Surgical Management of the Chiari Malformation
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Historical Descriptions

• Antoine Portal -1803 described the post-mortem spinal cord of a servant with ascending numbness and eventual paralysis who had a cavity within the cord
• Charles Prosper Ollivier d’Angers – 1827 coined term syringomyelia
• John Cleland – 1883 described the hindbrain anomalies in a spina bifida patient
• Hans Chiari – 1891 described three types of hindbrain anomaly and an expanded report in 1896 with 24 cases described and illustrated
History

John Cleland-1883

Hans Chiari-1891
The Chiari Malformation and Syringomyelia in Children
CMI is a disorder of the mesoderm and is thus inherently different from Chiari II and Chiari III malformations. The anomaly occurs sporadically but can be transmitted genetically in some families. The most constant feature of CMI is a volumetrically small PCF, which predisposes patients to hindbrain overcrowding. Displacements of CSF probably contribute to the symptoms.

The clinical syndrome of CMI is characterized by headaches, pseudotumor-like episodes, a Meniere's disease-like syndrome, lower cranial nerve signs, and spinal cord disturbances even in the absence of syringomyelia.

Diagnosis is established by MRI. Minimal evidence of hindbrain overcrowding consists of obliteration of the retrocerebellar CSF spaces in association with a meniscus sign at the lower pole of the cerebellar tonsils.

CINE-MRI can be helpful in demonstrating a disturbance of CSF velocity/flow at the foramen magnum in patients with tonsillar herniation of less than 5 mm.
<table>
<thead>
<tr>
<th>Symptoms and Signs</th>
<th>No. of Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>CMI (n = 126)</td>
</tr>
<tr>
<td><strong>Subjective</strong></td>
<td></td>
</tr>
<tr>
<td>Sensory</td>
<td></td>
</tr>
<tr>
<td>Paresthesia/hyperesthesia</td>
<td>40</td>
</tr>
<tr>
<td>Nonradicular segmental pain</td>
<td>29</td>
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<tr>
<td>Analgesia or anesthesia</td>
<td>24</td>
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<tr>
<td>Burning dysesthesia</td>
<td>15</td>
</tr>
<tr>
<td>Poor position sense</td>
<td>11</td>
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<tr>
<td>Impaired temperature sensation</td>
<td>3</td>
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<tr>
<td><strong>Motor</strong></td>
<td></td>
</tr>
<tr>
<td>Muscular weakness</td>
<td>51</td>
</tr>
<tr>
<td>Spasticity</td>
<td>16</td>
</tr>
<tr>
<td>Muscular atrophy</td>
<td>3</td>
</tr>
<tr>
<td>Trophic phenomena(^b)</td>
<td>15</td>
</tr>
<tr>
<td>Urinary incontinence</td>
<td>9</td>
</tr>
<tr>
<td>Impotence (75 adult male patients)</td>
<td>3/24</td>
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<tr>
<td>Fecal incontinence</td>
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</tr>
<tr>
<td><strong>Objective</strong></td>
<td></td>
</tr>
<tr>
<td>Sensory</td>
<td></td>
</tr>
<tr>
<td>Analgesia or anesthesia</td>
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<tr>
<td>Dissociated sensory loss</td>
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<tr>
<td>Impaired position sense</td>
<td>5</td>
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<tr>
<td><strong>Motor</strong></td>
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<tr>
<td>Muscular weakness</td>
<td>25</td>
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<tr>
<td>Impaired fine-motor function</td>
<td>48</td>
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<tr>
<td>Increased muscle tone</td>
<td>8</td>
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<tr>
<td>Muscular atrophy</td>
<td>1</td>
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<tr>
<td><strong>Reflexes</strong></td>
<td></td>
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<tr>
<td>Hyperreflexia</td>
<td>18</td>
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<tr>
<td>Clonus or Babinski sign</td>
<td>7</td>
</tr>
<tr>
<td>Hyporeflexia</td>
<td>1</td>
</tr>
<tr>
<td>Trophic abnormalities</td>
<td>4</td>
</tr>
<tr>
<td>Affected patients(^c)</td>
<td>83</td>
</tr>
</tbody>
</table>

\(^a\) CMI, Chiari I malformation; SM, syringomyelia.

