Morphologies of Chiari I deformity: what matters

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Chiari: malformations or deformities?

- Definitions relate to pathogenesis, not degree
  - Malformation
    - Chiari III: NTD (cranio-cervical encephalocele)
  - Deformities
    - Chiari II: secondary to NTD (myelomeningocele)
    - Chiari I: other chronic causes
  - Herniation: acute/subacute mass effect
- What makes no sense in this perspective
  - tonsillar descent cut-off of 5mm
  - Chiari 0 and Chiari 1.5
Chiari 1 deformity: possible processes

• Small cranial container
  – suture synostosis, mainly Crouzon
  – intracranial hypotension with closed sutures, thick skull, thick dura
  – other etiologies of thick skull (e.g. thalassemia)

• Large cranial content: fore- and/or hind-brain
  – NF1
  – macrencephaly, hemimegalencephaly
  – cerebellar hyperplasia
  – chronic supratentorial mass effect (i.e. chronic hydrocephalus)

• Small posterior fossa
  – synchondroses
  – CVJ malformations
Not a malformation but a deformity

Uncommonly, Chiari 1 deformity may be shown to appear or regress
Anatomical landmarks of posterior fossa

- Chamberlain, McGregor, McRae
- Wackenheim, clivo-cervical angle
- Osteo-neural landmarks
  - incisura → mid-midbrain
  - synchondrosis → mid-pons
  - basion → obex
  - dens → medulla/cord
- Patent cisterns
- Regular pentagon
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Common Chiari 1

- flat posterior fossa, low hindbrain, effaced cisterns
- clivo-cervical angle normal, asymmetric pentagon
Small posterior fossa

- short clivus, short supraoccipital, shallow PF with low & high hinbrain
- clivo-cervical angle normal, asymmetric pentagon
Small posterior fossa

- small posterior fossa, effaced cisterns, Chiari I features
- the vaults give way, cerebellar ascent, distorted pentagon
Small posterior fossa, retroverted dens

- short clivus, abnormal craniocervical angulation with long “retroverted” dens, basal invagination
- clivo-cervical angle decreased, flattened pentagon
E: Ebner fissure
- Centrum intersegmental
- Hypocentrum segmental; lost except O4 and C1
Malformations or craniovertebral junction

- **Aplasia**
  - vs lack of ossification

- **Hypoplasia**

- **Abnormal segmentation (homeobox)**
  - cranial shift
  - caudal shift
  - mosaic

- **For each:**
  - centrum, hypocentrum, lateral
  - symmetric or not
  - single or in combination
• cephalic flexion is at occipito-atlantal joint mostly
• Jugular tubercle + condyle: 25mm
• Lateral mass: 17mm
• pB-C2 < 9mm
Occipital hypoplasia and Chiari I

- Head flexed forward, basal invagination
- Relative ascent of the dens
- Main fulchrum for head flexion remains at occipito-atlantal joints, while dens part of fixed spine, brainstem part of moving head
Cranial segmentation shift: cervicalization

Prominent dens & short clivus
Associated hypoplasia of jugular tubercles-condyles
Neo-articulation with unknown ligamentous anatomy
Short clivus, basal invagination

Short clivus, small condyles

Short clivus, platybasia (>140°), hypoplastic condyle, long dens

Short clivus, basal invagination, long dens
Fused hypocentra with undivided O4, long dens and invagination, abnormal ligaments
Summary

• In assessing Chiari 1 deformity, need to dissociate
  1. cause and mechanism
  2. location of tonsils with impact on CSF flow and cord
  3. CVJ dynamics and osteo-neural relationships (no Chiari possible)
Conclusions

- Chiari I deformity results from mechanical processes (container/content), similar to classical tonsillar herniation.
- Accordingly, tonsillar dislocation is a feature of many different disorders.
- Tonsillar dislocation has its own pathology (CSF dynamics, local compression).
- CVJ malformations result in abnormal osteo-neural relationships (retroverted dens) and a specific pathology.
- Abnormal CVJ segmentation likely associated with specific but unknown anatomy of corresponding ligaments.