CHIARI MALFORMATION TYPE 1 – PATHOGENESIS, MANAGEMENT AND CONTROVERSIES

History

www.marvistavet.com

- Hans Chiari (1851–1916) was born in Vienna, Austria
- He was Professor of pathology at Prague, Czechoslovakia
- His initial work was published in Deutsche Medizinische Wochenscriff in 1891 and entitled “Concerning alterations in the cerebellum resulting from cerebral hydrocephalus.”

Other major contributors

- John Cleland (1883) - hind brain hernia in post mortem examination of a patient with myelodysplasia
- In 1894, Julius Arnold (Professor of pathologic anatomy) described an infant with spina bifida in whom there was, elongation of the hind part of the cerebellum, covering the fourth ventricle and extending into the cervical canal.
- Name of these professors often associated with the type II herniation
- Considering his detailed analysis - more often called *CHIARI MALFORMATIONS*

History

- Adult cases of Chiari I malformation not described until 1938, when Mc-Connell and Parker reported five cases, all with hydrocephalus and neurological symptoms
- Used the term "tonsils" to describe the prolapsed cerebellar tissue

McConnell AA, Parker HL: A deformity of the hind-brain associated with internal hydrocephalus. Its relation to the Arnold- Chiari malformation. Brain 1938; 61:415-429

Type	Definition
Chiari I	Caudal descent of *cerebellar tonsils* > 5mm below foramen magnum, *SYRINX seen in _50-70%* *HYDROCEPHALUS UNCOMMON*
Chiari II	Caudal herniation of *cerebellar vermis, brainstem and fourth* ventricle *ALMOST ALL HAVE HYDROCEPHALUS AND MYELOMENINGOCELE*
Chiari III	Chiari II and posterior fossa contents herniating into occipital/ high cervical encephalocele
Chiari IV	Cerebellar aplasia or hypoplasia with aplasia of tentorium cerebelli

Newer additions

Chiari 0 (described by Iskandar BJ et al)

- No hind brain herniation
- Syringo hydromyelia
- Marked improvement with posterior decompression

Chiari 1.5

- Somewhere between I and II
- Tonsillar herniation (I) and elongated caudally displaced brainstem and 4th ventricle (II)

How common is Chiari 1 ??

- 1/1280 in a series analyzed at Johns Hopkins

  Meadows et al. Asymptomatic chiari type I malformations identified on MRI 2000;92:920-26

  22,000 brain MRs reviewed.?? Referral bias

- Familial clustering has been established recently

- *Milhorat et al. Chiari I redefined: clinical and radiological findings for 364 symptomatic patients. 1999;44:1005-1117*

- When one member of an identical twin has CM I, the chances of the other having CM is higher

- *Speer et al .Chiari I with or without syringomyelia prevalence and genetics. Journal of Neurosurgery 2003;12:297-311*

Pathophysiology

1) Ratio of the posterior fossa neural element volume to posterior fossa cranial volume is increased. This is related to

- either an underdeveloped occipital somite originating from the paraaxial mesoderm, which results in a smaller posterior fossa,

- or to an overgrowth of the supratentorial component and consequent shallow posterior fossa.

2) The CSF flow across the foramen magnum is abnormal during systole and diastole and, thus, increased tonsillar velocity and pulsation occur.

The normal CSF flow in the spinal compartment is prevented.

Chiari I malformation

- Disorder of para-axial mesoderm (fourth occipital sclerotome)
- Subnormal posterior fossa volume but no reduction in infratentorial brain volume precipitates hindbrain herniation through FM **(Consistent with observed association seen with some hereditary connective tissue disorders like Ehlers-Danlos syndrome)**

Milhorat TH et al. Syndrome of occipitoatlantoaxial hypermobility, cranial settling, and chiari malformation type I in patients with hereditary disorders of connective tissue. J Neurosurg Spine 2007;7:601-9.

Theories of Syrinx

- Williams "Pressure differential" theory is currently the most accepted one
- Normally, with the valsalva maneuver; engorgement of epidural veins- compression of spinal subarachnoid space-pressure wave moving cephalad with no impedance
- When pressure returns to normal reverse flow occurs out of intracranial cavity
- Adhesions and tissue in foramen magnum interfere with equilibration of pressure

Theories

- Pressure differential is generated, opening up alternate pathways of decompression
- Spontaneous resolution in some cases supports the differential pressure concept
- William supported this theory by measuring the intraspinal and intracranial pressure gradients

Theories

- **Gardner** - **Hydrodynamic theory**

  Failure of rhomboid roof perforation or medial and lateral foramina development results in a noncommunicating hydrocephalus.

  Pulsations of CSF force open the central spinal canal through the patent obex.

- **Oldfield's theory**

  Hindbrain herniation prevents the CSF flow to and from the spinal compartment that otherwise occurs during the cardiac cycle.

  Obstruction at the cervicomedullary junction results in increased CSF pressure in the spinal compartment during systole, and fluid is forced into the spinal cord through its surface.

Causes

- May have genetic basis its association with known genetic disorders such as Achondroplasia and Klippel-Feil syndrome
- Some families suggest an autosomal dominant mode of inheritance
- Acquired CMI is reported to develop after lumboperitoneal or ventriculoperitoneal shunt placement

CSF flow

- During systole: increase cerebral flow

  → craniocaudal displacement.

