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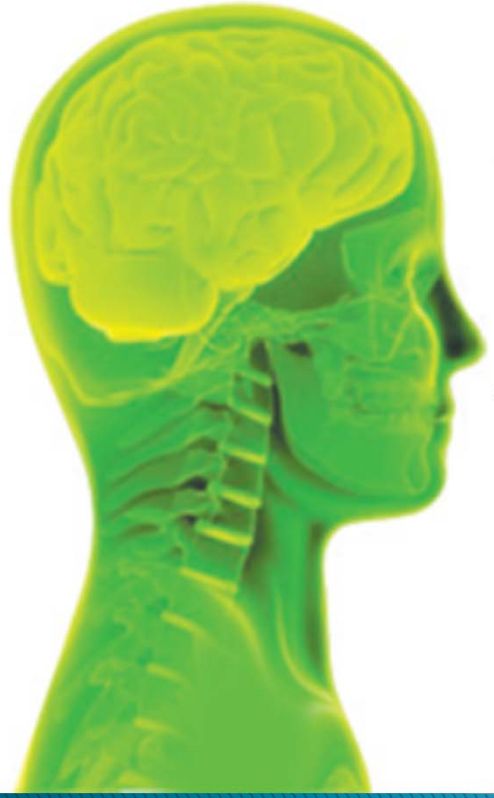

Chiari Malformations and Other Incidental Findings in the Age of MRI

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The Recent Literature

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Incidental Findings on Brain Magnetic Resonance Imaging of Children With Sickle Cell Disease

Lori C. Jordan, Robert C. McKinstry, III, Michael A. Kraut, William S. Ball, Bruce A. Vendt, James F. Casella, Michael R. DeBaun, for the Silent Infarct Transfusion Trial Investigators and John J. Strouse

Pediatrics 2010;126;53-61; originally published online Jun 14, 2010;
DOI: 10.1542/peds.2009-2800

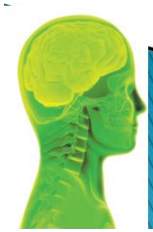
In this large cohort of children, incidental intracranial findings were identified on brain MRI
scans in 6.6% of cases, with potentially serious or urgent findings in 0.6%.

"These data should assist pediatricians in counseling families when incidental findings are encountered."

No Referral	
Cavum septum ,pellucidum, vergae, or velum interpositum	11
Choroidal fissure cyst	6
Gray matter heterotopia	5
Arachnoid cyst	4
Prominent perivascular (Virchow-Robin) spaces	3
Pineal cyst	3
Absent septum pellucidum	2
Arachnoid cyst vs prominent cerebrospinal fluid space	1
Occipital bone cyst, possibly epidermoid	1
Routine referral	
Chiari I malformation	20
Idiopathic ventriculomegaly	1
Temporal lobe cystic change	1
Cortical dysplasia	2
Arachnoid cyst, large	1
Rathke cleft cyst, purely intrasellar	1
Urgent referral	
Chiari I with spinal cord syrinx	2
Tumor	4
Total	68

What this talk is

- A guide to help understand the acuity of these common for us, rare for you, findings
- A way for pediatricians to open a dialogue about new findings with the families
- A roadmap to triage to subspecialty care
- A comprehensive list of what's really incidental and what's fortuitous



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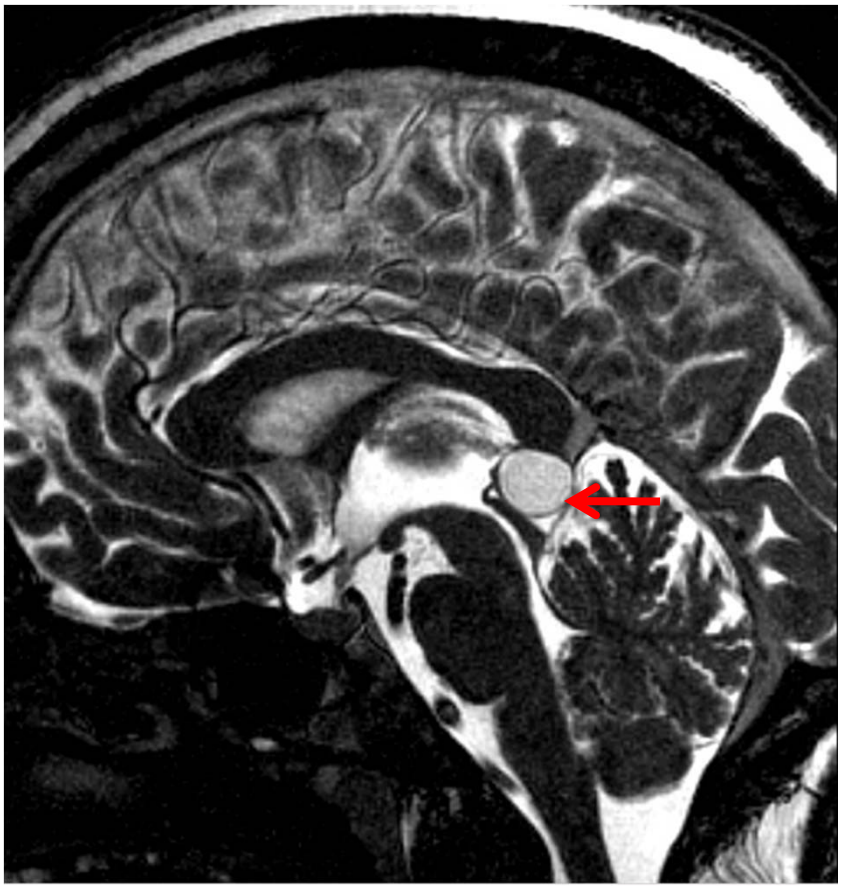
Incidental radiographic findings and normal variants

- The easy access to MRI scans (here in NYC)
- Image Gently campaigns
- Increased resolution and new sequences
- The medico-legal crisis and defensive medicine
- Extremely common reason for referral of a new patient to the pediatric neurosurgical office

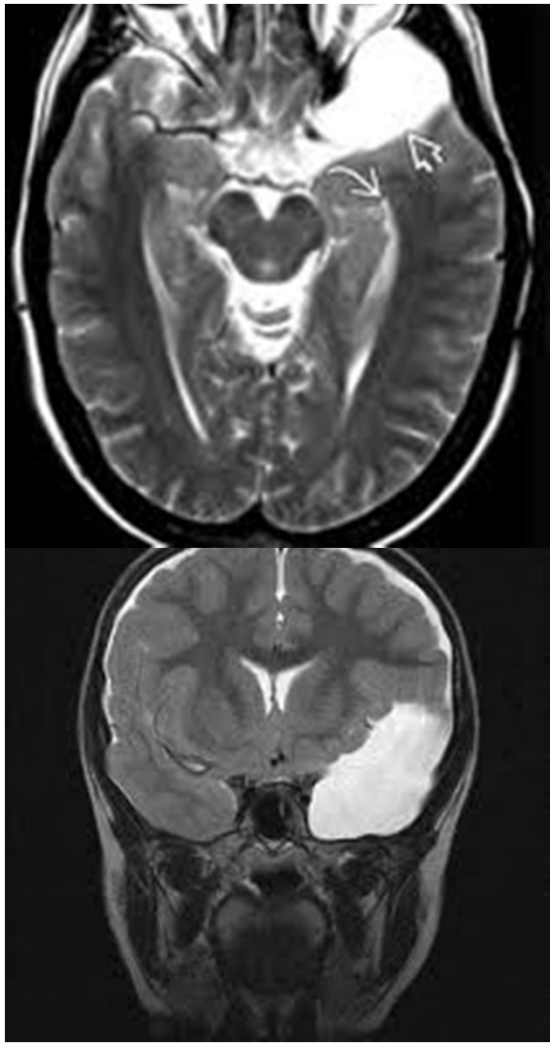
- Confusion for pediatricians
- Anxiety for parents
- Expensive for insurers

Incidental findings: pineal cyst



- A normal variant, occurs in 2-4% of MRI scans.
- Usually an incidental finding
- Present in large numbers of routine autopsies
- Mainly females, in puberty or midlife
- Rarely cause obstructive hydrocephalus or upgaze limitation

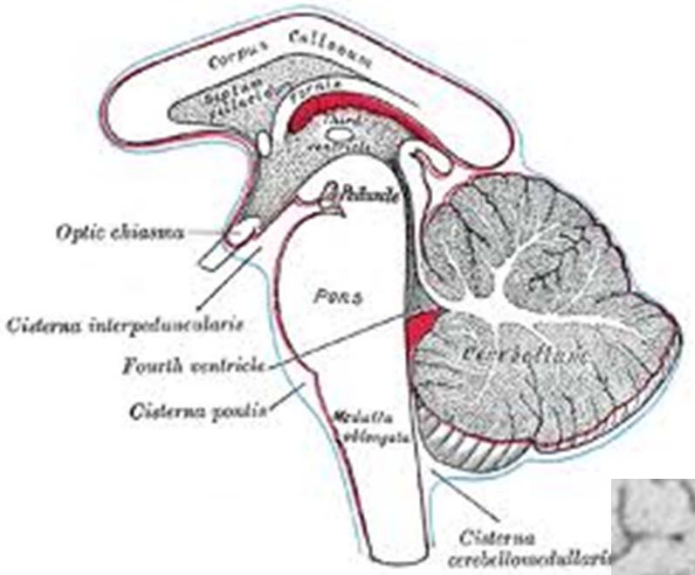
Incidental findings: middle fossa arachnoid cyst



- Found in 1/1000 individuals
- Usually an incidental finding.
- The exact cause of arachnoid cysts is not known.
- Developmental malformations from the splitting or tearing of the arachnoid.
- In some cases, arachnoid cysts occurring in the middle fossa are accompanied by underdevelopment (hypoplasia) or compression of the temporal lobe.

