Chiari malformations

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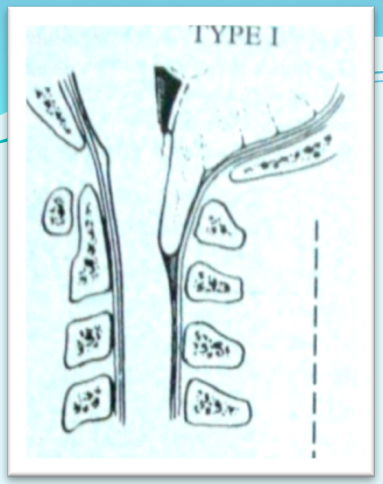
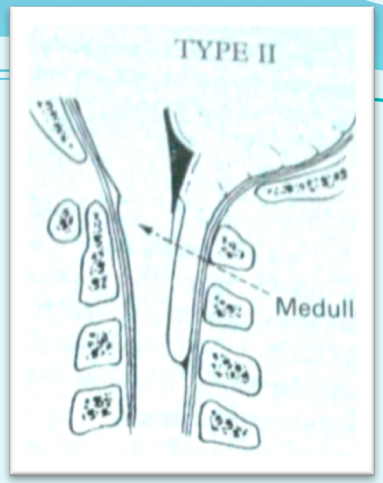
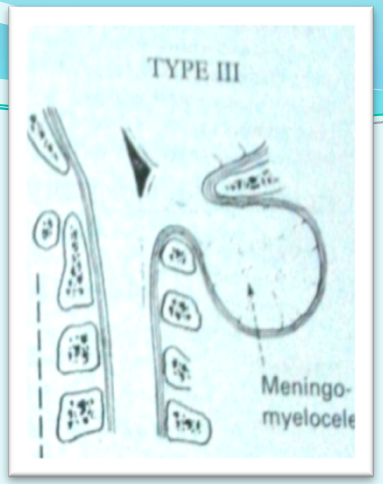
Introduction

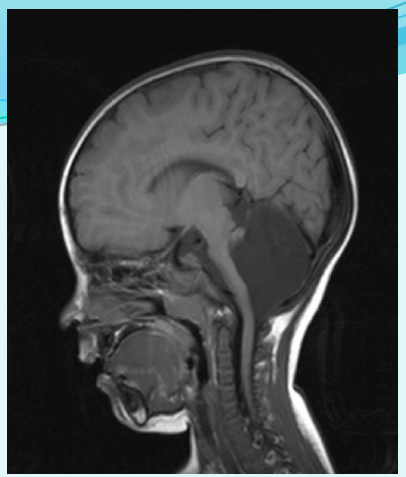
- **Hans Chiari** was Professor of pathology at Prague, Czechoslovakia
- 1891-coined Chiari I,II and III
- 1895-Chiari IV
- *Other professors:John Cleland (Glasgow) ,Julius Arnold, Nicholas Tulp*
- Adult cases: Mc-Connell and Parker(1938)
- Used the term "*tonsils*" to describe the prolapsed cerebellar 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 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

Other chiari malformations??

5) Chiari 0

- no hind brain herniation
- syringo hydromyelia
- marked improvement with posterior decompression

6) Chiari 1.5

- somewhere between I and II
- tonsillar herniation (I) and elongated caudally displaced brainstem and 4th ventricle(II)

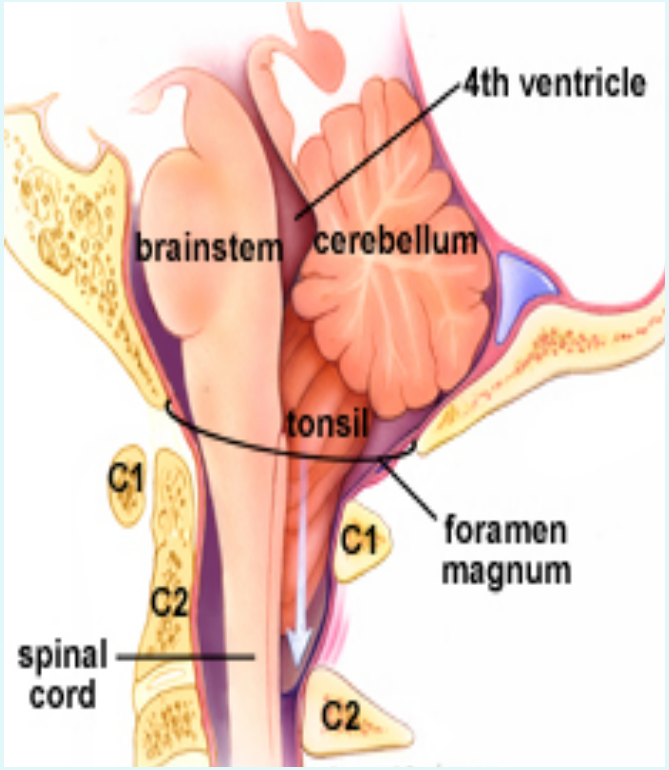
BASIC EMBRYOLOGY

Formation of brain +spinal cord : dorsal induction

2 stages of dorsal induction

- **Primary neurulation:brain +upper spine**
  - •*Chiari*
  - •Cephalocele
  - •MMC
- **Secondary neurulation: distal spine**
  - •Lipomeningocele
  - •Neuroenteric cyst
  - •Dermal sinus
  - •Caudal regression syndrome

