

Pediatric Neurosurgical Pearls for Primary Care

W. Bruce Cherny, MD

- Sacral dimples
- Craniosynostosis (or not)
- Scalp collections
- Macrocephaly and BEAFI, the **dark side**

Sacral Dimples

- Newborn exam:
 - “There’s a sacral dimple, it’s really deep and I can’t see the bottom”
 - “There’s a dimple. What imaging should I obtain?”
 - “There may be a few hairs too.”
 - “I got an ultrasound and it was ...”
 - “I am trying to avoid doing a sedated MRI.”

Goals

- How to recognize a benign sacral dimple, what to do with it (and what NOT to do with it)
- How to work up lumbosacral lesions of concern
- Achieve confidence in your decisions regarding these



What would you do?

Essential Pearls

Reassure Family

- Dimple over coccyx within intergluteal fold
- Solitary cutaneous hemangioma
- Isolated café-au-lait spot
- Small melanotic nevus
- Slate gray spot (old term Mongolian spot)



Essential Pearls

Hemangioma present - Take a Closer Look

- Careful exam for subcut. masses, dimples, puncta, abnormal intergluteal cleft as these can be assoc. with spina bifida occulta (SBO)
- Look for vertebral deformities, neurologic abnormalities/asymmetries of lower extremities
- As an isolated finding, seem to have no clinical significance unless they are large and confluent



Essential Pearls

Refer to another specialist

- Giant or hairy melanotic nevus
 - Not assoc. with SBO but are assoc. with...
 - Neurocutaneous melanosis: disseminated malignant melanoma throughout leptomeninges, possible hydrocephalus
 - Consult medical genetics, oncology, dermatology and poss. neurosurgery if hydrocephalus present

Essential Pearls

Refer to another specialist

- Multiple café-au-lait spots
 - Hallmark of NF1
 - Can be seen in other congenital & syndromic conditions
 - Medical genetics, peds neurology first; poss. ophthal., oncology, occ. peds neurosurgery

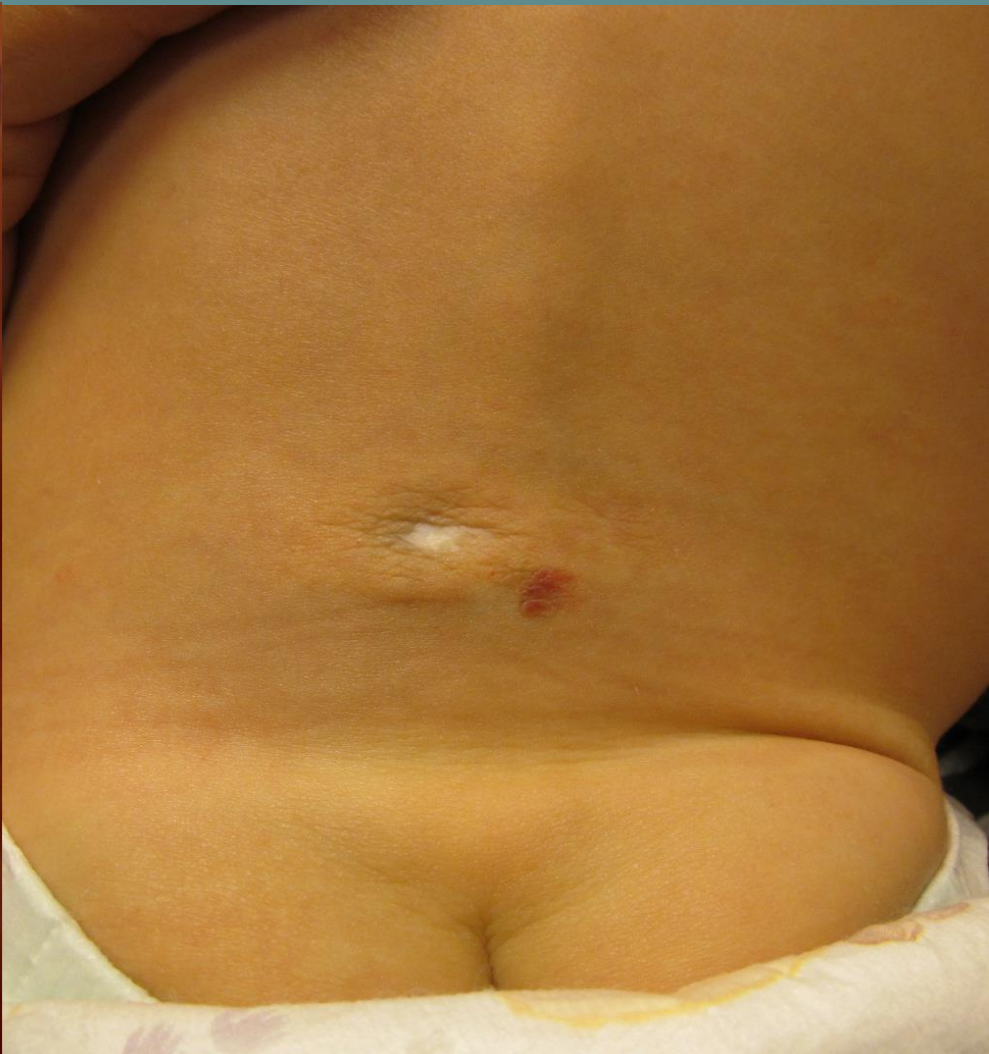
Essential Pearls

Image or refer to peds neurosurgery

- Dimple over lumbar spine visible ABOVE the intergluteal fold, without spreading the buttock
- Sacral clefts with dimples
- Midline or paraspinal subcut. lipoma
- Midline, subcut, fluid filled cyst
- Hairy patch over thoracic or lumbar spine
- Circumscribed atrophic skin “cigarette burn”







Benign Sacral Dimples

(sacrococcygeal pit)



Benign Sacral Dimples

(sacrococcygeal pit)



Benign Sacral Dimples

(sacrococcygeal pit)



- Within the intergluteal fold
- Palpates to the level of the coccyx
- “Tracks” downwards with skin tenting maneuver
- Completely lined with epidermis
- No assoc. with intraspinal pathology
- No imaging needed, doesn't matter how deep;
resist the urge!!
- No surgery needed unless recurrent infection
- Can be familial

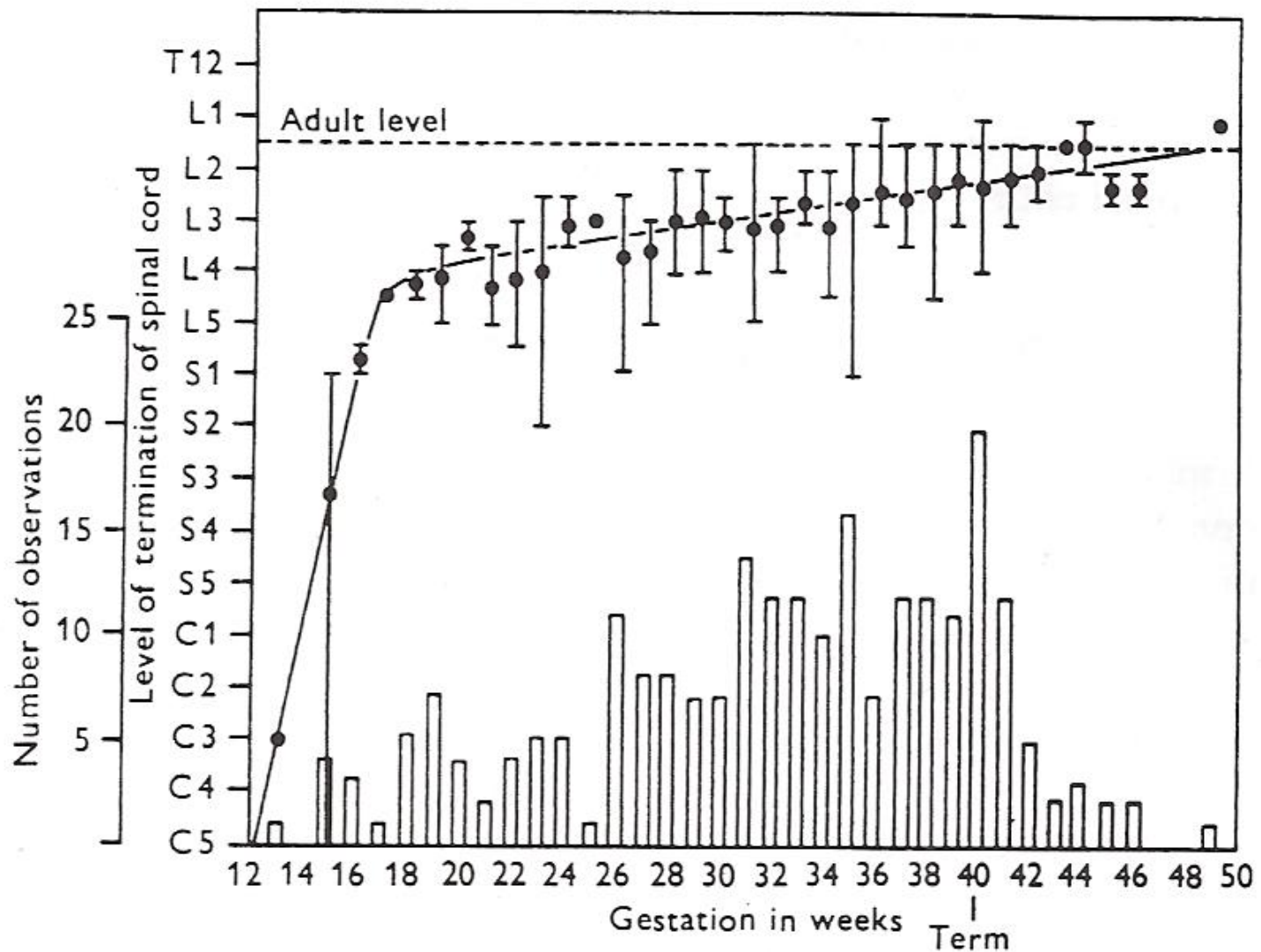


Figure 17-4. Results of an autopsy study of the position of the conus in asymptomatic infants and fetuses. The position of the conus is plotted against the gestation and age. (Adapted with permission.¹⁵)

Level of Conus with Age

- Final position achieved by 3 mos of age
 - Barson AJ: Journal of Anatomy 106:489-497, 1970

Borderline low conus medullaris on infant lumbar sonography: what is the clinical outcome and the role of neuroimaging follow-up?

Neil H. Thakur · Lisa H. Lowe

- 748 consecutive spinal sonograms seeking to identify infants with conus terminating between L2-L3 disc space and mid-L3 level without other findings of tethered cord.
- Indications for the initial lumbar spine sonograms included sacral dimple, hemangioma or tuft of hair in the majority of infants
- Isolated borderline low conus was found in 90 of 748 infants (12%) on sonography. Follow-up imaging in 11 children (10 MRI, 1 sonogram), showed change in conus position to “normal” level in 10, no change in 1, and no new findings within lumbar spine. Clinical follow-up was available in 50 children, with normal motor milestones met in all.

ORIGINAL ARTICLE

Borderline low conus medullaris on infant lumbar sonography: what is the clinical outcome and the role of neuroimaging follow-up?

