When It Is Not Just a Headache: Diagnosis and Management of Chiari Malformation

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Speaker Introduction

Dr. Teresa Whited is a certified pediatric primary care nurse practitioner who has practice for more than 15 years in primary care, pediatric cardiology and complex health care settings. Dr. Whited completed her BSN and MS at the University of Oklahoma and her DNP at Texas Christian University. She is interim associate dean of academic programs and director of the MNSc program for UAMS College of Nursing. She has a strong passion for educating future and current nurse practitioners, legislative issues and caring for all children but especially the most vulnerable like those with complex healthcare needs.

Disclosures

Teresa Whited receives speaker compensation for the Pediatric Primary Care Review Course from NAPNAP.

I will not be discussing any off label drugs in this presentation.

Learning Objectives

By the end of the session, participants will be able to:
• Identify the latest recommendations related to Chiari Malformation.
• Incorporate the key components of identifying and caring for children with Chiari Malformation.
• Discuss the nurse practitioner’s role in the care of children with Chiari Malformation.

What is Chiari Malformation

• A. Some foreign city that I am not sure I want to visit
• B. I don’t know but it doesn’t sound good
• C. A really bad headache
• D. A malformation of the brain associated with cerebellum

Why I care?

• Babies, children and Adolescent/Adults present differently
• Case study:
  • 8 month infant who presented with RSV and dysphagia
  • Newborns who have heart disease
  • Adolescents with Chiari Headache/Dizziness/Syncpe
What is Chiari Malformation?

- A group of complex brain abnormalities affecting the lower posterior skull
- Various Rhombencephalic anomalies of cerebellar tonsillar herniation
- Group of disorders characterized by abnormal morphology and positioning of the posterior fossa structures
- First described by Hans Chiari in 1890s
- Characterized by anatomical presentation:
  - Chiari I
  - Chiari II
  - Chiari III
  - Chiari IV

What is the most common form of Chiari Malformation?

- A. Chiari I
- B. Chiari II
- C. Chiari III
- D. Chiari IV

Chiari I

- Most common type:
  - Lower part of the cerebellum but no the brain stem extends into the opening at the base of the skull (foramen Magnum)
  - Caudal displacement of cerebellar tonsils 5mm below the foramen Magnum
    - 3-5mm by some sources
    - Especially if symptomatic

Chiari Type II

- Usually seen in children with Spina Bifida
- Also called Arnold-Chiari Malformation
  - Both the cerebellum and the brain stem extend into the foramen magnum
  - Sometime also involves the fourth ventricle

Chiari III

- Most serious form
- Protrusion or herniation of the cerebellum and brain stem through the foramen manum and into the spinal cord.
- Rare
- Causes significant neurologic deficits

Chiari IV

- Incomplete or poor development of cerebellum
- Rare type
- Associated with exposed parts of:
  - Skull
  - Spinal Cord
- May be significant deficits/incompatible with life
Some say there is:
• Chiari 0
  • Chiari 1.5
  ?
  Chiari 3.5

How often does Chiari I occur in the general population?
• A. ~15%
• B. ~1%
• C. ~7%
• D. ~30%

Epidemiology
• Prevalence was thought to be ~1% of the US population
• Female predominance (3:1 in adults)
• Increased incidence in Pacific Islanders
• Increase incidence with family history
  • Unknown genetic basis but thought to be linked as commonly recurs in families
• Estimated 215,000 Americans have Chiari I
• Asymptomatic at presentation:
  • 33-50% of children
  • 14-30% of adults
  • Median age of diagnosis in children
  • 8 years old

National Conquer Chiari Patient Registry Database
• Average time to diagnosis was 3.43 years (1st visit to diagnosis)
• Usual specialists seen:
  • Primary care
  • Neurology
  • Neurosurgeons
• Two most common misdiagnoses:
  • Psychological
  • Neurologic

Terminology/Associated conditions:
• Spina Bifida-Incomplete development of the spinal cord or its covering
• Syringomyelia-cysts develops in the spinal coards central canal (contains spinal fluid)
• Tethered spinal cord-the spinal cord is attached to the bony spine
• Hydrocephalus-Increased CSF fluid in the brain
• Scoliosis-abnormal curvature of the spine

