Nursing Management of Posterior Fossa Syndrome (PFS) in Pediatric Patients

Patricia Nikolski, BScN, RN & Venus Shyu, BScN, RN
What Is Posterior Fossa Syndrome (PFS)?

The Posterior Fossa Syndrome is a neuropsychological and neurological syndrome consisting of the heterogeneous presentation of cognitive, emotional, behavioral, motor, language- and speech-related symptoms. It is most commonly seen in children after the surgical resection of a posterior fossa mass.
Background and Context

CNS malignancies are the second most common types of pediatric cancer following leukemia.

Of these malignancies, 70% occur in the post fossa region.

Surgical resection of these tumours results in the development of Posterior Fossa Syndrome in 8 – 40% of cases.

Canadian Cancer Society (2014)
What Do PFS Symptoms Look Like?

- Mutism
- Dysarthria
- Ataxia Hypotonia
- Cognitive Emotional Behavioural Changes
A Glimpse of PFS in the Pediatric Population

**Incidence**
- Wide range in literature from 8% up to 40%

**Onset of Symptoms**
- Short period of normal post-op functioning last from a few hours to few days
- Mutism is usually the first symptom that becomes evident

**Duration of Symptoms**
- Duration of symptoms varies widely, with symptoms lasting from days, to months, to years
Various Definitions and Classifications

- Posterior Fossa Syndrome
- Cerebellar Mutism Syndrome
- Cerebellar Mutism or Transient Cerebellar Mutism
- Mutism and Subsequent Dysarthria
- Cerebellar Signs or Cerebellar Syndrome

Gudrunardottir, et al. 2011
Possible Causes of PFS?

Axonal Injury
- Surgical manipulation
- Release of the compressive force resulting in axonal distortion

Edema
- Correlates with delayed onset of symptoms

Vascular Disturbances
- Ischemia
- Vasospasm
Posterior Fossa Region

Dr. Robert Draoul, 2015
Cerebellum

Anterior Lobe: Motoric Function (Balance and Coordination)

Posterior Lobe: Cognitive Function

Left Hemisphere: Spatial and Executive Tasks

Right Hemisphere: Language

Vermis: Regulation of Affect

Posterior Fossa Society, 2015
Dentothalamocortical Pathways

- This pathway brings information from the cerebellar nuclei about speech and oromotor function to the cortex.
- Damage along this pathway can lead to symptoms like those seen in post fossa syndrome.
Pediatric Risk Factors for PFS

<table>
<thead>
<tr>
<th>Known risk factors</th>
<th>• Tumour type, midline location, brainstem involvement and pre-operative brain stem compression</th>
</tr>
</thead>
<tbody>
<tr>
<td>Inconclusive risk factors</td>
<td>• Vermal incision, size of the mass, radical resection, edema in the cerebellum, younger age at diagnosis</td>
</tr>
<tr>
<td>No correlation</td>
<td>• Hydrocephalus, shunt treatment, meningitis, post-operative CNS infections, gender, length of vermali incision, type of neurosurgeon (adult vs. pediatric)</td>
</tr>
</tbody>
</table>
# A Comparison of Pediatric vs. Adult PFS

<table>
<thead>
<tr>
<th></th>
<th>Pediatric</th>
<th>Adults</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Mean or %</td>
<td>SD</td>
</tr>
<tr>
<td>Age</td>
<td>7.4 years</td>
<td>3.2</td>
</tr>
<tr>
<td>Male/Female</td>
<td>59.2% / 40.8%</td>
<td></td>
</tr>
<tr>
<td>Lesion Site: Midline</td>
<td>90.7%</td>
<td></td>
</tr>
<tr>
<td>Latency of Mutism</td>
<td>1.5 days</td>
<td>1.7</td>
</tr>
<tr>
<td>Duration of Mutism</td>
<td>49.7 days</td>
<td>85.5</td>
</tr>
<tr>
<td>Dysarthria</td>
<td>98.8%</td>
<td></td>
</tr>
</tbody>
</table>

Mariën et al., 2013
Etiology of Pediatric vs. Adult PFS

Mariën et al., 2013
Previously healthy 6 year-old male, M.H., with two week history of mild headaches. Reported having trouble watching TV, prompting parents to take him to the optometrist where papilledema noted.

Taken to local hospital to an ophthalmology clinic where an MRI was done showing a tumour with obstructive hydrocephalus. Transferred to Sick Kids for further management.