- During diastole: elastic recoil → reversal

Morphometric study of the posterior cranial fossa

- PFBV/PFCV: significantly higher in patients with Chiari I
- Hence overcrowding of posterior fossa

- *Sabri Aydin et al., Chiari type I malformations in adults: a morphometric analysis of the posterior cranial fossa. Surgical Neurology 64 (2005) 237–241*

Acquired Chiari I malformation

- Acquired tonsillar ectopia (*Secondary Chiari*)
  - Due to reduced volume of cranial cavity (Rickets, Craniosynostosis, Pagets disease)
- Due to increase in volume of intracranial contents (acute hydrocephalus, tumor, cerebral edema, posterior fossa vascular malformations, posterior fossa masses,
- Lumbo peritoneal shunts (CSF hypotension syndrome)

Associated abnormalities

- Skull
  - Shortened clivus
  - Larger than normal foramen magnum
  - Empty sella
  - Clival concavity, platybasia
- Spinal cord
  - *50 to 75 %* have cavitation within the cord (syrinx)
  - Lower cervical and thoracic cord mostly involved
- Brain usually normal except for tonsillar abnormality
- Hydrocephalus associated in 3 to 10 % (in Chiari 1)

Associated abnormalities

- Spine
  - Platybasia, Basilar invagination
  - Klippel Feil deformity, Atlanto axial assimilation
  - Incomplete ossification of C1 ring (5%)
  - Spina bifida at the C1 level
  - Kyphosis
  - Increased cervical lordosis
  - Cervical rib
  - Retroflexed odontoid
  - Scolioisis (Levoscoliosis)

- Meninges
  - Elevated slope of tentorium cerebelli
  - Thickened arachnoid at foramen magnum level
  - Dural thickening at the level of arch of atlas

Platybasia, basilar invagination

- Abnormal flattening of the skull base
- When platybasia is associated with basilar invagination, or the inward and upward migration of the cervical spine through the foramen magnum, signs and symptoms of compression of the brainstem and upper cervical cord can result.

Clinical presentation

- Initially considered an "adult" disease presenting in 2nd and 3rd decade
- Now reported quite frequently in pediatric population - widespread MRI availability
- 3 causes of symptoms :

  a) due to *brainstem compression*

  b) due to syrinx

  c) due to *cerebellar involvement*

SYMPTOMATOLOGY

SPINAL CORD:SYRINX

- SCOLIOSIS
- DISSOCIATED SENSORY LOSS
- DYSETHESIAS
- WASTING OF ARMS/ HANDS
- SPASTICITY OF LEGS
- URINARY INCONTINENCE
- ARM/HAND WEAKNESS

BRAIN STEM

- NECK PAIN/HEADACHE
- DOWN BEAT NYSTAGMUS
- HOARSE VOICE
- PALATAL DYSFUNCTION
- TONGUE ATROPHY/ FASISCULATIONS
- DYSPHAGIA
- RESPIRATORY PROBLEMS
- FACIAL NUMBNESS
- DROP ATTACKS
- DYSARTHRIA

CEREBELLUM

- ATAXIA
- NYSTAGMUS

Neck pain/headache

- Nonradicular occipital/cervical pain frequently associated with dysesthesias of C2 dermatome

- Frequently brought on by coughing/sneezing (Valsalva induced)

Unusual presentations

- Glossopharyngeal neuralgia
- Cardiopulmonary arrest
- Transient quadriparesis after minor trauma

Imaging

- MRI brain + spine
- X rays (dynamic x-rays to rule out AAD)
- CT of CVJ
- Dynamic phase contrast cardiac gated Cine MRI

Imaging findings

- Obliterated cisterna magna
- Hydrocephalus
- Flattened spinal cord
- Tonsillar ectopia
- Peglike cerebellar tonsils
- Normally positioned fourth ventricle

Chiari I malformation - imaging

- MRI

Can show compression of brain stem at foramen magnum (common, and significant finding)

- Hydrocephalus can be present
- Syringomyelia
- Descent of cerebellar tonsils through foramen magnum
  - *Importance probably related to brainstem compression at foramen magnum; nevertheless, this is classic finding associated with type 1 Chiari.*

Chiari I malformation

- Traditionally >5mm descent of tonsils below the plane of foramen magnum
- Tonsils ascend with age, hence the length of descent may not be absolute
  - Abnormal for age
    - > 6mm in first decade
    - > 5 mm in second and third decade
    - > 4 mm in fourth through eighth decade
    - > 3 mm in ninth decade

Mikulis DJ et al. Variance of the position of thecerebellar tonsils with age: preliminary report.Radiology 1992;183(3):725–8

Chiari I malformation

- More important than absolute tonsillar descent may be
  - Peg like shape of tonsils
  - Attenuation of posterior fossa cisternal spaces
  - Posterior fossa volume
  - Suggestive clinical picture
  - Tectal beaking

Chiari I malformation - Imaging

- A review of 22,591 patients in whom MR imaging was performed.
- Tonsillar herniation extending more than 5 mm below the foramen magnum was found in 0.77%.
- Fourteen percent of those patients were asymptomatic (with 7–25 mm of ectopia) and 25% of those had peglike tonsils.
- Radiologically significant tonsillar ectopia may be completely asymptomatic
- There are patients in whom the tonsillar herniation extends less than 5 mm from the foramen magnum, whose clinical behavior is similar to the remainder of the Chiari population, including even those with radiological evidence of syringohydromyelia

Meadows J, Kraut M, Guarnieri M, et al: Asymptomatic Chiari Type I malformations identified on magnetic resonance imaging. J Neurosurg 92:920–926, 2000

Cine MRI

- A dynamic picture of brain that shows the movement of CSF around the cervicomedullary junction
  - Also shows piston like movement of tonsils
  - In patients who do not improve after decompression - useful tool to gauge if the obstruction to CSF flow persists
  - Quantitative CSF flow measurement

