Intrathecal chemotherapy can be administered through a lumbar puncture or a ventricular or lumbar reservoir (i.e., Ommaya reservoir) by a licensed physician or credentialed midlevel provider.
    - iii. Oral chemotherapy can be administered as a liquid, tablet, or capsule. The family can administer these medications in the home setting, and this may contribute to a sense of control for the patient or family.
    - iv. Nurses, all healthcare providers, and families should use safe-handling practices for any form of chemotherapy and its resultant hazardous waste products in accordance with clinical practice guidelines, including the use for adequate personal protective equipment, including gloves, gown, and goggles (Connor

& McDiarmid, 2006; Polovich & Martin, 2011). This is especially true for pregnant or lactating women.

#### **Nursing recommendations**

1. Nurses should demonstrate competency in the administration and handling of all forms of chemotherapy to ensure the highest level of safe care (Level 3).
  2. Nurses should follow safe-handling procedures and educate families to follow safe-handling procedures when administering or handling any form of chemotherapy or its waste products (Level 3).
- b. Basic classes of chemotherapeutic agents and mechanisms of actions (Ettinger & Rale, 2011)
- i. Classification is determined, in part, by whether or not the agent is
    - a) Cell cycle specific, which exerts effects during a specific phase of the cell cycle, with the maximal effect on rapidly dividing cells.
    - b) Cell cycle nonspecific, which acts on the cell regardless of cell cycle phase, and is dose-dependent with activity even on a cell that is slow growing or resting.

#### **Nursing recommendation**

1. Nurses should have the knowledge to classify chemotherapy and biotherapy into basic categories as necessary to understand the mechanisms of action, side effect profiles, and supportive care interventions (Level 3).
  2. Nurses should use this knowledge during administration of these medications to monitor for side effects during therapy (Level 3).
- c. Recurrences in pediatric neuro-oncology are often difficult to treat and cure. When conventional chemotherapy and RT fail to improve the prognosis for these children, providers will often explore more molecularly targeted treatment strategies as well as novel therapies (Pollack, 2011).
- d. Patients are often eligible for Phase I/II clinical trials with novel agents when other more conventional methods have failed (Wills, 2011).
- i. Phase I studies have a primary goal of determining the maximum tolerated dose and dose-limiting toxicities of an investigational drug or combination of drugs using a recommended administration schedule.

- ii. Phase II studies determine the efficacy of a new agent in the treatment of specific disease types and validate toxicity and dosage schedule. Information about drug administration, acute toxicity, and supportive care is also obtained during these trials.

#### **Nursing recommendations**

1. Nurses should recognize the goals of treatment, especially for patients who are relapsing/refractory, and use this knowledge to effectively communicate with patients and families, maintaining hope, but not offering false expectations. Nurses should reinforce that enrollment in a trial may or may not occur with intent to treat or cure disease (Level 3).
  2. Nurses should be able to distinguish goals for treatment when interacting with the patient and family and communicate and formulate interventions appropriately (Level 3).
- e. Nursing care of the child with a brain tumor
- i. Anticipatory guidance (Rodgers et al., 2012)
    - a) Chemotherapy-induced nausea and vomiting is a common distressing side effect of treatment. Among school-age children, 28%–60% report nausea, and 5%–38% report vomiting. The occurrence of delayed chemotherapy-induced nausea/vomiting is more prevalent than previously thought, and nurses often fail to assess for and appropriately medicate during this period.
    - b) Antiemetics for chemotherapy-induced nausea and vomiting may include oral or parenteral forms of ondansetron (Zofran), lorazepam (Ativan), phenothiazine (Phenergan), and dexamethasone (Decadron). Dexamethasone should be avoided as a premedication or prechemotherapy antiemetic in patients with brain tumors because of the ability of dexamethasone to decrease the permeability of the BBB, resulting in decreased efficacy of the chemotherapy (Hedley-Whyte & Hsu, 1986).

#### **Nursing recommendations**

1. Nurses must be cognizant of the need to assess for both immediate and delayed chemotherapy-induced nausea and vomiting and assess this issue often and thoroughly (Level 2).

<b>Table 7. Chemotherapeutic Agents Administered in the Treatment of Pediatric Brain Tumors and Common Side Effects</b>		
<b>Alkylating Agents</b>		
Commonly seen in the treatment of pediatric central nervous system tumors; myelosuppression is a common dose-limiting toxicity of all of these agents, as well as any other drug-specific toxicities listed below (Adamson, Bagatell, Balis, & Blaney, 2011).		
<b>Generic Name</b>	<b>Trade Name</b>	<b>Common Side Effects</b>
Cyclophosphamide	Cytosin	Hemorrhagic cystitis
Ifosfamide	Ifex	Hemorrhagic cystitis, seizures
CCNU	Lomustine	Nausea/vomiting
Cisplatin	Platinol	Most nauseating of the chemotherapy agents; hearing loss, renal dysfunction
Carboplatin	Paraplatin	Nausea/vomiting
Oxaliplatin	Eloxatin	Hypersensitivity reaction to cold (liquids, food, and environment)
Temozolomide	Temodar	Nausea/vomiting, alopecia
Procarbazine	Matulane	Nausea/vomiting, alopecia
<b>Antimetabolites</b>		
Structural analogs that inhibit synthesis of DNA and RNA; prolonged drug exposure that results from administering these agents by continuous infusion or by chronic daily dosing increases the chance of exposing a higher proportion of the tumor cell population to the drugs during active DNA replication (Adamson et al., 2011).		
<b>Generic Name</b>	<b>Trade Name(s)</b>	<b>Common Side Effects</b>
Methotrexate	Rheumatrex	Myelosuppression, mucositis
Thioguanine	Tabloid, 6TG	Myelosuppression, mucositis
Cytarabine	Cytosar, Ara-C	Nausea/vomiting, conjunctivitis
Gemcitabine	Gemzar	Flu-like symptoms, myelosuppression
<b>Antitumor Antibiotics (Anthracyclines)</b>		
Bind to and disrupt the shape of DNA (Adamson et al., 2011)		
<b>Generic Name</b>	<b>Trade Name(s)</b>	<b>Common Side Effects</b>
Doxorubicin	Adriamycin	Cardiac toxicity, pink/red urine, nausea/vomiting
Daunorubicin	Cerubidine	Cardiac toxicity, pink/red urine, nausea/vomiting
Dactinomycin	Cosmegen	Nausea/vomiting, skin photosensitivity
<b>PLANT PRODUCTS</b>		
Derived from naturally occurring plants. Some are known to disrupt microtubule formation and function and interrupt mitosis and cell function. Others are responsible for DNA strand breaks and cell death (Ettinger & Rale, 2011).		
<b>Vinca Alkaloids</b>		
<b>Generic Name</b>	<b>Trade Name(s)</b>	<b>Common Side Effects</b>
Vincristine	Oncovin	Peripheral neuropathy, constipation
Vinblastine	Velban	Myelosuppression, paresthesias, alopecia
Vinorelbine	Navelbine	Flu-like symptoms
<b>Epipodophyllotoxi</b>		
Etoposide	Toposar, Vespil	Myelosuppression, hypotension when administered intravenously
<b>Camptothecins</b>		
Topotecan	Hycamptin	Myelosuppression, alopecia
Irinotecan	Camptosar	Diarrhea, abdominal cramping, alopecia
<b>Taxanes</b>		
Paclitaxel	Taxol	Pain, swelling, alopecia, anaphylaxis
Docetaxel	Taxetere	Myelosuppression, fluid retention, rash