Neil H. Thakur · Lisa H. Lowe

- Initial sonogram was performed on day of life 14 showing conus at L2-L3 disc space level, with repeat sonogram performed at 6.6 months showing change in conus position to mid L2 vertebra level
- Follow-up MRI demonstrated change in conus position to a normal level above the L2-L3 disc space in 9 of 10 cases

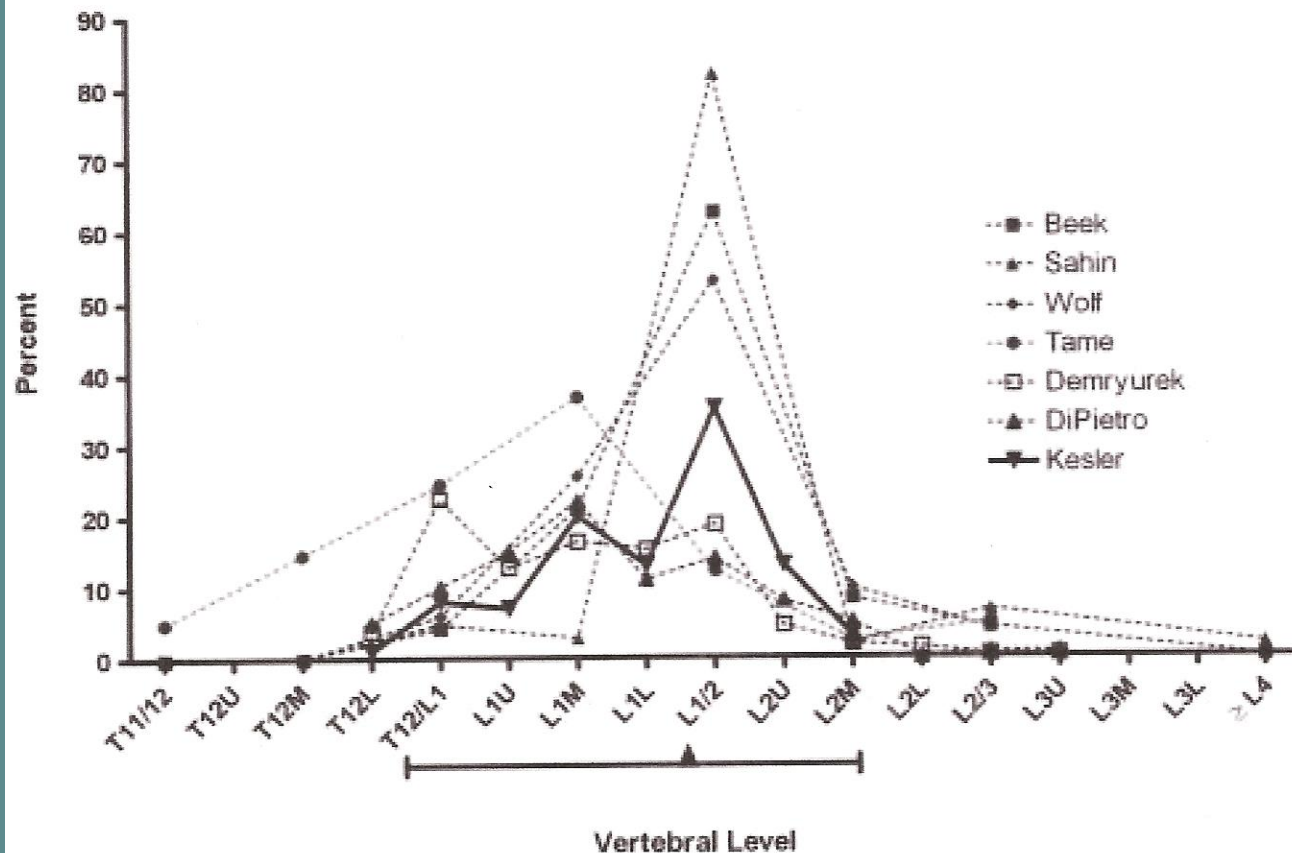
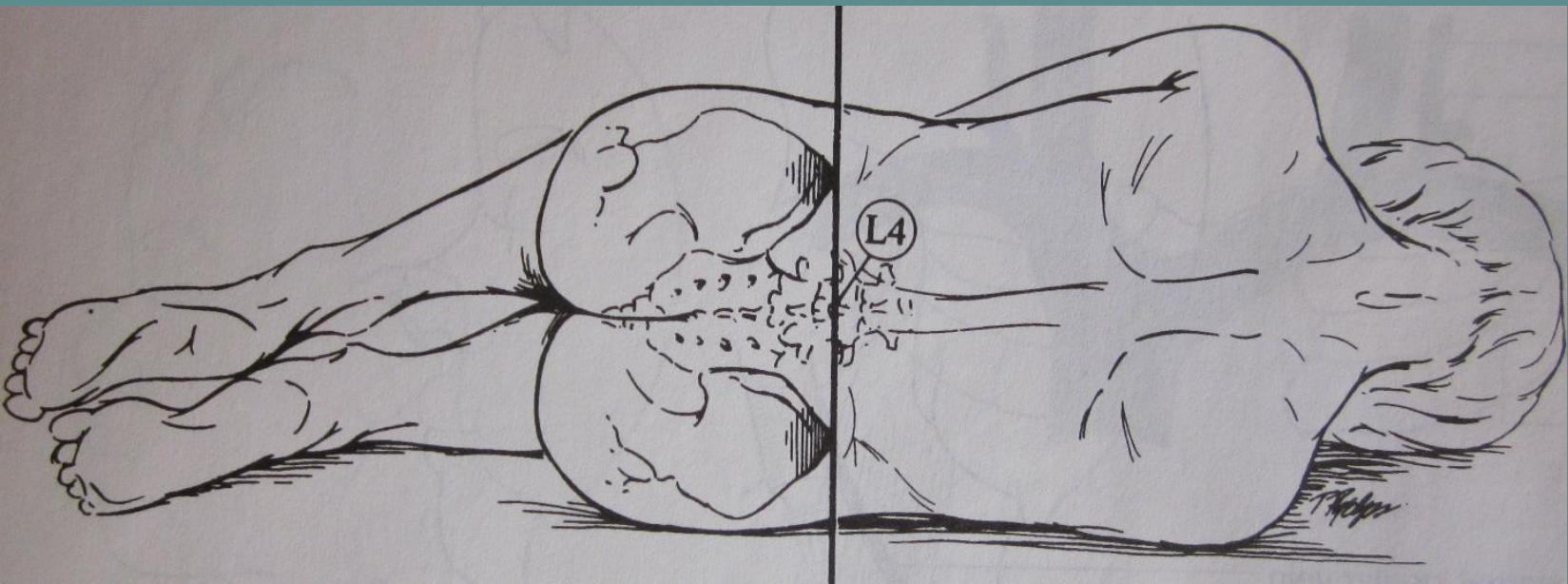


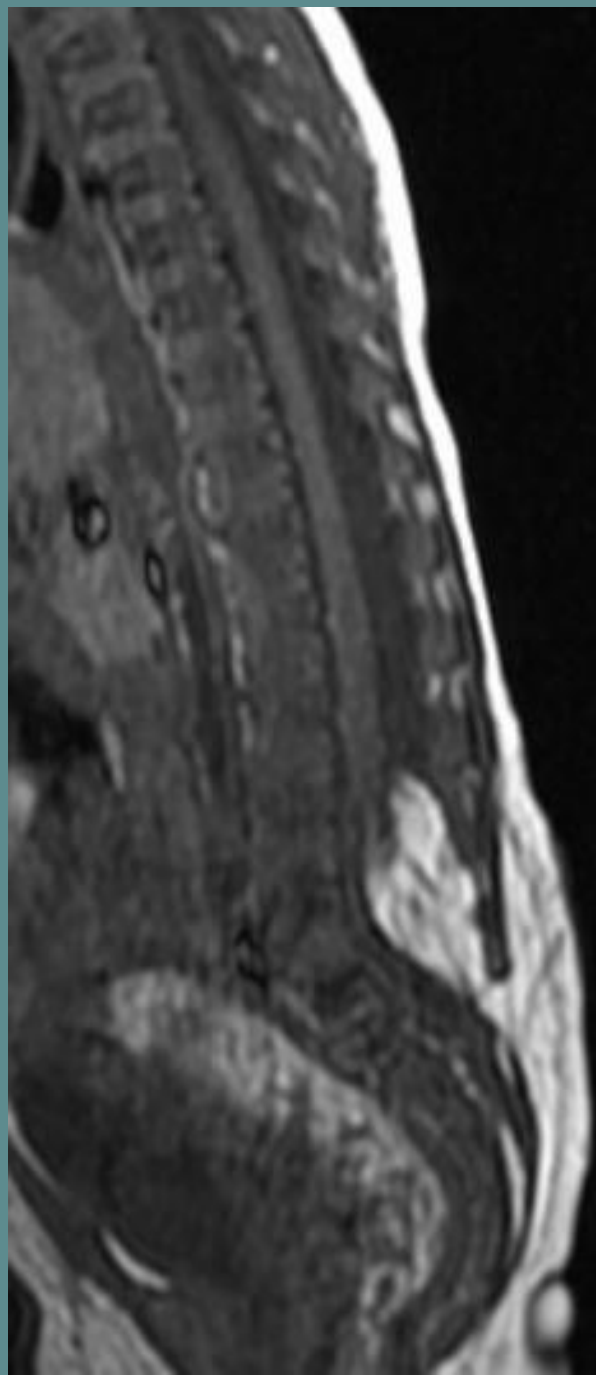
FIG. 4. Distribution of CM termination levels in published studies. Note the presence of the “tail” on the right side of the graph (representing findings caudal to the L1/2 disc space) in previous studies and the absence of such findings in the present study. The overall median value and 95% CI are indicated by the pointer and line located below the abscissa of the graph.

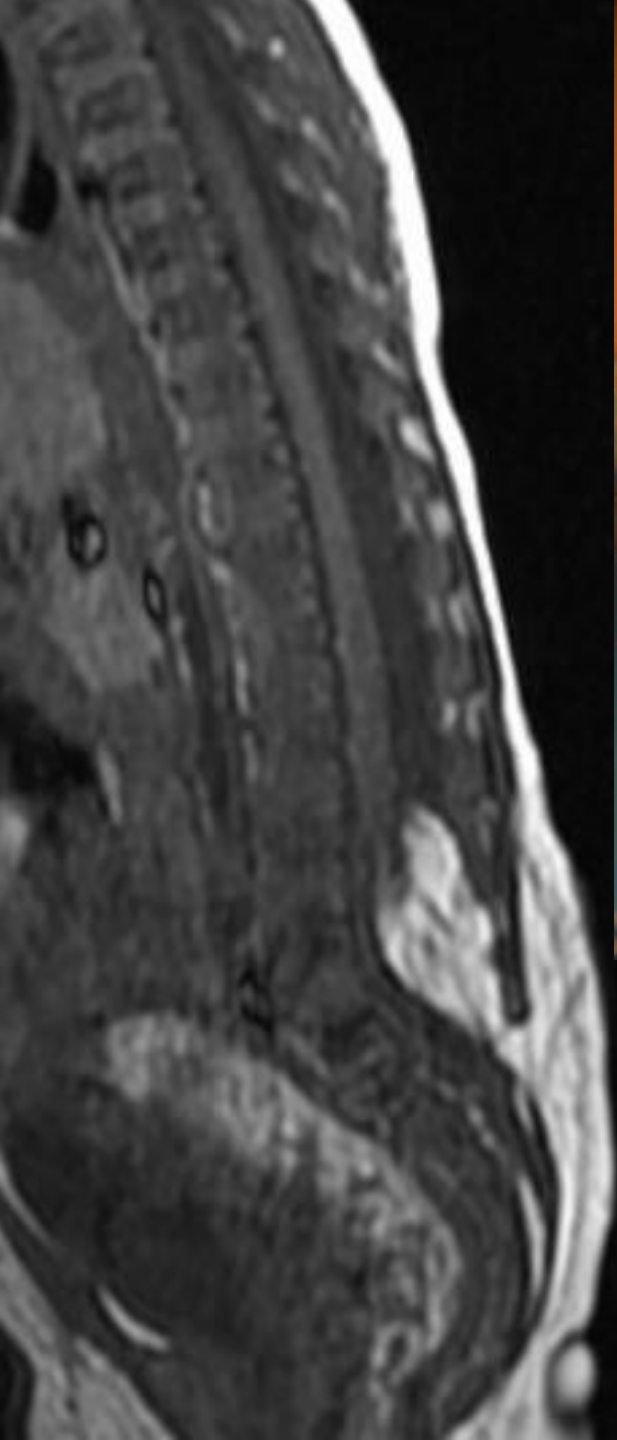
*Level of termination of the CM reported in previous imaging studies**