Role of CSF flow studies in Chiari I malformation

- Distinguish "obstructed CSF flow"
- Identify patients who will benefit from cranio-occipital decompression
- Limited effectivity of current imaging
  - Comparisons of symptomatic and asymptomatic Chiari I malformation lacking
  - Pressures not measured non-invasively
  - Velocities measurements typically at only one level

Technique for PC MR

- Cardiac gated PC MR images
- Flip angle 20°
- TR/TE 20/5 ms
- Slice thickness 5 mm
- Field of view 180 mm
- Matrix 256 x 256
- Encoding velocity (venc) 10 cm/s
- Location and projection according to reference
- Flow velocity for each time point and each voxel from the phase shift

Characterization of CSF flow in the foramen magnum

Normal subjects

- Relative preponderance of flow anteriorly
- Inhomogeneity of flow
- Presence of small jets
- Unidirectional flow

Patients with Chiari I

- Diminished flow posteriorly
- Greater preponderance of flow anteriorly
- Large jets in anterior subarachnoid space
- Synchronous bidirectional flow.

Interpretation of CSF flow studies

- PC MR images in axial or sagittal projection
- Examine for evidence of flow obstruction
  - Compare flow in anterior and posterior subarachnoid space
  - Evaluate homogeneity of flow
  - Evaluate for synchronous bidirectional flow