\(^b\) Includes cold or pale extremity, hyperhydrosis, glossy skin, and gangrene of toes or fingers.

\(^c\) Affected patients were defined as those with two or more symptoms.
The Chiari Malformation and Syringomyelia in Children

The Vomiter

The Scratcher
This 3 year old boy had vomited almost every day since his first week of life. Although gaining weight, he had multiple GI workups including endoscopy and radiological workups that had been repeated at various intervals. An MRI revealed a Chiari Malformation with abnormal somatosensory evoked potentials but a normal exam. After decompressive surgery, he ate in the recovery room, and 11 years later has been asymptomatic. His only complaint is his neurosurgeon will not let him play football.
This 6 year female complained of neck pain and occipital headache off and on for years. She was treated expectantly with Tylenol. Due to years of headache, and vague complaints of dizziness, an MRI was performed. The only finding on exam was an area of constant excoriation of the skin on her left lateral calf from what she described as constant fire ants or mosquitoes biting her all the time. The neurological exam was normal. She was dismissed three days after decompression of the brainstem. At one year the cutaneous findings on the leg had resolved as had her headaches and the syrinx on MRI. Exactly one year after her surgery she was hit in the back of the head with a baseball at her operative site. An immediate CT scan of the head was negative. Due to recurrence of the leg complaints, the rash, etc., she had a repeat MRI. The syrinx had returned. She underwent another exploration of the area, scar tissue was removed, a dura graft placed, and at one year everything had normalized.
Pre and Postoperative MRI
Syringomyelia Mechanisms
Syringomyelia Mechanisms

• Gardner- outlet obstruction at Magendie and resultant misdirection of flow
• Williams- added that epidural venous pulsations created pressure gradients that exacerbated fluid movement within the syrinx
Syringomyelia Mechanisms

Ball, Daylan, Alouker-
Spinal Subarachnoid
pressure is higher than
that in syrinx, driving
CSF into the cavity.
Does this trans-
ependymal egress
eventually result in
gliosis?
Prior to the CT and MRI era, patients with this entity usually underwent diagnostic studies only if symptoms and exam findings warranted performing invasive procedures such as air myelograms, angiograms, ventriculograms, and eventually intrathecal contrast computed tomography. These procedures had small but significant morbidity.

“Three types of pneumoencephalograms; diagnostic, therapeutic, and punitive” – J. Yeakley, MD May 1, 2009
Air Ventriculography

From 1918 until the 1970’s, about 50 years, neuroimaging involved injecting air and eventually contrast media (oil based, then water soluble) into the CSF. Coupled with the then evolving cerebral angiography and the HISTORY AND EXAMINATION, decisions were made as to whether patients should undergo neurosurgical procedures. One might think ‘exploratory brain surgery’ should be a better term, however the accuracy of said scenarios cannot be underestimated.
Contemporary

MRI, the current imaging modality of choice for most initial neurological testing will readily delineate tonsillar descent below the foramen magnum. The incidental finding of this entity often is the cause of much distress and debate not only for the parent but for the doctors.
Incidence

Kaiser-Permanente estimate from California
Personal Communication – Conquer Chiari Research Symposium 2007,
University of Illinois at Chicago

1 per 1000
The question then becomes what to do if anything in the asymptomatic patient
The clinical presentation, exam, and neurological evaluation

Ian J. Butler, MD
CHIARI MALFORMATIONS

CLINICAL FEATURES

1) HEADACHES
2) HYDROCEPHALUS – Luschka & Magendie foramina
3) CHIARI
4) MIGRAINE
5) PSEUDOTUMOR CEREBRI
CHIARI MALFORMATIONS
ANATOMY OF CLINICAL FEATURES