Author(s) & Year	Population	Imaging Modality	Average	Range
Wilson et al., 1989	184 infants	MRI	L1/2	T12-L2/3
Hill & Gibson, 1994	103 infants	ultrasound	L2U	T12/L1-L3M
Wolfe et al., 1992	114 infants	ultrasound	L1/2	T12-L4
Sahin et al., 1997	64 infants	ultrasound	L1/2	T12/L1-L2/3
Tame & Burstal, 2003	45 children	MRI	L1	T11/12-L2/3
DiPietro, 1993	161 children	ultrasound	L1	T10/11-L3U
Demiryurek et al., 2002	639 adults	MRI	L1M	T11/12-L3U
Lee et al., 2004	210 adults	MRI	L1L	T12/L1-L2U
Saifuddin et al., 1998	504 adults	MRI	L1L	T12M-L3U

* The vertebral terminology used in this paper is explained in Materials and Methods. Abbreviations: U = upper; M = middle; L = lower.









Ultrasonographic screening in infants with isolated spinal strawberry nevi

J Neurosurg (Spine 3) 98:247–250, 2003

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AND NATHAN R. W. SELDEN, M.D., PH.D.

Division of Pediatric Neurosurgery, Department of Neurological Surgery, Oregon Health & Science University and Doernbecher Children's Hospital, Portland, Oregon; and Neurological Surgery and Pediatrics, MCP Hahnemann University and Section of Neurosurgery, St. Christopher's Hospital for Children, Philadelphia, Pennsylvania

- 20 pts: 15 isolated, 5 had addl. stigmata
- All 15 “isolated” had normal U/S; 3 of these also had a benign coccygeal dimple
- 4 of 5 with other stigmata were abnormal
- When both U/S & MRI done, U/S had 40% false negative rate (2/5 pts)

Use of lumbar ultrasonography to detect occult spinal dysraphism

J Neurosurg Pediatrics 9:274–279, 2012

Clinical article

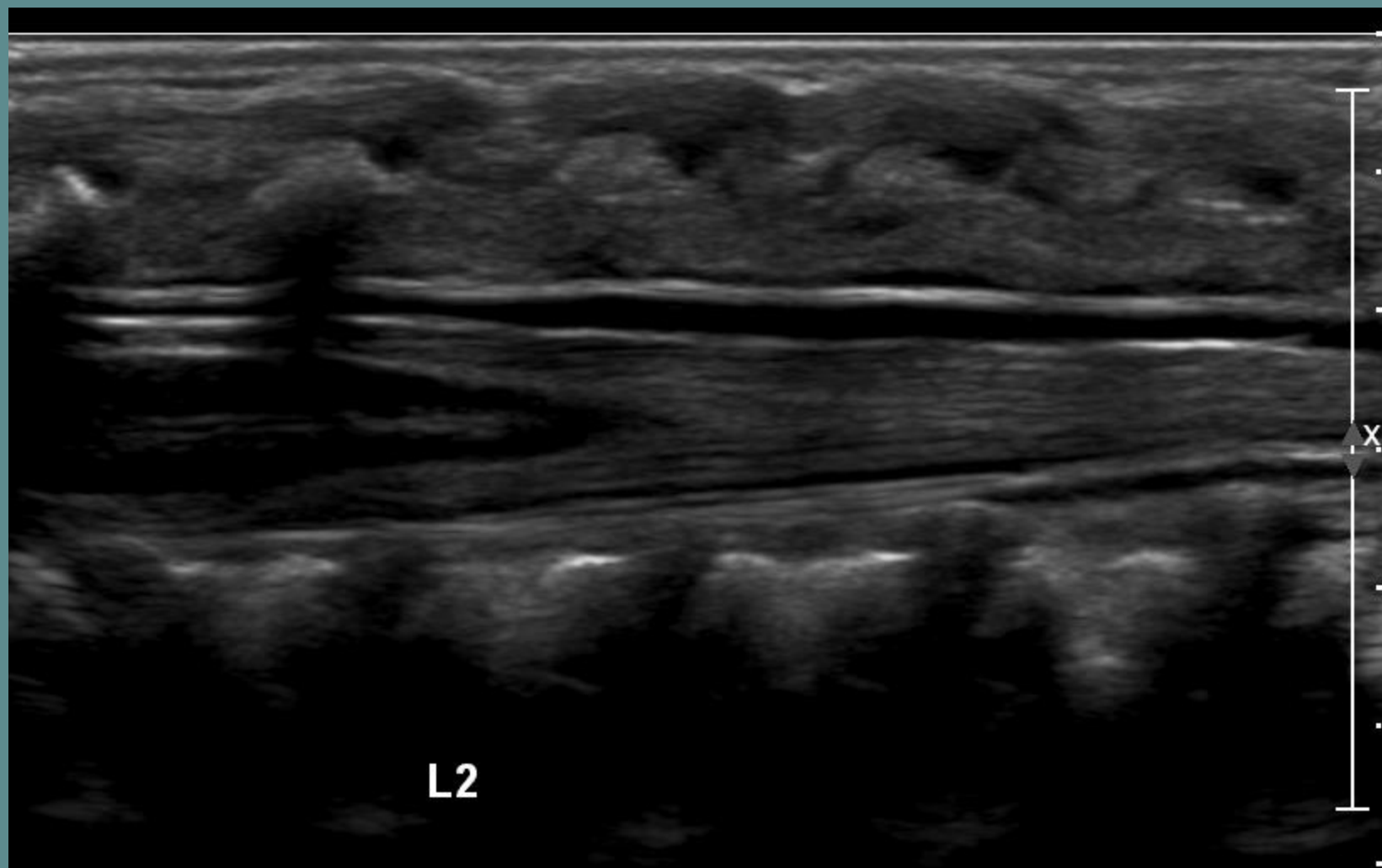
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- 1273 pts over 5 years (~250/yr!!)
- 943 with cutaneous stigmata only
- Considered a conus position below L2-3 as abnl
- 68% imaged for sacral dimple; 9% hemangioma
- Cutaneous stigmata only: 6% abnl U/S (58 pts)
- All 58 underwent MRI revealing that ...
- U/S had 48% false positive rate

St Luke's Boise 2013

- 18 of 21 spinal U/S performed for sacral dimple



FINDINGS:

Spinal cord terminates at the L2-L3 level and there is minimal movement of the cauda during quiet respiration, most consistent with a tethered cord. Nerve roots of the cauda equina are normally distributed in the thecal sac. The dimple is at the level of the coccyx and there is no evidence of a sinus tract. No evidence of intraspinal mass.

CONCLUSION:

1. Findings most consistent with a tethered cord with the conus terminating at L2-L3.
2. Otherwise, unremarkable spinal canal ultrasound with no evidence of an intraspinal mass.

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***** ADDENDUM #1 *****

Addendum: The conus terminating at L2-L3 in a newborn is at the lower range of normal.

Recent U/S on a 3 yo

- **FINDINGS:** Evaluation of the spinal canal in this patient is limited due to advanced osseous development. The spinal cord appears to terminate at approximately L2 which is borderline low lying. There are is a thin echogenic linear focus extending from the L2 region to the sacrum within the canal suspicious for a thickened filum. Evaluation of the canal is otherwise limited by ultrasound. This warrants better characterization with MRI.
- **IMPRESSION: SIGNIFICANTLY LIMITED EXAM DUE TO OSSEOUS DEVELOPMENT AND FOR VISUALIZATION OF THE SPINAL CANAL. SOMEWHAT LIMITED FINDINGS ARE SUSPICIOUS FOR TETHERED CORD IN THIS WARRANTS FURTHER EVALUATION WITH LUMBAR SPINE MRI.**

Imaging - Ultrasound

- Role is unclear (in my opinion)
 - My rec: use rarely if ever
- Very operator dependent
- High false positive rate; potentially high false negative rate as well
- Can only visualize well in newborns & infants
- Can potentially evaluate cord motion
- Can look at conus level
- But that doesn't tell you if it's tethered because tethering is a *clinical* diagnosis



So now, what would you do?



Nothing...Location, location, location

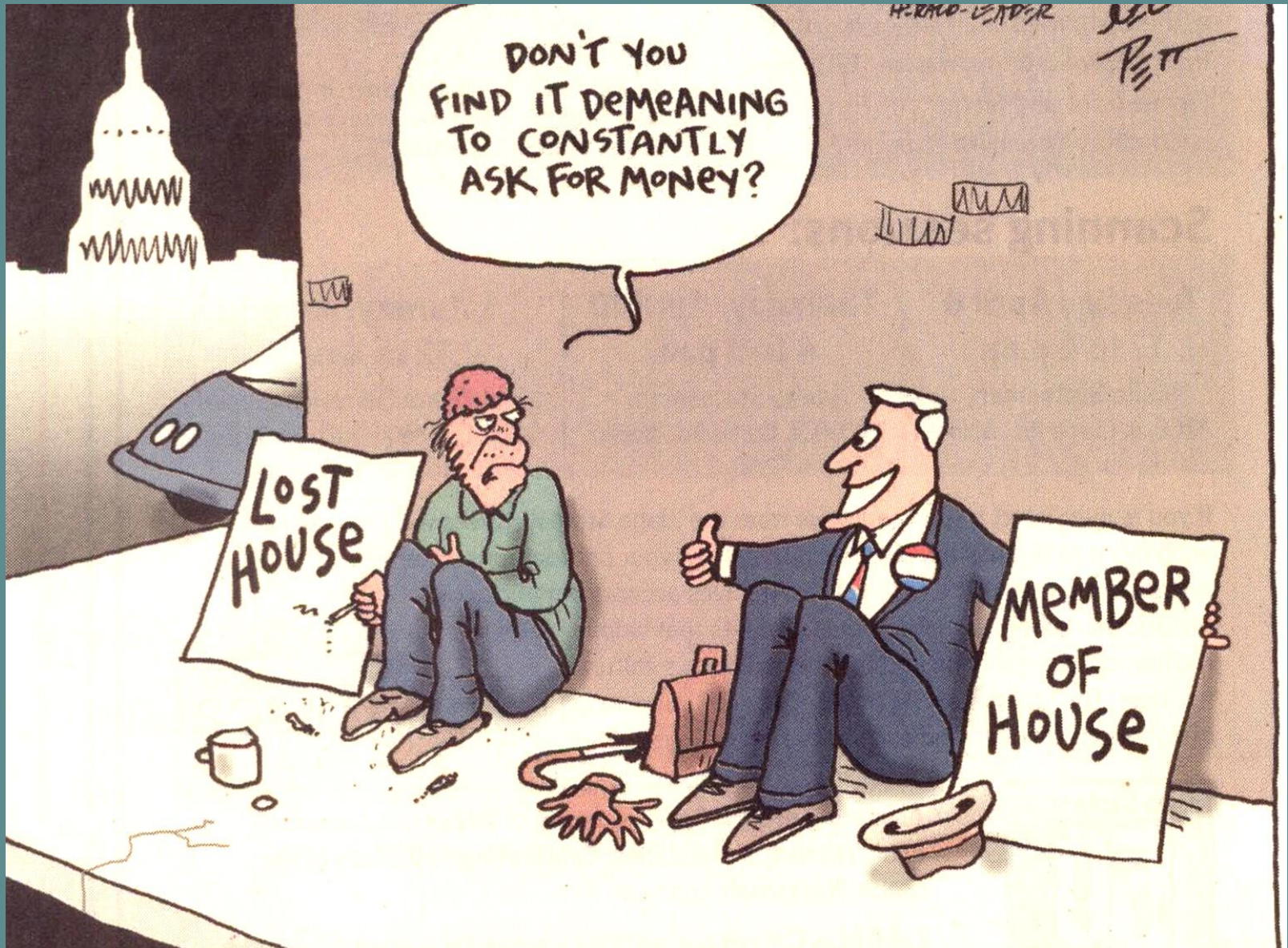
Essential Pearls

Imaging

- Newborn U/S: I have not found it to be particularly useful & would like to discourage its use (*esp. in sacral dimples*)
- There is no rush to image a normal exam
- MRI w/o contrast is the modality of choice.
- Best done after 3 months of age, after 6 months is even better