Chiari I malformation

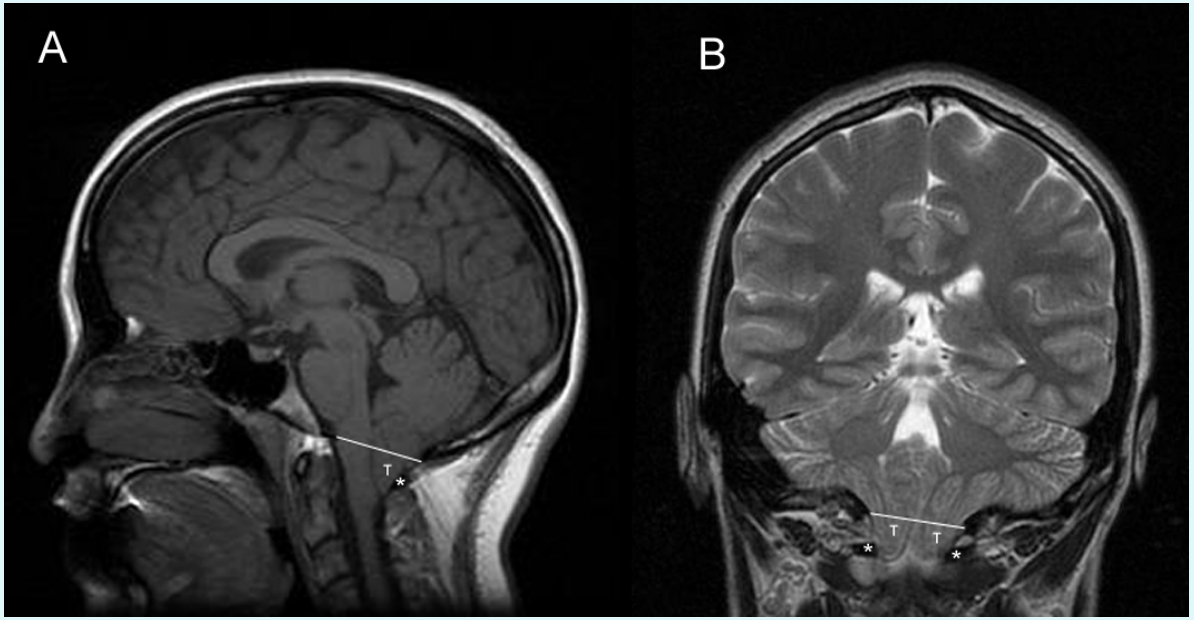


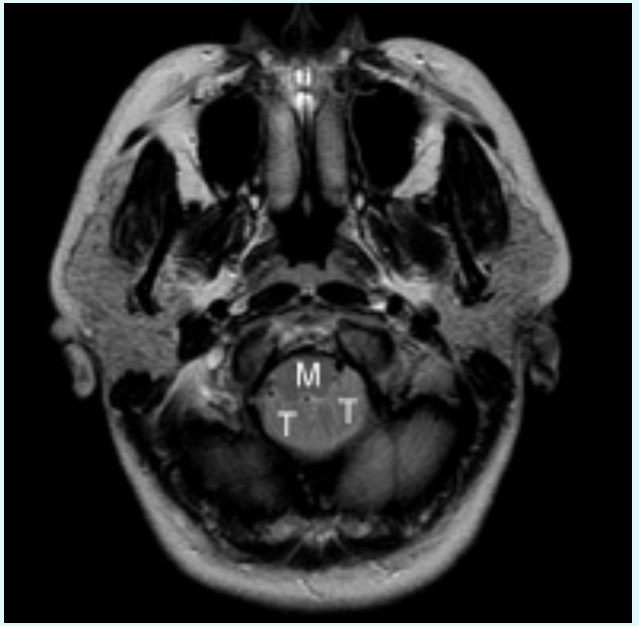
- Traditionally >5mm descent of tonsils below the plane of foramen magnum
- Tonsils ascend with age, hence the length of descent may not be absolute
  - Abnormal for age
    - > 6mm in first decade
    - > 5 mm in second and third decade
    - > 4 mm in fourth through eighth decade
    - > 3 mm in ninth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

  Mikulis DJ et al. Variance of the position of the cerebellar 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
  - Post. fossa volume
  - Suggestive clinical picture





HOW COMMON IS IT ???

1/1280 in a series analyzed at Johns Hopkins

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

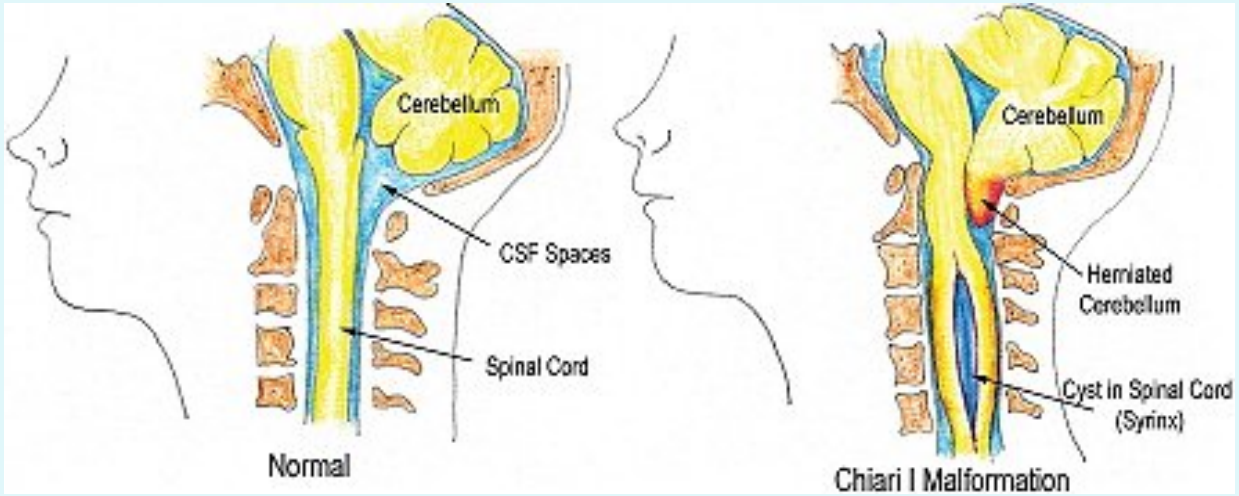
22,000 brain MRs reviewed.?? Referral bias

Familial clustering has been established

Milhorat et al.Chiari I redefined: clinical and radiological findings for 364 symptomatic patients. Neurosurgery 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 genetic counseling 2003;12:297-311



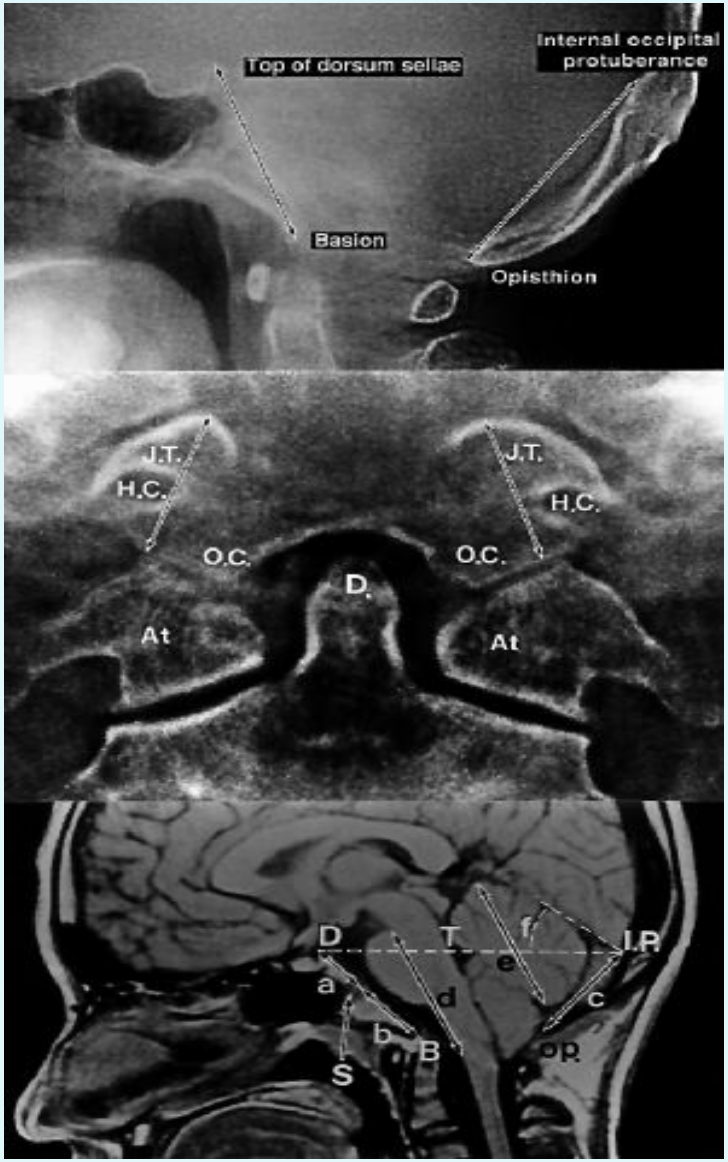
Chiari I malformation

- **Subnormal posterior fossa volume but no reduction in infratentorial brain volume precipitates hindbrain herniation through FM**

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.

