The Chiari Malformation: What It Is and How to Manage It

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The Chiari Malformation - Overview

- History
- Classification
- Demographics
- Presentation

- Evaluation
- Treatment
- Outcomes
- Acquired variety
History

- 1883 - First described by Cleland.
- 1891 - Hans Chiari (Austria) developed classification system. (Julius Arnold was a colleague.)
- 1932 - First report of surgery.
- 1940 - Association with syringomyelia.
Classification

- Spectrum of hindbrain/upper cervical anomalies
- “Chiari malformation” typically used for type I
- “Arnold-Chiari malformation” usually refers to type II
Type I

- Herniation of cerebellar tonsils
- Brainstem typically normal
- 30-70% with associated syrinx
- Young adult presentation
Type II

- Caudal displacement of cervicomedullary junction
- Small posterior fossa
- Beaking of tectum
- Commonly with myelomeningocele and hydrocephalus
- (Many other possible features)
Others

- Chiari 0 - “Chiari-like” without tonsillar descent
- Chiari III - Cerebellum into cervical canal, often with encephalocele
- Chiari IV - Cerebellar hypoplasia without herniation
Hear ye, Hear ye--

- Henceforth, we will focus only on type I, “the” Chiari malformation
Demographics

- Prevalence slightly less than 1:1000
- Average age – 41 (range 12-73)
- Female: male 1.3:1
- Symptoms for 3.1 years average (range 1 mo – 20 yrs)
- Many individuals with radiographic Chiari are asymptomatic
Presentation

- Brainstem compressive symptoms
- Hydrocephalus
- Syringomyelia
- Association with behavioral/psychiatric issues
Presentation

- Headache
- Weakness
- Numbness
- Loss temperature sensativity
- Burning
- Balance issues
- Diplopia
- Dysphagia
- Tinnitus
- Vomiting
- Dysarthria
- Sleep apnea
- Syncope
Presentation

- Suboccipital headache
- Typically worsens with valsalva maneuvers
- Can be positional
Evaluation

- MRI
- Others
  - X-rays
  - Cine MRI (CSF flow study)
  - Myelography
  - CT
Evaluation

- Degree of tonsillar herniation
- Tonsils ascend with age
- Various criteria for radiographic Chiari I
- Ultimately, symptoms/signs (not some number) predict response to treatment
Evaluation

- Syringomyelia
  - From alteration of CSF flow
Treatment

- **Goals**
  - Decompression
  - Normalize CSF dynamics

- **Indications**
  - Symptomatic
  - Syringomyelia
  - Hydrocephalus

- **Techniques**
  - Suboccipital craniectomy
  - ± Cervical laminectomy
  - ± Duraplasty
  - ± Separation of tonsils
  - ± Shrinking tonsils
  - VP Shunt if needed
  - ± Spinal stabilization
Treatment
Outcomes
Outcomes

- Complications
  - CSF leak
  - Respiratory depression
  - Meningitis
  - Vascular injuries
  - Craniocervical instability
  - Cerebellar herniation
  - Inadequate decompression
Acquired Chiari Malformation

- Radiographically can be identical to idiopathic
- Can have positional headaches
- Standard Chiari treatment could be disastrous
- Treatment is to address primary problem

Common causes

- Intracranial hypertension
  - Hydrocephalus
  - Tumor
- Intracranial hypotension
  - CSF fistula
  - Lumboperitoneal shunting
Take-Home Points for Primary Care Providers

- Patients with discovered Chiari’s should be referred for neurosurgical evaluation.
- Patients with symptoms should be worked up for Chiari’s.
- Treatment is based on symptoms and associated findings, rather than extent of tonsillar herniation.
- Work-up should include MRI brain and cervical spine.
- Referral can typically be non-emergent, though some exceptions exist
  - Hydrocephalus
  - Apnea
- Outcomes tend to be better if surgery performed sooner after symptom onset.
Comparative Anatomy
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Thank you.

Questions?