Take Home Message

- Don't image benign sacral dimples
- Don't image isolated hemangiomas unless large and confluent
- Don't send to these neurosurgery (or anyone)
- Don't worry that you are missing something, you're NOT
- You'll know when to image anatomically
- You'll know when to image clinically
- If imaging is needed, it'll be an MRI, but later most likely
- The lower limit of normal for the conus is probably L2-3
- Tethering is a **clinical not anatomical** diagnosis
- Call us for questions



DON'T YOU
FIND IT DemeanING
TO CONSTANTLY
ASK FOR MONEY?

Lost
House

MEMBER
OF
HOUSE

HERROD-ANDERSON

Pett

Craniosynostosis (or not)

On a newborn or infant exam:

- “I no longer can (or never could) feel the fontanel” “It’s closed”
- “This child’s head is small”
- “I feel a ridge”
- “The sutures are overlapping.”

And then the parent says:

- “Well he looks just like . . .”

Craniosynostosis (or not)

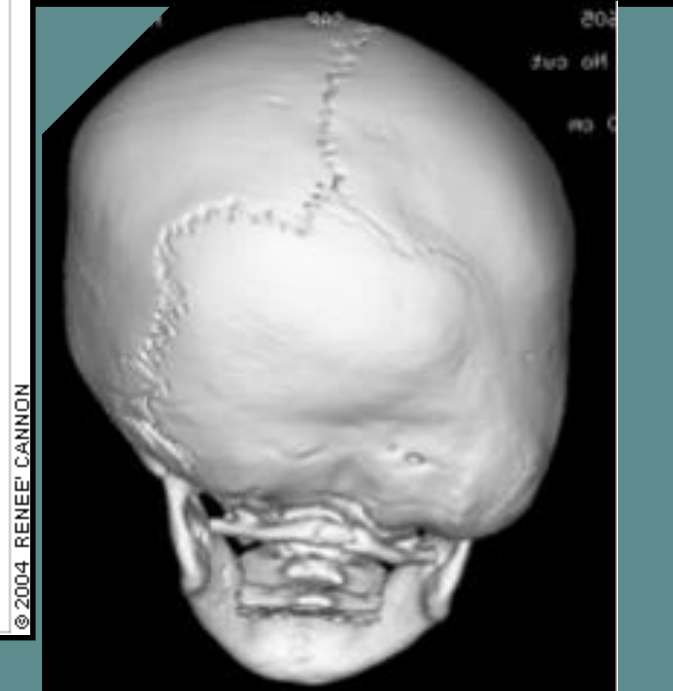
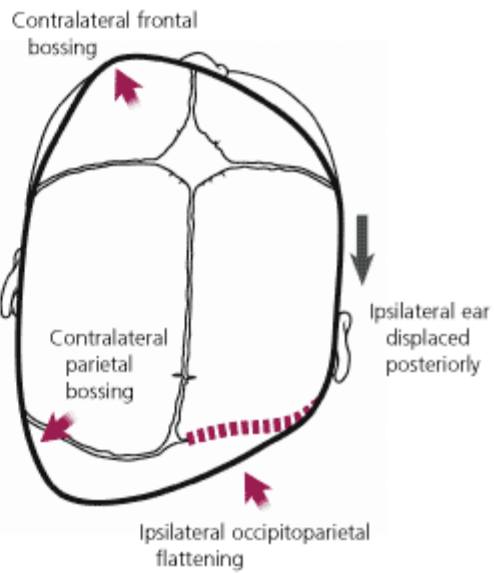
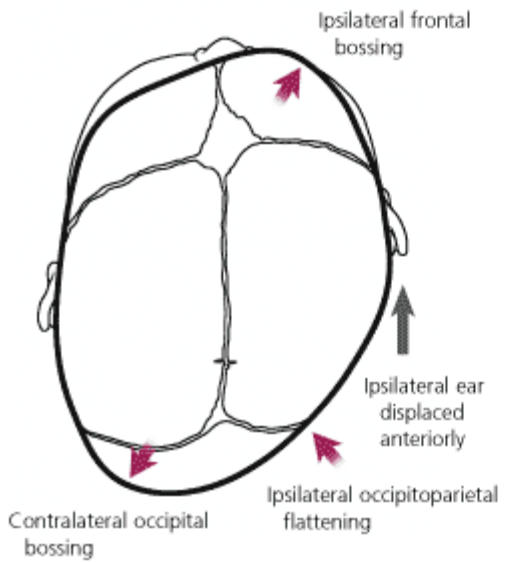
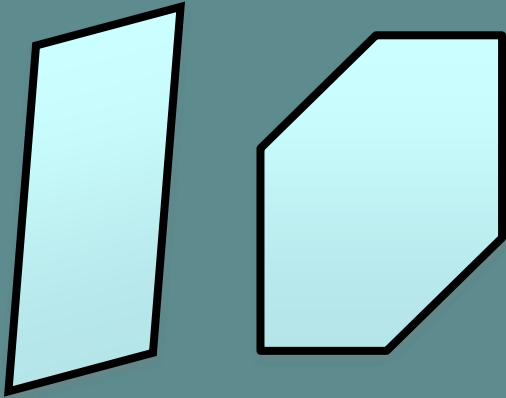
- Never allows for a normal shape of skull
- Creates classic skull shape abnormalities that differentiate it from positional plagiocephaly

Thus:

- Palpably absent fontanel with normal shape is not synostosis
- Small head with normal shape is not synostosis
- Small (or “absent”) fontanel by itself has no significance
- Ridging/elevation of any suture with otherwise normal shape is benign and self-limited; it’s where the highest rate of bone deposition occurs; metopic is most common
- The sole presence of a symmetrical occipital shelf/protuberance is a benign normal variant

Positional Plagiocephaly "rhomboid"

Lambdoid Synostosis "trapezoid"



The Anterior Fontanel

- The “nl” age of closure is highly variable, 3-24 mos (median 13.8 mos)
- Most “palpably absent” fontanelles are not actually closed; physiologically open
- A small or closed fontanel does not result in microcephaly; but, the opposite may be occur (microcephaly \longrightarrow closed fontanel)

MIKE
PETERS

HERE,
GRIMMY

WAIT, IS THIS
VEGETARIAN?

YES

GLUTEN-
FREE?

UH-
HUH

HIGH-FRUCTOSE
CORN SYRUP?

NO

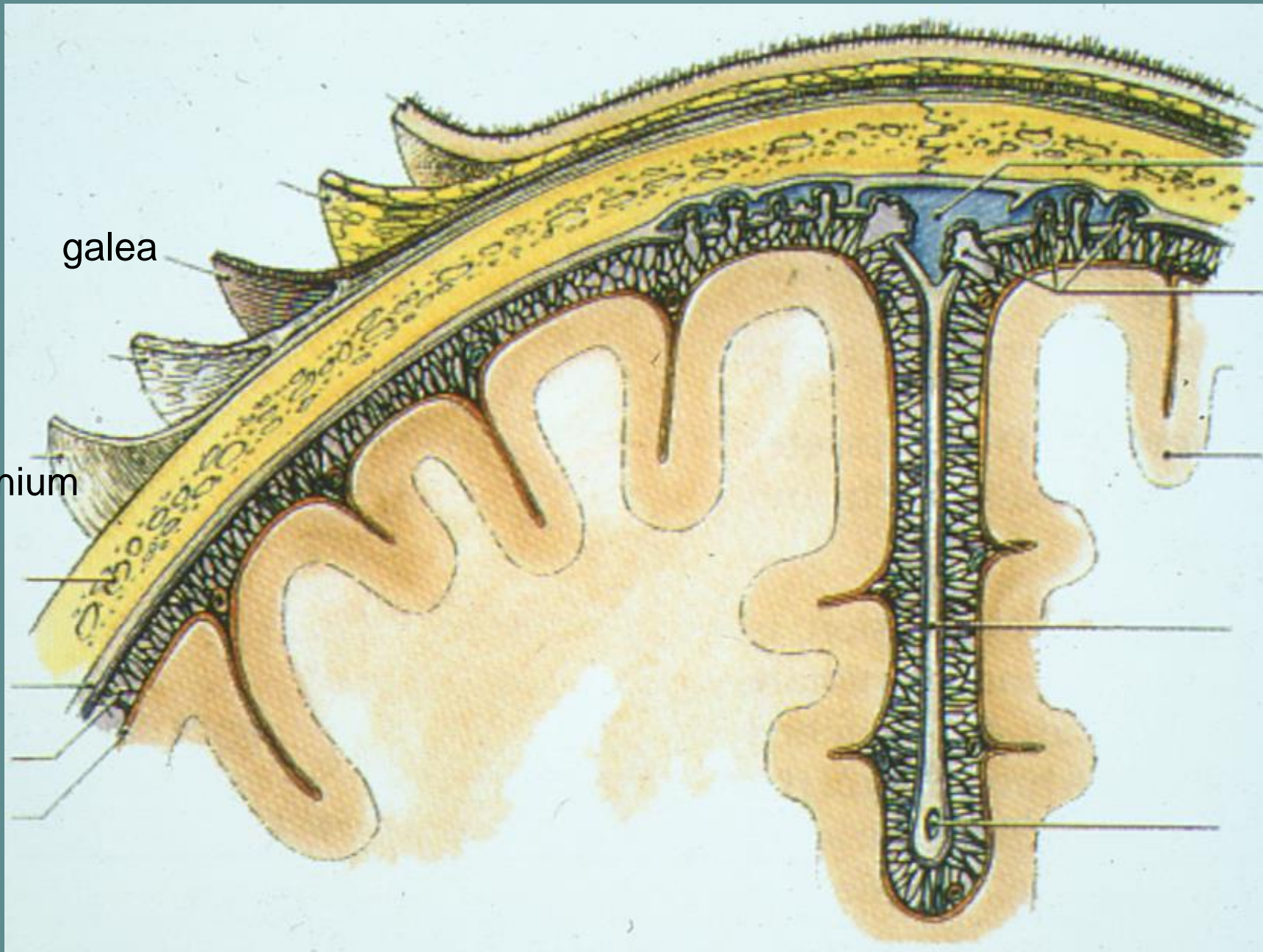
LACTOSE-
FREE?