• HYDROCEPHALUS
• CEREBELLAR- Ataxia
• BRAINSTEM- Vomiting, abnormal respirations
• CRANIOCERVICAL JUNCTION
  – -Posterior columns (topography)
  – -Corticospinal tracts
• SPINAL CORD- SYRINGOMYELIA/ TETHERED CORD
  – -Posterior column- gait
  – -Sensory- pain, temperature
  – -Anterior horn cells- focal wasting
  – -Corticospinal- spasticity
  – -Autonomic- bladder, bowel, vascular
CHIARI MALFORMATIONS
DIAGNOSTIC STUDIES

- MAGNETIC RESONANCE IMAGING (MRI)
  - BRAIN
  - CRANIOCERVICAL JUNCTION- CSF flow analysis
  - SPINAL CORD
- SOMATOSENSORY EVOKED RESPONSES- SEPs
  - Both arms and legs
- BRAIN STEM EVOKED RESPONSES- BAER
- CEREBROSPINAL FLUID- pressures changes
- COMPUTERED TOMOGRAPHY- bone changes
CHIARI MALFORMATIONS MANAGEMENT
“Conservatively aggressive”

• MIGRAINE
  – Prophylaxis with two (2) medications (propranolol, amitriptyline)
• PSEUDOTUMOR CEREBRI
  – Post-operative leaking
  – Acetazolamide
• SURGERY
  – Hydrocephalus, Chiari malformation, syringomyelia, tethered cord
• NEUROIMAGING
  – Serial brain and craniocervical junction and spinal cord
• EDUCATION- genetics
What we do not know

• Natural history of Chiari I malformation
• Best operation if needed
• Inheritance pattern
Trauma Chiari Data

• 18 patients with Chiari I malformation discovered as a consequence of either an MRI of the brain or cervical spine performed in conjunction with a trauma evaluation

• None had symptoms suggestive of what we consider ‘Chiari complex’ prior to the injury

• One a 15 year old had surgery elsewhere after a second injury, baseball injury, caused neurological symptoms

• A twin female fell from a high chair and sustained a cerebellar contusion. So far no surgery but off and on complaints. The twin has a Chiari also.

• The rest have had no further complaints or evaluation

• Follow-up from 5 to 13 years.
What we know

• **Tonsillar herniation** has been known to spontaneously regress in kids, probably many factors, but growing skull and spine probably play a big role

• **Tonsillar herniation** can be associated with entities that cause elevated intracranial pressure, tumors, craniosynostosis, lumbar drains, overshunting, undershunting, postoperative craniotomies, tethered spinal cord, and a variety of other entities that alter the dynamics of the neuraxis.

• Clinical presentations, when due to the Chiari malformation, vary tremendously

• Strong association of Chiari with Syringomyelia, less than previously because of larger denominator of MRI studies being done

• Hydrocephalus when present complicates the picture, and often once treated may still not change the clinical picture, further complicating decision making.

• Pedi Neurosurgery / UT Pedi Neurology
  
  Three sets of twins with Chiari
  
  One set of quadruplets, two with, two not studied, mom with Chiari
  
  Multiple pts and mothers with Chiari
The clinical picture may be due to any of the following factors or combinations of these:

- Direct compression of the brainstem, or cranial nerves
- Constant (pseudotumor cerebri like states) or transient alterations of CSF flow
- Repetitive trauma due to positioning or motion
- Constant or transient ischemia
- Traction on the brainstem from tethering of the spinal cord
Surgical Options

• There is no data to support a particular surgical technique as having an advantage over others. However general neurosurgical principles regarding this entity would dictate that it is important to:
• Alleviate the patients’ complaints if present (what about prophylactic operations)
• Relieve pressure on the brainstem
• Improve CSF flow/dynamics around the brainstem with a return to ‘normal’ state
• Prevent instability of the cranio-cervical junction if that is a potential concern
• Provide a physiologic milieu that enhances the resolution of syringomyelic cavities and symptoms if present.
• Dr Manish Shah spearheading study to assess the best technique
Therefore, what is the best operation

