THE CHIARI MALFORMATIONS: DIAGNOSIS AND MANAGEMENT

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Initially described three malformations
5 years later he revised the second and added a fourth
Arnold described a single case with Chiari II features
The chiari II malformation also known as Arnold-Chiari malformation.
Series of hindbrain anomalies
No anatomical or embryological correlation between them
Four types
<table>
<thead>
<tr>
<th>Type</th>
<th>Definition</th>
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<td>Chiari I</td>
<td>Caudal descent of <em>cerebellar tonsils</em> &gt; 5mm below foramen magnum. Hydrocephalus uncommon.</td>
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<tr>
<td>Chiari II</td>
<td>Caudal herniation of <em>cerebellar vermis, brainstem and fourth ventricle</em>. Almost all have hydrocephalus and myelomeningocele.</td>
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<td>Chiari III</td>
<td>Chiari II and posterior fossa contents herniating into occipital/ high cervical encephalocele.</td>
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<td>Chiari IV</td>
<td>Cerebellar aplasia or hypoplasia with aplasia of tentorium cerebelli.</td>
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Chiari I malformation

- Traditionally defined as > 5mm tonsillar descent below the foramen magnum.
- Tonsils ascend with age
- 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.
more important than absolute tonsillar descent may be
- Peg like shape of tonsils
- Attenuation of posterior fossa cisternal spaces
- Suggestive clinical picture

Primarily chiari I is manifestation of underdevelopment and malformation of occipital cranium

Cerebellar ectopia due to reduced posterior fossa volume and crowding of contents
Other manifestations of hypodeveloped post fossa

- Increased slope of the tentorium
- Reduced height of the supraocciput
- Reduced length of the clivus
- Retroflexion of odontoid process
- **Acquired tonsillar ectopia**
  - Due to reduced volume of cranial cavity (Rickets, Craniosynostosis, Pagets disease)
  - Due to increase in volume of intracranial contents (acute hydrocephalus, tumour, cerebral edema)
Chiari I malformation
Associated findings

- **Skull**
  - Shortened supraocciput
  - Shortened clivus
  - Larger than normal foramen magnum
  - Empty sella
  - Clival concavity, platybasia, basilar impression
- **Spine**
  - Klippel-flail deformity and atlanto axial assimilation
    - Retroflexed odontoid process
    - Thickened ligamentum flavum
    - Scoliosis
- **Meninges**
  - Elevated slope of tentorium cerebelli
  - Thickened arachnoid at foramen magnum level
  - Dural thickening/ at the level of arch of atlas
- Spinal cord
  - 50 to 75% have cavitation within the cord (syrinx)
  - Lower cervical and thoracic cord mostly involved.
  - Segment of cord caudal to 4th ventricle may be spared from cavitation.
- Brain usually normal except for tonsillar abnormality
- Hydrocephalus described in 3 to 10%.
Pain is the most common complaint
- Occipital and cervical region pain aggravated by Valsalva, cough-laugh headaches

Signs and symptoms related to brainstem / cranial nerve and cerebellar compromise
- Ataxia, downbeating nystagmus, incordination, dizziness
- Dysphagia, dysarthria, hiccoughs, glossal atrophy
- Impaired gag, facial numbness
- Extreme cases – “cerebellar fits”
- **Signs and symptoms related to syrinx**
  - Dissociative sensory loss, upper limb weakness and thinning, lower limb spasticity.
  - Neuropathic pain in the extremities.
  - Neuropathic joints in upper extremities.
  - Uncommonly, JPS loss leading to sensory ataxia.

- **Presentation usually in the 2nd and 3rd decades with a female preponderance**
Theories explaining syringomyelia

- Gardner’s hydrodynamic theory
  - Blocked fourth ventricular outlet
  - Pulsatile CSF pressure transmitted to central canal through obex “Water hammer”

- William’s craniospinal dissociation theory
  - Valve like obstruction to free flow of CSF between cranial and spinal subarachnoid space at FM
  - Equalisation of CSF pressure between cranial and spinal compartments hindered
  - CSF sucked into the syrinx.
Oldfield’s theory

- Systolic downward motion of the tonsils creates a piston effect on the cervical spinal cord
- Interstitial fluid driven into the central canal distending it.