MADE IN A
FACTORY THAT
PROCESSES
PEANUTS?

IT'S
WATER,
YOU IDIOT

Scalp Collections

- Cephalohematomas
- Subgaleal hematomas
- Scalp hematoma
- Newborn office follow-up exam:
 - “This is the largest one I’ve ever seen”
 - “This one crosses midline”
 - “It started go away and then it became hard”
 - “It just feels funny”
 - “The parents are anxious”



galea

pericranium

Cephalohematomas

- You are actually the experts here, not me; you see many more of them than I do
- There are no references, sections or paragraphs regarding these in either of the 2 most respected pediatric neurosurgery texts or the 2 general neurosurgery texts; I have no magical insight or special knowledge
- They never need surgery or drainage
- May calcify
- Remodel & absorb into growing skull as expands with age



- They can be big, but they do not require a neurosurgical consultation
- Reassure the parents
- Though reported in literature, I have not seen this assoc. w/ subsequent growing fx



Subgaleal Hematomas/Fluid

- Resorbable collections beneath galea and above pericranium in loose connective tissue layer; self-limited & resolve
- Can cross sutures and midline
- Trauma, birth, surgery with scalp reflection
- Can migrate with gravity, dependent



Scalp Hematoma

- Contused skin layers of scalp
- Often generalized to include subgaleal variety
- Self-resolve, no surgery, rarely necrose

Infantile CSF Leak into Scalp

- Rare event, etiology unknown
- Assoc w/ macrocephaly (BEAFI), suction or forceps delivery, wormian bones
- Usually begins 1-3 months after birth and after any prior scalp collections have resolved
- Can enlarge for several weeks once starts to collect
- Classically at vertex and midline
- Often full but never tense; fluid rests dependently
- Ballotable, a tap creates a wave/ripple
- Asymptomatic
- Self-resolve over 2-6 weeks and never need surgery; they do not come back

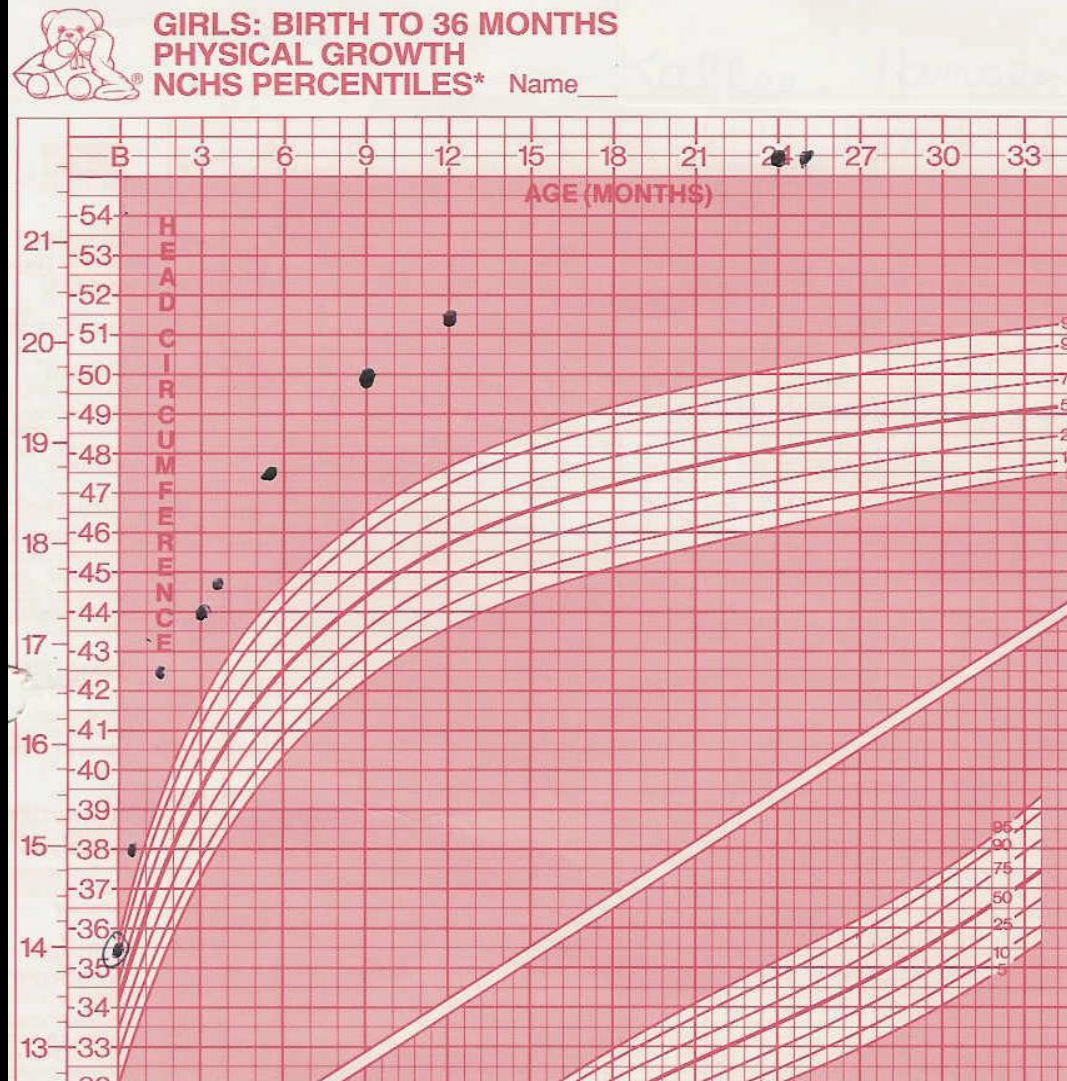
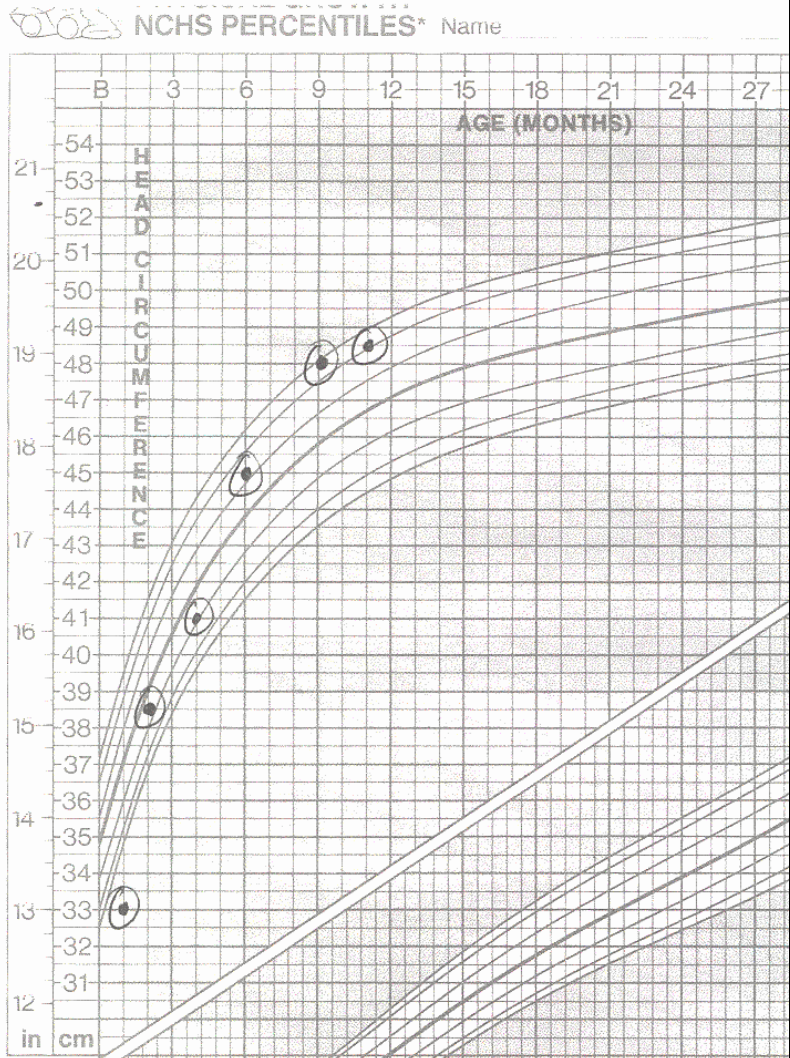