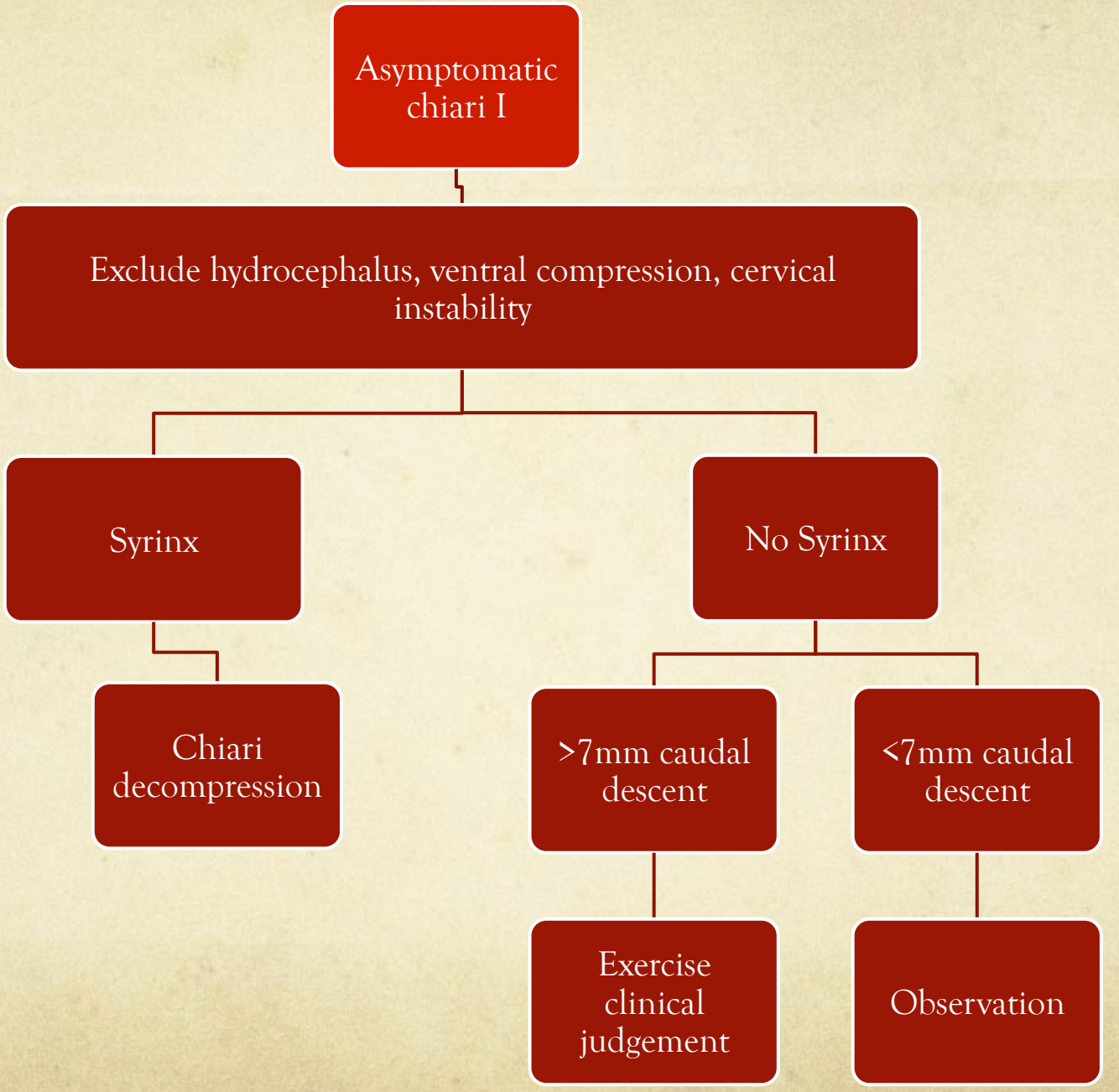
Approach to treatment of chiari I

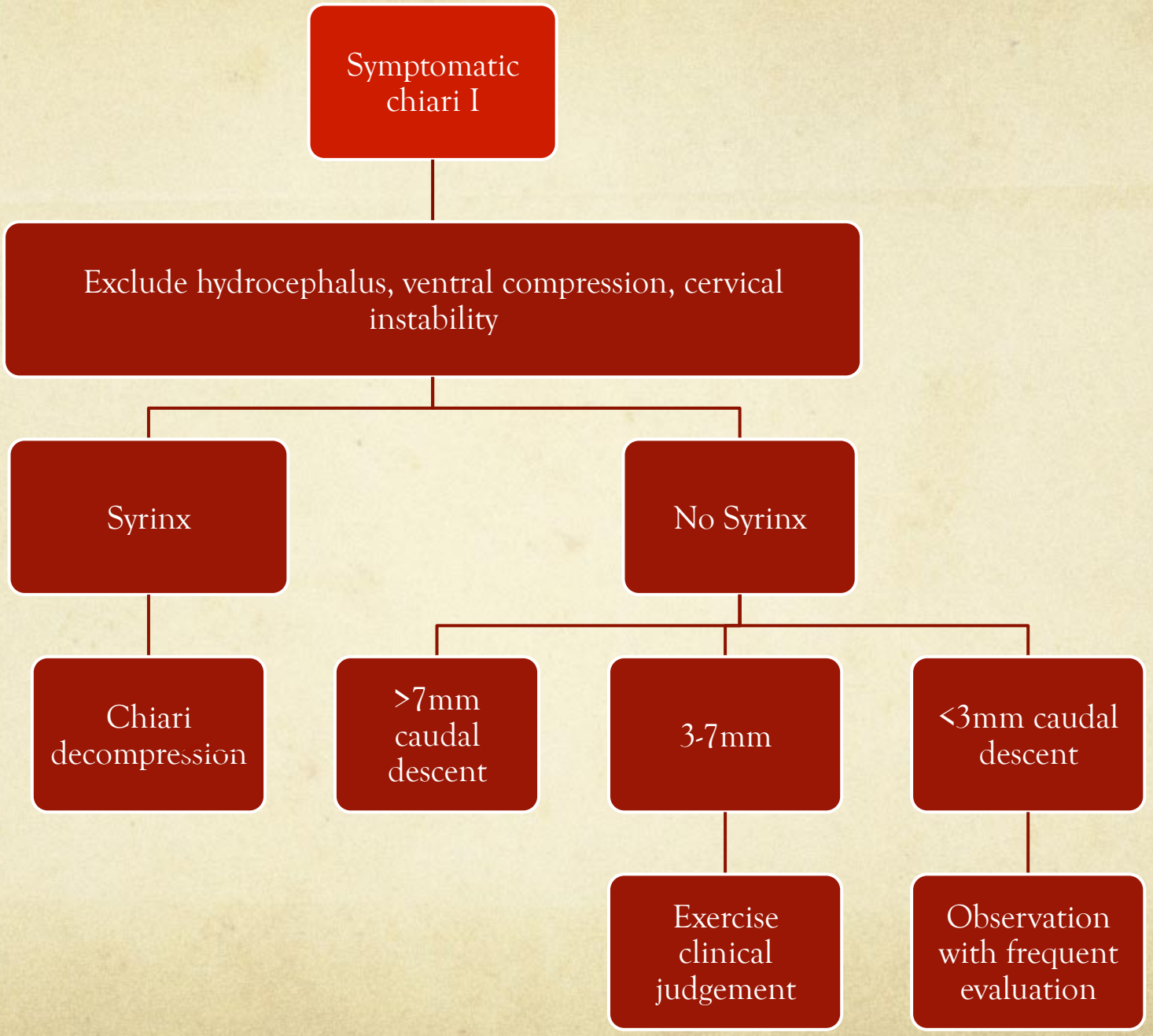
- Initial workup includes examination of supratentorial structures to ensure the absence of a mass lesion or hydrocephalus.
- In a patient in whom a shunt of any kind is in place, it is important to determine that the device is functioning adequately.
- It is important to evaluate spinal stability, especially in the setting of neck pain or spinal dysfunction.
- In patients with significant ventral brainstem compression an anterior decompressive procedure may be warranted prior to treating the malformation