*continued*

**Table 7. Chemotherapeutic Agents Administered in the Treatment of Pediatric Brain Tumors and Common Side Effects** *continued*

Biologically Targeted Therapies/Agents	Interrupt activation of signal-transduction pathways that regulate growth, differentiation, adhesion, metabolism, and apoptosis (Adamson et al., 2011; Nageswara Rao & Packer, 2012).	
Drug Category	Example(s)	Common Side Effects
PDGFR inhibitors	Imatinib (Gleevec)	Most biologically targeted agents can cause skin toxicity (rash) and potential for hypersensitivity (allergic) reaction.
VEGF inhibitors	Bevacizumab (Avastin) sorafenib (Nexavar) Sunitinib (Sunitinib)	
EGFR inhibitors (monoclonal antibodies and small-molecule inhibitors)	Cetuximab (Erbix) Nimotuzumab (Theracim) Erlotinib (Tarceva) Gefitinib (Iressa) Lapatinib (Tykerb)	
Protein kinase inhibitors/C-B inhibitors	Thalidomide (Thalomid) Lenalidomide (Revlimid)	
mTOR	sirolimus (Rapamune) Temsirolium (Torisel) Everolimus (Afinitor)	

*EGFR, epidermal growth factor receptor; mTOR, mammalian target of rapamycin; PDGFR, platelet-derived growth factor receptor; VEGF, vascular endothelial growth factor.*

*Sources.* "General Principles of Chemotherapy," by P. C. Adamson, R. Bagatell, F. M. Balis, & S. M. Blaney, 2011, In P. A. Pizzo, D. G. Poplack, P. C. Adamson, S. M. Blaney, & L. J. Helman (Eds.), *Principles and practice of pediatric oncology* (6th ed.). Philadelphia: Wolters Kluwer/Lippincott Williams & Wilkins. "Chemotherapy: Agents and classifications," by A. G. Ettinger & M. L. Rale, 2011, In N. E. Kline (Ed.), *The pediatric chemotherapy and biotherapy curriculum* (3rd ed., pp. 45–63). Glenview, IL: Association of Pediatric Hematology/Oncology Nurses. "Impact of molecular biology studies on the understanding of brain tumors in childhood," by A. A. Nageswara Rao, & R. J. Packer, 2012, *Current Oncology Reports*, 14(2), 206–212.

2. Nurses should appraise the need for antiemetics and familiarize themselves with the types of antiemetics needed in the anticipatory, acute, and delayed stages of chemotherapy-induced nausea and vomiting (Level 2).
- f. Management of chemotherapy side effects and related patient and family education (Table 7)
  - i. Hair loss is a common side effect of chemotherapy. Caregivers play an important role in determining a child's response to the loss of their hair. Family perceptions of hair loss as well as distraction with mind/body control help alleviate psychological distress associated with alopecia (Williams et al., 2006).
  - ii. Family caregiver reports of chemotherapy-induced nausea and vomiting often correlate with pediatric patient reports (Rodgers et al., 2012).
  - iii. Fever during chemotherapy-induced neutropenia may be the only indication of severe underlying infection because signs and symptoms of inflammation are typically attenuated (Freifeld et al., 2011).
    - a) Patients and families should be taught to monitor for fever (temperature higher than 38.3° C) during periods of neutropenia (absolute neutrophil count [ANC] lower than 500 cells per microliter).
    - b) Laboratory studies including ANC, blood cultures, and urine cultures should be collected in appropriate quantities prior to administration of any antibiotic therapy and in a timely manner after initial presentation (Sung, Phillips, & Lehrnbecher, 2011).
    - c) Antibiotics should be administered within a 60-minute period of initial presentation of fever for patients with known or suspected neutropenia (Volpe et al., 2012).

**Nursing recommendations**

1. Nurses should incorporate family caregivers into anticipatory guidance for common adverse effects and recognize the vital role family caregivers play in the recognition and alleviation of symptomatic distress (Level 3).
2. Nurses should have knowledge and recognize the impact of a child's developmental stages on behavior and response to treatment (Level 3).
3. Nurses should instruct patients and families that fever and neutropenia are treated as a medical emergencies (Level 3).
4. Nurses should triage patients accordingly to obtain laboratory studies and blood cultures and administer antibiotics within 60 minutes of initial presentation with fever and neutropenia, as ordered by the licensed provider, to minimize potential complications associated with severe bacterial infection (Level 3).
- g. Palliative care and end of life

- i. Entry into palliative care
    - a) Palliative care for children begins when the illness is diagnosed and continues regardless of whether or not a child receives treatment directed at the disease (Johnston et al., 2008; Nelson et al., 2000).
    - b) Parents value accurate and reliable communication across the care continuum.
    - c) Parents also desire a combination of curative and palliative programs rather than having to decide between curative and palliative care.
    - d) Parents desire accurate prognosis and realistic expectations of the trajectory of illness up front (Heinze & Nolan, 2012).