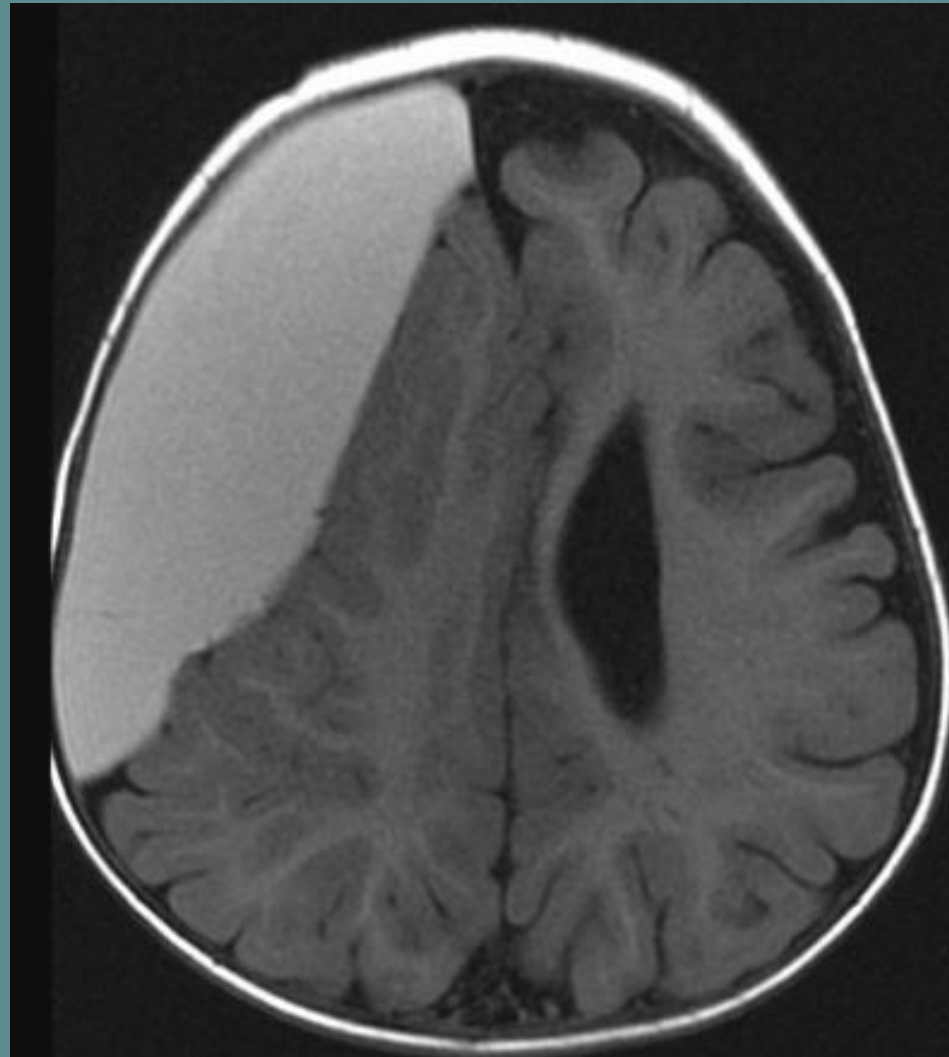
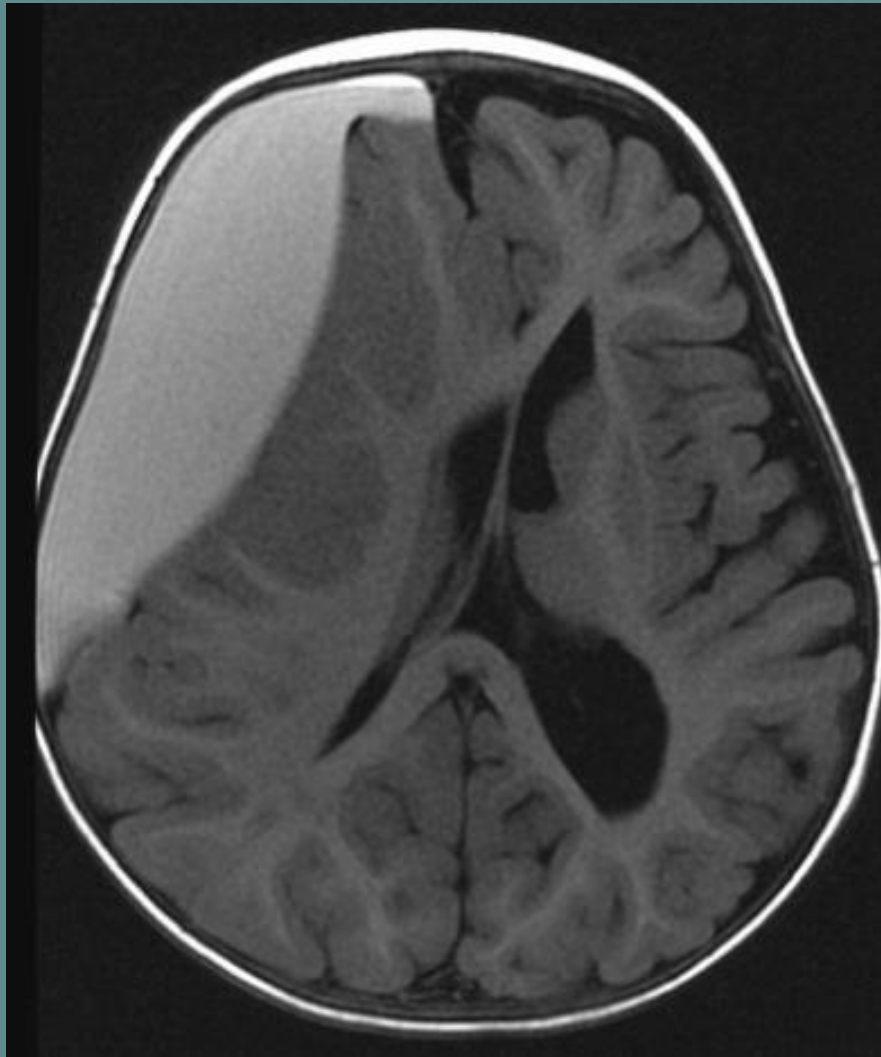
YEAH Program Nightmare

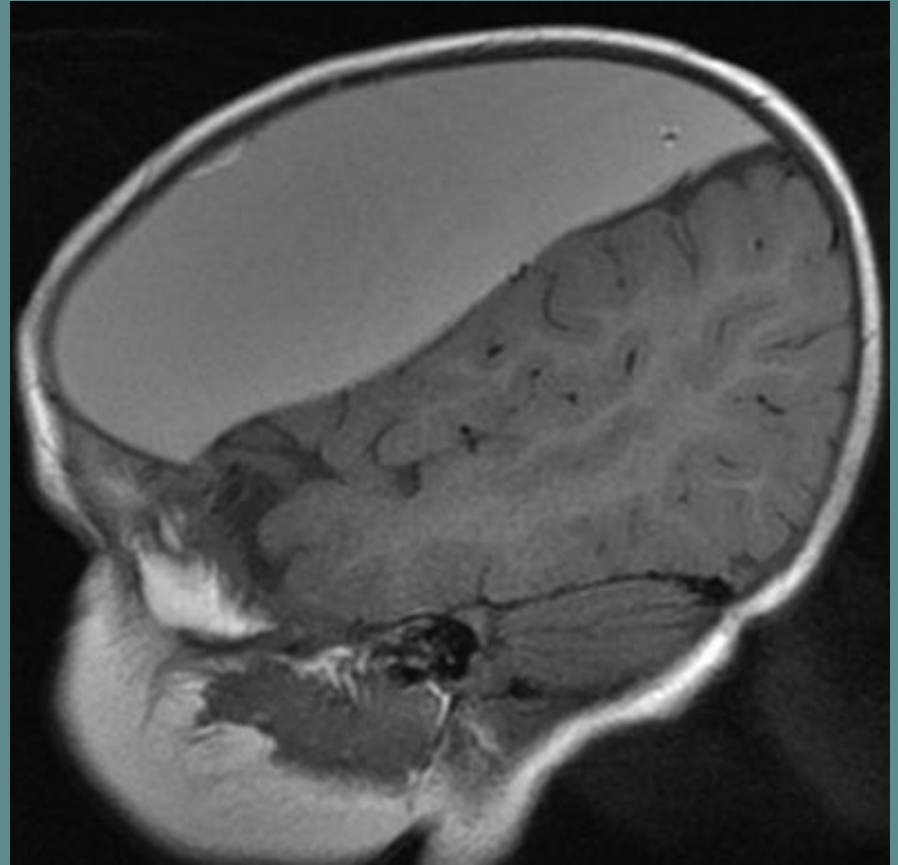
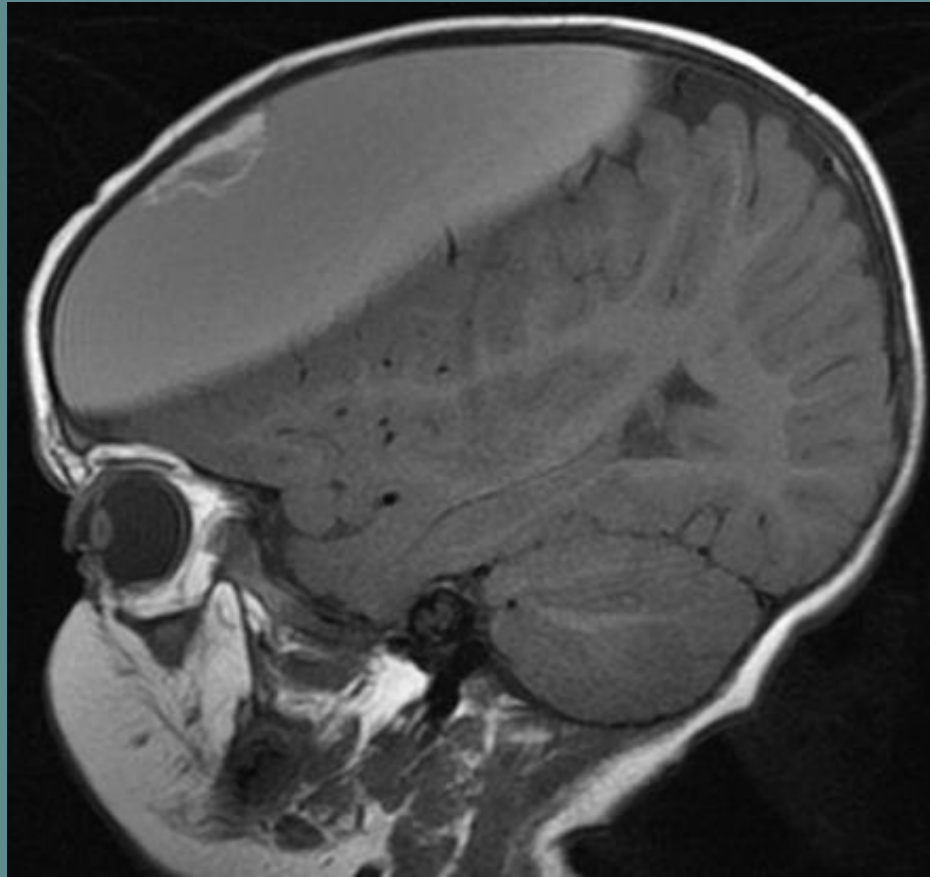
BEAFI

- Benign Extra-Axial Fluid of Infancy
- Historically: External hydrocephalus
- Head is large, possibly at birth
- Begins to cross percentile lines in first 3-6 months, can continue through 18 mos
- Runs in families
 - (Measure the OFC of parents & siblings)