No theory however, successfully explains all observations
Approach to treatment of Chiari I
asymptomatic chiari I

Exclude hydrocephalus, ventral compression, cervical instability

syrinx
Chiari decompression

No syrinx
>7mm caudal descent
Exercise clinical judgement

<7mm caudal descent
Observation
Symptomatic Chiari I

Exclude hydrocephalus, ventral compression, cervical instability

Syrinx

Chiari decompression

No syrinx

>7mm caudal descent

3-7mm

<3mm caudal descent

Exercise clinical judgement

Observation with frequent evaluation
Chiari I decompression

- Aim of the surgical procedure
  - Establishment of normal CSF outflow from the ventricles
  - Increasing posterior fossa volume
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
  - Dura left intact
  - Dura inelastic hence, decompression inadequate
  - 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 craniocervical junction, division of thick dural band
- Opening the arachnoid
  - Required when significant tonsillar descent with syringomyelia
  - Arachnoid bands divided
  - Arachnoid pegged to the dural edges
  - Augmentation duraplasty
- Tonsil reduction
  - Subpial coagulation
  - Subpial resection when tonsils gliotic
- 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 redundant
Complications

- Aseptic meningitis (most common)
- Wound dehiscence, pseudomeningocele
- CSF leak
Chiari II malformation

- Defining features
  - Caudal descent of vermis, fourth ventricle and brainstem
  - Almost always associated with hydrocephalus and associated anomalies
  - Seen in almost all patients with myelomeningocele
  - 0.02% of all births with female preponderance
Chiari’s theory

- Hydrocephalus leading to secondary chiari
- 10-20% may not have hydrocephalus
- Associated anomalies not explained
- Chairi II features precede hydrocephalus
- Small post fossa, low lying tentorium, upward cerebellar herniation not explained
Cleland’s theory
  - Primary dysgenesis of the hindbrain
  - Fails to explain supratentorial anomalies

Induced small posterior fossa
  - Due to CSF leaking out from the open spinal cord defect
  - Fails to explain associated anomalies
- Penfield’s traction theory
  - Traction by tethering of cord at the site of myelomeningocele pulls the post fossa contents
  - Traction effect however dissipated four spinal levels rostral
  - Fails to explain associated cranial deformities
Unified theory of Mclone and Knepper

- 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
Radiological findings

- skull
  - Luckenschadel/craniolacunia
  - Frontal bone scalloping “lemon sign”
  - Scalloping of petrous bone and jugular tubercles
  - Concavity of the clivus
  - Low inion, small post fossa
  - Enlarged foramen magnum
  - Clival concavity
  - Basilar invagination and atlas assimilation
Frontal bone scalloping - “Lemon sign”

Normal fetus
Scalloped petrous bones in Chiari II

Normal skull in 50 yr old man
- Spine
  - Cervical spinal canal enlarged.
  - Scalloping of the odontoid process
  - Incomplete posterior arch of C1
  - Klippel-Feil deformity
Ventricle and cistern

- Hydrocephalus seen in 90%.
- Fourth ventricular outlet obstruction responsible
- Aqueductal stenosis uncommonly responsible for hydrocephalus
- Medial pointing of the inferior margins of floor of lateral ventricles
- “colpocephaly”
- Fourth ventricle typically small, flat and elongated
- Lateral recesses not well defined
colpocephaly  
Pointed frontal horns  
Slit like fourth ventricle
“bat wing” appearance
- **Meninges**
  - Tentorium cerebelli usually widened heart shaped
  - Low lying, hypoplastic
  - Falx cerebri fenestrated/hypoplastic