- Occasionally associated with headache or seizures
- Risk of bleeding with trauma
- Treatment is fenestration or shunt when symptomatic

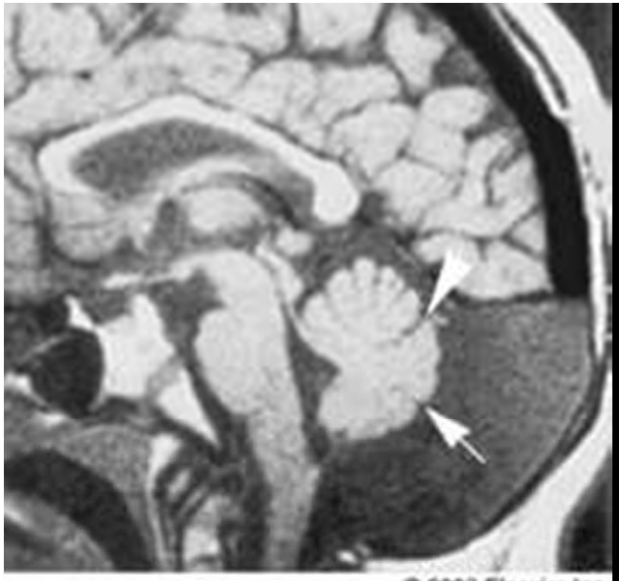
Incidental finding: mega cisterna magna

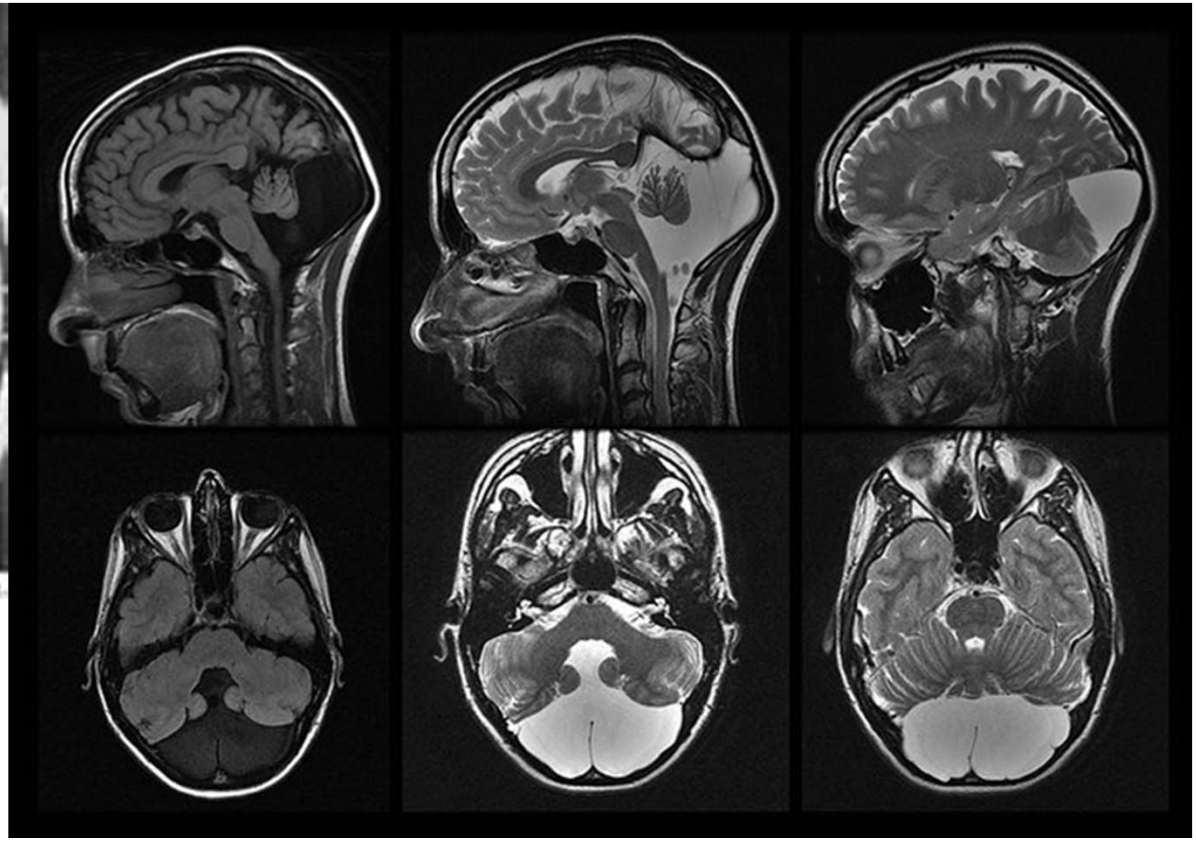


- Distinguish from Dandy-Walker and posterior fossa arachnoid cyst
- Dandy Walker Malformation: Agenesis of the vermis
- No mass effect, no hydrocephalus; communicates with 4th ventricle

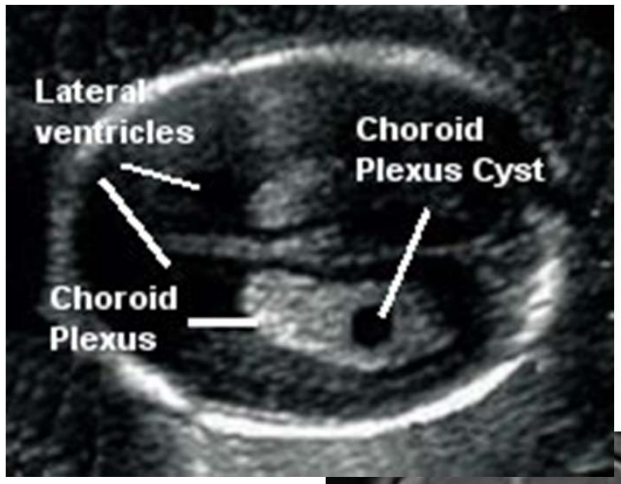
Incidental finding: Mega Cisterna Magna vs Dandy Walker

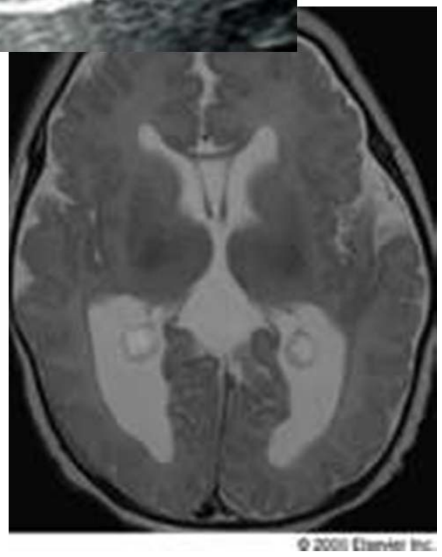
Hypoplasia of cerebellar vermis, elevated position of the torcula and tentorium, often associated with hydrocephalus due to obstruction of CSF flow





Incidental findings: Choroid Plexus Cysts





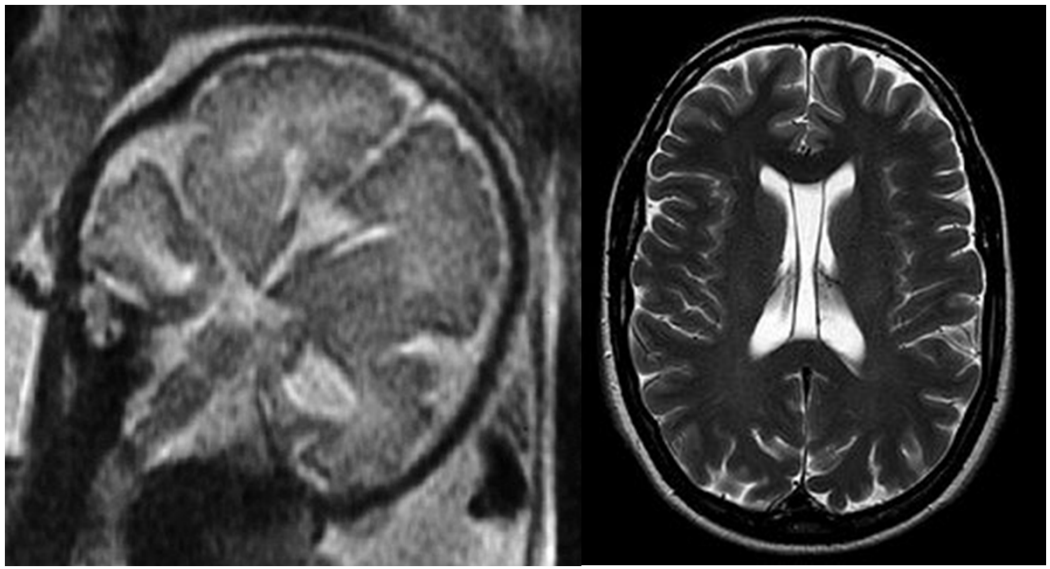
- The choroid plexuses are structures in the ventricles (spaces) of the brain that produce the cerebrospinal fluid
- Occasionally fluid becomes trapped and forms pockets in the choroid plexus. These pockets of fluid are called choroid plexus cysts (CPC).
- Seen during 1% to 3% of all mid-trimester prenatal ultrasound examinations.
- No effect on fetal development.