The cause of Chiari?
• A. Genetic defect
• B. Lack of Folic Acid
• C. Unknown
• D. Infection
**Pathophysiology/Causes:**

- Unclear cause (genetic basis, environmental components)
- Discrepancy in neuronal contents of the forebrain/hindbrain and skeletal deformities of the cranial vault and cranio-cervical junction
- Secondary to insufficiency of the paraxial mesoderm after neural tube closure and underdevelopment of occipital somites
- Disruption in flow of CSF at the craniocervical junction
- May lead to syrinx formation or symptoms of Chiari
- Can be acquired:
  - Secondary to hydrocephalus
  - CSF leak or shunts (lumboperitoneal cause negative downward pressure
  - Spinal cord tethering pulling downward
  - Increased ICP

**Precipitating Events**

- Car accident (especially with whiplash injury)
- Falling
- Pregnancy (especially with difficult vaginal delivery)
- Head trauma

**Symptomatology**

- Can be asymptomatic
- 1–4% of patients will have Chiari identified on MRI of brain
- Unless significant concerns
- Follow clinically
- Monitor for symptoms
- Consider repair if worsening findings or symptoms

**Common symptoms of Chiari Malformation I (CMI)**

<table>
<thead>
<tr>
<th>Common Symptoms Infant/Young Child</th>
<th>Common Symptoms Older CMI/Adult</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sleep apnea</td>
<td>Headache (occipital) worsened by Valsalva maneuver/cough</td>
</tr>
<tr>
<td>Oropharyngeal dysfunction</td>
<td>Neck pain</td>
</tr>
<tr>
<td>Cranial nerve dysfunction</td>
<td>Syncope</td>
</tr>
<tr>
<td>Failure to Thrive</td>
<td>Dizziness</td>
</tr>
<tr>
<td>GERD</td>
<td>Ear pain, tinnitus, hearing loss, tinnitus</td>
</tr>
<tr>
<td>Strabismus</td>
<td>Eyes blurred vision, double vision, strabismus, sensitivity to light</td>
</tr>
<tr>
<td>Irritability</td>
<td>Bladder problems</td>
</tr>
<tr>
<td>Atypical crying</td>
<td>Sleep apnea</td>
</tr>
<tr>
<td>Weakness/gait dysfunction</td>
<td>Weakness/Sensory issues in upper back and arms</td>
</tr>
</tbody>
</table>

**Most common Symptom of Chiari 1?**

- A. Headache
- B. Sensory deficits
- C. Dizziness
- D. Gait disturbance

**Less common symptoms in Chiari I**

- Brain Fog
- Chronic Pain
- Depression
- Anxiety
- Hiccups
- Severe snoring
- Facial numbness
- Memory problem
What test should be ordered for diagnosis?

- A. Spine MRI
- B. Brain MRI
- C. Brain CT
- D. EEG

Algorithm for Chiari (BMJ, 2019)

Diagnostic studies: How to get the diagnosis??

- Many patients undergo a variety of studies:
  - X-ray
  - CT of brain
  - MRI of neck
  - Mylogram
  - Tilt test
  - Echocardiogram/EKG
  - Psychiatric testing
  - Neurologic exam

Gold standard for Diagnosis:

- MRI of the brain is the gold standard for diagnosis
  - Allows visualization:
    - Brain
    - Brainstem
    - Ventricles
    - Cerebellar tonsils
    - McRae Line
      - Imaginary line drawn at foramen magnum

Management of Chiari Malformation

- Conservative Management
  - Medical management of pain and other symptoms
    - NSAIDs
    - Muscle relaxers
  - No intervention
    - If asymptomatic
    - Follow by neurology annually

The NP is seeing an 8 year old female with Chiari 1 malformation found incidentally on MRI of brain following a car accident. She is asymptomatic at time or presentation. What is the most appropriate management for this patient?

- A. Monitor clinically with intervention if symptomatic
- B. Refer to neurosurgeon for surgical correction
- C. Refer to chiropractor for manual manipulation
- D. Refer to OT/PT for improvement in ROM
Indications for Surgery

- Syrinx
  - Symptomatic or worsening
- Hydrocephalus
- Crowding of the posterior fossa causing brainstem compression
- Severe and short lived intensity of headache worsened by VM
- Increase in headache days per month
- Abnormal neurologic examination
  - Sleep apnea
  - Feeding difficulties
  - Ataxia
  - Etc.