The Dark Side of BEAFI

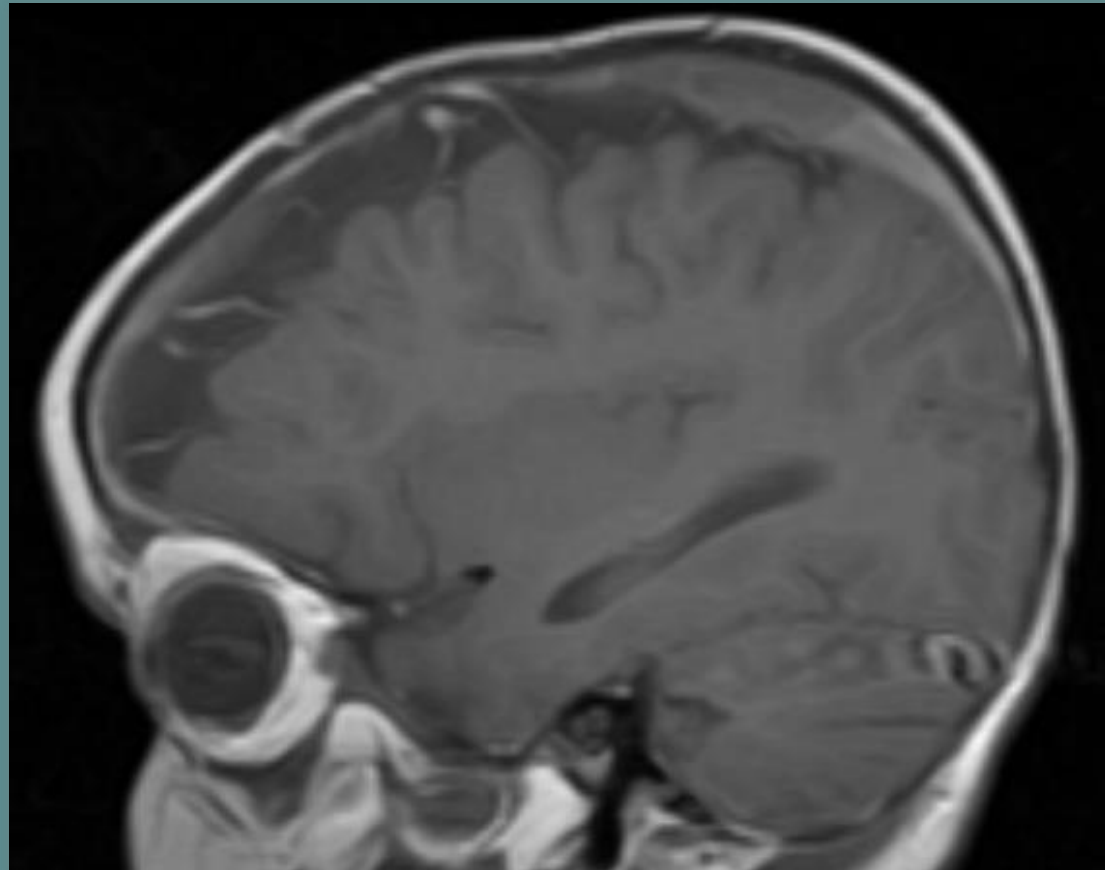




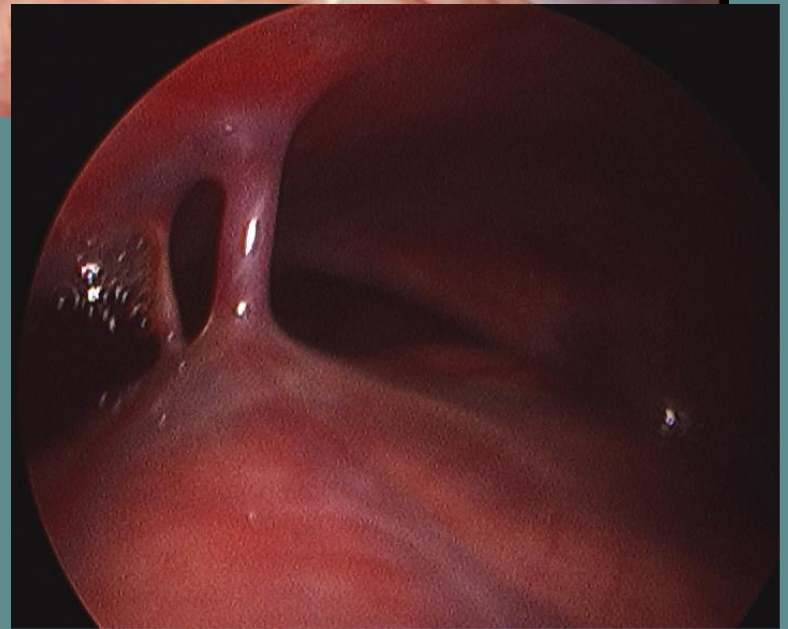
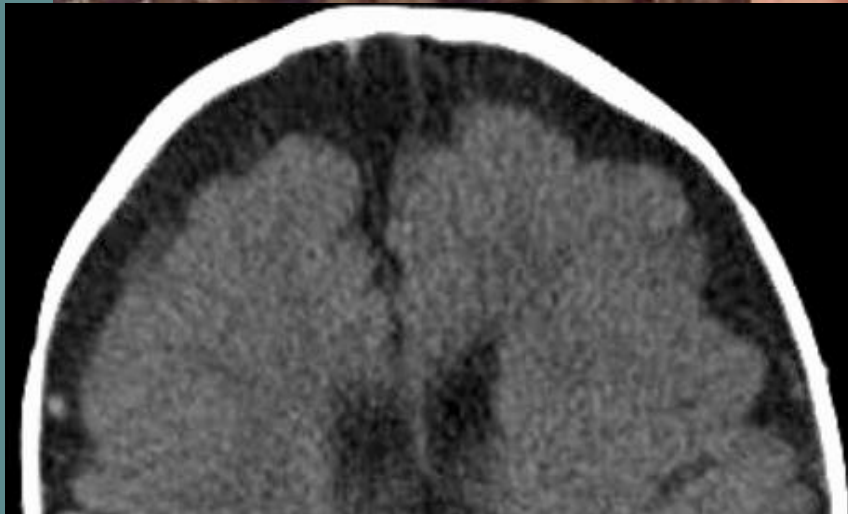
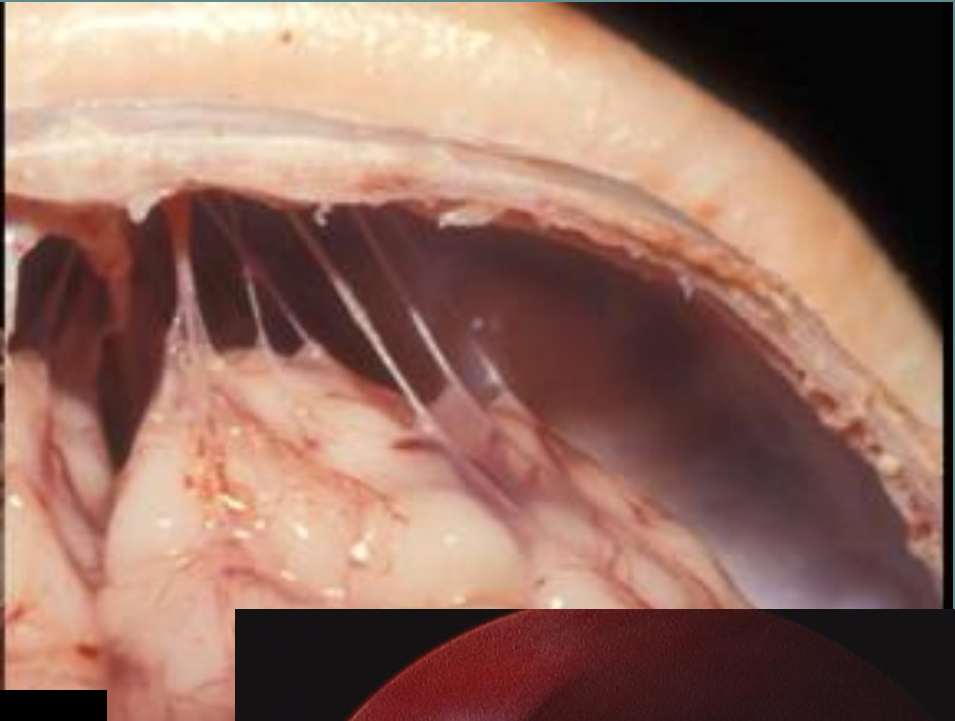


BEAFI & SUBDURAL HEMATOMA

- Bridging veins traverse from the cortical surface of the brain to the dura and draining venous sinuses

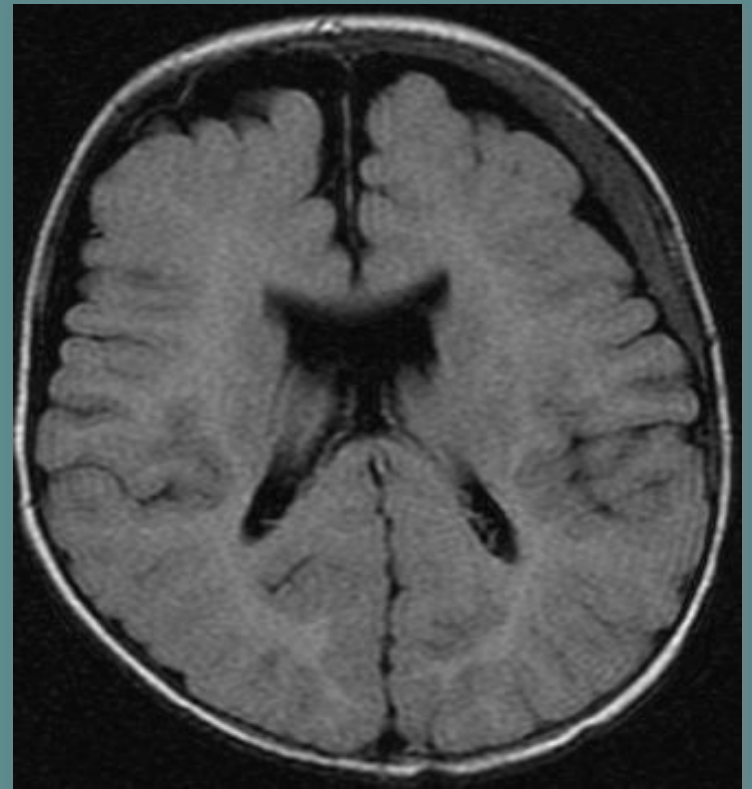


Bridging Veins – Subdural Space



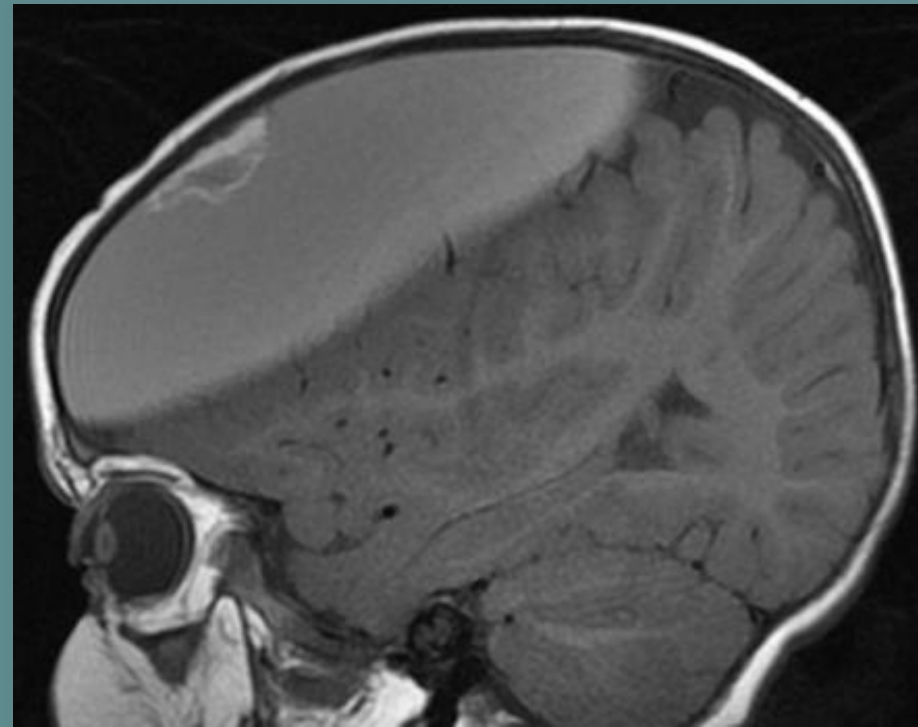
BEAFI & SUBDURAL HEMATOMA

- With trauma, there is more room in the cranial cavity for the brain to shift, putting the cortical veins on stretch
- This can lead to vessel leakage or rupture, accumulating blood in the subdural space