Approach to treatment of chiari I

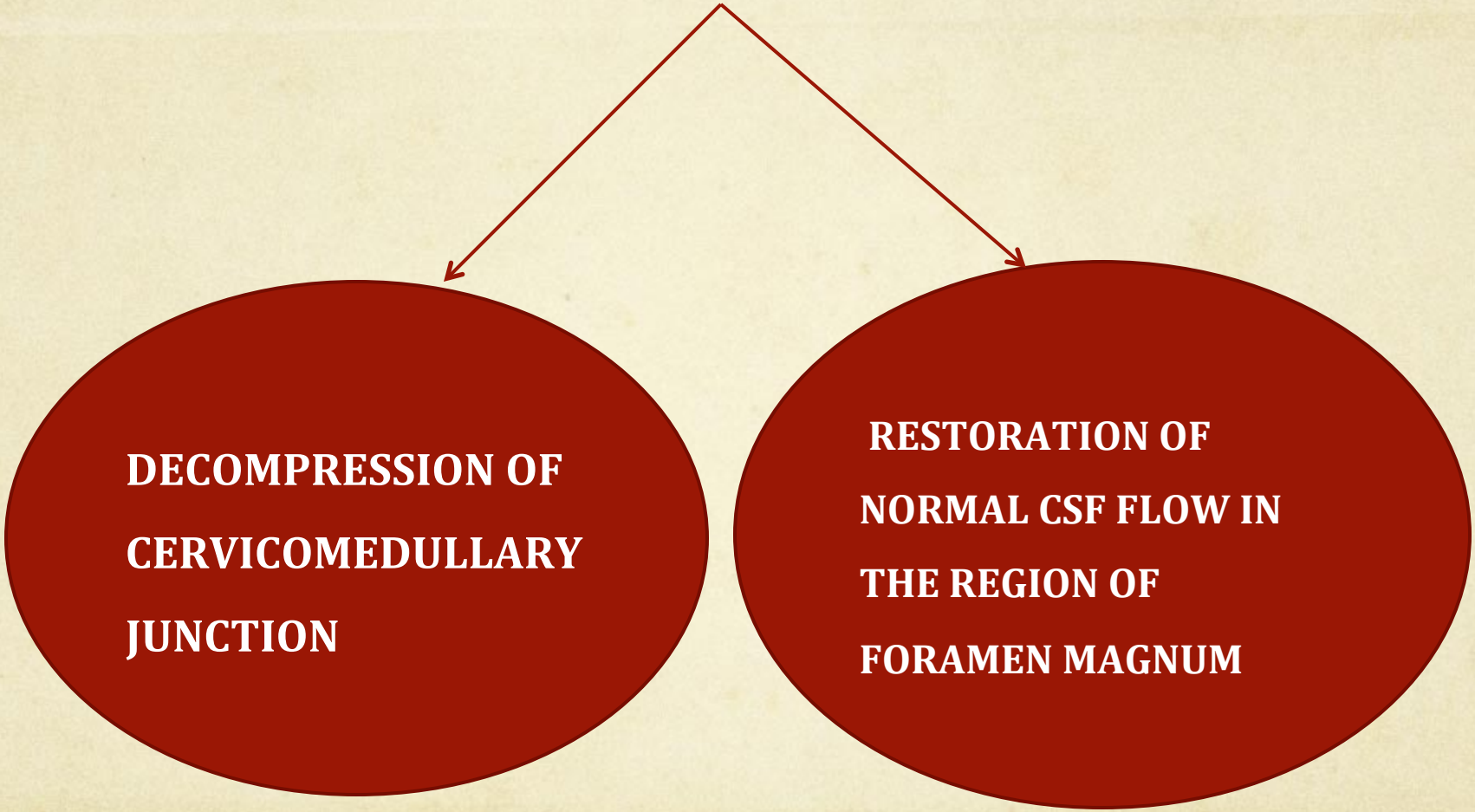
- There is no role for prophylactic treatment in an asymptomatic patient with an incidental CMI.

- All symptomatic patients require surgical treatment.


Asymptomatic chiari I
Exclude hydrocephalus, ventral compression, cervical instability
Syrinx
No Syrinx
Chiari decompression
>7mm caudal descent
<7mm caudal descent
Exercise clinical judgement
Observation


Symptomatic chiari I
Exclude hydrocephalus, ventral compression, cervical instability
Syrinx
No Syrinx
Chiari decompression
>7mm caudal descent
3-7mm
<3mm caudal descent
Exercise clinical judgement
Observation with frequent evaluation

Goals of treatment



Chiari decompression

- Surgical options
  - Suboccipital bone removal
  - Dural opening with or without closure
  - Arachnoid opening and hitching
  - Tonsillar reduction and opening of fourth ventricular outlet
  - Fourth ventricular shunting

- **Suboccipital bone removal + C1 laminectomy**
  - 3 X 3 cm suboccipital craniectomy
  - Division of thick dural band

    Dural band pathology in syringomyelia with Chiari type I malformation. Nakamura et al Neuropathology 2001;20:38-43
  - Dura left intact
  - Larger craniectomies with dural opening result in cerebellar ptosis

- **Dural opening with or without closure**
  - Options
    - Only superficial layer divided
    - Durotomy with intact arachnoid
    - Augmentation duraplasty
    - Williams procedure – dural edges sutured to the muscle
    - At CVJ, division of thick dural band

Intradural procedures

- When significant tonsillar descent with syringomyelia
- Arachnoid bands divided
- Arachnoid pegged to the dural edges
- Arachnoidal adhesions between the 2 cerebellar tonsils and between each tonsil and the medulla divided to mobilize the tonsils supero-laterally and to expose the obex and floor of the 4th ventricle
- Extreme care taken to avoid injury to the tonsillar segments of the PICA
- Augmentation duraplasty

Intradural procedures

- **Tonsil reduction**
  - Subpial coagulation
  - Subpial resection (when tonsils gliotic)

- **Tonsillar hitching**

- **Fourth ventricular shunting**
  - When tonsils encased in dense arachnoid scar
  - Shunt tubing inserted under USG guidance into the fourth ventricle and communicated to cervical subarachnoid space

- **Obex plugging** : not used now

Operative Results

- Patients with pain as primary complaint respond best to surgery; weakness less responsive, but overall ~80% of patients report favorable results.
- Patients presenting with pain (mainly headache and neck pain) & weakness without associated atrophy – best results.
- Brain stem and cerebellar syndromes - good recovery
- Cranial nerve dysfunction – moderate recovery

CHIARI I MALFORMATION

- Sensory recovery poor.
- The presence of central cord syndrome due to a syrinx-indicative of poor recovery.
- Presence of muscle atrophy, ataxia, scoliosis and duration of symptoms >2 yrs all associated with poorer outcome.
- Neuropathic pain carries poor prognosis

Controversies

- Many different approaches have been described in the literature.
- Plugging of the obex, placement of a stent in the fourth ventricle, extensive posterior fossa craniectomy, and multiple cervical laminectomies have been suggested
- Simple decompression is advocated based on histological analysis of the dural band.
- The success of the decompressive procedure can be confirmed using intraoperative ultrasonography
- Further decompression can be achieved by scoring the dura's outer leaf and thus promoting expansion.