More than 90% of choroid plexus cysts resolve spontaneously by 28th weeks' gestation .

Incidental findings:choroidal fissure cysts

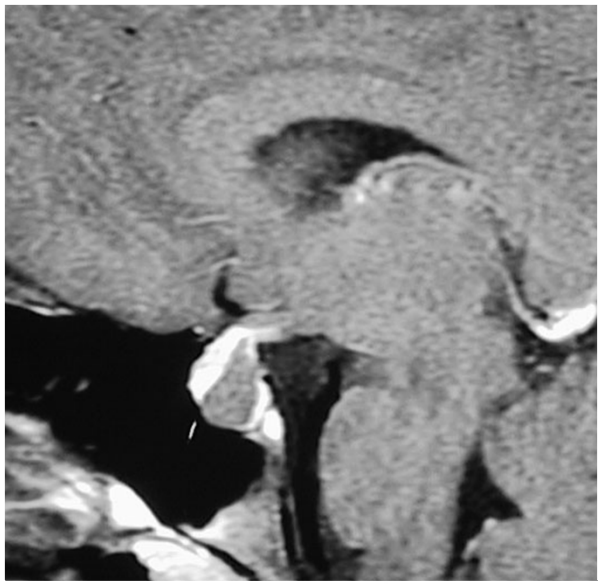
- A neuroepithelial cyst containing CSF in the medial temporal lobe adjacent to the choroid fissure.
- The choroidal fissure is a CSF space between the fimbria of the hippocampus and diencephalon
- Choroidal fissure cysts are usually asymptomatic and discovered incidentally.
- It is typically small and has minimal mass effect. It should not be mistaken for a pathologic lesion.

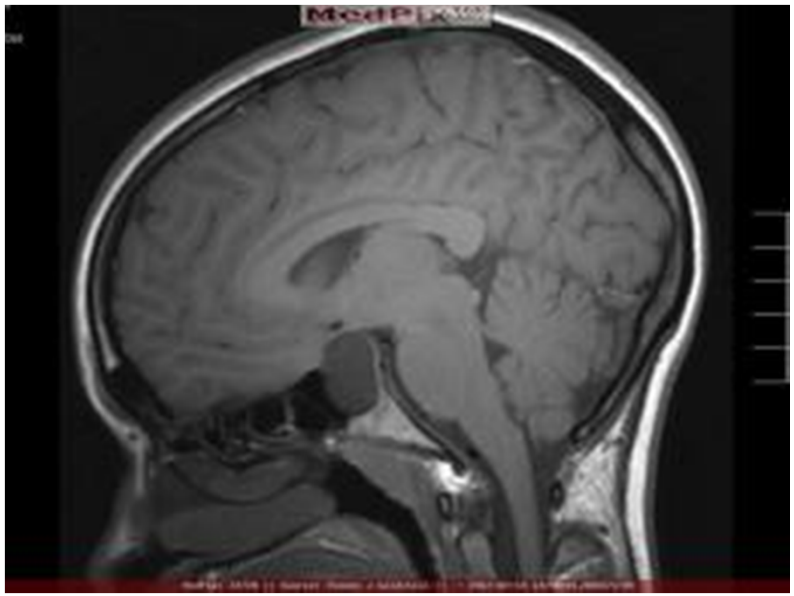
Incidental finding:cavum septum pellucidum

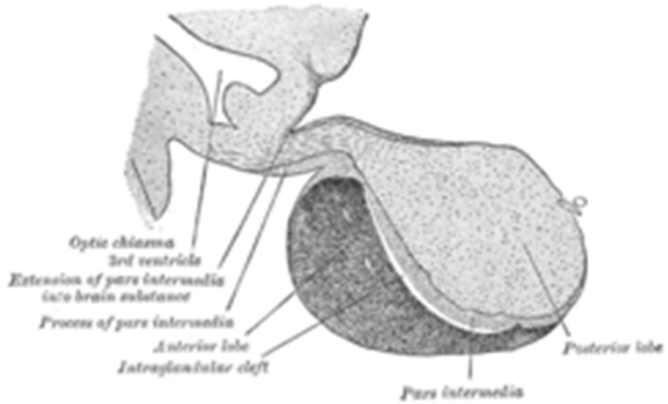


- Almost universally present at birth
- Persistent normal variant in 20% of normal population
- Unless ventricles obstructed, no further management or followup

Incidental findings: pars intermedia cyst





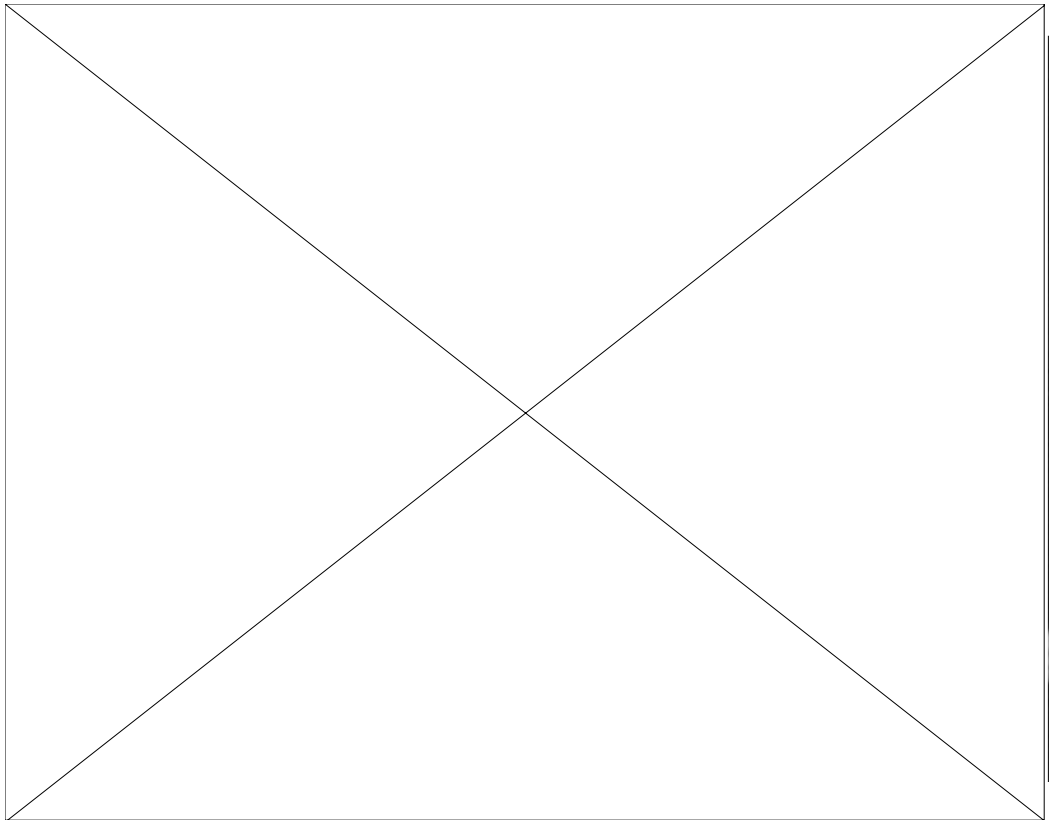


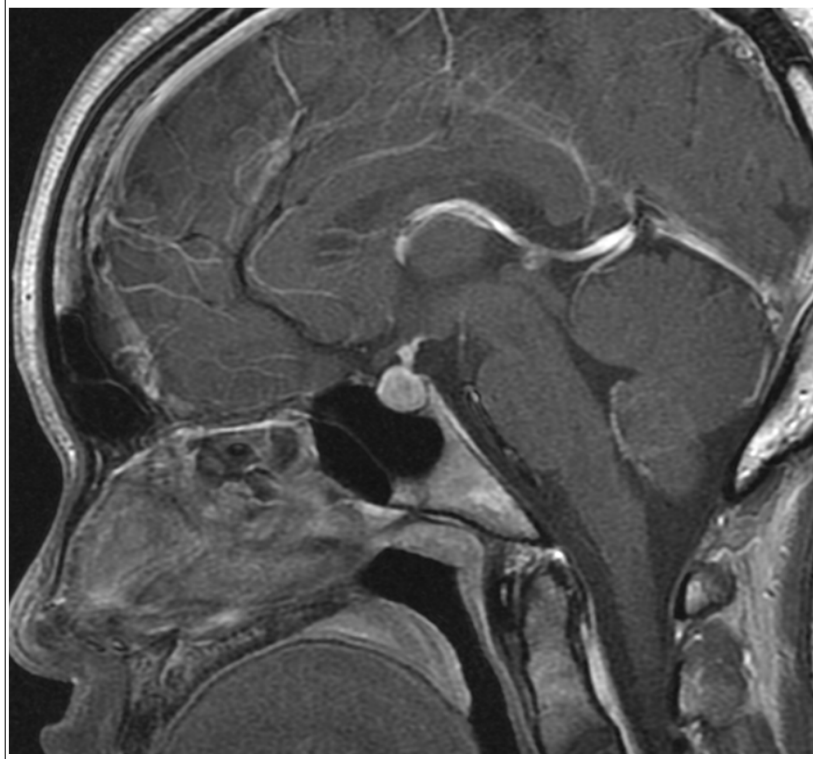
Non-neoplastic intrasellar cysts include:

- Rathke's cleft cysts,
- Pars intermedia cysts,
- Craniopharyngiomas
- Arachnoid cysts,
- Mucoceles, “Empty sella.“
- Cystic pituitary adenoma