Morphometric study of the posterior cranial fossa

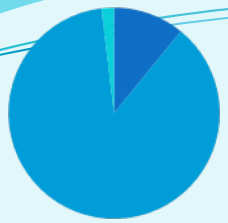
- PFBV/PFCV: significantly higher in patients with Chiari I
- Hence overcrowding of posterior fossa



Nishikawa et al. Pathogenesis of Chiari malformation: a morphometric study of the posterior cranial fossa.
Neurosurgery Focus 1996

Associated abnormalities

- Skull
  - Shortened clivus, clival concavity
  - Larger than normal foramen magnum
  - Empty sella
- Spinal cord
  - 50 to 75 % have cavitation within the cord (syrinx)
- Brain usually normal except for tonsillar abnormality, HCP= 3 to 10 %
- Spine
  - Klippel-feil deformity , BI
  - Scoliosis(levoscoliosis)
- Meninges
  - Elevated slope of tentorium cerebelli
  - Thickened arachnoid at foramen magnum level
  - Dural thickening at the level of arch of atlas



SYMPTOMATOLOGY

SPINAL CORD:SYRINX

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

BRAIN STEM

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

CEREBELLUM

- ATAXIA
- NYSTAGMUS

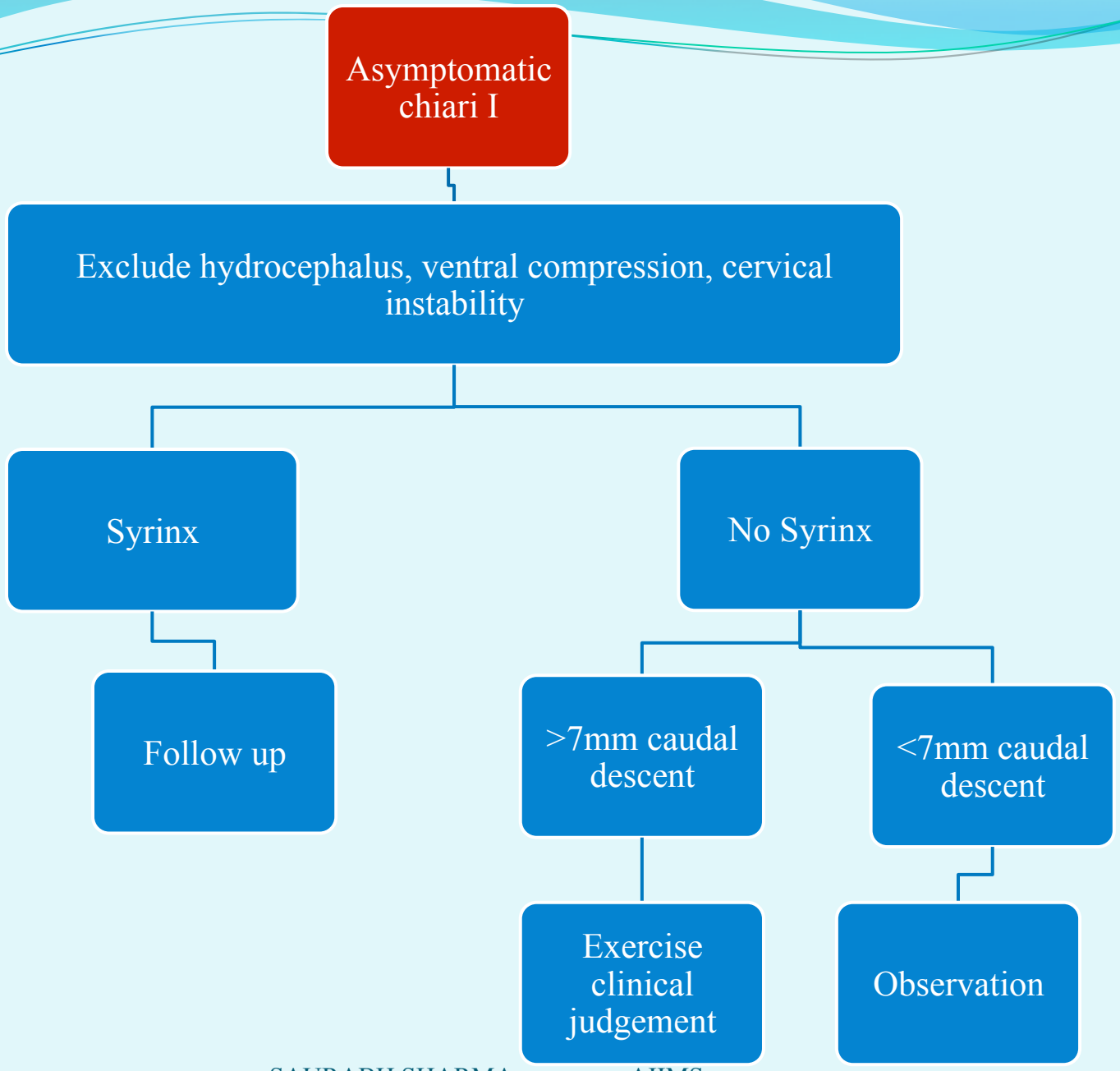
Imaging

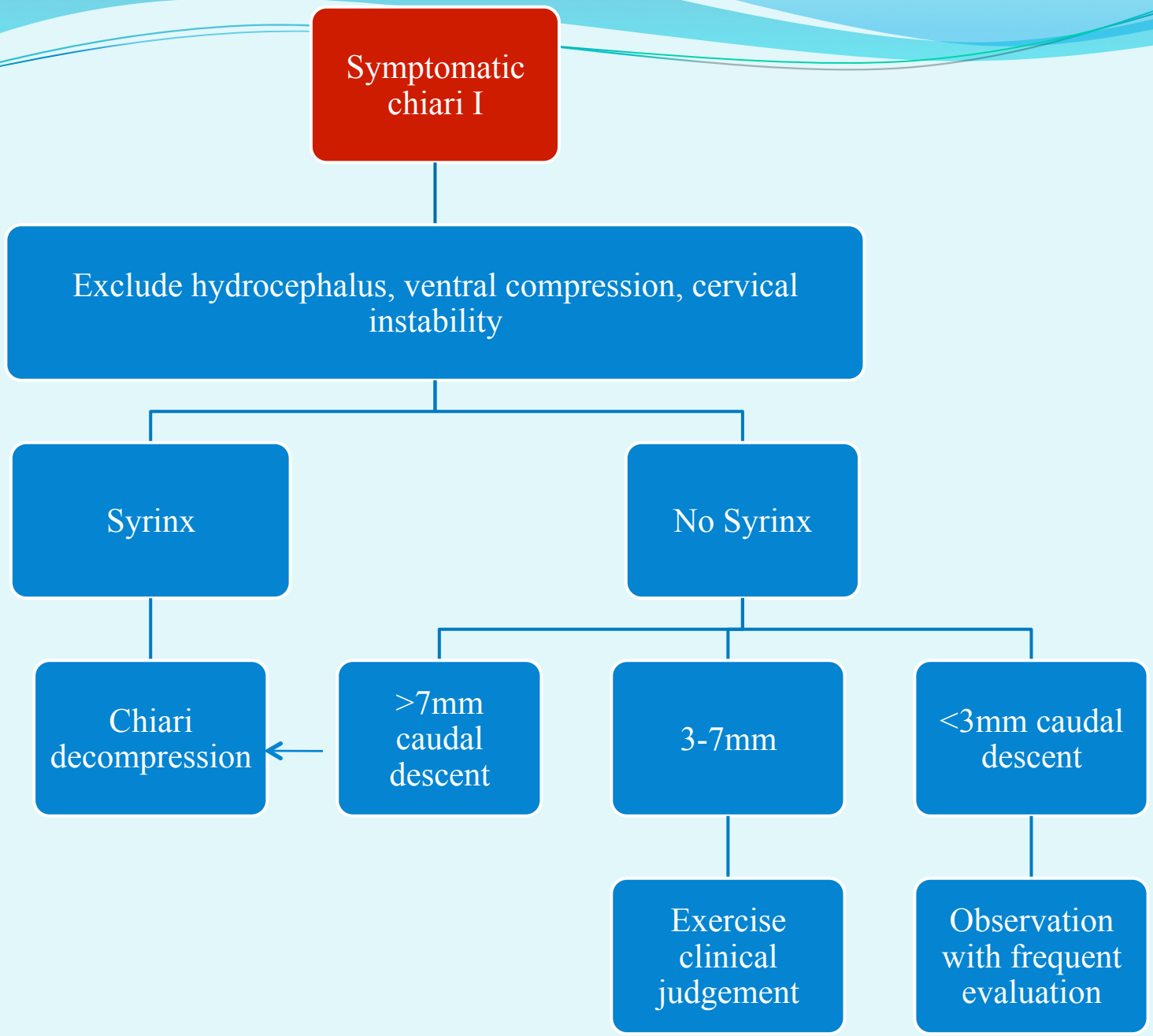
- X rays (dynamic x-rays to rule out AAD)
- CT scan myelography
- CT head for hydrocephalus + CT of CVJ
- MRI (plain) brain + spine
- Dynamic phase contrast cardiac gated Cine MRI
- Intra op USG