**Nursing recommendations**

    1. Nurses should expect introduction to palliative care in the setting of pediatric brain tumors to occur at diagnosis and facilitate this service with patients and families (Level 2).
    2. A team approach should be coordinated by the nurse and accurate and reliable expectations should be conveyed by the healthcare team (Level 3).
  - ii. Pain control (Gregoire & Frager, 2006; **Table 8**)
    - a) Multiple studies conducted during the last several years report that the incidence of pain in children dying of a cancer-related diagnosis can be as high as 90%.
  - b) Nonpharmacological and integrative methods of pain management such as physical comforts, distraction, and cognitive-behavioral methods can be used for pain management at end of life.
  - c) Pharmacological interventions are often necessary to relieve pain and are best used in combination with other integrative methods. WHO proposed guidelines for a stepwise analgesic approach that is appropriate for use in children at end of life, with an emphasis on step 1 (nonsteroidal anti-inflammatory drugs and acetaminophen) and step 3 (opioids; WHO, 1998).
  - d) When swallowing becomes difficult toward end of life, pain medications can be administered via alternative routes such as transdermal, sublingual, subcutaneous, rectal, and IV.
  - e) Neuropathic pain is a frequent indication for adjuvant pain relief in pediatric palliative care. Anticonvulsants such as gabapentin (Neurontin) and tricyclic antidepressants such as amitriptyline (Elavil) have shown analgesic efficacy (Friedrichsdorf & Nugent, 2013).
- Nursing recommendations**
1. Nurses should possess the knowledge to appropriately assess pain in children through the use of age-appropriate pain scales and awareness of nonverbal pain cues

**Table 8. Integrative Pain Management Methods Across Developmental Ages**

Age	Physical comforts	Distraction	Cognitive behavioral
Infants: 0–1 years	Rocking, swaddling, kangaroo care, pacifier, sucrose, decrease light and noise, massage, therapeutic touch	Music, singing, soothing and familiar voice, bubbles, pacifier, mobiles, lullabies and other rhyming patterns	Parent support and guided teaching on how to increase an infant's comfort
Preschoolers: 2–5 years	Rocking and cuddling, pacifier, sucrose, decreased light and noise, massage, TENS, therapeutic touch, positioning for heat/cold packs, acupressure, comfort, physical therapy	Familiar songs, music, pop-up books, puppets, videos, bubble-blowing, stories, stories on tape, clowning, pet visits	Art and music therapy, imagery and hypnosis, therapeutic play, relaxation games (such as rag doll), participation in favorite stories, simple explanations, parent support and guidance
School age: 6–11 years	Comfort rocking, cuddling, decrease light and noise, massage, TENS, therapeutic touch, positioning for comfort, heat/cold packs, acupressure and acupuncture, physical therapy	Familiar songs, music, pop-up books, puppets, favorite toys and games, videos, bubble-blowing, stories, stories on tape, clowning, pet visits	Art and music therapy, imagery and hypnosis, relaxation games (e.g., rag doll, belly breathing), participation in favorite stories, information, biofeedback, psychotherapy, parent support and guidance
Adolescents: 12–18 years	Massage, TENS, therapeutic touch, positioning for comfort, heat/cold packs, acupressure and acupuncture, physical therapy, adjust environments to teen's preference	Favorite music, games, stories on tape, videos, pet visits, books read aloud	Imagery and hypnosis, art and music therapy, relaxation and deep breathing, information, biofeedback, psychotherapy, parent support and guidance

*TENS, transcutaneous electrical nerve stimulation*

Source: Table 22.1 (p.332) from chapter "Pain—an Integrative Approach" by Leora Kuttner, in *Oxford Textbook of Palliative Care for Children* edited by Ann Goldman, Richard Hain & Stephen Liben (2006). © 2006 by Oxford University Press. Reprinted by permission of Oxford University Press.

in dying children, such as wincing, grunting, and agitation (Level 3).

2. Nurses should advocate for patient use of age-appropriate pharmacologic and nonpharmacologic therapy to treat pain (Level 3).
3. Nurses should possess proficiency in the administration of analgesics via various routes (Level 3).

### C. Seizures in palliative care patients (Krouwer, Palagi, & Graves, 2000)

1. The fear of seizures is very real for most dying patients with brain tumors and their family members.
2. Despite AED treatment, 50%–75% of patients with primary brain tumors will have recurrent seizures.
3. Chemotherapy and glucocorticoids can alter the bioavailability and effectiveness of AEDs; however, glucocorticoids can help with seizure activity if the seizure is related to peritumoral edema.
4. AEDs should be selected based on several factors (Wusthoff, Shellhaas, & Licht, 2007):
  - a. Seizure type
    - i. Generalized epilepsy: Broad spectrum AEDs
      - a) Clobazam (Frisium, Onfi)
      - b) Clonazepam (Klonopin)
      - c) Lamotrigine (Lamictal)
      - d) Levetiracetam (Keppra)
      - e) Phenobarbital
      - f) Topiramate (Topamax)
      - g) Valproate (Depakote, Depakene)
      - h) Zonisamide (Zonegran)
    - ii. Focal epilepsy: Narrow or broad-spectrum AEDs (examples of narrow-spectrum provided)
      - a) Carbamazepine (Tegretol)
      - b) Gabapentin (Neurontin)
      - c) Oxcarbazepine (Trileptal)
      - d) Phenytoin (Dilantin)
      - e) Pregabalin (Lyrica)
    - iii. Syndrome-specific epilepsy: Syndrome-specific AEDs
      - a) Absence seizures: Ethosuximide (Zarontin)
  - b. Formulation availability
    - i. Oral
    - ii. Rectal
    - iii. IV
    - iv. Buccal/intranasal
  - c. Special considerations
    - i. Ability to swallow
    - ii. Coexisting pain
    - iii. Level of anxiety
    - iv. Presence of mood disorder

- v. Level of alertness

### Nursing recommendation

1. Nurses should educate family members regarding the likelihood of seizures at end of life, first-aid management of seizures, and administration of different formulations of AEDs (Level 3).

### D. Hospice care (Hendricks-Ferguson, 2008)

1. Hospice care is based on a clinical approach to foster a peaceful death, freedom from pain, and enhanced quality of life.
  - a. End-of-life symptom-management strategies used by pediatric hospice programs to support children and their families are not well described in the literature.
  - b. Approximately one-third of parents report their children's breathing difficulties caused them significant distress during the last week of their lives.
    - i. Presence of fatigue and loss of motor function were the second most common concerns.
    - ii. Other end-of-life symptoms include changes in heart rate and fever, changes in optic function, disturbing body movements, pain, epistaxis, and adverse reactions to pain medication.
  - c. According to a survey of parents regarding symptoms during the last week of their children's lives, 35% reported their child had no end-of-life symptoms or concerns on the last day of their life (Hendricks-Ferguson, 2008). This suggests that parents believed their child's symptoms were managed effectively when receiving end-of-life care from hospice.

### Nursing recommendations

1. Nurses should evaluate parents' overall concerns related to their child's end-of-life trajectory and address those concerns when planning care (Level 3).
2. Nurses must educate parents about symptoms that may be observed during their child's end of life, especially signs of possible impending death (Level 3).

### E. Nursing care

1. Distinguishing whether the goal is curative vs. palliative is helpful in managing symptoms throughout the course of treatment. Palliative chemotherapy may be administered without intent to cure to alleviate symptoms and/or to extend life (Waldman & Wolfe, 2013).
2. At the end of life, assessment and appropriate management of end-of-life symptoms is of great value to the parents of children dying from brain tumors (Hendricks-Ferguson, 2008).
3. Progressive neurologic deterioration influences psychological status both positively and



negatively. Treatments that build on positive experience and minimize negative symptoms are vital to reduce child and family suffering and optimize quality of life (Cataudella & Zelcer, 2012).

#### **Nursing recommendations**

1. Nurses should differentiate between curative and palliative chemotherapy when planning nursing care and interventions, especially as a child approaches end of life (Level 3).
2. Nurses should assess and appropriately manage both physical and psychological symptoms toward end of life to reduce both the patient's and family's suffering (Level 3).

#### **F. Patient and family education**

1. Children often express their own awareness of their impending death in an indirect manner. Parents who do not discuss the terminality of their child's diagnosis often later express regret in not talking with their child more openly about their impending death (Cataudella & Zelcer, 2012).
2. Parents who had conversations with their care providers regarding end-of-life decisions prior to the time when it actually became necessary to make those decisions were more likely to decide against resuscitation during end of life (Hechler et al., 2008).

#### **Nursing recommendations**

1. Nurses should recognize the importance of clear, effective communication with the patient and family throughout the disease process, especially nearing end of life, and work to provide clear and effective communication (Level 3).
2. Nurses should provide clear, accurate information; patients and families value honesty (Level 3).

### **VII. Long-Term Effects**

#### **A. Long-term neurocognitive effects**

1. The location of a brain tumor and the type of treatment received (e.g., surgery, cranial irradiation, chemotherapy) will influence the long-term effects of childhood brain tumor survivors (Ashley et al., 2012; Geenen et al., 2007; Mabbott et al., 2011; Wolfe, Madan-Swain, & Kana, 2012).
2. Neurocognitive functioning
  - a. Neurocognitive functioning (cognitive functions associated with distinct areas of the brain and/or neural pathways) can have a significant effect on childhood cancer survivors (Geenen et al., 2007; Robinson et al., 2010).
  - b. Posterior fossa tumors exhibit the most devastating cognitive and behavioral problems and can affect brain development (Sands et al., 2012).
  - c. The impact on intellectual changes will be different for each survivor (Oeffinger et al., 2006).

- i. Neurocognitive dysfunction includes problems with
  - a) Decline in IQ (Edelstein et al., 2011; Ris et al., 2008; Sands et al., 2012; Zuzak et al., 2008)
  - b) Verbal learning (Robinson et al., 2010; Sands et al., 2012)
  - c) Nonverbal abilities (Sands et al., 2012)
  - d) Executive functioning: attention, memory, processing speed, organizational skills, planning, and problem solving (Edelstein et al., 2011; Ellenberg et al., 2009; Kahalley et al., 2013; Mabbott et al., 2011; Robinson et al., 2010; Sands et al., 2012; Wolfe et al., 2012; Zuzak et al., 2008). Kahalley et al. (2013) suggest that slow processing speed may inhibit childhood cancer survivors from realizing academic gains that may cause grade retention. Academic accommodations for cancer survivors may support scholastic achievement.
  - e) Visual learning/processing (Mabbott et al., 2011; Robinson et al., 2010; Sands et al., 2012)
  - f) Visual-motor functioning (Sands et al., 2012)

#### **Nursing recommendations**

1. Nurses should educate patients and families regarding the potential for neurocognitive delay or regression (Level 2).
  2. Nurses should work as active members of a multidisciplinary team to complete a comprehensive neurocognitive assessment evaluating cognitive functioning and academic achievement (Level 1).
  3. Nurses should work with educators to encourage the provision of academic accommodations (e.g., allowing students more time to process information, one-on-one instruction, and reduced stimulation; Level 2).
  4. Nurses should make appropriate referrals to case managers or social services if a patient is not in a long-term program (Level 2).
- d. Risk factors for neurocognitive dysfunction may be influenced by
    - i. Tumor location and/or type (Mabbott et al., 2011; Ris et al., 2008; Sands et al., 2012)
    - ii. Female gender, which demonstrated lower verbal intellectual abilities and lower quality-of-life scores than male gender (Ellenberg et al., 2009; Oeffinger et al., 2006; Sands et al., 2012).

- iii. Younger age at diagnosis (Mabbott et al., 2011; Sands et al., 2012)
- iv. Craniospinal irradiation (CSI) versus focal cranial RT (CRT)
  - a) In a prospective trial in 2012, focal CRT provided no significant decrease in either cognitive or motor functioning 4 years after treatment. The study reports, however, that the children were developmentally impaired, indicating that surgery and/or the effects of the tumor have more influence on neurocognitive dysfunction than formerly identified (Ashley et al., 2012).
  - b) CSI is associated with slower processing speed and general reasoning ability (Ellenberg et al., 2009; Geenen et al., 2007; Kahalley et al., 2013; Mabbott et al., 2011; Neglia et al., 2006).
- v. Higher doses of radiation (Gajjar et al., 2006; Mabbott et al., 2011; Neglia et al., 2006)
- vi. Hydrocephalus with shunt placement (Ellenberg et al., 2009; Hardy, Bonner, Willard, Watral, & Gururangan, 2008; Mabbott et al., 2011; Turner et al., 2009)
- vii. A cerebrovascular event (Ellenberg et al., 2009)
- viii. Comorbid conditions such as inherited genetic syndromes (e.g., neurofibromatosis type 1, Down syndrome, mental retardation, low birth weight, or learning disabilities; Ullrich & Embry, 2012)

#### **Nursing recommendations**

1. Nurses should be aware of the multiple risk factors for neurocognitive dysfunction and collaborate to provide multidisciplinary care (Level 2).
  2. Nurses must have a thorough knowledge of a patient's chemotherapy and/or radiation regimen history as they educate patients and families and assist in interventions that will support symptom management of potential acute and long-term side effects (Level 2).
  3. Nurses should proactively coordinate referrals to disciplines such as physical and occupational therapy for restorative and compensatory strategies to address muscle weakness and low physical performance (Level 2).
- e. Interventions for neurocognitive dysfunction
- i. Individual education plan (IEP): A multidisciplinary team including schoolteachers, parents, patients, nurses, and a neuropsychologist should provide input

- into an IEP to promote school achievement (Robinson et al., 2010)
  - ii. School systems may be able to offer support and resources.
  - iii. Special education will be required (Turner et al., 2009).
  - iv. Barriers exist to addressing all cognitive-behavioral interventions in the clinic setting such as time, transportation, and financial concerns (Patel, Katz, Richardson, Rimmer, & Kilian, 2009).
- f. Cognitive rehabilitation
- i. Remediation programs consisting of cognitive-behavioral interventions (Patel et al., 2009; Wolfe et al., 2012)
  - ii. Home-based, computerized cognitive training programs (Hardy, Willard, & Bonner, 2011; Patel et al., 2009; Wolfe et al., 2012)
  - iii. Assistive technology (Ellenberg et al., 2009)
- g. Pharmacological (Conklin et al., 2010; Wolfe et al., 2012)
- i. Methylphenidate (Ritalin)
    - a) Psychostimulant
    - b) Used for attention-deficit disorders
  - ii. Donepezil (Aricept), an acetylcholinesterase (AChE) inhibitor (Castellino et al., 2012)
    - a) Improves executive function
    - b) Enhances memory performance

#### **Nursing recommendations**

1. Nurses should collaborate with the multidisciplinary team to inform parents and teachers about the cognitive sequelae associated with CNS cancer (Level 1).
2. Nurses should work as part of the multidisciplinary team to promote cognitive-behavioral training appropriate for childhood cancer survivors within the school systems as needed (Level 3).
3. Nurses should refer or coordinate childhood cancer survivors to long-term follow-up programs for early social and educational intervention (Level 1).
4. Nurses should encourage the use of early intervention by using home-based, computerized cognitive training programs (Level 1).

#### **B. Long-term medical effects**

1. Many studies report that childhood cancer survivors have one or more long-term medical effects after treatment (Crom et al., 2010; Hudson et al., 2013).
2. Neurological consequences
  - a. Headache (Johnson, Jordan, & Mazewski, 2009; Turner et al., 2009)
  - b. Hearing impairment (Oeffinger et al., 2006), especially when treatment includes platinum-based chemotherapy (Orgel et al., 2012).

- c. Hydrocephalus/shunt dependence (Hardy et al., 2008; Morris et al., 2007; Turner et al., 2009)
  - d. Neurogenic bowel/bladder (Wilson, Oleszek, & Clayton, 2007)
  - e. Neuropathy including numbness and tingling in the extremities, which has been reported when treatment includes platinum-based chemotherapy (Albers, Chaudhry, Cavaletti, & Donehower, 2011).
  - f. Reduced loss of vision/visual field deficits (Oeffinger et al., 2006; Turner et al., 2009; Winkfield et al., 2011)
  - g. Seizures can occur more than 5 years after treatment. An increased risk of seizures may occur with RT and in younger children (Oeffinger et al., 2006; Turner et al., 2009; Wells, Gaillard, & Packer, 2012).
  - h. Sensory/motor deficits can develop after surgery including focal extremity weakness, truncal/appendicular ataxia, sensory deficits, and cranial nerve deficits (Turner et al., 2009).
  - i. Stroke is a risk factor after RT (Bowers et al., 2006; Morris et al., 2007; Oeffinger et al., 2006).
3. Nephrologic effects (Oeffinger et al., 2006) can occur, particularly after administration of platinum-containing chemotherapy (Geenen et al., 2007).

#### **Nursing recommendations**

1. Nurses should emphasize the necessity of long-term ongoing health assessment to patients and their families because childhood cancer survivors often have one or more long-term medical effects (Level 2).
2. Nurses should assist patients and families in organizing long-term follow-up care (Level 2).
3. Nurses should monitor for seizures, provide first responder care, and notify the provider if seizures occur (Level 2).
4. Nurses should provide patient/family education on seizure precautions, including how to keep a seizure diary, and, if appropriate, medication instruction (Level 3).
5. Nurses should assess risk for health problems and create interventions to enhance patients' health status (Level 3).
6. Nurses should coordinate the referral of patients at risk for hearing loss for audiometry testing at baseline, during treatment, and during follow-up visits (Level 2).
7. Nurses should recognize and aid in the diagnosis and management of headaches and counseling of patients/families on preventative measures (Level 3).
8. Nurses should coordinate referral to a headache clinic or long-term cancer survivor clinic

for patients with headaches who are off of treatment (Level 3).

#### **C. Secondary CNS tumors**

1. An increased risk for developing a secondary CNS tumor resulting from cranial and spinal radiation is related to the dose of RT and younger age at diagnosis (Armstrong et al., 2009; Neglia et al., 2006).
  - a. Male gender prevalence has been reported (Vinchon et al., 2011).
2. A secondary cancer can occur 10 or more years after initial radiation (Armstrong et al., 2009; Geenen et al., 2007; Neglia et al., 2006; Reulen et al., 2011).

#### **Nursing recommendations**

1. Nurses should monitor for signs and symptoms of secondary tumors (Level 2).
2. Nurses should educate patients and families on secondary tumors and stress the importance of long-term follow-up care (Level 2).

#### **D. Cardiovascular disease (Morris et al., 2007; Oeffinger et al., 2006)**

1. Cardiovascular disease in childhood brain tumor survivors is consistent with early aging, causing needs comparable to the elderly in 30- to 40-year-old adults (Edelstein et al., 2011; Trachtenberg et al., 2011).
2. Cardiovascular abnormalities may be a result of a growth hormone deficiency (Lipshultz et al., 2012).
3. Cardiac complications can occur years after receiving RT (Geenen et al., 2007).
  - a. Risk factors include dose of radiation, female gender, concomitant RT, and age.
  - b. Higher risk applies to younger children at age of treatment (Trachtenberg et al., 2011).
4. Cardiovascular risks include hypertension, dyslipidemia, obesity, and insulin resistance (Edelstein et al., 2011; Lipshultz et al., 2012; Trachtenberg et al., 2011).
5. Treatment includes regular screening (Trachtenberg et al., 2011)
  - a. Imaging with echocardiography or radio-nuclide angiography and monitoring serum biomarkers
  - b. Management of the factors (medications, dietary modification, and physical activity)

#### **Nursing recommendations**

1. Nurses should recognize and assess for signs and symptoms of cardiovascular disease in long-term survivors (Level 2).
2. Nurses should educate patients on diet modification and physical activity as appropriate (Level 1).

#### **E. Endocrine deficiencies**

1. Children with CNS tumors are at risk for endocrinopathies (Crom et al., 2010; Geenen et

- al., 2007; Morris et al., 2007; Shalitin et al., 2011; Turner et al., 2009).
2. Common endocrine morbidities (Morris et al., 2007)
    - a. Adrenocorticotrophic hormone (ACTH)
      - i. Monitoring of brain tumor survivors for ACTH deficiency must continue beyond 10 years following irradiation.
      - ii. Monitoring should preferably include an insulin tolerance test and glucagon stimulation test.
    - b. DI (Crom et al., 2010)
    - c. Diabetes mellitus/hyperinsulinism (Shalitin et al., 2011)
    - d. Growth hormone deficiency
      - i. Childhood brain tumor survivors are at increased risk for late endocrine effects (Crom et al., 2010; Karachaliou, Simatos, Batika, Michalacos, & Kaldrymides, 2009), especially children treated with cranial RT and younger than 3 years of age (Shalitin et al., 2011).
      - ii. The frequency of hormonal deficits increases with time and warrants lifelong surveillance (Karachaliou et al., 2009).
        - a) Monitoring of stature every 6 months prior to fusion of growth plates
        - b) Monitoring of growth hormone secretion if there is a decrease in growth rate
        - c) Monitoring of pubertal status and thyroid function every 6 months
    - e. Adrenocorticotrophic hypogonadotropic hypogonadism may lead to infertility in some patients (Crom et al., 2010; Oeffinger et al., 2006).
    - f. Hypogonadism secondary to gonadal dysfunction may require sex hormone replacement of testosterone injections for males and estrogen and progesterone pills for females (Shalitin et al., 2011).
    - g. Hypothyroidism: Treat with thyroxine (Shalitin et al., 2011)
    - h. Obesity/overweight (Armstrong et al., 2011; Crom et al., 2010; Karachaliou et al., 2009; Oeffinger et al., 2006; Winkfield et al., 2011)

#### **Nursing recommendations**

1. Nurses should monitor and evaluate for endocrine effects, especially in children treated with cranial radiation and those in whom the condition is diagnosed when they are younger than 16 years of age (Level 3).
2. Nurses should educate patients and families regarding compliance with hormone replacement therapy and improvement in their diet (Level 3).

3. Nurses should stress the importance of long-term follow-up because these disorders may not become evident until years after treatment (Level 2).
4. Nurses should coordinate referral of patients/families to their oncologist to discuss the risk of infertility and the potential for harvesting eggs and sperm before treatment begins (Level 2).

#### **F. Posterior fossa syndrome**

Posterior fossa syndrome (cerebellar mutism) usually occurs hours after surgery. The effects may linger for months to years and, in some cases, residual deficits may be permanent.

1. Most frequently observed deficits include ataxia, speech and language dysfunction.
2. May also exhibit varying degrees of global intellectual disability (Robertson et al., 2006).

#### **Nursing recommendation**

1. Nurses should have a heightened knowledge of long-term consequences for patients with posterior fossa syndrome and make appropriate referrals if a patient is symptomatic (Level 2).

#### **G. Musculoskeletal effects**

1. Decreased bone mineral density (BMD; Cohen et al., 2012; Karachaliou et al., 2009; Oeffinger et al., 2006)
  - a. Cause for decreased BMD in long-term survivors is often multifactorial in nature (Cohen et al., 2012); factors include
    - i. The disease itself
    - ii. The treatment
    - iii. Poor nutrition
    - iv. Lack of exercise
    - v. Development of endocrinopathies.
2. Scoliosis (Wilson et al., 2007)

#### **Nursing recommendations**

1. Nurses should monitor body composition, bone density, and lipid levels periodically (Level 2).
2. Nurse should encourage appropriate dietary calcium and vitamin D intake, weight-bearing exercises, and avoidance or cessation of smoking (Level 3).

#### **H. Long-term physical effects**

1. Physical performance
  - a. Childhood cancer survivors usually have muscle weakness and poor exercise tolerance contributing to overall physical performance limitations (Turner et al., 2009). These limitations are associated with poor school performance and restricted home-life participation (Ness et al., 2010).
  - b. Muscle strength and performance scores for childhood brain tumor survivors (young to middle-aged adults) are comparable with those of elderly people.
  - c. Poor physical tolerance increases risk for medical problems such as osteoporosis,

cardiovascular disease, and obesity from inactivity (Ness et al., 2010).

- i. Risk factors for poor physical tolerance (Ness et al., 2010)
  - a) Younger age at diagnosis (strongest indicator)
  - b) Segment-specific radiation in a specific anatomical segment (frontal cortex, temporal lobe, posterior fossa, or parietal or occipital cortex.
  - c) Treatment with vincristine or platinum

#### **Nursing recommendations**

1. If muscle weakness and low physical performance are present, nurses should refer survivors of childhood brain tumors to appropriate disciplines such as physical and occupational therapy for restorative exercises and compensatory strategies (Level 1).
2. Nurses should educate pediatric brain tumor survivors and caregivers that aerobic and resistance-training interventions may be of benefit to survivors with cognitive and other neurologic deficits and encourage patients to actively participate in these activities (Level 2).
2. Sleep disturbance
  - a. Brain tumors involving the hypothalamus, thalamus, and brain stem can affect sleep (Gapstur, Gross, & Ness, 2009; Greenfeld, Constantini, Tauman, & Sivan, 2011; Rosen & Brand, 2011).
  - b. Risk factors for sleep disturbance include
    - i. Radiation (higher risk; Gapstur et al., 2009; Mulrooney et al., 2008)
    - ii. Extensive surgery (higher risk; Gapstur et al., 2009; Greenfeld et al., 2011)
    - iii. Age younger than 5 years at diagnosis (Gapstur et al., 2009)
    - iv. Diagnosis of craniopharyngioma (Gapstur et al., 2009)
    - v. Chemotherapy (Gapstur et al., 2009; Greenfeld et al., 2011)
    - vi. Cancer-related fatigue (Mulrooney et al., 2008)
    - vii. Pain (Gapstur et al., 2009)
    - viii. Seizures (Greenfeld et al., 2011)
    - ix. Obesity (Mulrooney et al., 2008)
    - x. Endocrinopathies (Gapstur et al., 2009)
    - xi. Heart failure (Mulrooney et al., 2008)
    - xii. Blindness (Greenfeld et al., 2011)
    - xiii. Medication (Greenfeld et al., 2011)
    - xiv. Anxiety (Gapstur et al., 2009; Greenfeld et al., 2011)
    - xv. Depression (Mulrooney et al., 2008)
  - c. A significant number of childhood cancer survivors report excessive daytime sleepiness (EDS), disrupted sleep, and fatigue,

even among survivors making a full recovery (Gapstur et al., 2009; Greenfeld et al., 2011).