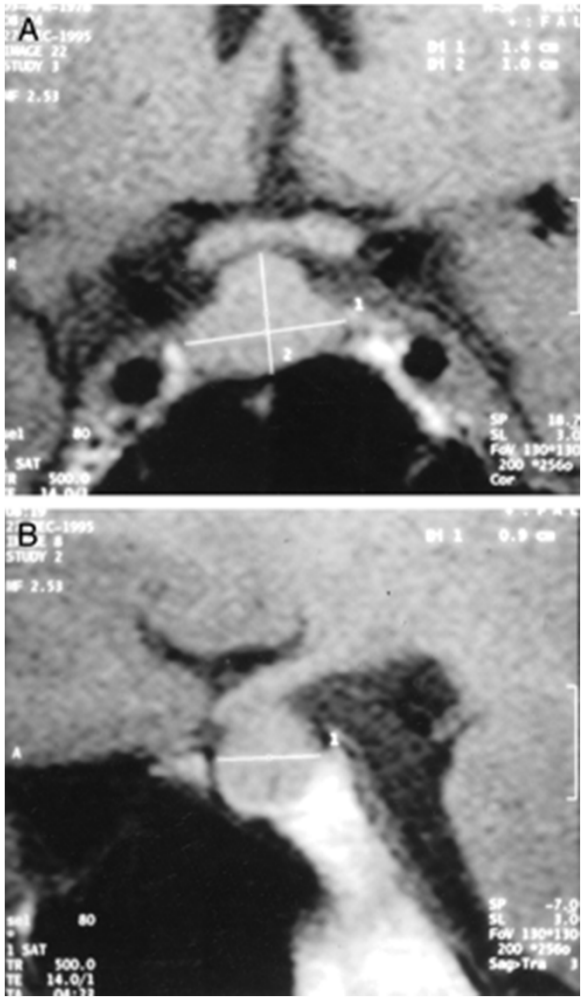
Question: This 15 year old girl presents with headaches for 3 months and her neurologist decides to get a scan.

Exam: non-focal neurologically c/o tender breast exam





Incidental findings: Normal adolescent pituitary



Homogeneously enhances

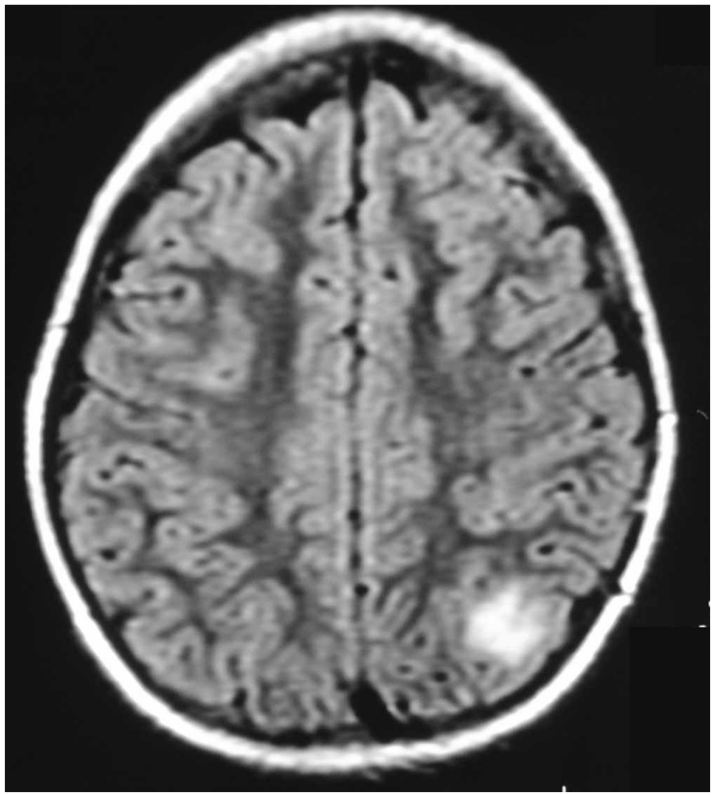
Often misread as tumor in adolescent females

In neuroradiological series, 25–50% of healthy women who are 18–35 yr old have a convex superior pituitary contour, but pituitary height exceeds 9 mm in less than 0.5% of cases.

Physiological pituitary hypertrophy is confirmed by normal baseline pituitary function and extensive hormonal tests.

Identification of these patients is important to avoid unnecessary pituitary surgery

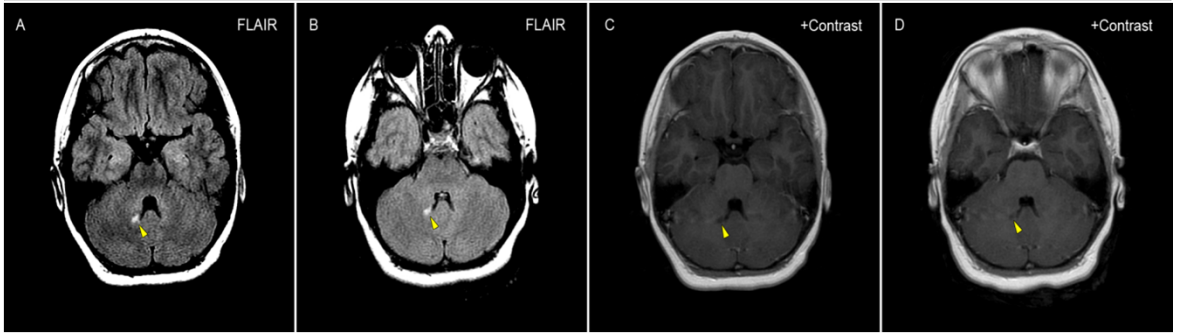
Incidental findings: FLAIRoma



- Often incidental finding
- No enhancement
- Seen on FLAIR and T2 sequences
- Differential: tuber, UBO, migraine,lyme, demyelination,
- Low grade glial tumor
- Treatment: surveillance scans vs biopsy

MSKCC Incidental Brain Tumor

MRI of Patient with Incidental, Asymptomatic PF Lesion over 14 months



Introduction

- The management of incidental MRI lesions is uncertain
- There is no published experience pertaining to pediatric, incidental PF lesions

Methods

Records of all patients (pts) seen by the pediatric neuro-oncology service at MSKCC with PF lesions from July 1, 1993 to January 22, 2010 were reviewed

Results

- 393 pts were identified
- 81 pts were excluded due to incomplete records
- 7 of 312 pts were found to have asymptomatic, incidental PF lesions
- 5 pts underwent gross total resection, 2 due to asymptomatic tumor progression
- 2 patients did not have surgery
- Median of 9 months between initial scan and surgery
- All pathology was low grade
- Median of 17 months since initial scan, all pts doing well

Conclusions

- No pts had high grade disease and none required adjuvant therapy, post-operatively
- This small series suggests asymptomatic PF lesions can be managed with clinical and radiographic observation
- Neurologic intervention may be deferred in this select population without adverse affect in outcome

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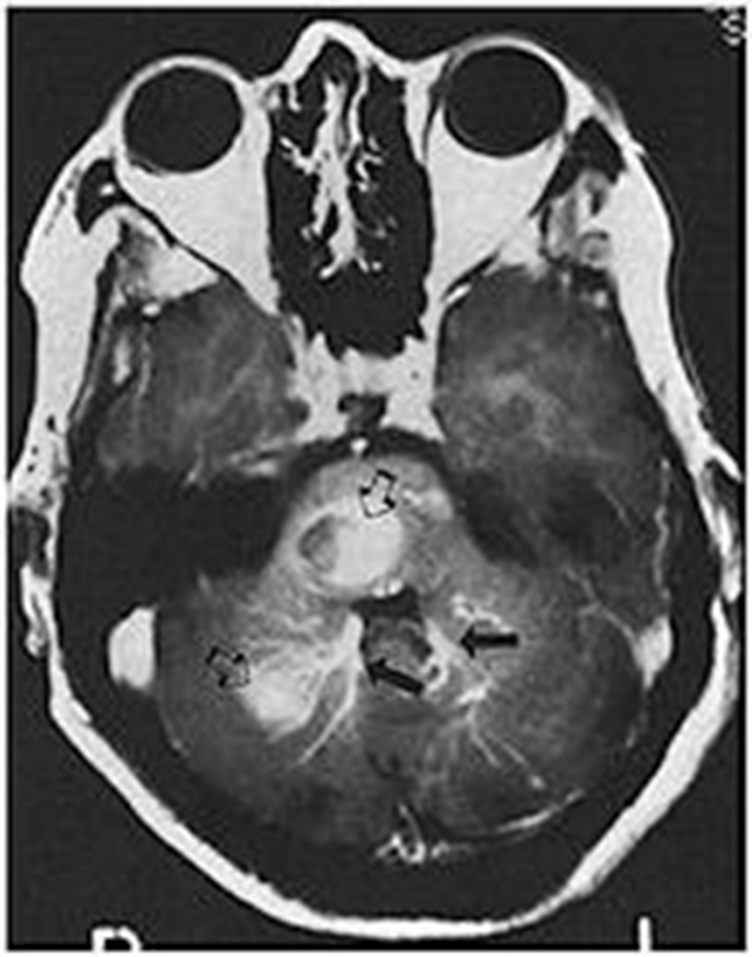
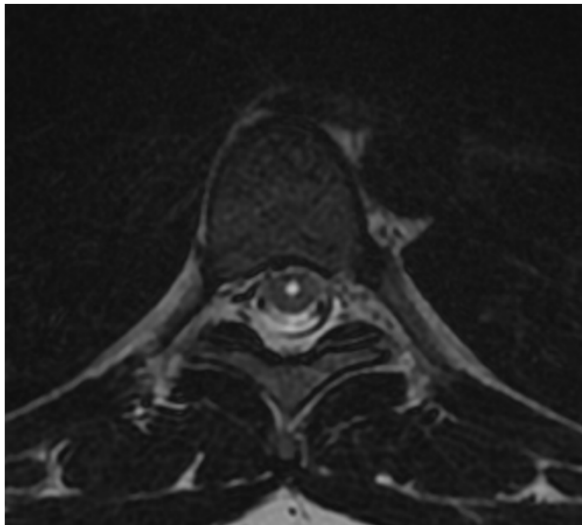
Incidental findings: venous angioma



