Management of Chiari I Malformation or Anomaly

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I have no conflicts of interest related to this presentation.
The Chiari I Malformation

An Unfortunate Name That Inevitably Leads To Confusion
Part of the Problem Relates to Definition

Chiari I

Normal with Cistern

Normal No Cistern
The Cisterna Magna

- A potential space filled with spinal fluid behind the cerebellar vermis
- A point of mixing of CSF from the fourth ventricle, spinal subarachnoid space and cortical subarachnoid space
What Constitutes Success
Why The Name Is So Terrible

• Giving a condition the name X malformation implies that it
  • Has a single definable cause
  • Began before birth
  • Does predictable things to the patient or the patient’s brain.
Arnold-Chiari malformation or often simply Chiari malformation is a malformation of the brain. It consists of a downward displacement of the cerebellar tonsils through the foramen magnum.

Chiari malformations are structural defects in the cerebellum. They can be caused by defects in the spinal cord and brain that occur during fetal development.

http://webclipart.guide@about.com
Almost everything is controversial

- Definition of Chiari I
- Causes of Chiari I
- Is it malformative
- What makes the patient a candidate for surgery?
- Relationship to Fibromyalgia
- What is the appropriate surgical treatment?

What defines success or failure?
Chiari Malformation Diagnosis

There is general agreement among neuroradiologists that the diagnosis of Chiari I involves a descent of the Cerebellar tonsils greater than 5 mm. Why 5 mm? What causes the problem?
Part of the Problem
Relates to Definition

Chiari I

Normal with Cistern

Normal No Cistern
What’s Missing

Cisterna Basilica Istanbul
From Memet Ozek
Why is the Cisterna Magna Important?

• Serves as a hydraulic capacitor
• Stores CSF for its critical role in the distribution of intracranial pressure transients
• Buffers the brain and spinal cord from compression at the time of neck movement
Hindbrain Herniation

Begins with a hole that is incompletely covered

Next comes a force pushing or pulling something through the hole
Step 1

- Skin incision from overlying the spinous process of C2 to vertex well above the inion
- Care is taken to preserve the pericranium above the nuchal line
- Harvest the pericranial patch about 4 cm in diameter
Why the Pericranium

- Use of pericranial patch requires the incision to extend above the nuchal line
- The tissue is more elastic and thicker than available manufactured patches
- Since changing to the pericranium there has been a dramatic decline in chemical meningitis and a measureable decline in pseudotmeningocele
Step 2

• Cranietomy of Squamous portion of occipital bone. 3X3 CM at Foramen Magnum

• C1 Laminectomy

• Open Dura from just above C2 to above the Marginal Sinus then create a “Y”
Caution

• In order to stop bleeding in sinuses, even the marginal sinus it is essential to compress the two leaves of the dura that create the sinus together.

• In operating on babies the posterior fossa dura may be a large venous lake
Remember
Manufactured Patches
Derived from Biological Materials Must be Soaked for 30 minutes
The Patch

• Use braided suture (I prefer 4-O Neurolon)
• Place stay sutures in the three corners of the dural opening.
• Running suture
• Multiple valvsalva maneuvers
The Chiari Plate

• Place three 2-0 Ethibond sutures in the patch and leave long

• Bend the Chiari plate to create a dome

• Screw the plate to the edges of the bone to cover the decompression

• Bring the Ethibond through the interstices of the plate and fix the patch to the back of the plate
Why the Plate

- The dome creates a space behind the cerebellar tonsils (Neo cisterna magna)
- It prevents the nuchal muscles from pushing into the repair
- Prevents pressure on posterior fossa in recumbancy
- You don’t have to “shrink” the tonsils
My Surgical Approach

• If you decide upfront that the goal of surgery is to recreate a cisterna magna then it is a simple thing to analyze the outcome
• Limited suboccipital craniectomy (2X3 CM)
• C1 Laminectomy
• Always open the dura unless there is a clear boney cause of the herniation
• Generous pericranial patch
• Posterior fossa reconstruction with plate
Posterior Fossa Reconstruction
6 months postoperative
Conclusions

• All hindbrain hernia conditions have a root cause
• While sometimes the cause is still obscure it is essential to look for it as the management differs with different causes
• We will eventually be able to determine the cause in all cases
Conclusions 2
What is success?

Successful management of the hindbrain hernia requires the creation or recreation of a cisterna magna.