- d. Symptoms of sleep dysfunction include
  - i. EDS (most common; Gapstur et al., 2009; Greenfeld et al., 2011; Mulrooney et al., 2008)
  - ii. Insomnia (Gapstur et al., 2009; Greenfeld et al., 2011)
  - iii. Limb movement disorders (Gapstur et al., 2009)
  - iv. Increased nighttime awakenings (Gapstur et al., 2009; Greenfeld et al., 2011; Mulrooney et al., 2008)
  - v. Sleep-disordered breathing (Gapstur et al., 2009; Greenfeld et al., 2011).
- e. Interventions to encourage sleep (dependent on cause of sleep disturbance)
  - i. Avoid daytime naps and exposure to bright light upon awakening; focus on weight reduction and treatment of obstructive sleep apnea, appropriate medication use, and controlling endocrine deficiencies (e.g., hypothyroidism and cortisol deficiency; Rosen & Brand, 2011)
  - ii. Pharmacological
    - a) Melatonin
      - (1) A deficiency can be induced by the tumor or treatment (Gapstur et al., 2009).
      - (2) Should be administered at night
    - b) Stimulant medications (Conklin et al., 2010)
      - (1) Methylphenidate (Ritalin) or amphetamines
      - (2) Modafinil (Provigil), a nonamphetamine, is being trialed in children to promote wakefulness.
      - (3) Should be given earlier in the day to avoid insomnia
    - c) AChE inhibitor (Donepezil) has been shown to improve executive function and memory in a small pilot study (Castellino et al., 2012).

#### **Nursing recommendations**

1. Upon initial assessment of pediatric brain tumor survivors, nurses should ask about sleep-wake disturbances, especially EDS (Level 1).
2. Nurses should screen for potential sleep-wake problems, especially EDS, as part of the routine follow-up of cancer survivors (Level 1).
3. Nurses should provide sleep hygiene education as appropriate and include timing of medications to provide best effect (Level 3).

4. Nurses should coordinate referral of patients for evaluation to a sleep center as appropriate (Level 1).

#### **I. Oral/dental effects**

1. Septicemia, nutritional compromise, significant pain, and long-term morbidity are recognized oral complications of cancer therapy (Fulton, Middleton, & McPhail, 2002; Leggott, 1990; Simon & Roberts, 1991).
2. Negative effects include mucositis; viral, bacterial, and fungal infections; xerostomia; and dental abnormalities including root stunting, microdontia, hypodontia, and overretention of primary teeth (Belfield & Dwyer, 2004; Fulton et al., 2002).
3. Meticulous oral hygiene is the most important measure in the prevention and treatment of oral complications of pediatric cancer treatment (Belfield & Dwyer, 2004; Bonnaure-Mallet et al., 1998).
4. Dental care must be an integral part of cancer care (Simon & Roberts, 1991).

#### **Nursing recommendations**

1. Nurses should recognize the effects of cancer therapy on oral and dental health and educate patients and families on the importance of good oral hygiene (Level 2).
2. Nurses should facilitate referral to dental services for both preventative and treatment services (Level 2).
3. Nurses should recognize that oral and dental health is a multidisciplinary responsibility and encourage the entire healthcare team to include it in their daily assessment and plan of care (Level 2).

#### **J. Neurobehavioral effects**

1. Social adjustment, social performance, and social skills are decreased in childhood brain tumor survivors after treatment (Boydell, Stasiulis, Greenberg, Greenberg, & Spiegler, 2008; Edelstein et al., 2011; Quillen, Crawford, Plummer, Bradley, & Glidden, 2011; Schulte & Barrera, 2010; Turner et al., 2009).
2. Pediatric brain tumor survivors are at increased risk for anxiety, depression, social isolation, and behavior problems such as conduct disorder (Boydell et al., 2008; Edelstein et al., 2011; Quillen et al., 2011; Schulte & Barrera, 2010; Turner et al., 2009).
3. Executive functioning in areas of judgment, planning, and inhibition control (Boydell et al., 2008; Edelstein et al., 2011; Quillen et al., 2011; Schulte & Barrera, 2010; Turner et al., 2009)
4. Social development and autonomy are milestones that may take longer to achieve with varying degrees of success for childhood cancer survivors progressing to adulthood compared with their peers (Boydell et al., 2008; Edelstein

et al., 2011; Quillen et al., 2011; Schulte & Barrera, 2010; Turner et al., 2009).

5. Childhood cancer survivors may have more than one medical or psychological issue after treatment (Geenen et al., 2007; Turner et al., 2009).
6. More research is needed to better understand deficits at the levels of social performance and social skills to uncover more specific sources of social difficulties and to provide individualized interventions tailored to the cancer survivor's social adjustment deficit (Geenen et al., 2007).

#### **Nursing recommendations**

1. Nurses should advocate for the survivors' educational needs within the school system and for the necessity of long-term follow-up (Level 2).
2. Nurses should coordinate referrals for continued assistance for academic needs, psychological assessment, and long-term follow-up (Level 2).
3. Nurses should assess for suicidal ideation as part of the patient's psychological assessment. Many childhood cancer survivors have one or more medical or psychological issues (Level 3).
7. A worse outcome for social adjustment is associated with a longer time since diagnosis (Schulte & Barrera, 2010).
  - a. Posterior fossa tumors result in the most devastating cognitive and behavioral problems and can affect brain development (Sands et al., 2012).
  - b. There is no standard for psychological evaluation of neurobehavioral effects. Neuro-oncology long-term follow-up or general long-term follow-up programs are more likely to evaluate patients than centers without long-term follow-up programs (Bowers et al., 2006).

#### **Nursing recommendation**

Nurses should be aware that childhood brain tumor survivors experience decreases in social adjustment following treatment and should refer survivors for neuropsychological testing as appropriate (Level 1).

8. Childhood survivors and parents of children with low-grade cerebellar astrocytoma report fewer social issues than other childhood brain tumor survivors and their parents.
  - a. In a study of similar comparison groups of patients with astrocytomas and medulloblastomas, survivors of astrocytomas had better improvement rates than medulloblastoma survivors (Roncadin, Dennis, Greenberg, & Spiegler, 2008; Zuzak et al., 2008).

- b. Methylphenidate has been found to be beneficial in improving social skills, depression, and behavior problems (Conklin et al., 2010).

**Nursing recommendation**

Nurses should educate patients and families regarding potential medication (such as methylphenidate) options for the treatment of social adjustment issues and coordinate referral to a licensed independent care provider for further evaluation as appropriate (Level 2).

- c. Continued support for patients and families after the acute care phase of treatment is needed (Pruitt, Ayyangar, Craig, White, & Neufeld, 2011). Support should include siblings and friends of the patient and family.

**Nursing recommendations**

1. Nurses should educate patients and families regarding the necessity of continued support after treatment and the need to include siblings and friends (Level 1).
2. Nurses should coordinate and encourage referrals to comprehensive pediatric brain tumor treatment programs as needed for emotional support and resources for educating family, friends, and teachers about their illness and treatment plan (Level 1).

