Impairments Associated with Pediatric Brain Tumors and Implications for Physical Therapy

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Objectives

• Describe the most commonly seen pediatric brain tumors and their pathology
• Understand the medical interventions used to treat different brain tumors based on pathology and location
• Recognize common impairments seen as a result of brain tumor location, pathology and their necessary medical/surgical interventions
• Identify appropriate tests and measures for this population
• Choose appropriate treatment interventions along with equipment/orthoses prescription

Epidemiology/Pathology

Epidemiology

• 2nd most common pediatric malignancy
– About 4.5 cases/100,000 children
– > 3,000 new cases/year in US
– Incidence
• 5-year survival rate has increased from 35% to 65% over past 40 years
• Leading cause of cancer death


Epidemiology

Prognosis
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Etiology and Pathogenesis

- Immunodeficiencies
- Ionizing radiation
- Inherited familial syndromes

Clinical Presentation

Generalized Signs and Symptoms

- Increased Intracranial Pressure (ICP)
  - Headache, nausea, vomiting, fatigue
  - Decreased upgaze, 6th Cranial Nerve (CN) palsy, papilledema
  - Infants: Macrocephaly, Failure To Thrive (FTT), developmental delay

Localizing Symptoms

Cerebral Hemisphere
- Glioma, Ependymoma, Primitive Neuroectodermal Tumor (PNET)
  - Seizure
  - Hemiparesis
  - Increased ICP
  - Visual Field Cut
  - Change in Behavior

Pineal Region
- Germ cell tumor, Glioma, Pineoblastoma
  - Pannaud's Syndrome
    - Impaired upgaze
    - Conversion nystagmus
    - Light/near dissociation
  - Increased ICP

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**Cerebellum**
- Pilocytic Astrocytoma, Medulloblastoma, Ependymoma
  - Increased ICP
  - Ataxia
  - Dysmetria

**Brainstem**
- Glioma
  - CN deficits
  - Hemiparesis
  - Increased ICP

**Suprasellar**
- Craniopharyngioma, Glioma, Germ cell tumor, Pituitary tumor
  - Visual field deficit
  - Endocrinopathy/Diabetes Insipidus
  - Weight gain
  - Increased ICP
  - Diencephalic Syndrome
    - FTT
    - Euphoria
    - Increased appetite
    - Emesis

**Spinal Cord**
- Astrocytoma, Ependymoma
  - Back pain
  - Extremity weakness
  - Sensory changes
  - Bowel/bladder dysfunction

**Brain Tumor Metastases**
- Spread via leptomeninges
  - PNET
  - Germ cell tumors
  - Ependymoma
  - Atypical teratoid/rhabdoid tumor (ATRT)
  - Glioblastoma Multiforme (GBM)

**Medical and Surgical Interventions**
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Variation of Treatment
- Pathology
- Staging
  - Dissemination
  - Extent of resection
- Patient age

Medical Treatment
- Radiation
- Chemotherapy

Types of Radiation
- X-ray/Photon Therapy
  - Total brain irradiation
  - Deposit energy in small packets along their path through tissue
  - Can damage normal cells as well as cancer cells
- Proton Therapy
  - Targets the tumor at a higher intensity without damaging surrounding areas
  - Fewer short-term and long-term effects
  - Decreases the likelihood of secondary malignancies

Radiation Therapy: Risk Factors
- Neurocognitive
  - Age, location, and dose-related
    - Severity of intellectual decline is inversely related to age at time of XRT
- Endocrinopathies
- Stroke
- Hearing loss

Proton vs Photon
- Type
- Size
- Location
- Age
- Proximity to normal tissues that are sensitive to radiation
- Distance through the body the radiation needs to travel
- Patient’s general health and medical condition
- Other types of treatment the patient will have

Dose Distribution

Proton therapy precisely targets tumors, reducing the radiation dose to healthy tissue compared with X-rays.
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Chemotherapy

- Risk factors
  - Leukemia
  - Hearing Loss
  - Fatigue
  - Nausea and Vomiting
  - Pain
  - Anemia/Neutropenia
  - Nerve/Muscle effects

Surgical Treatment

- Gross total resection
  - Large tumor borders
  - Multiple surgeries
- Partial resection
  - Tumor location
  - Tumor type
- Risk factors
  - CVA
  - Physical and cognitive impairments

Common Pediatric Brain Tumors

Primitive Neuroectodermal Tumor (PNET)/ Medulloblastoma (MB)

- Most common CNS malignancy in childhood
- 20% of primary CNS tumors
- Mean age: 3-4 years
- More common in male
- Very Invasive
- Location:
  - 72% are in posterior fossa
  - 12% are hemisphere
  - 7% are pineal