Cine MRI

Cine MRI pre and post surgery. Return of CSF flow behind the cerebellar tonsils post decompression can be seen (white arrows)

- 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


Asymptomatic chiari I
Exclude hydrocephalus, ventral compression, cervical instability
Syrinx
No Syrinx
Follow up
>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

DECOMPRESSION OF CERVICOMEDULLARY JUNCTION

RESTORATION OF NORMAL CSF FLOW IN THE REGION OF FORAMEN MAGNUM

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(Y-shaped) with intact arachnoid
    - Augmentation duraplasty
    - Williams procedure – dural edges sutured to the muscle
    - At CVJ, division of thick dural band

Intradural procedures

- **Tonsil reduction :** *unrestricted outflow of CSF from 4th ventricle*
  - Subpial coagulation (Bertrand)
  - Subpial resection when tonsils gliotic
- **Tonsillar hitching**

Intradural procedures

- **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

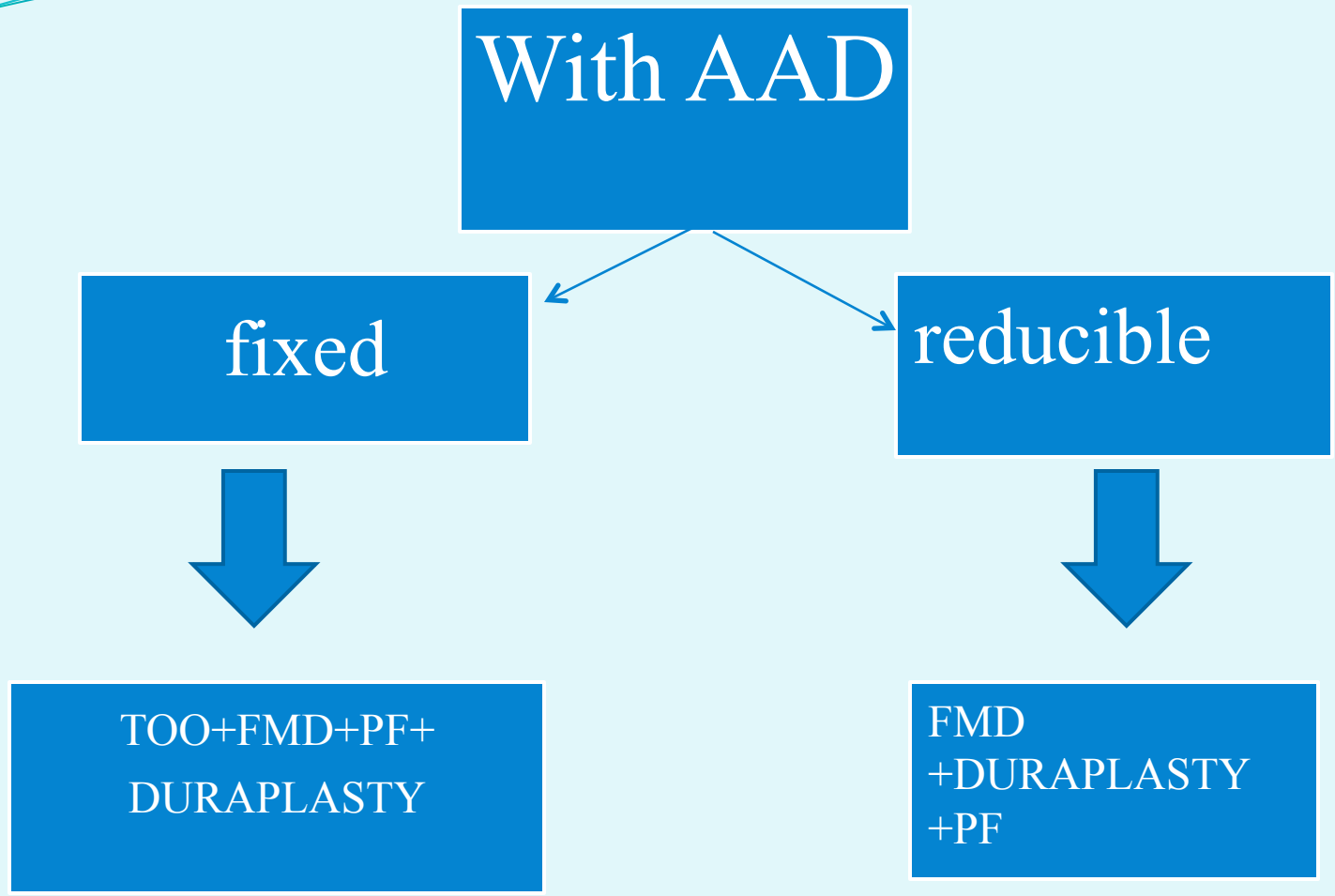
Complications

- Aseptic meningitis (most common)
- Wound dehiscence, pseudomeningocele
- CSF leak
- Muscular pain

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

- Two prospective (n=154) and five retrospective (n=428)
- **Posterior fossa decompression without duroplasty-higher rates of re-operation**
- **post. fossa decompression with duraplasty group**
  - **significantly lower rate of re-op.**
  - **higher rate of postop CSF-related complications**
- No significant difference between the groups was found in rates of clinical improvement (four studies) or post. op decrease in syrinx

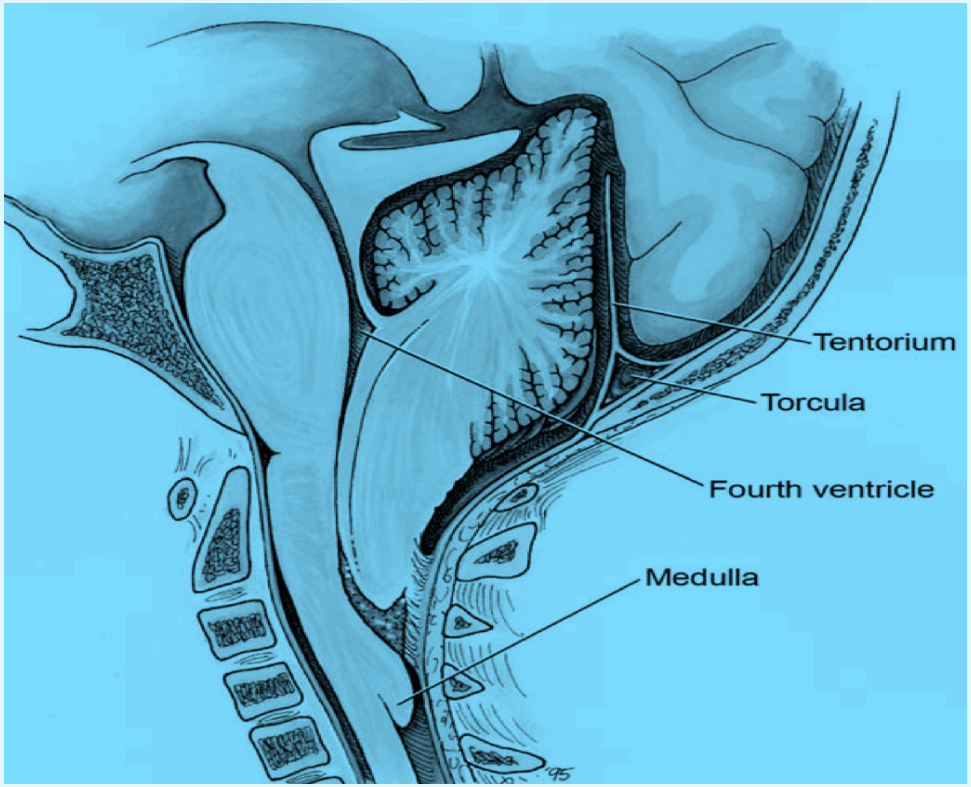
Chiari I with bony CVJ anomaly


With AAD
fixed
reducible
TOO+FMD+PF+
DURAPLASTY
FMD
+DURAPLASTY
+PF

Chiari I with BI : pre and post op

Chiari II malformation

- Defining features
  - Caudal descent of vermis, fourth ventricle and brainstem
  - *Almost* invariably associated with myelomeningocele and hydrocephalus
- Mortality rate is 15% in the first year of life
- Long-term mortality rates as high as 50%



Theories

1) Chiari's theory : *Hydrocephalus leading to secondary chiari*

10-20 % may not have hydrocephalus

Associated anomalies not explained

Chairi II features precede hydrocephalus

2) Cleland's theory : *Primary dysgenesis of the hindbrain*

Fails to explain supratentorial anomalies

3) Induced small posterior fossa(Osaka):*Due to CSF leaking out from the open spinal cord defect*

4) Penfield's traction theory

Traction by tethering of cord at the site of MMC pulls the post. fossa contents

Fails to explain associated cranial deformities/upward cerebellar movement through the tent.