**K. Quality of life**

1. Childhood brain tumor survivors report increased rates of impaired health vs. their siblings or even childhood leukemia survivors (Oeffinger et al., 2006; Sands et al., 2012).
2. Survivors, compared with their siblings, describe increased rates of depression, somatization, fatigue, and daytime sleepiness (Oeffinger et al., 2006; Sands et al., 2012).
3. Cancer survivors with a history of radiation may experience more fatigue (Oeffinger et al., 2006; Sands et al., 2012).
4. Females report lower physical quality of life and more symptoms of depression than males (Oeffinger et al., 2006; Sands et al., 2012).

**Nursing recommendations**

1. Nurses should play a role in improving the quality of life of cancer survivors by providing education on patient medication, sleep hygiene, exercise, and depression and by making referrals to long-term follow-up centers specializing in childhood survivors of cancer (Level 2).
2. Nurses should provide early assessment and interventions for physical therapy and psychosocial services, especially for female survivors (Level 2).
3. Nurses should be aware that most cancer survivors have at least one significant long-

term health problem and assess the patient accordingly (Level 2).

5. Quality of life for childhood cancer survivors may be affected by sexuality, fertility, and body image issues resulting from permanent changes such as visible scars (Geenen et al., 2007).

**Nursing recommendations**

1. Nurses should educate cancer survivors about reproductive information before cancer treatment begins (Level 3).
2. Nurses should coordinate and encourage referral of cancer survivors to “survivorship clinics” or long-term clinics specializing in survivors of cancer to improve their health status and quality of life (Level 3).
3. Nurses should be aware that late effects experienced by childhood cancer survivors involve physical, medical, social, emotional, behavioral, and neurocognitive functioning, and it is crucial that nurses perform a holistic baseline assessment and provide appropriate interventions (Level 3).

**L. Resource assessment**

1. Caregivers of children with CNS cancer face economic hardships along with the devastating diagnosis of a brain tumor (Turner et al., 2009).
2. Expenses not covered by insurance include travel to appointments and parking (Bowers et al., 2006; Park et al., 2012).
3. Job-related changes include decreasing the number of hours worked to care for the child with a brain tumor.
4. Access to health care in terms of specialists available and geographic locations of health services (Bowers et al., 2006; Park et al., 2012)
5. Survivors of childhood cancer report more long-term healthcare needs, and lack of insurance may be a barrier to obtaining follow-up health care (Park et al., 2005).

**Nursing recommendations**

1. Nurses should recognize that caregivers of cancer survivors are working through complex issues each day, and assess both the patient and family for the presence and effect of these stressors and provide appropriate interventions (Level 3).
2. Ease all aspects of patient and family complex issues through coordination of care and appropriate referrals.
3. Assess needs and refer caregivers to the appropriate resources because financial concerns may contribute to missed follow-up appointments (Level 2).
4. Nurses should have knowledge about community resources that will help long-term survivors obtain needed resources, but they should exercise caution to avoid providing

- false hope regarding the accessibility of these resources (Level 2).
6. Survivors of childhood cancer have economic challenges as they transition into adulthood.
    - a. Significantly lower amounts of affordable health insurance coverage are available compared with insurance available to their siblings (Park et al., 2005).
    - b. A knowledge deficit exists regarding the details of their insurance coverage (Park et al., 2012).
    - c. If uninsured, a knowledge deficit exists about eligibility and how to obtain Medicaid (Park et al., 2012).
    - d. Yearly income is \$20,000 or less for 40% of survivors (Park et al., 2005).

#### **Nursing recommendations**

1. Nurses should recognize that transitioning from pediatric facilities to adult care involves unique concerns related to patients with chronic problems, and they should prepare patients and families for this transition (Level 3).
2. Nurses should assess patient and family knowledge regarding health insurance and refer cancer survivors to the appropriate resources and provide appropriate recommendations and interventions (Level 3).
7. Cancer survivors are at risk for a decline in their ability to live independently (financially, developmentally, or both) and interact in interpersonal relationships; they may also experience a decline in emotional functioning and in their health status. Each of these factors also influences a patient's quality of life (Armstrong et al., 2011; Armstrong et al., 2009; Turner et al., 2009).
8. Text messaging, video games, Web-based chat rooms, and social media websites are used to educate and increase compliance with taking medication; these technologies are also used to communicate follow-up appointment reminders and connect patients and families to other survivors of childhood cancer (Crom et al., 2010).

#### **Nursing recommendations**

1. Nurses should perform a comprehensive assessment of cancer survivors to determine needs (cognitive, emotional, physical, and financial) and educate on compensatory strategies including assistive technology (Level 2).

2. Nurses should be actively involved in designing the digital technology used to improve a patient's quality of life (Level 3).
3. Nurses should coordinate and encourage referrals to transitional facilities to promote independent living, job placement, and coaching, which will enhance quality of life for CNS malignancy survivors (Level 2).
4. Nurses should coordinate and encourage referrals as appropriate to multidisciplinary healthcare professionals such as social workers and neuropsychologists (Level 2).

#### **M. Follow-up care**

1. Cancer survivors are at risk for mortality, secondary cancers, and other long-term effects (Morris et al., 2007). Survivors of CNS tumors require less frequent follow-up than other cancer survivors, which may lead to missed follow-up appointments (Armstrong et al., 2009; Barakat, Schwartz, Szabo, Hussey, & Bunin, 2012).
2. Cancer survivors likely will be followed by general physicians and internists because most of these patients are not followed at a long-term cancer treatment center (Oeffinger et al., 2006).

#### **Nursing recommendations**

1. Follow-up care and periodic screening based on assessment, tumor location, and type of treatment are a necessity, and nurses should educate patients and families and make referrals to long-term cancer centers or clinics (Level 2).
2. Nurses should educate patients and families of the critical importance of follow-up (Level 2).
3. Nurses should collaborate within organizations to place an effective reminder system in place targeting specific populations of cancer survivors because brain tumor survivors are less likely to continue long-term follow-up care (Level 2).
4. Nurses should educate childhood cancer survivors and their families about the increased risk for secondary tumors and other health conditions for more than 5 years after treatment (Level 2).
3. Brain tumor survivors are living longer and transitioning into adult care.
  - a. Many childhood cancer survivors are first treated at pediatric institutions that are not set up to continue long-term follow-up care or to seamlessly transition pediatric patients into the adult setting.



- b. This places the transitioning patient at risk for being lost to follow-up (Armstrong et al., 2009)

**Nursing recommendations**

1. Nurses should be aware that because of high risk for mortality, subsequent cancers, and other medical conditions, survivors of pediatric brain tumors will require specialized care and screening in a cancer center or a clinic specifically intended for long-term follow-up of pediatric patients with brain tumors, and that it is critical to encourage and coordinate referrals as necessary (Level 2).
2. Nurses should use appropriate interventions during each phase of care, adapting and changing them to effectively address the varying needs of patients (Level 2).

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