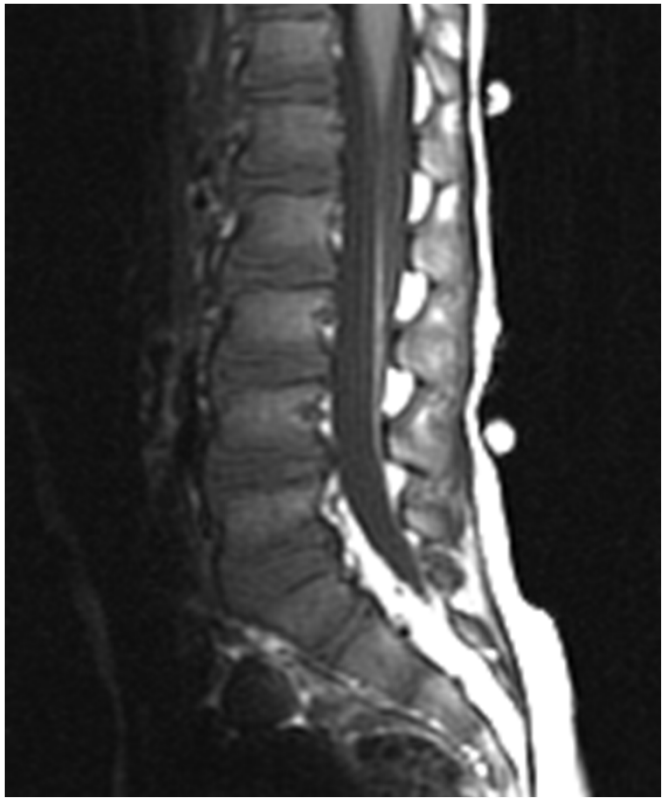
- Spider like “caput medusae” veins seen on MRI/MRA
- Unlikely to bleed
- Represent needed venous drainage pathways
- May be associated with other types of malformations (cavernomas), but do not require treatment.

Incidental findings: Visible Central Canal

- Frequently reported as “syrinx”
- Found in 1.5% of MRI
- Thoracic level in 69% of cases
- Defined as 4mm or less in width
- No treatment or follow-up is necessary; not the cause of pain or scoliosis



Incidental finding: fatty filum

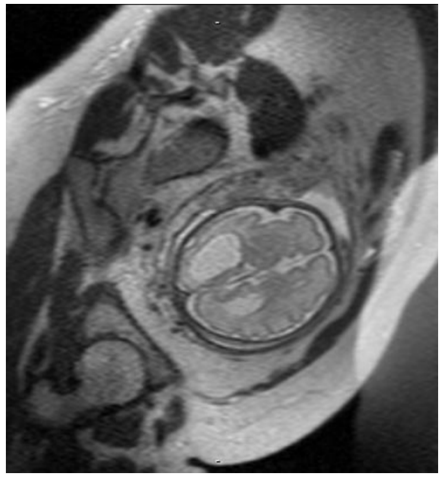


Fat in the filum terminale is not an infrequent occurence, seen in 4 - 6% of individuals , and is especially easily detected on MRI (especially T1

It is usually an incidental finding of no clinical concern.

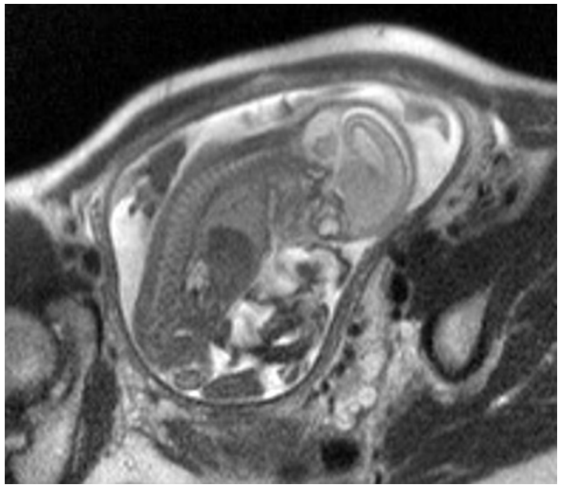
In some individuals however it is associated with spinal dysraphism, thickening of the filum terminale (>3 mm) and tethering of the spinal cord.

Fetal studies



Agenesis of corpus callosum

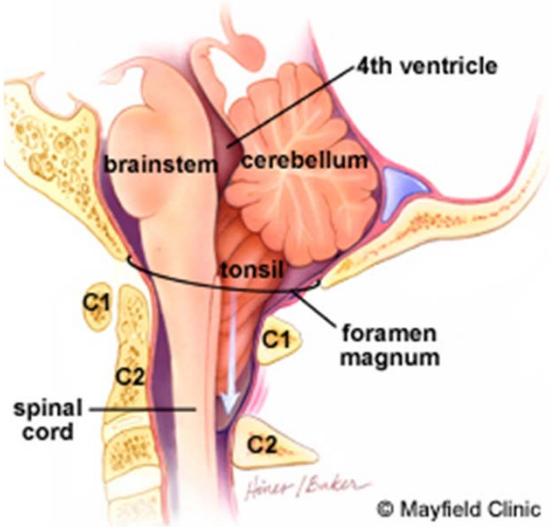
Cerebellar hypoplasia and DWC

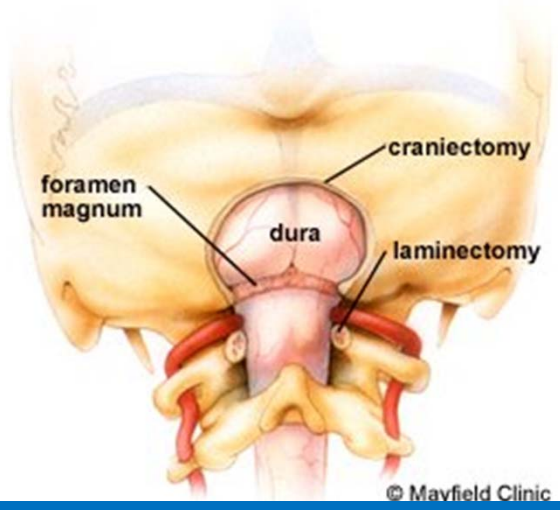


Incidental findings: Chiari I malformation

Posterior Fossa

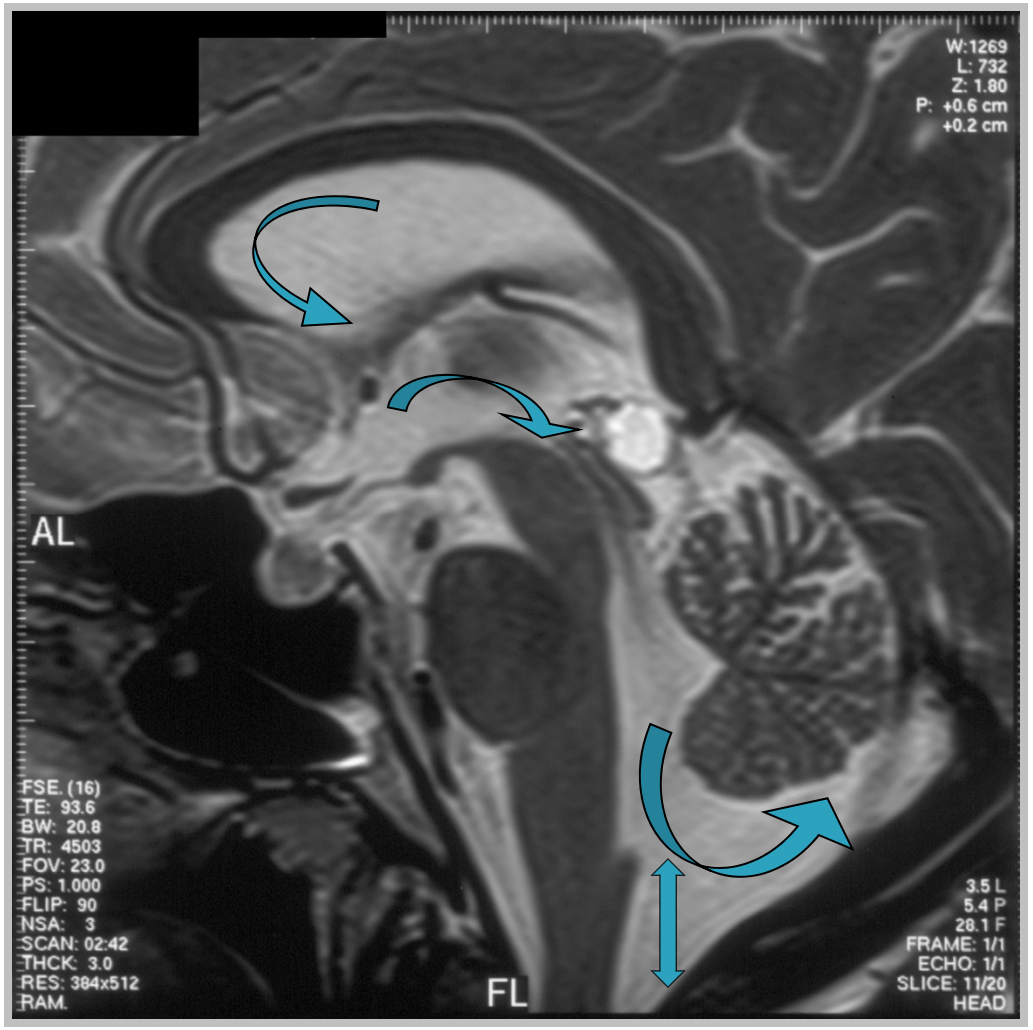
- Subunit of the intracranial compartment
- Non-distensible
- Contents
  - Cerebellum
  - Brain stem & cranial nerves
  - 4th ventricle, cisterns
  - Posterior circulation
- Limits & Borders
  - Superior: tentorium, incisura
  - Inferior: occipital bone, foramen magnum
  - Lateral: occipital bone
  - Anterior: petrous bones, clivus
  - Posterior: occipital bone