- Tokuno, et al., attempted to address the small posterior fossa by conducting an expansive suboccipital cranioplasty.
- Durotomy without duraplasty
- Other authors have emphasized that duraplasty is essential for the prevention of scar formation and recurrent symptoms.
- Debate still exists as to whether, once the dura is opened, arachnoid dissection should be performed.

- Resection of the cerebellar tonsils has been advocated as a way to improve the volume mismatch and to increase communication between the fourth ventricle and the spinal compartment.
- Use of a syringosubarachnoid shunt in the treatment of patients with Chiari I malformation and syringomyelia.

Controversies

- There is no consensus on the procedural steps that lead to a consistently favorable outcome.

  Posterior fossa decompression and reconstruction in adolescents and adults with the Chiari I malformation.

  Oró JJ *et al.* ***Neurol Res. 2011 Apr;33(3):261-71***

- Controversies in the surgical management of chiai 1 malformations: What is the surgical procedure of choice? To open dura or not to open dura?

  Karin M muraszko et al,

  Clinical Neurosurgery, Vol 51,2004

- Controversies in Chiari 1 malformations

  Jamie Baisden

  Surgical neurology International: Spine 2012

Duraplasty Or Not

Against duraplasty

- This preliminary experience indicates that in children 2 years of age or younger, posterior fossa bony decompression without duraplasty can be effective treatment for Chiari malformations

  Posterior fossa decompression without duraplasty in infants and young children for treatment of Chiari malformation and achondroplasia.

  Yundt KD et al Pediatric Neurosurg. 1996 Nov;25(5):221-6

- While PFD was associated with a higher rate of recurrent symptoms requiring repeated decompression, this may be justified by the significantly lower morbidity rate.

  Decompression of Chiari malformation with and without duraplasty: morbidity versus recurrence.

  Ian S. Mutchnick et al, J Neurosurg Pediatrics 5:000–000, 2010

- There was improvement in conduction through the brainstem after bony decompression and division of the atlanto-occipital membrane, ratehr than the dural opening.

  Chiari I malformation: potential role for intraoperative electrophysiologic monitoring. Anderson RC, *et al.* J Clin Neurophysiol. *2003 Feb;20(1):65-72*

Only outer layer of dura was opened

- Foramen magnum decompression by our surgical technique is advantageous because all the procedures are extradural and there are, therefore, fewer postoperative complications.

  Foramen magnum decompression with removal of the outer layer of the dura as treatment for syringomyelia occurring with Chiari I malformation.

  Isu T et al., Neurosurgery. 1993

- This series seems to demonstrate that even a simple extradural surgical approach, with a lower rate of postoperative complications and short stay in hospital, is sufficient to arrest the disease and to improve the symptomatology in a high percentage of cases (97.2%), which is comparable to that achieved with other, more aggressive, procedures.

  Chiari type I anomalies in children and adolescents: minimally invasive management in a series of 53 cases.

  Genitori L et al, Childs Nerv Syst. 2000 Nov;16(10-11):707-18

Transverse incisions of

- These results suggest that foramen magnum decompression combined with transverse microincisions of the outer layer of the dura 1) is an effective and safe treatment option for syringomyelia and Chiari I malformation
- **Transverse microincisions of the outer layer of the dura mater combined with foramen magnum decompression as treatment for syringomyelia with Chiari I malformation.**
- Gambardella G et al. Acta Neurochir (Wien)

Favour duraplasty

- PFD, C1 laminectomy, and duraplasty for the treatment of Chiari I malformation may lead to a more reliable reduction in the volume of concomitant hydromyelia, compared with PFD and C1 laminectomy alone.

  Effects of posterior fossa decompression with and without duraplasty on Chiari malformation-associated hydromyelia.

  Munshi I et al, Neurosurgery. 2000 Jun;46(6):1384-9; discussion 1389-90.

- After intradural exploration, nearly all patients with Chiari I malformation experienced clinical improvement and CSF flow profiles, paralleling those of normal volunteers, were shown.

  Toward a rational treatment of Chiari I malformation and syringomyelia.

  Ellenbogen Rget al. Neurosurg Focus. 2000 Mar 15;8(3):E6.

- Two prospective (n=154) and five retrospective (n=428)

- No significant difference between the groups was found in rates of clinical improvement (four studies) or post. op decrease in syrinx

- Post. fossa decompression with duraplasty group
  - significantly lower rate of re-operation (2.1% vs. 12.6%)
  - higher rate of postop CSF-related complications (18.5% vs. 1.8%)

Durham et al. Comparison of posterior fossa decompression with and without duraplasty for the surgical treatment of Chiari malformation Type I in pediatric patients: a meta-analysis. Journal of Neurosurgery Pediatrics 2008; 2(1): 42-49

REVIEW PAPER

Duraplasty or not? An evidence-based review of the pediatric Chiari I malformation

Childs Nerv Syst (2011) 27:35–40

- There is no level 1 or 2 a study

- Selection of the most appropriate surgical technique for CMI is not yet guided by evidence that is stronger than level IIb and the balance of the evidence is level III. Consistent with this is the fact that there is considerable debate regarding which operative technique is the most appropriate for children with CMI.

Role of intraoperative ultrasound to determine the need of duraplasty

Intra op USG

- Following a standard suboccipital craniectomy and one or more laminectomies, USG used to evaluate the decompression

  Decompression considered adequate

  if CSF space anterior to the brainstem and dorsal to the tonsils seen without evidence of abnormal tonsillar piston activity

Against usg

- In the setting of moderate-to-severe tonsillar CM-I, intraoperative ultrasonography demonstrating decompression of the subarachnoid spaces ventral and dorsal to the tonsils may not effectively select patients in whom bone decompression alone is sufficient. Duraplasty may be warranted in cases of tonsillar herniation that extends below the C-1 lamina regardless of intraoperative ultrasonography findings.