On arrival patient was ataxic and complained of intermittent double-vision. Vitals otherwise stable. GCS = 15.
## Case Study – Diagnostic Imaging and OR

<table>
<thead>
<tr>
<th>MRI</th>
<th>OR</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Post fossa lesion, centrally located, no midline shift</td>
<td>• No intraoperative complications</td>
</tr>
<tr>
<td>• Infiltration of medulla, and possible invasion of the vermis and cerebellum</td>
<td>• 90% of tumour was resected</td>
</tr>
<tr>
<td>• Minimal edema</td>
<td>• EVD placed</td>
</tr>
<tr>
<td>• Significant mass-effect on adjacent structures</td>
<td>• Verbally appropriate immediately post-op</td>
</tr>
<tr>
<td>• Displacement and compression of brainstem and cerebellum</td>
<td>• GCS = 14-15</td>
</tr>
</tbody>
</table>
### Nursing Implications – Post-op Care

Expected post-op complications are made more difficult by PFS

<table>
<thead>
<tr>
<th>Condition</th>
<th>Implication</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mutism</td>
<td>Pain → Unable to verbalize pain and discomfort</td>
</tr>
<tr>
<td>Dysarthria</td>
<td>Nausea/Vomiting → Higher risk for aspiration</td>
</tr>
<tr>
<td>Ataxia/Weakness</td>
<td>Incisional Leakage/Infection → Can prolong hospital stay and delay rehab and recovery</td>
</tr>
<tr>
<td>Behavioural Changes</td>
<td>Presence of EVD → Difficult to manage when the patient is irritable and uncooperative</td>
</tr>
</tbody>
</table>
# Case Study – Post-Op Days 1-3

<table>
<thead>
<tr>
<th>Day One</th>
<th>Day Two</th>
<th>Day Three</th>
</tr>
</thead>
<tbody>
<tr>
<td>Verbally responding</td>
<td>Decreased use of words</td>
<td>Drowsy</td>
</tr>
<tr>
<td>GCS = 12–13, moaning, minimal words</td>
<td>Weakness to all 4 limbs</td>
<td>No OT swallowing assessment due to decreased LOC</td>
</tr>
<tr>
<td>Further drop in LOC, GCS = 7–9</td>
<td>High EVD output → replacements started 1:1</td>
<td>Having fevers, query infections → antibiotics started</td>
</tr>
</tbody>
</table>
Nursing Interventions – Mutism and Speech

**Inability to Communicate**
- Support alternate use of communication such as word boards, signals and body language
- Involve family in the development of a communication system that is individualized

**Patient Frustration**
- Encourage patient to constructively express feelings through the use of age-appropriate therapeutic play
- Utilize reward system as positive reinforcement for appropriate behaviour

**Primary Resource:** Speech Language Pathologist
## Case Study – Mutism and Speech

<table>
<thead>
<tr>
<th>Week 1 Post-op</th>
<th>Week 2 Post-op</th>
<th>Week 3 Post-op</th>
</tr>
</thead>
<tbody>
<tr>
<td>Grunting and moaning</td>
<td>Using feet to communicate</td>
<td>Minimal improvement to speech, laughing/moaning</td>
</tr>
<tr>
<td>Diagnosed with PFS</td>
<td>Still only able to grunt or moan</td>
<td>Unable to use communication board due to ataxia</td>
</tr>
<tr>
<td>SLP started</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Communicating through body language</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Laughing and smiling appropriately</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

- **Week 1 Post-op**
  - Grunting and moaning
  - Diagnosed with PFS
  - SLP started
  - Communicating through body language
  - Laughing and smiling appropriately

- **Week 2 Post-op**
  - Using feet to communicate
  - Still only able to grunt or moan

- **Week 3 Post-op**
  - Minimal improvement to speech, laughing/moaning
  - Unable to use communication board due to ataxia
Nursing Interventions – Dysarthria

**Aspiration Pneumonia**
- Assess swallow and gag reflexes
- Initiate NG tube feeds when necessary
- Manage secretions through oral suctioning and medication

**Malnutrition**
- Monitor weight and calorie intake
- Provide NG teaching to families

**Primary Resources:** Occupational Therapist and Dietician
# Case Study – Dysarthria

## Week 1 Post-op
- **Drooling – requiring suctioning**
- **NG inserted and feeds started**
- **Mom attempting oral feeds**
- **Taste-stimulation started**

## Week 2 Post-op
- **NG still in place**
- **More alert**
- **Minimal improvement with oral movement**
- **Decreased management of oral secretions**

## Week 3 Post-op
- **NG teaching started**
- **Improvement in saliva management**
- **Still not ready for PO**
- **Thrush on tongue**
## Nursing Interventions – Ataxia and Hypotonia

| Skin Breakdown          | • Ensure positional support and frequent turning  
<table>
<thead>
<tr>
<th></th>
<th>• Assess for need for pressure relief mattresses</th>
</tr>
</thead>
</table>
| Pneumonia               | • Encourage deep breathing exercises            
|                       | • Promote activity                               
|                       | • Initiate chest physiotherapy if needed         |
| Muscle Wasting and Decreased Independence | • Encourage ROM exercises                        
|                       | • Use TEDS or anticoagulation as needed          
|                       | • Assess for appropriate mobility aids           |