Theories

5) Marin-Padilla : *small posterior fossa theory* related to a mesodermal deficiency

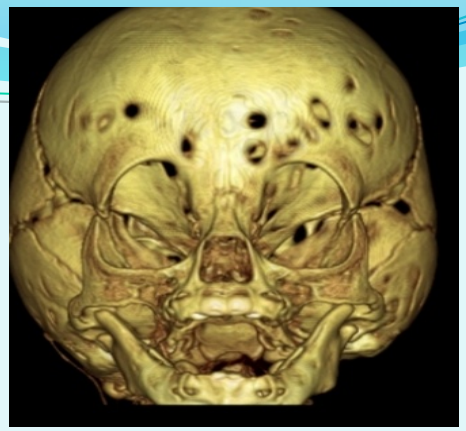
6) Unified theory of Mclone and Knepper 1

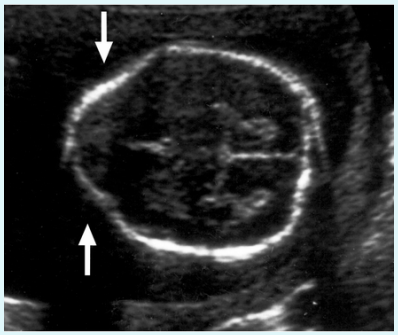
- **Currently most accepted**
- Both the open neural tube defect and incomplete occlusion of central canal responsible
- Temporary occlusion of the neural tube (day 23-32) mandatory for upstream ventricular distension
- Post. fossa not fully developed due to inadequate ventricular distension
- Rapid growth of hindbrain later leads to herniation

1 McLone DG, Knepper PA: The cause of Chiari II malformation: a unified theor Pediatr Neurosci 1989; 15:1-- 12

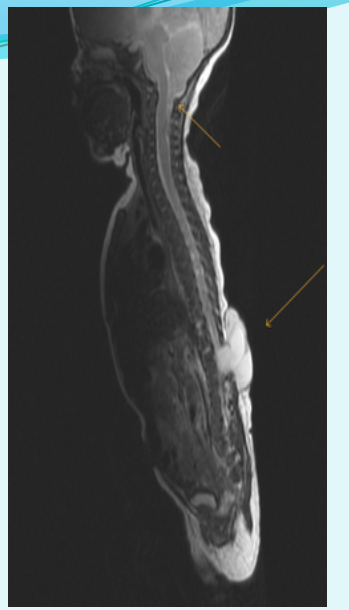
Associated findings

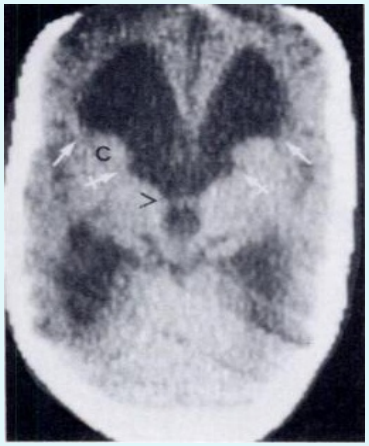
- Skull
  - Luckenschadel/ craniolacunia (disappears by 6m)
  - Frontal bone scalloping "*lemon sign*"(usg)
  - Scalloping of petrous bone
  - Low inion, small post. fossa
  - Enlarged foramen magnum
  - Clival concavity

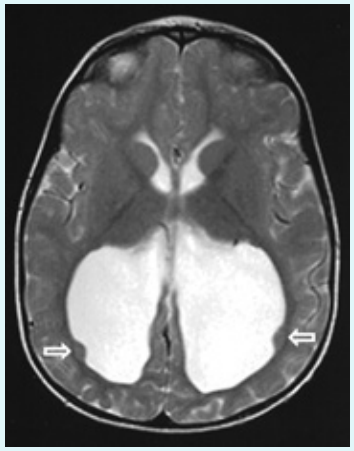


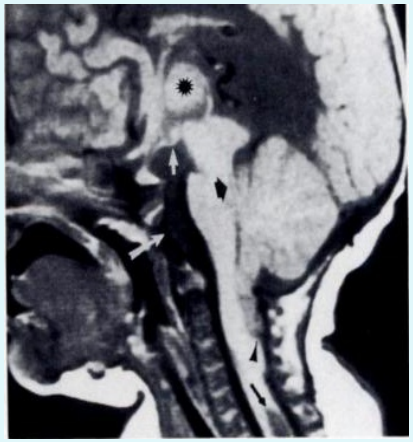


- Spine
  - Syrinx,Klippel-Feil deformity,mmc
  - Medulla
    - Medulla may be elongated and flattened
    - Protuberance just caudal to gracile and cuneate tubercles(70%)usually between **C2-C4, cervicomedullary kink/spur**
- Ventricles and cisterns
  - Hydrocephalus seen in 90%
  - Pointed frontal horns
  - "*colpocephaly*"-enlarged occipital horn and atrium
  - 4th ventricle typically small, flat and elongated, slit like



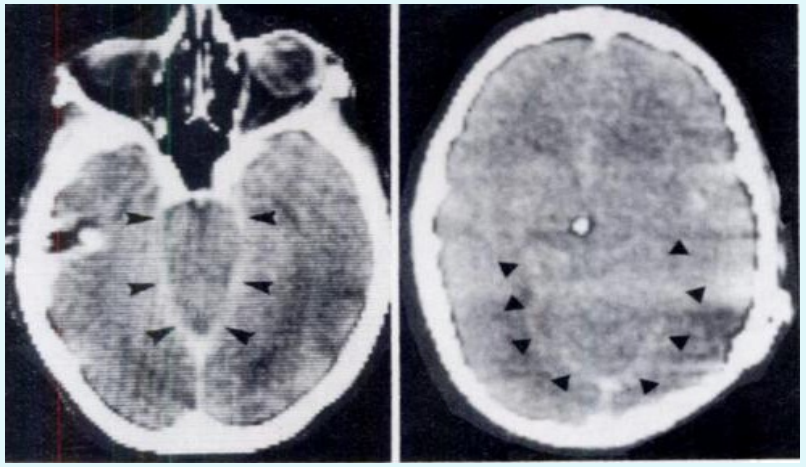


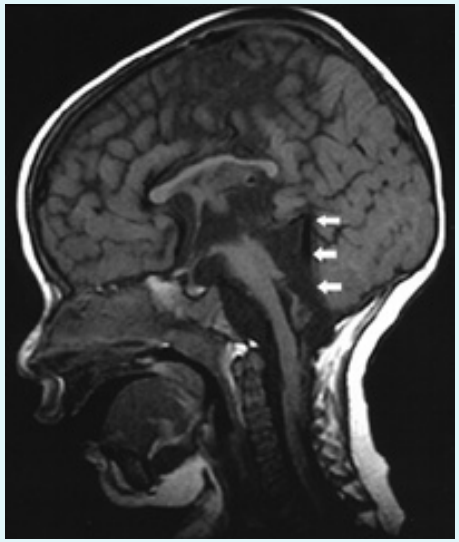




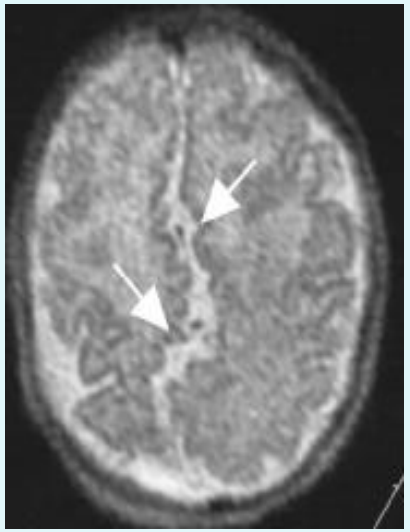
- Meninges
  - Tentorium cerebelli usually widened heart shaped
  - Low lying hypoplastic tent
  - Falx cerebri fenestrated/ hypoplastic