PNET/ Medulloblastoma

- Supratentorial PNET
  - Tend to be more aggressive
- Medulloblastoma
  - Posterior fossa PNET
  - Must be found in the cerebellum

PNET: High Risk Features

- Residual tumor: >1.5cm^2
- Patient age: <3 y/o
- Location: Supratentorial
- Metastases

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PNET/MB Therapy

- Surgery
- Radiotherapy
  - Local and craniospinal
- Chemotherapy
  - Vincristine

Astrocytomas

- Pilocytic and Fibrillary
  - Pediatric
  - Low Grade
- Anaplastic and Glioblastoma Multiforme
  - Adult
  - High Grade

Astrocytomas

- Most common pediatric brain tumor
- Locations
  - Cerebellum
  - Cerebral hemispheres
  - Deep midline structures
  - Optic pathway/hypothalamus

Glioblastoma Multiforme

- 11% of pediatric brain tumors
- Surgery: Aggressive resection
- Radiotherapy: Focal for survival prolongation
- Chemotherapy: Modest impact on long-term outcome

Importance of the Cerebellum to Physical Therapists

Astrocytomas

- 11% of pediatric brain tumors
- Surgery: Aggressive resection
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Importance of the Cerebellum to Physical Therapists

Neuroanatomy Cerebellum

- 3 lobes
- Located at the base of the cortex

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Role of Cerebellum

- Motor control
- Language
- Memory
- Mental Imagery


Role of Cerebellum

- Spinocerebellum
  - Repetitive movement
  - Muscle tone
- Vestibulocerebellum
  - Postural control
  - Maintenance of equilibrium
  - Coordination of eye movements
- Cerebrocerebellum
  - Regulates skilled and complex movements


Dysfunction of Cerebellum

- Still able to produce movement
- Movement is erratic and uncoordinated
  - Ataxia
  - Dystonia
- Vestibulocerebellum Damage
  - Impaired equilibrium
  - Abnormal gait mechanics
- Spinocerebellum Damage
  - Errors in force, direction and speed of movement
  - Gross motor impairments
- Cerebrocerebellum Damage
  - Errors in force, direction and speed of movement
  - Fine motor control impairments


Posterior Fossa Syndrome (PFS)

- 8-25% children operated with a cerebellar tumor
- Occurs 1-2 days after surgery
- Comprised of:
  - Mutism
  - Emotional lability
  - Neurobehavioral abnormalities
  - Severe ataxia
  - Dysphagia
  - Axial hypotonia


Posterior Fossa Syndrome


- 63 patients with posterior fossa tumor
- PFS =29%
- Predictors of PFS
  - Brainstem invasion
  - Midline tumor location
  - Younger age
  - Absence of radiographic residual tumor


Posterior Fossa Syndrome


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Tests and Measures

- **Musculoskeletal**
  - Manual muscle testing
  - Cervical PROM/AROM
- **Neurological**
  - CN testing
  - Sensory
  - Coordination
  - Balance
  - Tone
- **Cardiovascular**
- **Integumentary**

Outcome Measures

- Pediatric Balance Scale
- Berg Balance Scale
- 6 minute walk test/Energy Expenditure Index
- Timed Up and Go
- Dynamic Gait Index
- Bruininks-Oseretsky Test of Motor Proficiency Second Edition
- Ataxia Rating Scales

PT Examination

- **Musculoskeletal**
- **Neurological**
  - CN testing
  - Sensory
  - Coordination
  - Balance
  - Tone
- **Cardiovascular**
- **Integumentary**

PT Interventions for Motor and Non-Motor Symptoms

Non-Motor Symptoms

- Poor executive function
- Impaired spatial cognition
- Linguistic difficulties
- Disinhibited behavior

Motor Symptoms

- Incoordination
- Dysmetria
- Hypotonia
- Weakness
- Balance dysfunction
- Gait dysfunction
- Oculomotor dysfunction

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**Locomotor training**


- Case report of 13 y/o female with cerebellar ataxia
- Lite Gait over treadmill 5x/wk for 4 wks then 4 months at home
- Outcomes
  - 152m using U-Step walker with S
  - Wee FIM: change of 9 points in motor domain

**Compensatory Approaches**

- Single joint movements
- Visual/verbal cues
- Assistive technology
- Assistive Devices and Orthoses
- Weights and Lycra garments


**Restorative Approaches**

- Repetition of task
- Stepping and targeting
- Gait training
- Balance training
- Ocular exercises


**References**