The risk of complications form Chiari Surgery is?

- A. 20%
- B. 50%
- C. 10%
- D. 3-5%

Risks/possible complications of Surgery

- Neurologic compromise
- CSF leakage
- Infection
- Vocal cord or swallowing difficulties
- Severe blood loss
- Infection
- Death
- Lack of resolution of symptoms
- 3-5% risk of complication

Management of Chiari Malformation

- Surgical Management
  - Bony Decompression of the posterior fossa
    - With or without duraplasty (controversial)
  - May increase risk of complications or not
  - Goal of surgery is to restore CSF flow at the level of the foramen magnum
  - 80-95% show improvement in headache
  - More variable when other symptoms involved but overall improve outcomes
- Syrinx
  - Children more likely to resolve

Surgical Management

- Surgery performed by:
  - Neurosurgeon
  - Patient placed supine for initial induction
  - 3 point head holder rotated to prone position
  - Incision made from occipital bone to C2
  - Posterior occipital bone and foramen magnum/C1 region enlarged
  - Duraplasty
    - Tonsils are spread apart
    - 4th ventricle to subarachnoid space-free communication of fluid
    - Dural graft to close defect (gore-tex, pericranium)

Medical management during and following surgery:

- Preoperative antibiotics
  - Cefazolin 1gm IV
  - Vancomycin 1gm IV
- Postoperative
  - Monitored in neurosurgical ICU or intermediate care unit for 1 night
    - Frequent neuro checks for first 8 hours
    - Looking particularly for lower brainstem, CN dysfunction
    - Poor gag/swallowing
    - Nystagmus, difficulty hearing
    - Respiratory depression
    - Hypotension
Medical management during and following surgery

- Pain management
  - Headache/posterior neck pain expected
- 1st day
  - Opioid management
  - PCA pump
- 2nd day
  - Oral opioid management
  - Other analgesics
- Two weeks
  - Other analgesics
  - Occasional opioid at bedtime

Medical Management during and following surgery

- Incision Care
  - Monitor dressing for CSF leak
  - If healing well, removal of dressing with 3-5 days
  - Open to air
  - No submerging in bath until fully healed
  - Don’t let shower beat on incision
- Diet
  - If swallowing safe, clear to full diet advance
  - Prevention of pneumonia and DVT
  - Activity-up on side of bed evening of surgery, advance to walking
  - Avoid bending, straining or lifting more than 10lbs for 6 weeks.

Medical Management during and following surgery

- Discharge
  - Average hospital stay is 2-4 days
  - No driving while on narcotics
  - No staying alone for several days following procedure
  - Return to work/school 2-6 weeks post-operative
  - Follow up for wound evaluation/post-operative care in 1-6 weeks

The NP is seeing a 12 year old girl who underwent surgical repair of Chiari 1 malformation 6 weeks ago. Mother asks how long the patient has to be followed. The correct response would be?

- A. Until asymptomatic for 1 year
- B. Until postoperative for 2 years
- C. Lifelong
- D. Until asymptomatic for many years

Long Term Implications

- Follow up in 3 months with repeat MRI
  - Evaluate resolution of symptoms if going to occur then will be resolved by then
  - Evaluate structural change and syrinx
- Follow up until asymptomatic for several years
  - If symptomatology, may need re-intervention
  - Slightly higher risk without duraplasty
- May need OT/PT and other multi-disciplinary team
  - Associated conditions
  - Rehab from surgery/symptoms

Latest research

- Upright MRI
  - Gravity might reveal tonsillar displacement that was not seen in traditional MRI
- CSF space in the craniocervical junction
  - Computation of this in relation to systole
- Potential Subgroups of Chiari
  - O, 1.5, 3.5, additional types of Chiari 1
- Surgical Techniques
  - Identify best outcomes with dura vs non-dura
- Genetic markers
  - Identify genetic component or other markers that may lead to family transmission
Follow up on baby:

- Bony Decompression without duraplasty
- OT/ST for feeding difficulty
- Repeat dysphagogram scheduled
- PT/OT for developmental delays
- Repeat sleep study scheduled
- Parents report

References: