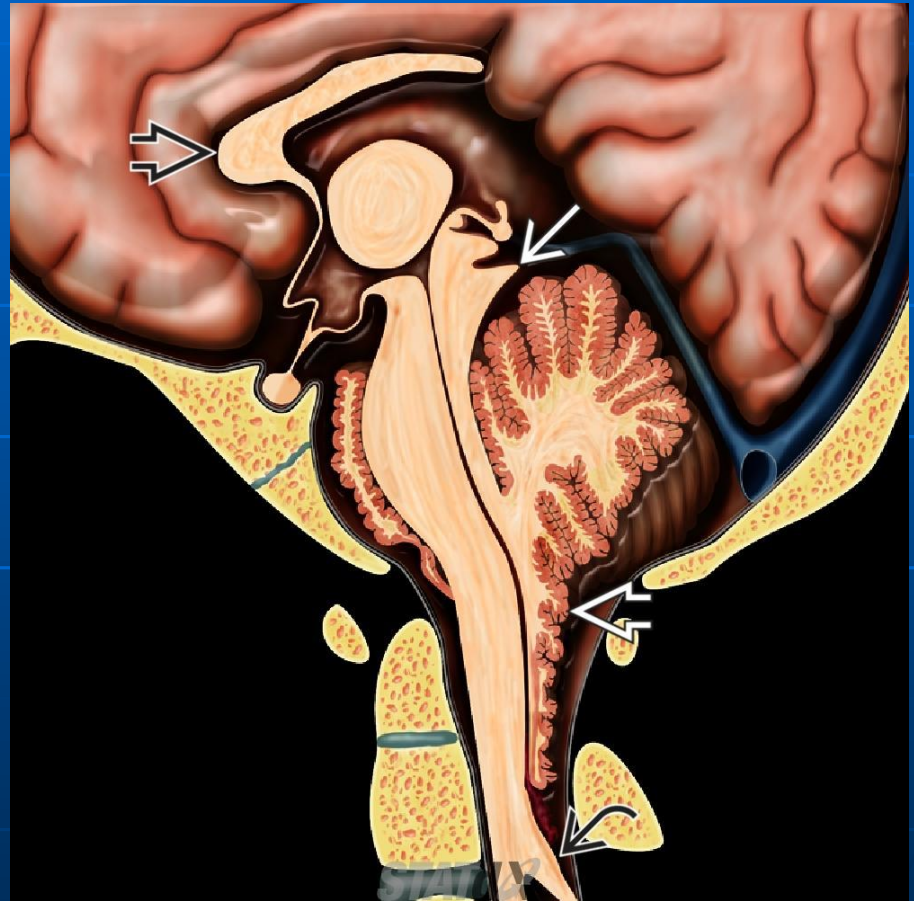


# Chiari 2 malformation

- Callosal dysgenesis (black open arrow)
- Tectal beaking (white solid arrow)
- Small posterior fossa, vermian ectopia (white open arrow)
- Medullary kinking (black curved arrow).



# Chiari 2 malformation

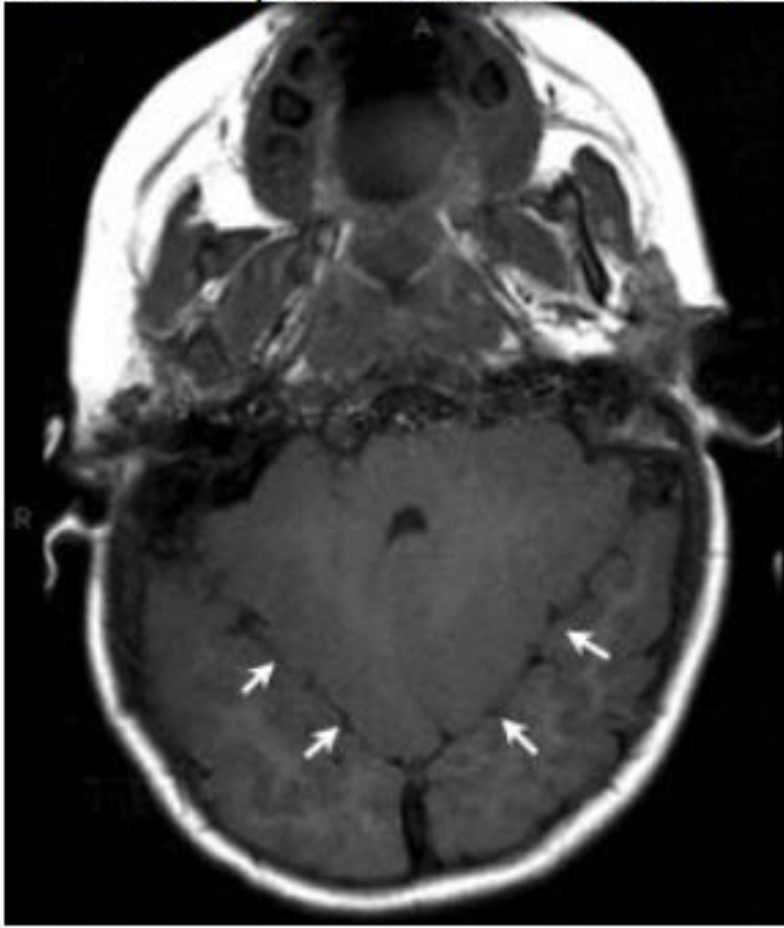
- Complex hindbrain malformation
- Virtually 100% associated with neural tube closure defect (NTD), usually lumbar myelomeningocele (MMC)
- Imaging
  - Cerebellum "wraps" around medulla and "towers" through incisura, with "beaked" tectum and heart-shaped midbrain
  - Virtually 100% have neural tube closure defect (NTD)
  - Cerebellum "wraps" around medulla and "towers" through incisura, with "beaked" tectum and heart-shaped midbrain

# Pathology

- Secondary to sequelae of CSF leakage through open spinal dysraphism during gestation (4th fetal week)
- Methylene tetrahydrofolate reductase (MTHFR) mutations → abnormal folate metabolism
- Spine- and brain/skull-associated anomalies common

# Chiari 2

Heart Shaped Tentorial Incisura



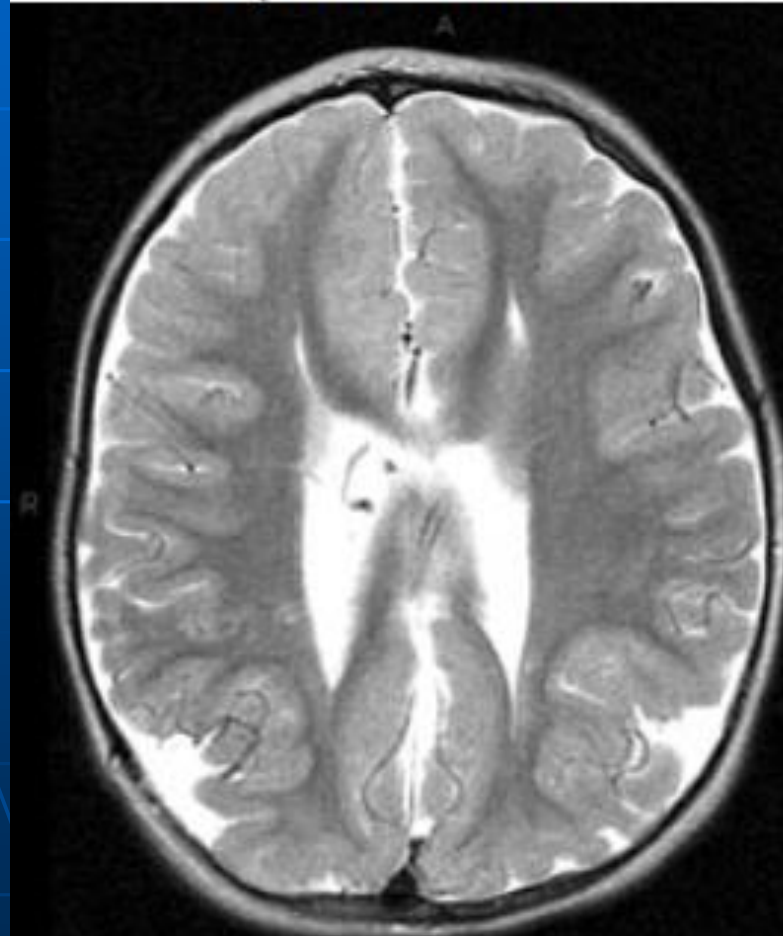
Towering Cerebellum





# Chiari 2

Dysgenesis of the  
corpus callosum



Fenestrated falx