AANS SURVEY

Bone, cervical laminectomy, duraplasty  JNS 1998

Bone decompression alone with or without duraplasty based on electrophysiology intraoperatively. Fletcher, Butler, Mancias, et al. BJNS 11.08, Abstracts from Syringomyelia and Chiari research conference, Rugby England.
The Syrinx

• Classic Capelike distribution of sensory defects or more importantly variations on that theme.

• Often normal exam even in patients with a large syrinx
Surgical Management

- Bone decompression
- Removal of skull base and lamina
- Dural graft
- Fusion in some cases
University of Texas Neurology and Neurosurgery
Chiari / Syringomyelia

• April 1, 2003 to 2009
  – 1364 patient *encounters* for Chiari I, Syringomyelia, or a combination of the two
  – 98 operations, 2 Surgeons
    • 97 craniectomy and laminectomy (classic chiari surgery)
    • 2 Syrinx to Subarachnoid shunt
Primary complaints in 98 surgeries

• 3 - Syncope with negative cardiac workup
• 5 – Vomiting with negative GI workup
• 2- Thoracic radiculopathy and back pain
• 1- arm pain and weakness
• 1- leg pain and weakness
• 5 – toe walking
• 81 headache, headache variant , and/or other
Chiari and Associated Conditions that factor in on causation and potential treatments

- Hydrocephalus
- Migraine headache and variants
- Pseudotumor Cerebri and other causes of general elevated intracranial pressure
- Cranial base abnormalities
- Previous trauma
- Craniosynostosis
- Retardation of skeletal growth
Diagnostic Adjuncts

- Cervical Spine Films
- Scoliosis Survey
- MRI of entire spine
- SSEP’s and ABR’s
- Sequential histories, exams and all of the above
- Lumbar Puncture
- Intracranial Pressure Monitoring
- CT of Skull in selected cases?
Investigative Tools

• Volumetric Studies-Elswick, C (Wayne State) and Hochauser, L  UTHSCH
• Configuration studies of the posterior fossa
• Electrophysiology
  – Somatosensory Evoked Potentials
  – Auditory Brainstem Responses
  – Sleep Studies
Surgical Adjuncts

• Endoscopic approaches
• Intraoperative Ultrasound
• Intraoperative SSEP’s and ABR’s
Surgical Complications

• CSF leak
• Chemical Meningitis
  – Dural graft issues
• Cervical spine Instability
• Stroke
• Arachnoiditis
• Pseudomeningocele
Syringomyelia Resolution

- Decompression of Chiari establishes better flow
- May explain the decrease in size of syrinx
- Walls of cavity may have become stiff however and gliotic, preventing collapse of syrinx and improvement of symptoms
Conquer Chiari Research Symposium II. NIH/NINDS

Proposals for NIH funding for research especially those interested in cooperative programs include:

Chiari registry for natural history studies
Best operative procedure
Chronic pain management studies for pts with syringomyelia
Pathophysiology of Syringomyelia
Future Surgical Investigations

- Volumetric Classification with Configuration Categories to select best operative treatment
- Minimally invasive surgeries – advantage?
- Lamina sparing operations – prevention of deformities?
Diagnostic and Therapeutic Research

• Genetic studies underway at Duke others
• Molecular biology of various bones of the skull base - England
• Deformational studies of spinal cord models in syringomyelia – Australia, University of Illinois
• Outcome studies/best operation- Dr. Shah, UT
• Associations of Chiari and Autism Spectrum and other entities
References

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Patient Education Sites

• Conquer Chiari- Conquerchiari.org
• American Syringomyelia Alliance Project- ASAP.ORG