Intraoperative ultrasonography as a guide to patient selection for duraplasty after suboccipital decompression in children with chiari malformation type i

Matthew j. Mcgirt et al. J Neurosurg Pediatrics 2:52–57, 2008

For USG

- Color Doppler ultrasonography imaging is an important technological advance that permits the neurosurgeon to tailor the steps of Chiari surgery according to patient-specific variables. The success of this technique depends on the mastery of a new and sophisticated monitoring modality.

Tailored operative technique for Chiari type I malformation using intraoperative color Doppler ultrasonography.

Milhorat TH *et al.* *Neurosurgery. 2003 Oct;53(4):899-905; discussion 905-6*

Intra op USG

- In a series of 130 patients, *Yeh et al.*

Intraoperative ultrasonography used to determine the extent of surgery necessary during posterior fossa decompression in children with Chiari malformation type I. J Neurosurg 2006;105(1Suppl):26–32

91 underwent a bony decompression and duraplasty

39 underwent bony decompression alone

Surgical failures

4 in the bone decompression group and

2 in the duraplasty group

Complications

0 in bone decompression group

12 patients in the duraplasty group

Arachnoid opening or not

Arachnoid opening or not

- Cranio-cervical decompression associated with an enlargement duroplasty with preservation of the arachnoid membrane, achieved the best results with minimal complications and side-effects.

  Cranio-cervical decompression for Chiari type I-malformation, adding extreme lateral foramen magnum opening and expansile duroplasty with arachnoid preservation. Technique and long-term functional results in 44 consecutive adult cases - comparison with literature data.

  Sindou M et al. Acta Neurochir (Wien). 2002 Oct;144(10):1005-19.

- Arachnoid-preserving posterior fossa decompression is a safe and effective treatment for patients with CMI with associated syringomyelia.

  Surgical results of arachnoid-preserving posterior fossa decompression for Chiari I malformation with associated syringomyelia

  Hyun Seok Lee et al. Journal of Clinical Neuroscience 19 (2012) 557–560

Clinical Study

Surgical results of arachnoid-preserving posterior fossa decompression for Chiari I malformation with associated syringomyelia

Hyun Seok Lee, Sun-Ho Lee *, Eun Sang Kim, Jong-Soo Kim, Jung-Il Lee, Hyung Jin Shin, Whan Eoh

Department of Neurosurgery, Samsung Medical Center, Sungkyunkwan University, School of Medicine, 50 Ilwon-dong, Gangnam-gu, Seoul 135-710, South Korea

Journal of Clinical Neuroscience 19 (2012) 557–560

- Retrospective review
- 25 patients
- Duraplasty in 20 patients and outer layer of dura opened in 5 patients
- Syrinx regressed in 17 (68%) patients
- Stabilized in another 7 (28%)
- Progressed in only one patient
- This study shows that arachnoid-preserving posterior fossa decompression is a safe and effective treatment for patients with CMI with associated syringomyelia.

Size of craniotomy

Size of craniotomy

An efficacy analysis of posterior fossa decompression techniques in the treatment of Chiari malformation with associated syringomyelia

Journal of Clinical Neuroscience 18 (2011) 1346–1349

- Decompression size:

  Large craniotomy group - 5 cm 6 cm

  Small craniotomy group - 3 cm 4 cm

- Of these 132 patients, 69 received extended PFD (large craniotomy group), and the other 63 patients received only local PFD (small craniotomy group).
- At the short-term postoperative evaluation (1–4 weeks) the extended PFD appeared to be more effective than the local PFD ($p < 0.05$).
- However, there was no significant difference in long term analysis (6 months–11 years) ($p > 0.05$).
- Patients who had undergone local PFD exhibited more obvious radiological improvement of SM ($p < 0.05$) and fewer postoperative complications compared to patients undergoing extended PFD ($p < 0.05$).
- Therefore, local PFD is preferable for the surgical treatment of CM with associated SM.

Graft material / Dural substitutes

- Autologous pericranial grafts
- Cadaveric substitutes (Dura, pericardium and fascia lata)
- Synthetic dural substitutes (silastic, gortex, polyglactin mesh)
- Combined with collagen matrix (microfibrillar collagen)

Dural substitutes

Cadaveric dura

- Diseases like CJD
- Failure to restore normal CSF dynamics

Biological and synthetic grafts

- Arachnoid adhesions leading to recurrence of symptoms

- Duraplasty utilizing **ePTFE graft** was associated with improved maintenance of hindbrain space, accelerated syringomyelia improvement, and a trend toward decreased treatment failure versus pericranial autograft.

  Suboccipital decompression for Chiari 1 malformation: outcome comparison of duraplasty with expanded PTFE dural substitute versus pericranial autograft

  Child Nervous System (2009) 25:183-190

- The **synthetic dural substitutes DuraGen and AlloDerm** provide a suitable alternative duraplasty with comparable complication rates. DuraGen requires a significantly shorter operative time than AlloDerm.

  Experience with acellular human dura and bovine collagen matrix for duraplasty after posterior fossa decompression for Chiari malformations Shabbar f. Danish et al. J Neurosurg (1 Suppl Pediatrics) 104:16–20, 2006

- Duraplasty using a collagen matrix is safe and effective in the posterior fossa, and is easy to use and time efficient.