CSF Circulation

- Intraventricular
  - Lateral (2)
  - Third
  - Fourth
- Craniospinal
  - Cisterna magna
  - Spinal subarachnoid space

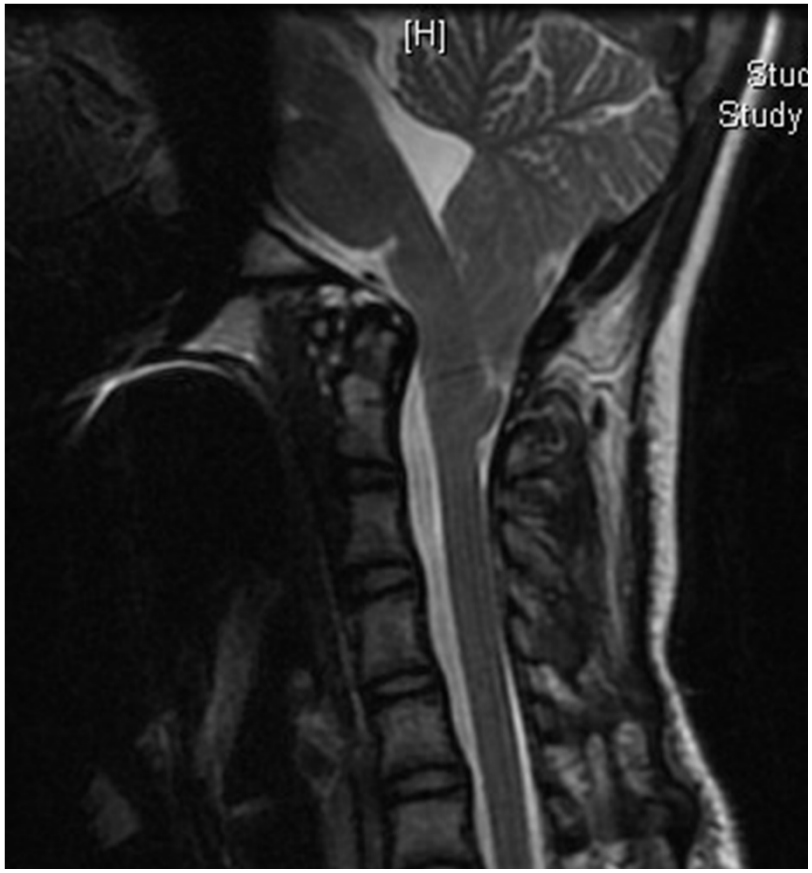


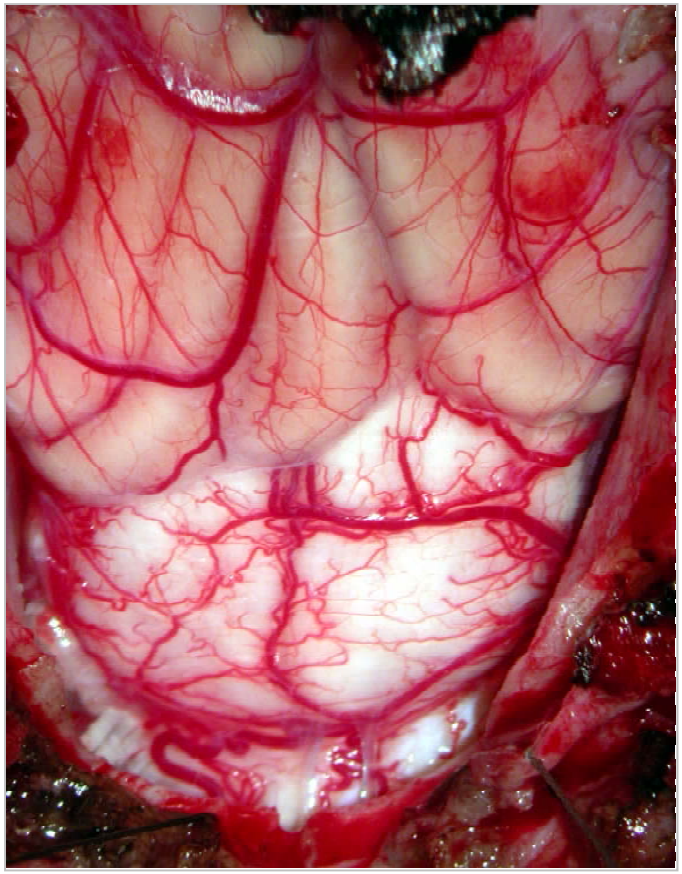
Chiari Malformations

Type	Features
Chiari 0	No descent of rhombencephalon, syringomyelia, reduced PFV
Chiari 1	Descent of cerebellar tonsils ($\geq$ 5 mm), +/− syringomyelia, CV junction abnormalities, infrequent hydrocephalus
Chiari 1.5	Chiari 1 with medullary kinking, descent of the obex, no open neural tube defect (NTD)
Chiari 2	Descent of cerebellar vermis and brain stem, spina bifida aperta, hydrocephalus, composite of multiple CNS anomalies
Chiari 3	Herniation of cerebellum and brain stem into occipital encephalocele, hydrocephalus, cerebral malformations common
Chiari 4	Aplasia/hypoplasia of the cerebellum

Chiari Malformation Type 1.5

Chiari 1 with medullary kinking, descent of the obex, no open neural tube defect

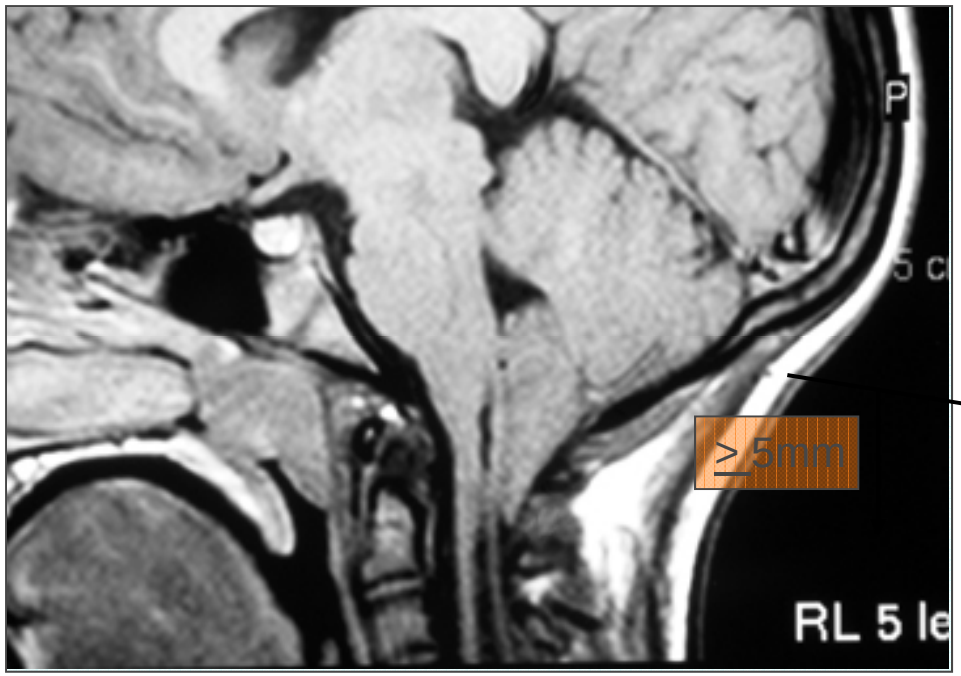


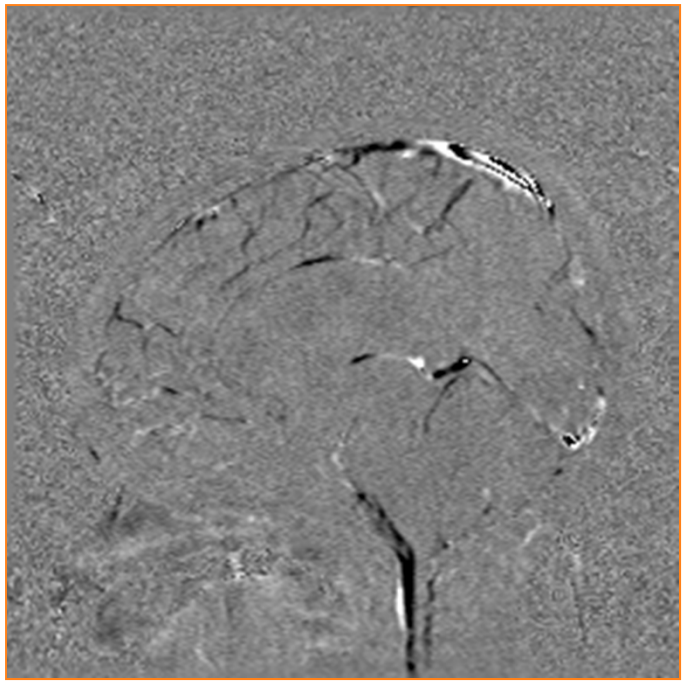


CM-I: Radiologic Findings

Tonsillar herniation below the basion-opisthion line

Tonsillar ectopia > 0 mm and ≤ 3 mm
Borderline > 3 mm and ≤ 5 mm
CM-I ≥ 5 mm





Phase contrast (2D PC) MRI (CINE) imaging for definition of flow surrounding the foramen magnum