Normal hiatus Hiatus in chiari II



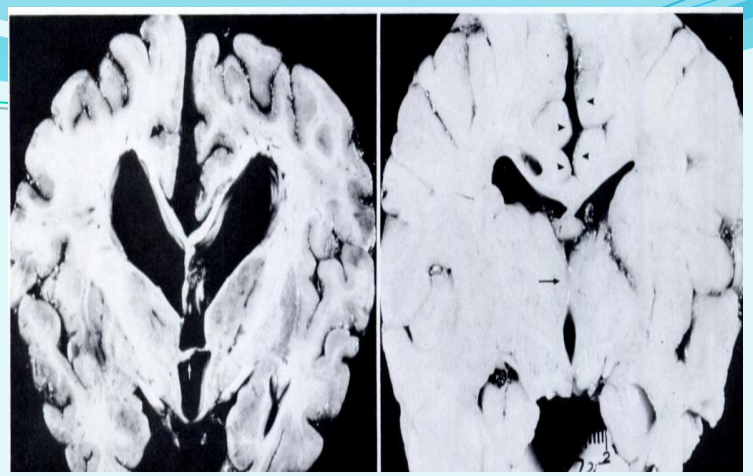


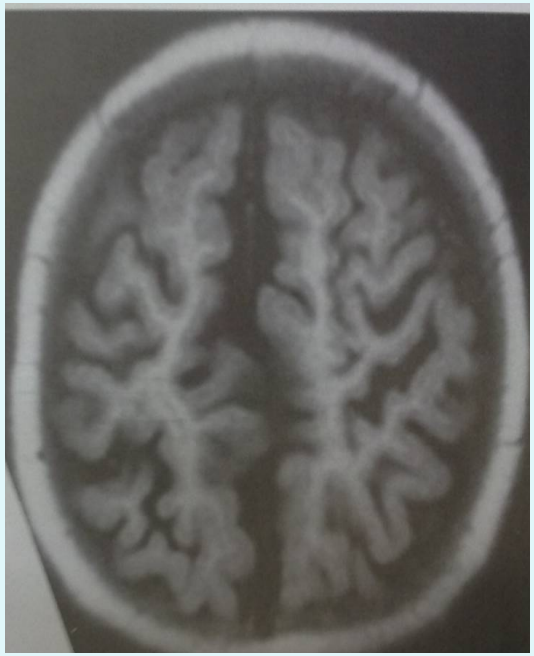
Low lying tent



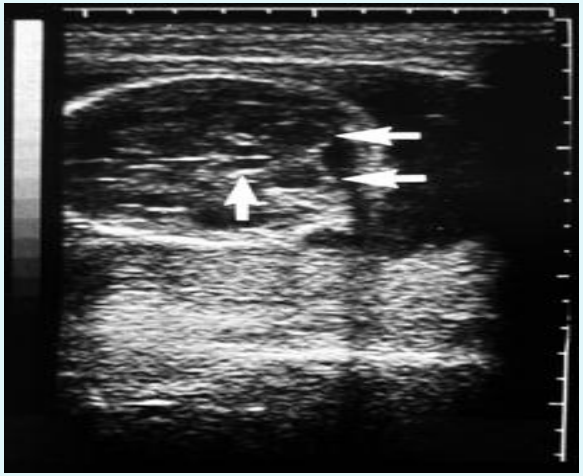
fenestrated falx

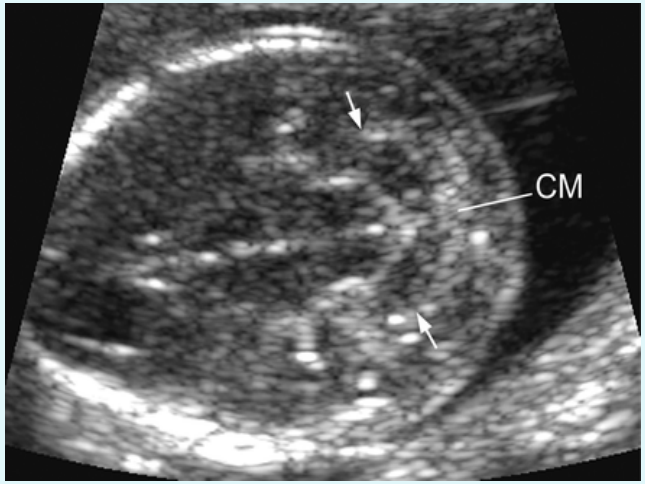
- Enlarged Massa intermedia
- Shark tooth 3rd ventricle
- Tectal beaking due to fusion of the colliculi
- vertically oriented straight sinus
- Aqueduct may be stenotic, stretched, posteriorly kinked or forked
- "*chinese lettering* "- interdigitation of occipital /parietal lobes





- Cerebellum grossly smaller and may tower above tentorium
- Cerebellum may be displaced laterally spreading around the brainstem –"*banana sign*"
- Pons elongated and flattened





Signs and symptoms

Respiratory distress and impaired swallowing (71%)

Inspiratory stridor (59%)

Episodic apnea (29%)

Weak or absent cry (18%)

Aspiration (12%)

Nystagmus

Pain in the upper and lower extremities

Weakness or spasticity of the upper and lower extremities (53%)

Depressed or absent gag reflex

Fixed retrocollis

Palsy of the seventh cranial nerve

Scoliosis

Worsening of bladder and/or bowel function

Respiratory problems-most common cause of death

- **PEAC(prolonged expiratory apnea with cyanosis)**
  More than half of patients with CM II and apnea during infancy can be expected to die of PEAC
- PEAC leads to opisthotonus, decreased heart rate and eventually death

- **Symptom type and age are closely correlated.**

a) Newborns generally asymptomatic

b) Infants ~ brainstem dysfunction(dysphagia, dysarthria)