  Collagen matrix duraplasty for posterior fossa surgery: evaluation of surgical technique in 52 adult patients

  Pradeep K. Narotam et al. J Neurosurg 111:380–386, 2009

- At median 8 months postoperatively,all (100%) patients with ePTFE graft maintained physiological CSF flow/decompressed hindbrain CSF space on cine-MRI versus 32 (79%) patients receiving pericranial autograft ($p<0.05$).
- Radiographic syrinx improvement occurred in 80% of patients with ePTFE graft and 52% of patients with pericranial autograft (median time to improvement: 5 vs 12 months, respectively, $p<0.05$).
- At median 16 months postoperatively, four (10%) patients with pericranial autograft required revision decompression versus 0 (0%) patients with ePTFE graft ($p=0.090$).
- Duraplasty utilizing ePTFE graft was associated with improved maintenance of hindbrain space, accelerated syringomyelia improvement, and a trend toward decreased treatment failure versus pericranial autograft.

Experience with acellular human dura and bovine collagen matrix for duraplasty after posterior fossa decompression for Chiari malformations

- The authors evaluated 56 patients who underwent duraplasty with a synthetic collagen matrix (Dura Gen) and 45 patients in whom the dural closure involved acellular human dermis (AlloDerm).

- *Conclusions.* The synthetic dural substitutes DuraGen and AlloDerm provide a suitable alternative duraplasty with comparable complication rates. DuraGen requires a significantly shorter operative time than AlloDerm.

Optimal suture material

- Optimal suture material for sewing dural graft is 7 O Gore tex suture (PTFE monofilament)
- Needle is smaller than the suture itself

Dural sealants

- Cellulose strips
- Collagen sheets
- Fibrin glue
- Fibrin sealants

- Tisseel (fibrin glue) has seen wide adoption in "off-label" use. DuraSeal, which is FDA approved, was associated with two instances of paralysis. Alternatively, BioGlue was described as neurotoxic even by the manufacturer

  Dural repair with four spinal sealants: focused review of the manufacturers' inserts and the current literature.

  Epstein NE, Spine J. 2010 Dec;10(12):1065-8.

- The hydrogel sealant applied to collagen-based dural grafts significantly reduced CSF leakage and functioned as an adhesion barrier. Such technology could be an important tool for cranial surgery.

  Application of a hydrogel sealant improves watertight closures of duraplasty onlay grafts in a canine craniotomy model

  Mark c. Preul et al. J Neurosurg 107:642–650, 2007

The Spine Journal 10 (2010) 1065–1068

Clinical Study

Dural repair with four spinal sealants: focused review of the manufacturers' inserts and the current literature

Nancy E. Epstein, MD[a,b,*]

- BACKGROUND CONTEXT: Deliberate or traumatic dural fistulas are typically augmented by a "sealant" or "fibrin glue" to enhance the strength of dural closure

- CONCLUSION: Despite the lack of FDA approval, Tisseel (fibrin glue) has seen wide adoption in "off-label" use. DuraSeal, which is FDA approved, was associated with two instances of paralysis. Alternatively, BioGlue was described as neurotoxic even by the manufacturer

Chiari I Malformation Associated with Syringomyelia and Scoliosis

A Twenty-Year Review of Surgical and Nonsurgical Treatment in a Pediatric Population

SPINE Volume 27, Number 13, pp 1451–1455

Twenty-five patients were identified, ranging in age from 19 months to 16.5 years. Nineteen patients (76%) had associated scoliosis.

Eleven of 19 patients with scoliosis (58%) underwent fusion.

Eight of 19 (42%) patients have not undergone fusion: 3 have experienced progress, 1 remains in a stable condition, and 4 have experienced improvement of curvature since undergoing decompression.

Early decompression of Chiari I malformation with syringomyelia and scoliosis resulted in improvement or stabilization of the spinal deformity in 5 cases.

Each of these patients underwent decompression before 8 years of age and before the curve was severe.

Outcomes of Chiari I-associated scoliosis after intervention: a meta-analysis of the pediatric literature

Childs Nerv Syst (2012) 28:1213–1219

- One hundred and twenty patients were identified in 12 studies, of them, 37 % were male.
- The mean age at the time of surgery was 9.7±4.1 years.
- After surgical intervention, curve magnitude improved in 37 % of patients (n042); there was no change in 18 % (n020), and curves progressed in 45 % (n0 51).
- Age (p00.0097) and presence of surgical intervention (foramen magnum decompression [p 0.0099] and syrinx shunting/drainage [p00.0039]) were statistically associated with improvement of the scoliotic curve. **Surgical decompression of the foramen magnum had the greatest impact on the scoliotic curves.**
- Data accrued from our analysis suggest that curve magnitude will improve after surgical treatment of the Chiari malformation in one third of patients, and curve progression will stabilize or improve in one half.

Chiari malformation associated with craniosynostosis

Neurosurg Focus 31 (3):E2, 2011

- The authors reviewed 383 consecutive patients treated for craniosynostosis at a single institution
- A total of 29 patients (8%) with both CM and craniosynostosis were identified.
- Children with single-suture lambdoid synostosis ($p < 0.001$) or multisuture craniosynostosis ($p < 0.001$) were much more likely to have associated CM
- 16 had craniosynostosis repair as well as CM decompression, and 13 patients did not undergo CM decompression.
- Of the 7 patients in whom craniosynostosis repair alone was performed, 5 had decreased tonsillar ectopia postoperatively and 5 had improved CSF flow studies postoperatively.
- Both patients with a spinal syrinx had imaging-documented syrinx regression after craniosynostosis repair.
- In 5 cases, de novo CM development was documented following craniosynostosis repair at a mean of 3.5 years after surgery.

Thank you