**Primary Resource:** Physical Therapist and Respiratory Therapist
# Case Study – Ataxia and Hypotonia

<table>
<thead>
<tr>
<th>Week 1 Post-op</th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>No upper limb movement</td>
<td>Weak to legs</td>
<td>Poor trunk control</td>
<td>Diapered</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Week 2 Post-op</th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Improvement to balance when sitting</td>
<td>Increase in spontaneous and voluntary movements</td>
<td>Minimal improvement to limb strength</td>
<td>Fatigues quickly</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Week 3 Post-op</th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Small movements to hands</td>
<td>Increased head and neck control</td>
<td>Improvement to upper extremities</td>
<td></td>
</tr>
</tbody>
</table>
### Nursing Interventions - Behavioural and Emotional Changes

<table>
<thead>
<tr>
<th>Category</th>
<th>Interventions</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Emotional Lability</strong></td>
<td>• Use various activities the child enjoys as distraction and redirection&lt;br&gt;• Touch and hold child for comfort&lt;br&gt;• Prevent over-stimulation</td>
</tr>
<tr>
<td><strong>Personality Changes</strong></td>
<td>• Support families to help maintain hope and accepting all emotions&lt;br&gt;• Engage the patient in normalizing activities</td>
</tr>
<tr>
<td><strong>Family Frustration and Guilt</strong></td>
<td>• Involve families in care of child&lt;br&gt;• Take active interest in family well-being and coping</td>
</tr>
</tbody>
</table>

**Primary Resource:** Child Life Specialist, Clown and Pet Therapy Program
Case Study – Behavioural and Emotional Changes

Week 1 Post-op
Inconsistently following commands | Frustrated with inability to speak

Week 2 Post-op
Responding well to dog therapy

Week 3 Post-op
Laughing and smiling appropriately | Engaging with family
Nursing Interventions – Social Support

Coping
- Link with appropriate community resources and support groups
- Support families in accepting all emotions
- Encourage use of credible sources of information

Financial Aid
- Assist families in accessing additional financial resources
- Explore alternate accommodations

Caring for Siblings
- Explore extended support systems
- Encourage parents to spend time with other family members
- Provide respite care, assess need for PSWs or volunteers

Primary Resource: Social Worker and Chaplaincy
# Case Study – Social Support

<table>
<thead>
<tr>
<th>Week 1 Post-op</th>
</tr>
</thead>
<tbody>
<tr>
<td>Persistent fevers, cultures negative</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Week 2 Post-op</th>
</tr>
</thead>
<tbody>
<tr>
<td>Another discussion with staff MD</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Week 3 Post-op</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dad discussing own finding in literature</td>
</tr>
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</table>
Case Study – Overall Trajectory

**Acute hospital admission lasted 22 days**
- Throughout his admission, M.H. had recurrent fevers and leaks from the old EVD site
- At time of discharge, M.H. was still hypotonic, fully NG fed and not using any words

**Rehabilitation hospital admission lasted 170 days**
- During this time his NG tube was removed, he started PO feeds
- Speech progressed from a few words to using full sentences
- Slowly regained strength
Prognosis and Long Term Outcomes

One year after diagnosis, 92% had ataxia, 66% had speech and language dysfunction and 59% had some degree of global cognitive impairment (Robertson et al., 2006)

One long term study (Korah et al., 2010) which followed the recovery of patients with PFS showed that 78% of affected children still have persistent deficits after 18 years

Global language and cognitive skills were the most affected (Korah et al., 2010)
Case Study – Where is M.H. Now?

- Home and attending school with the aid of an education assistant
- Seen by SLP once per week, speech clearer with faster reaction time
- Has private physiotherapy at home to address ataxia
- Walking some steps with minimal support
- Undergoing 70 weeks of chemotherapy (weekly) treatments as an outpatient
- Tumor is stable, no further growth seen
<table>
<thead>
<tr>
<th>Take Home Messages</th>
</tr>
</thead>
<tbody>
<tr>
<td>With appropriate knowledge, skills and judgement, nurses can play an instrumental role in caring for children and families affected by PFS.</td>
</tr>
<tr>
<td>After first experiencing a diagnosis of a posterior fossa mass, families are now faced with complex and varied presentation of this debilitating syndrome.</td>
</tr>
<tr>
<td>Nurses are in a unique position to be able to provide care and advocate for the patient and their family and coordinate efforts of the multidisciplinary team.</td>
</tr>
<tr>
<td>As front line staff, nurses are also key players in furthering research about this poorly understood syndrome.</td>
</tr>
</tbody>
</table>
Nursing Management of Posterior Fossa Syndrome (PFS) in Pediatric Patients

Patricia Nikolski, BScN, RN & Venus Shyu, BScN, RN
Hospital for Sick Children, Toronto


Discussion Questions

What terms are used in your area of practice?

How do your patients present at your centres?

What symptoms do your patients typically have and how do you manage them?

Consent and disclosure of risk of PFS pre-op?