Low lying tent  

Normal hiatus  

Hiatus in Chiari II
CECT head showing interruptions in falx blush suggestive of falx fenestrations
- **Spinal cord**
  - Myelomeningocele always associated with Chiari II
  - Syringomyelia in 20-95%
  - Shortened cervical cord

- **Telencephalon**
  - Complete partial agenesis of corpus callosum/septum pellucidum
  - Polygyria
  - “chinese lettering” - interdigitation of occipital / parietal lobes
- Gray matter heterotopia

- Agenesis of olfactory tract/bulb/cingulate gyrus
- Diencephalon
  - Enlarged massa intermedia
- **Mesencephalon**
  - Tectal beaking due to fusion of the colliculi
  - Midbrain typically elongated
  - Cranial nerve nuclei may be malformed
  - Aqueduct may be stenotic, stretched, posteriorly kinked or forked
- Metencephalon
  - Cerebellum grossly smaller and may tower above tentorium
  - Cerebellum may be displaced laterally spreading around the brainstem – "banana sign"
  - Lateral cerebellar edges may touch brainstem and basilar artery – ”cerebellar inversion”
  - Pons elongated and flattened
Banana sign

Normal fetus
• “Cerebellar inversion”

• Chinese lettering
- Myelencephalon
  - Medullary kinking, elongation and flattening
  - Pyramidal decussation more cephalad than normal
Most common is with open neural tube defects
Symptomatic chiari II is the most common cause of death in children <2 yrs of age with MMC.
Symptomatic patients can be classified according to age at presentation
Whatever be the age of child, hydrocephalus/shunt malfunction should be excluded
Age at presentation less than 2yrs

- Most frequent symptoms related to brain stem and cranial nerve dysfunction
- Symptomatic chiari is a neurosurgical emergency in this group.
- Most commonly inspiratory stridor and PEAC (prolonged expiratory apnea and cyanosis)
- PEAC = apneic spell+opthistonic posturing and cyanosis
- Laryngoscopy may reveal impaired vocal cord abduction
- Downbeat nystagmus, fixed retrocollis
Other signs and symptoms

- Impairment of gag
- Dysphagia, chronic aspiration, nasal regurgitation
- Quadriplegia, nystagmus, developmental delay
- Weak cry.
Presentation in older age group
- Less serious and rarely an emergency
- Hallmark is cervical myelopathy
- Weakness and spasticity in upper limbs
- Suboccipital headache
- Ataxia
- Hand weakness, atrophy
- Syringomyelia and associated symptoms
- Ophthalmic problems common in adults
Chiari II
Approach to management
asymptomatic chiari II

Exclude hydrocephalus/verify shunt function and cervical stability

syrinx

No syrinx

Verify shunt function

Small syrinx

observation

Large syrinx

Consider cervical laminectomy and syringo pleural shunt

observation
symptomatic chiari II

Exclude hydrocephalus/verify shunt function and cervical stability

Large syrinx
- Chiari decompression plus syringopleural shunt

No syrinx/small to moderate syrinx
- Cervical laminectomy with limited posterior fossa decompression
Chiari decompression

- Surgical technique basically similar to that in chiari-I
- Cervical laminectomy should expose the inferior margin of the herniated cerebellum
- Limited suboccipital craniectomy
- Constricting dural bands divided
- Key objective – finding the outlet of fourth ventricle
- Choroid plexus may serve as a guide
- Obstructive vermian tissue may be divided/perforated to encourage CSF flow out of the ventricles.
- Medullary kink not to be confused with vermis.
- Arachnoid clipped to dura with augmentation duraplasty
- Complications of this procedure similar to that for Chiari I.
Chiari III and IV

- **Chiari III**
  - Very rare
  - Occipital or cervical encephalocele along with chiari II anomalies

- **Chiari IV**
  - No hindbrain herniation
  - Cerebellar hypoplasia or aplasia
Chiari zero
- CSF equilibrium changes at cranio-cervical junction
- No hindbrain herniation
- Syringohydromyelia
- Post fossa decompression leads to dramatic improvement
- Other causes of syrinx must be excluded.
Thank you