Syringomyelia and CM-I

- Incidence, 50-80%
- Frequently spares C1
- Septations are common but seldom compartmentalize
- Result of dissociation between the cranial and spinal subarachnoid spaces

- Primary
  - Chiari: embryonic hydromyelia
- Aquired
  - *Hydrodynamic* theory (Gardner): Persistent opening between 4th ventricle and central canal
  - *Craniospinal Pressure Dissociation*

Presentation of CM-I

- Association of signs and symptoms is critical

- Presenting symptoms
  - Headache/neck pain
  - Paresthesia
  - Scoliosis*
  - Apnea/drop attacks, “cerebellar fits”*
  - Diplopia (oculomotor or abducens dysfunction)*
  - Stridor, dysphagia, hypohonia (lower cranial neuropathy)*
  - Trauma

Headache and Chiari Malformation I

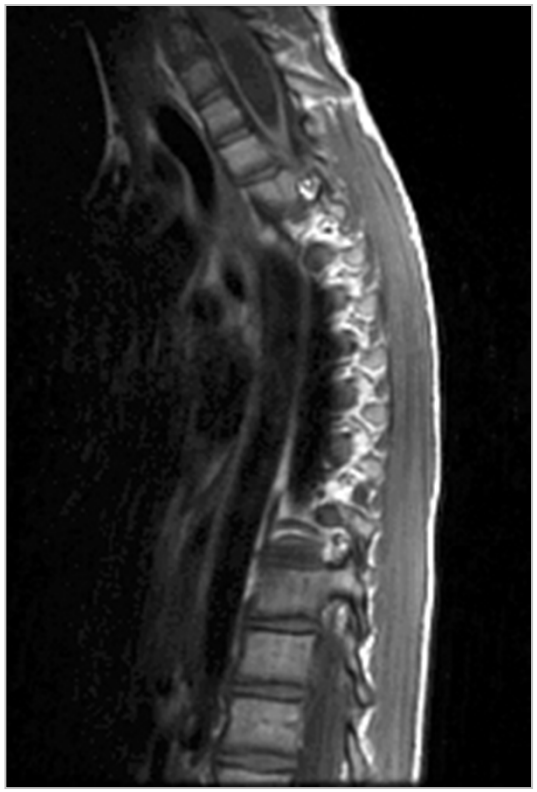
- Most frequent complaint in children being evaluated for CMI
  - Ranges from 15-75%
  - Headache/neck pain in 38% of treated patients[1]

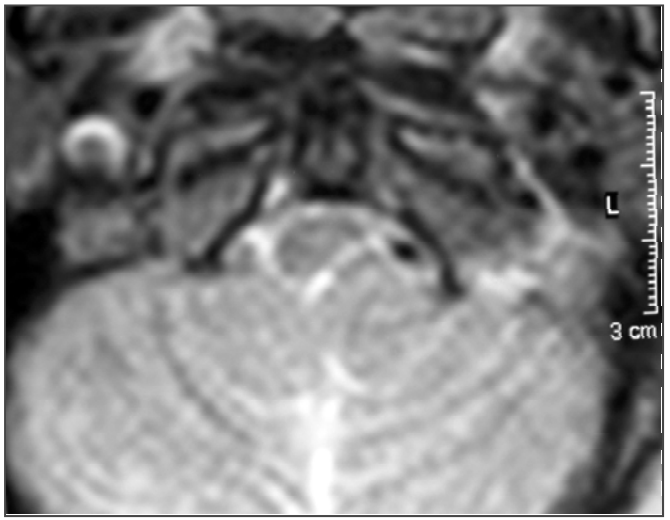
  Manifestations are age dependent
  - Irritability, opisthotonic posturing, and head grabbing in younger age
- Anatomical distribution: Occipital (greater occipital nerve dysesthesia), nape of neck, frontal
- Frequency: Highly variable
- Duration: Transient with spontaneous resolution
- Provocation
  - Raised intracranial pressure (exertional, tussive events, Valsava maneuvers)

CM I and Scoliosis

Possible etiologic factors:

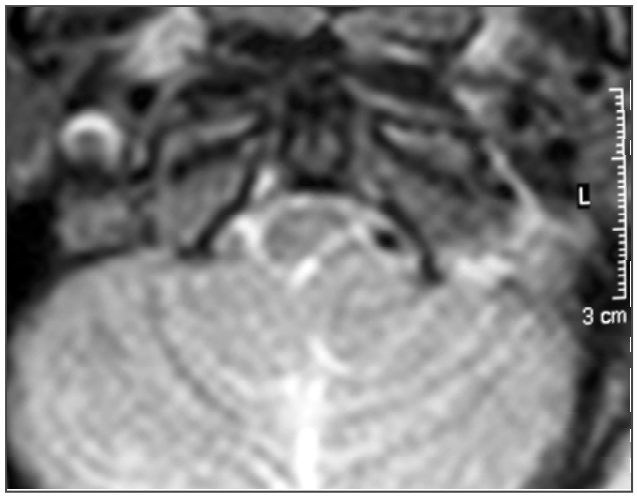
1. Direct interruption of anterior horn cells with syrinx formation
2. Asymmetric tonsillar impaction on cervical spinal cord

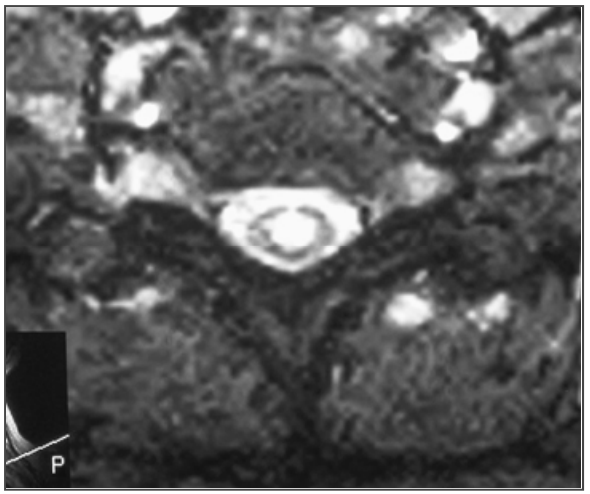




Chiari Malformation Type I: Physical Findings

- Cranial nerves & Brain Stem
  - Lower cranial neuropathy
- Cerebellar
  - Nystagmus
  - Ataxia
- Spinal cord
  - Myelopathic signs (spasticity, hypertonia, clonus, sensory loss)
  - Scoliosis