c) Infants to adulthood-cerebellar/spinal cord dysfunction

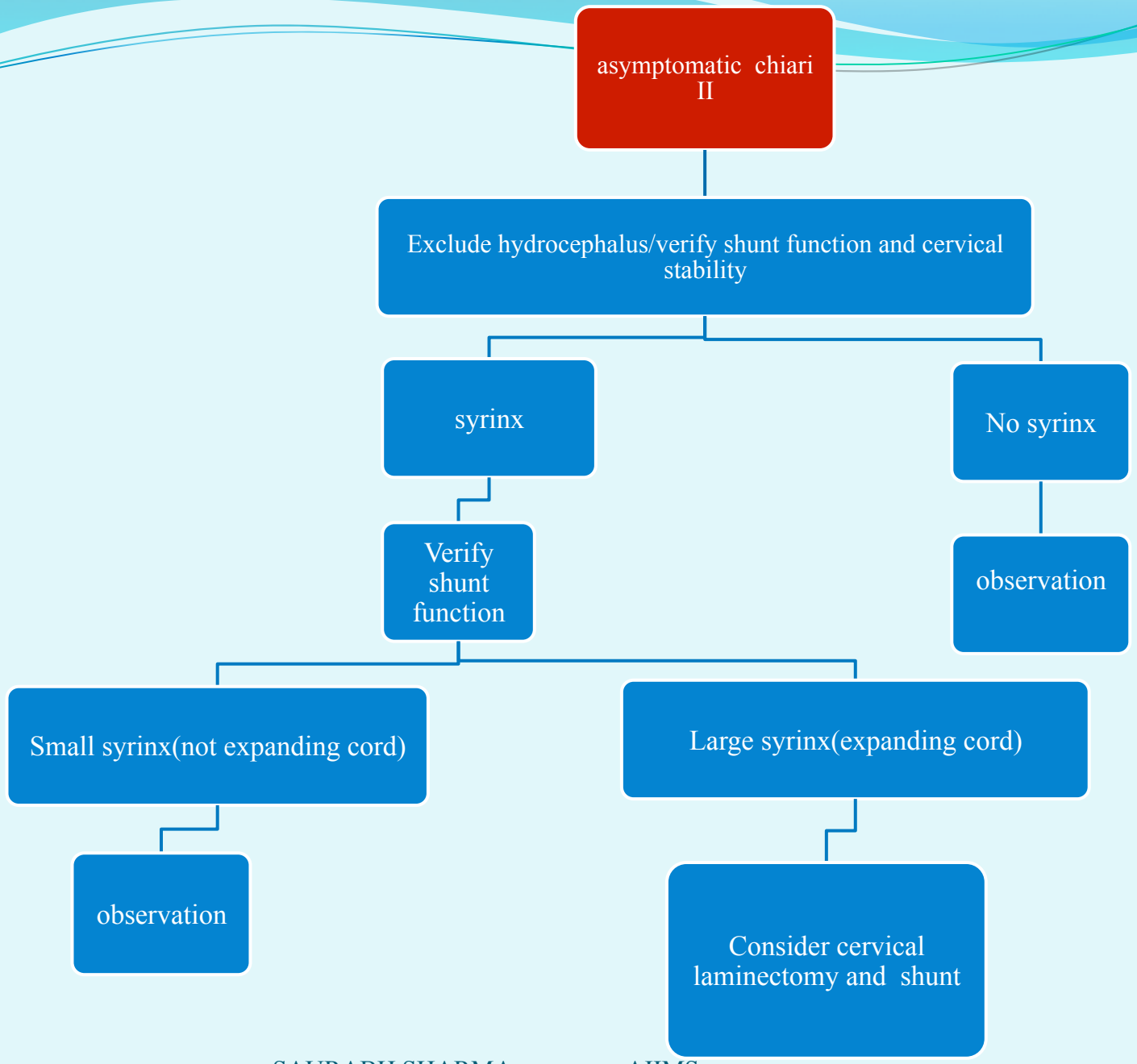
d) Adults-opthalmic problems common(strabismus/ nystagmus)

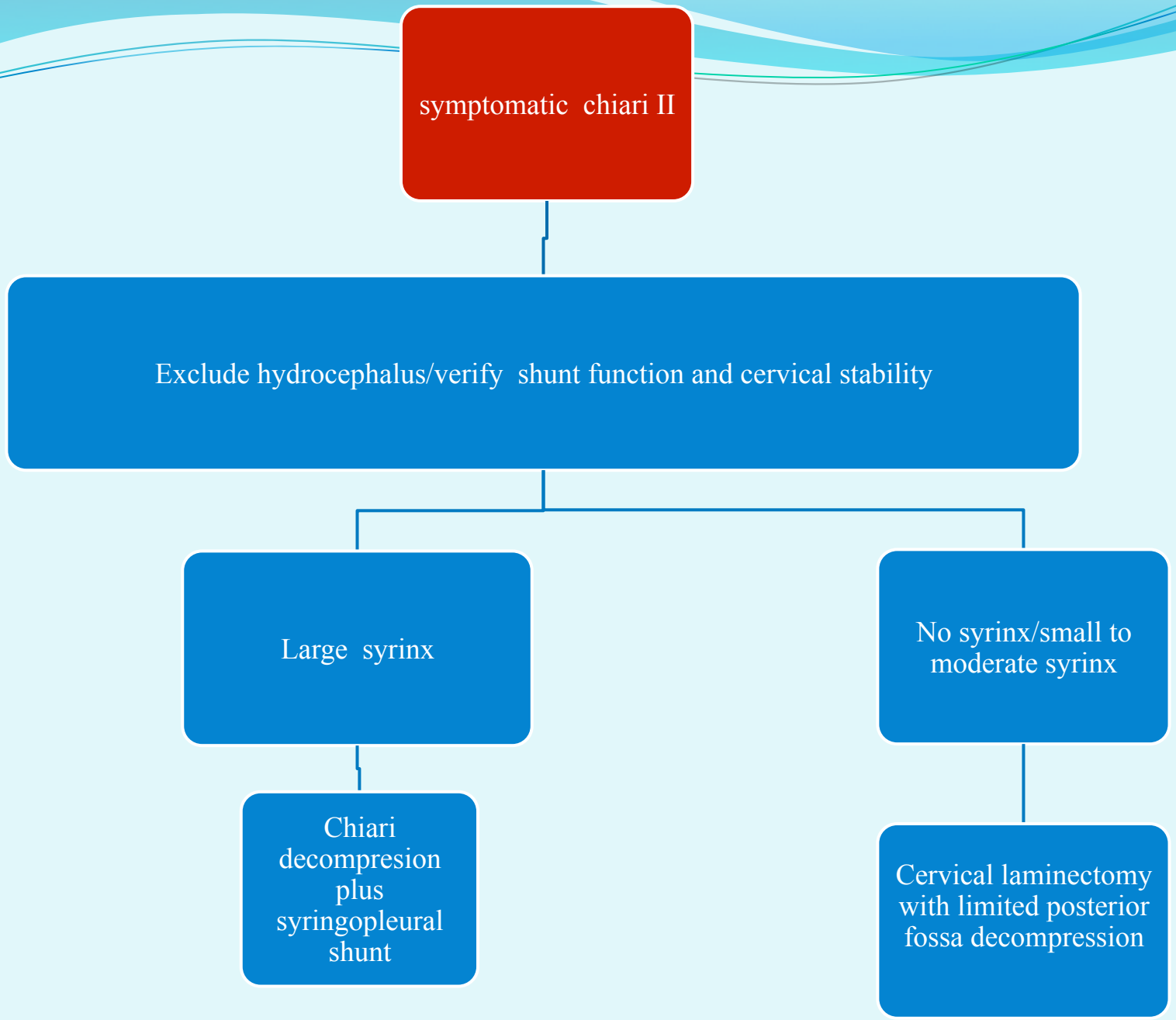
- ***Neurologically stable –if any change then suspect***