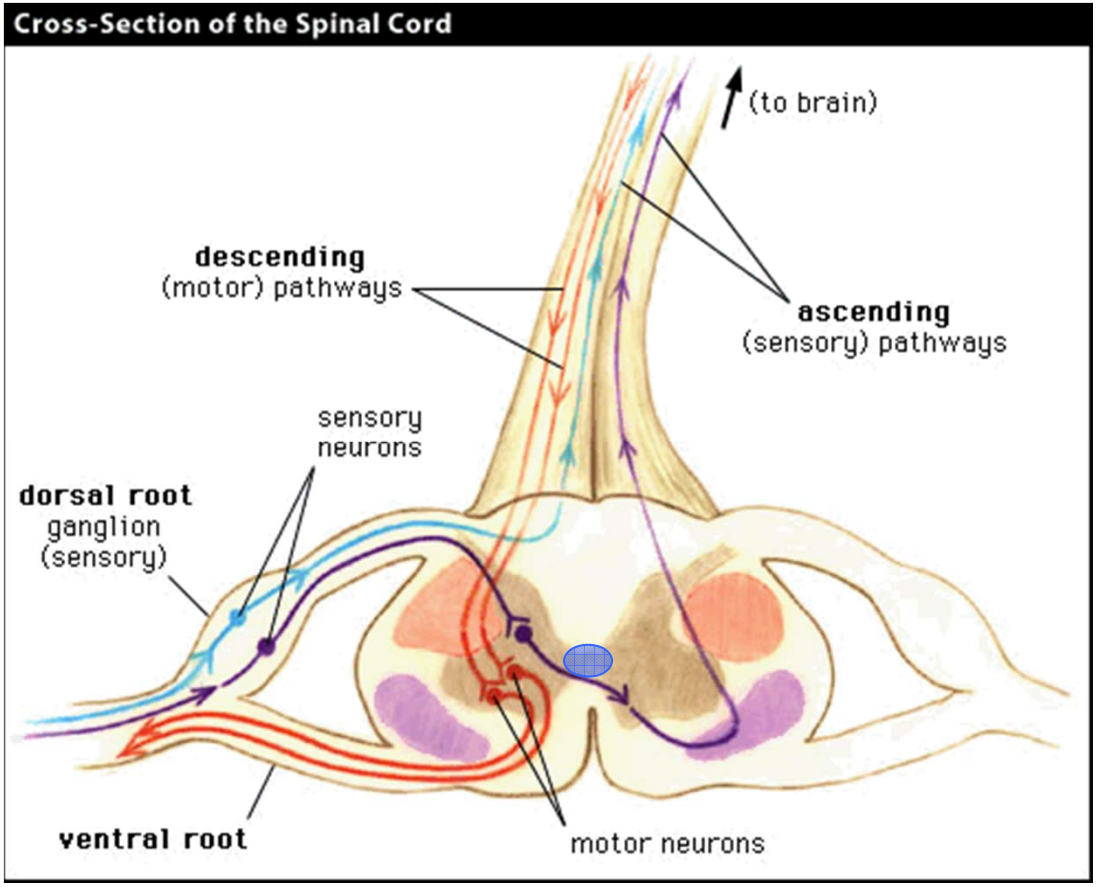


CM-I : Very Young

- Speech disturbance (40%)
  - Delay
  - Dysarthria, hypophonia
- Headache (35%) [1]
  - Head banging
  - Violent outbursts
- G.I. symptoms (20% – 72%)
  - Failure to thrive, gastroesophageal reflux, emesis, aspiration/dysphagia, etc.
  - Constipation (hesitancy to strain)
  - Nissen fundoplication, duodenal biopsy
- Sleep apnea
  - velopharyngeal incompetence, vocal cord paralysis

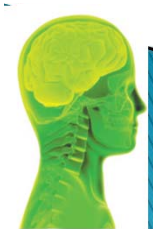
Sensory Disturbance in CM-I

- Distal extremity paresthesiae
- Syrinx formation and *dissociated sensory loss*



Current Clinical Practice

- Questionnaire to neurosurgeons worldwide on the management of CM1 with/without syringomyelia
- 246 questionnaires distributed
- 76 (30.8%) returned
  - Asymptomatic CM1 without syrinx 8%
  - Asymptomatic CM1 with syrinx
    - 2mm 28%
    - 8mm 75%
  - CM1 with syrinx and scoliosis 97%
  - CM1 without syrinx and scoliosis 58%



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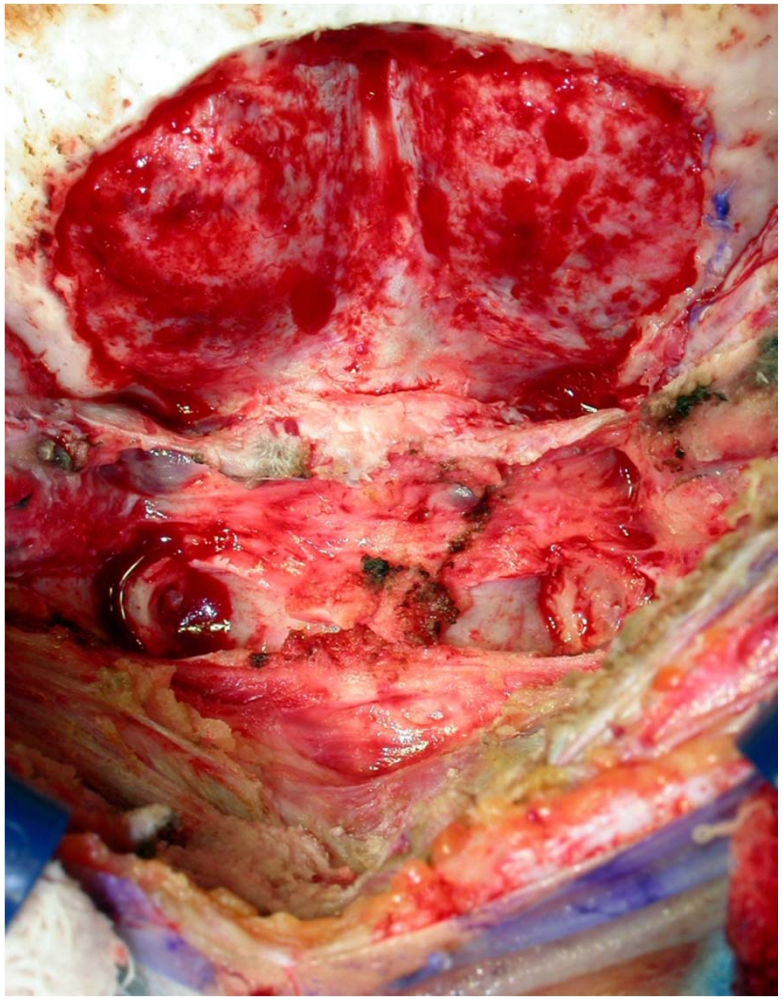
Osseous Decompression

A. Suboccipital Craniectomy

B. Cervical Laminectomy



Surgical Treatment CMI

Postoperative MRI demonstrates restoration of the subarachnoid space and tonsillar ascent and syrinx resolution

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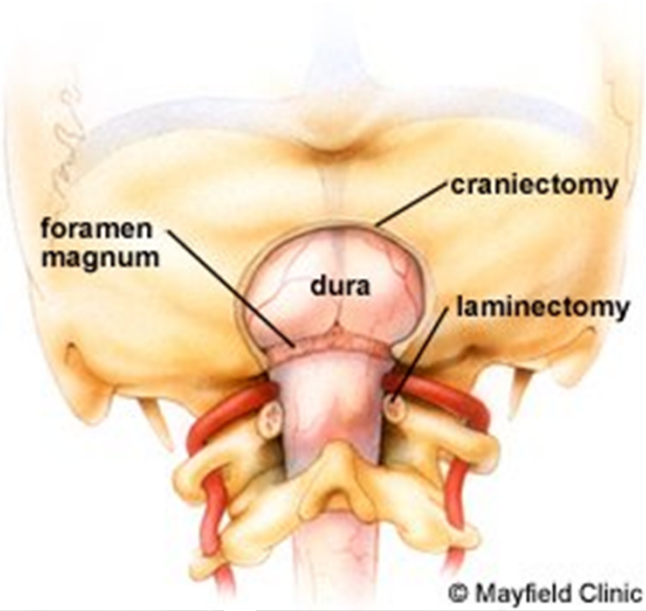
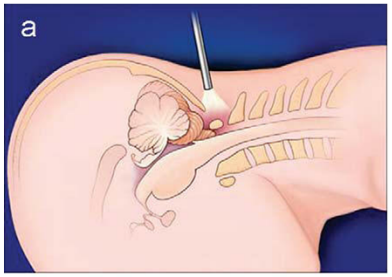
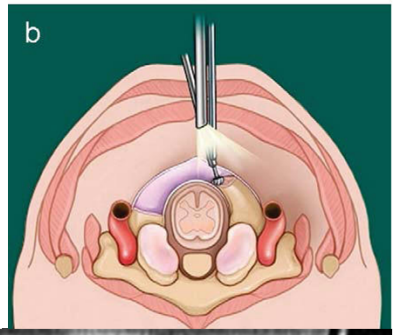
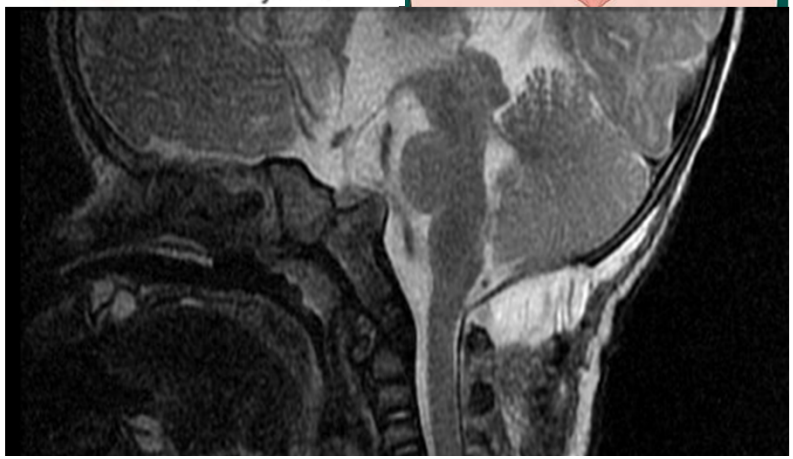
Dural Sparing/Minimally Invasive

- Infants
- Syndromic craniosynotosis
- Achondroplasia
- Obese





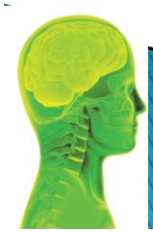




Incidental findings: Chiari I malformation: Who will benefit from surgery?

Chiari Take Home Message

- Most MRI's where CMs are found are in patients who are asymptomatic and *will not require treatment*
- Definition is subjective and it is essential to take the clinical spectrum into consideration
- *Requires a Pediatric Neurosurgical Evaluation*
- Diagnostic workup entails complete neuro-imaging
- Treatment results in resolution of radiographic and clinical symptoms in most cases (85-90%)



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Incidental Findings

- If you are not comfortable discussing the incidence or relevance of a finding or can't be sure if it is causing any symptoms – refer.
- Vast majority do not require follow-up
- It is likely that over-imaging exists
- It is likely that many incidental findings are discovered with unnecessarily ordered imaging
- It is often less than obvious to a primary care practitioner where on this spectrum any given MRI finding may lay.