a) Tethering at mmc closure site

b) raised ict

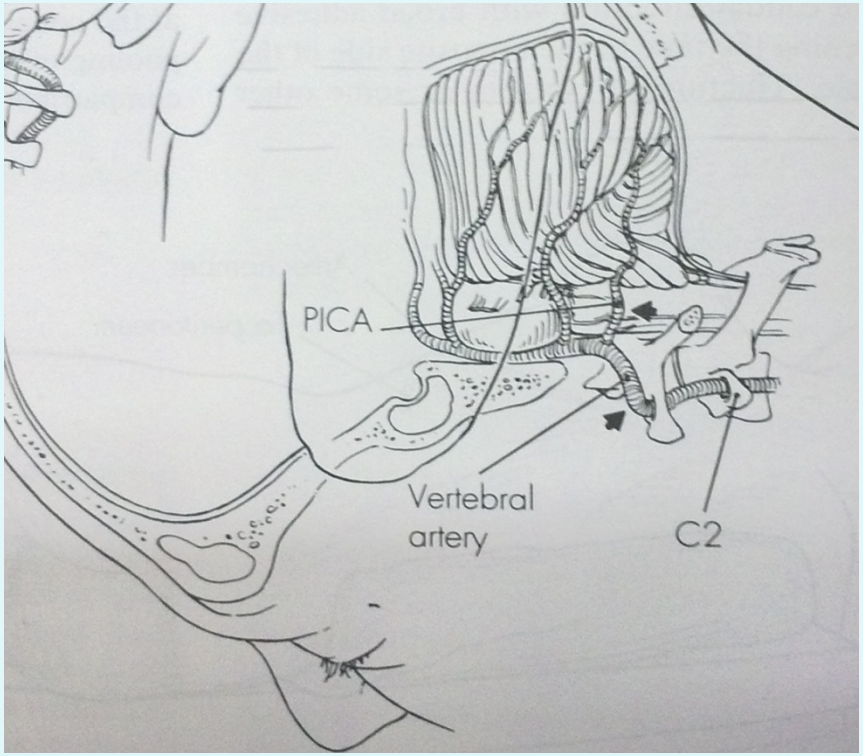
c) syrinx


asymptomatic chiari II
Exclude hydrocephalus/verify shunt function and cervical stability
syrinx
No syrinx
Verify shunt function
observation
Small syrinx(not expanding cord)
Large syrinx(expanding cord)
observation
Consider cervical laminectomy and shunt


symptomatic chiari II
Exclude hydrocephalus/verify shunt function and cervical stability
Large syrinx
No syrinx/small to moderate syrinx
Chiari decompresion plus syringopleural shunt
Cervical laminectomy with limited posterior fossa decompression

Chiari decompression-OPERATIVE CONSIDERATIONS

1. Cervical laminectomy to expose the inferior margin of the herniated cerebellum
2. Limited suboccipital craniectomy (torcula may be low)
3. foramen magnum typically larger than normal in CM II does not often play a role in compression of the brainstem
4. **Key objective – finding the outlet of fourth ventricle**
5. Choroid plexus extraventricular outside the 4th ventricle may serve as a guide
6. Malformed vermis and medulla look strikingly similar- great care should be taken in identifying the two structures
7. PICA can be present in the surgical field- seek, identify, protect



complications

- Blood loss, infection, vascular and CNS injury, persistence of symptoms
- Cervical instability and kyphosis
  - radiologically as high as 90%
- Recurrence
  - shunt malfunction
  - inadequate initial decompression
  - bone regrowth
  - epidural scarring with band like compression
  - syrinx formation
  - cervical spondylolisthesis

Fetal Surgery and CM II:MOMS TRIAL

- The MOMS trial (mx of mmc study) was closed for efficacy in December 2010 based on comparing outcomes after prenatal and postnatal repair in 183 patients.
- **Dr. N. Scott Adzick**, MD, Surgeon-in-Chief and Director of the Center for Fetal Diagnosis and Treatment at The Children's Hospital of Philadelphia.
- *The trial demonstrated that outcomes after prenatal spina bifida treatment are improved to the degree that the benefits of the surgery outweigh the maternal risks.*
- the study found that prenatal repair resulted in:

- ***Reversal of the hindbrain herniation component of the Chiari II malformation***
- ***Reduced need for ventricular shunting***
- ***Reduced incidence or severity of potentially devastating neurologic effects caused by the spine's exposure to amniotic fluid, such as impaired motor function***

Chiari III

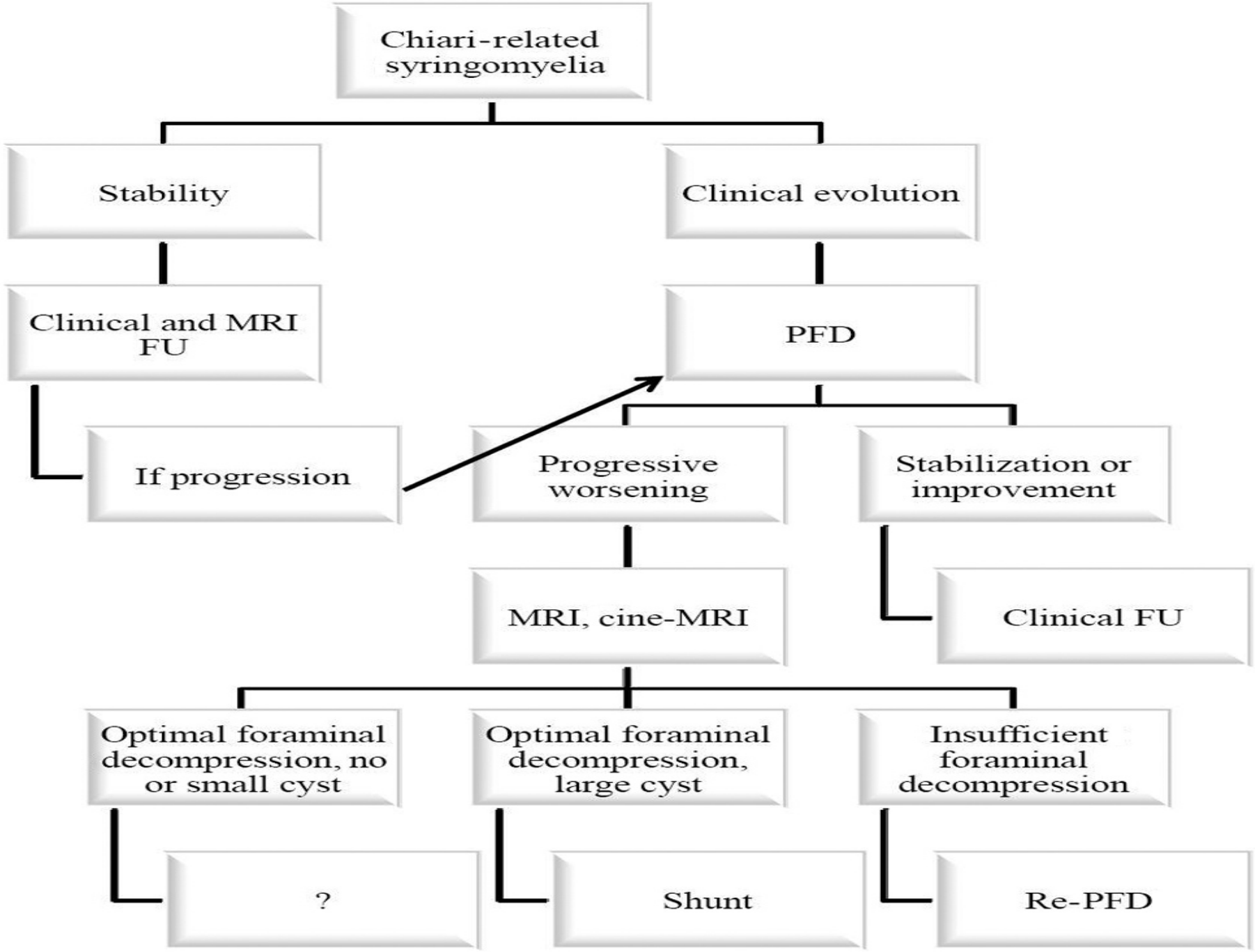
- Chiari described only *one case* in his series (very rare)
- Occipital/high cervical encephalocele with other anomalies seen with the type II
- Encephalocele contains varying amounts of dysmorphic neural elements
- Patients have ***severe neurological defects and a poor prognosis***

"Chiari IV" or primary cerebellar agenesis. tiny portion of residual quadrangular lobule just caudal to the tectum and a normal sized posterior fossa is seen.

CHIARI IV

(Extremely rare)

- No hind brain herniation
- Cerebellar a/ hypoplasia with tentorial hypoplasia
- Normal sized Post fossa


Chiari-related syringomyelia
Stability
Clinical evolution
Clinical and MRI FU
PFD
If progression
Progressive worsening
Stabilization or improvement
MRI, cine-MRI
Clinical FU
Optimal foraminal decompression, no or small cyst
Optimal foraminal decompression, large cyst
Insufficient foraminal decompression
?
Shunt
Re-PFD

Thank you !