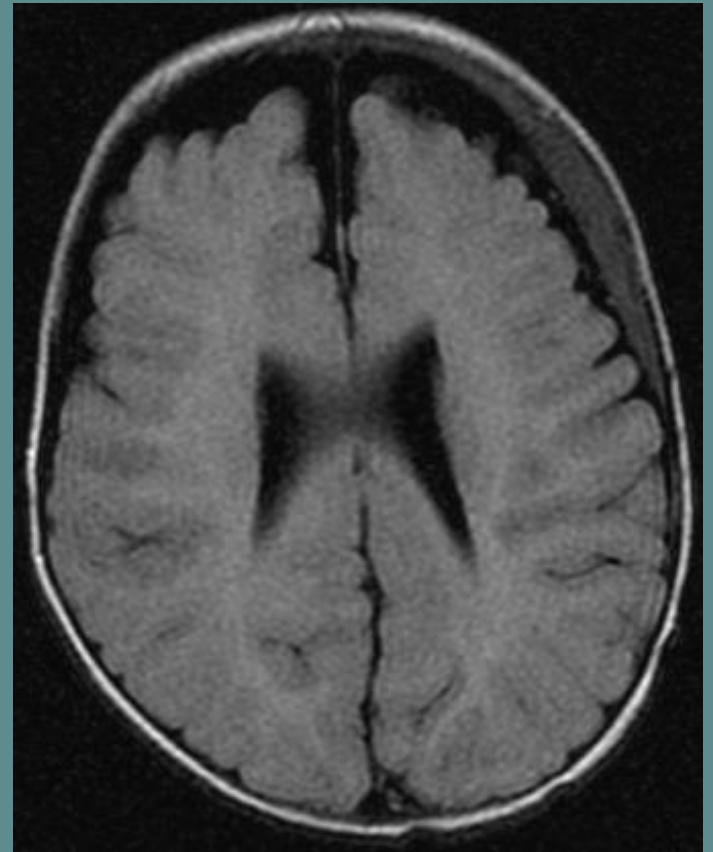
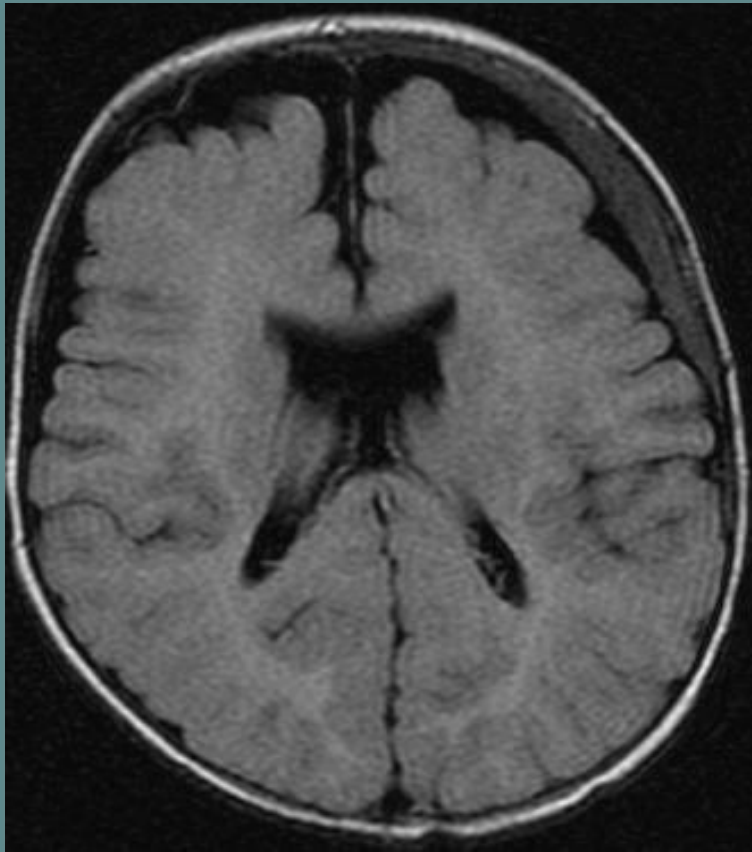
BEAFI & SUBDURAL HEMATOMA

- The danger is the blood has little resistance to spread (there is already a generous fluid space), so a significant leak can result potentially in a massive subdural hematoma
- Thankfully, this is rare



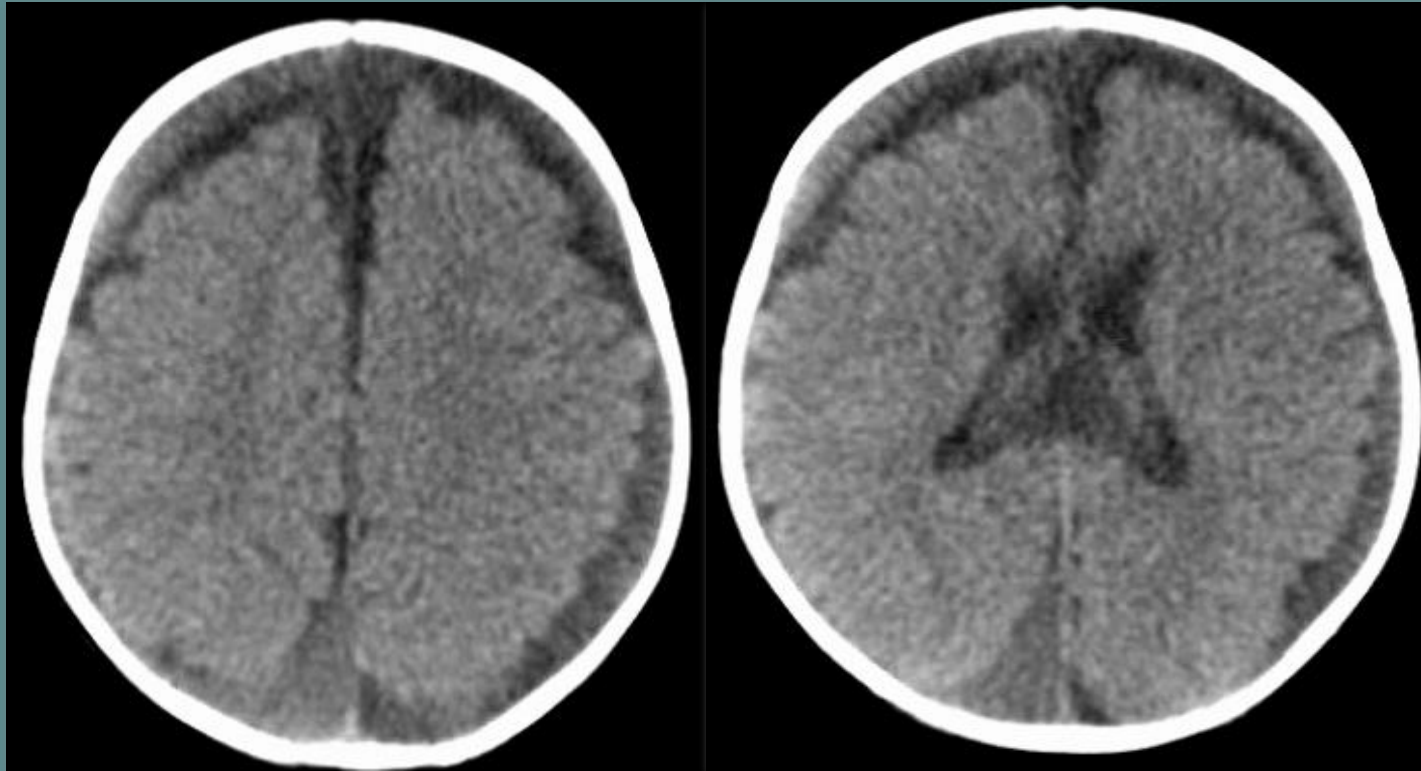
BEAFI & SUBDURAL HEMATOMA

- Only a small number with BEAFI bleed
- Most bleeds are small and do not require intervention surgically



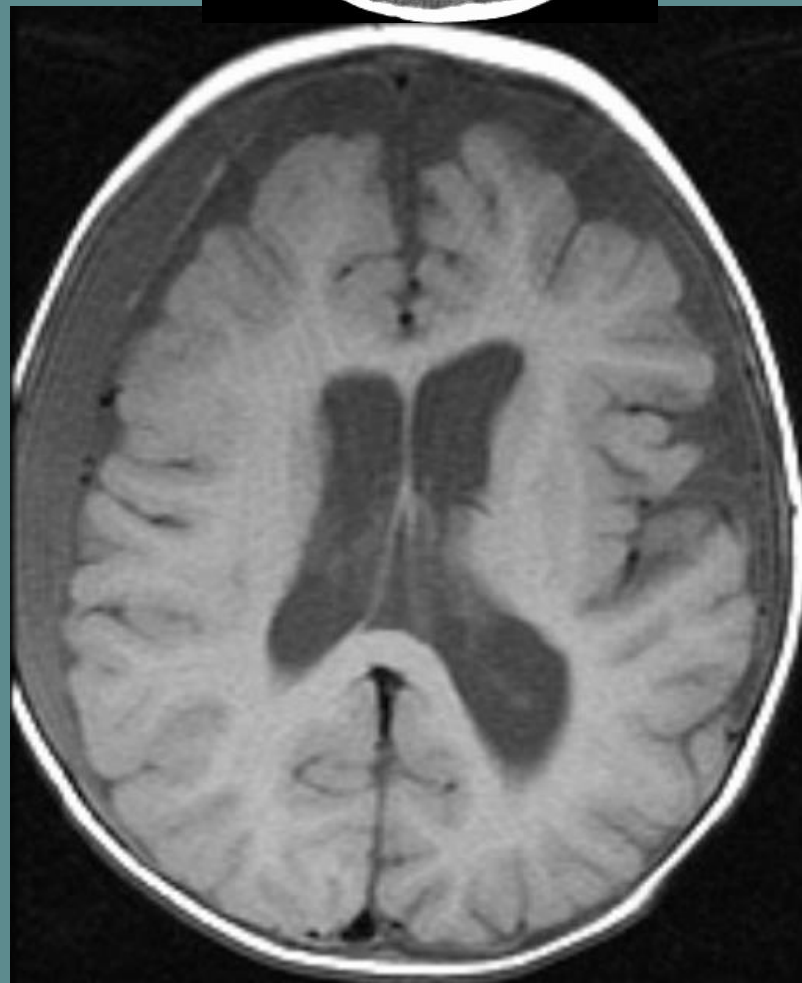
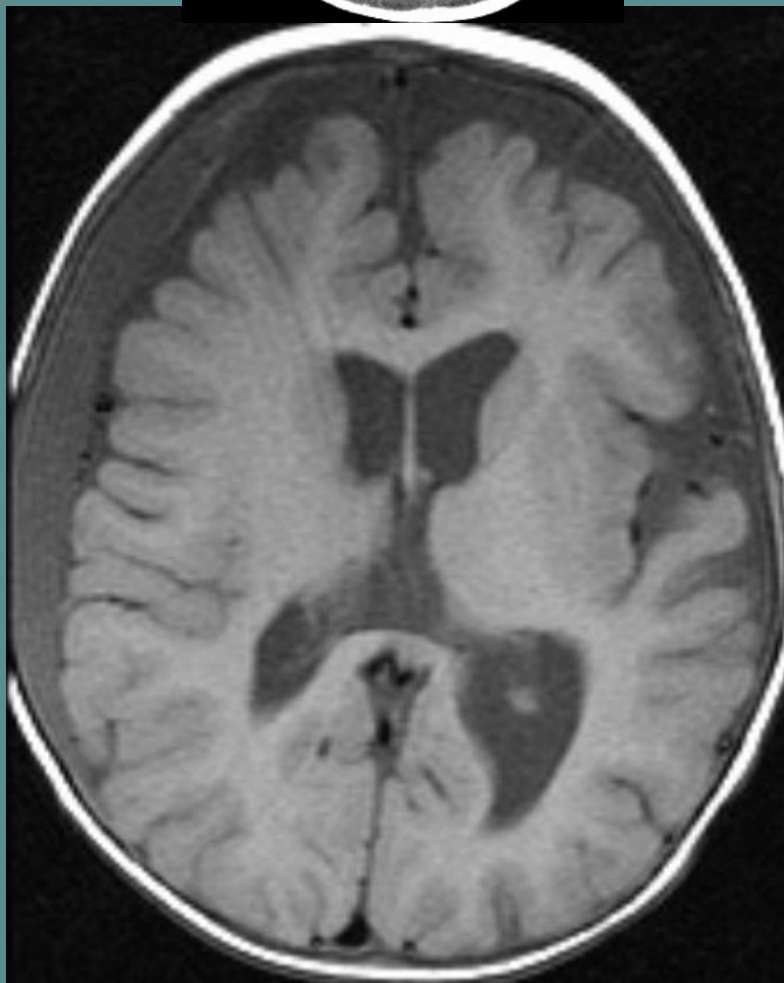
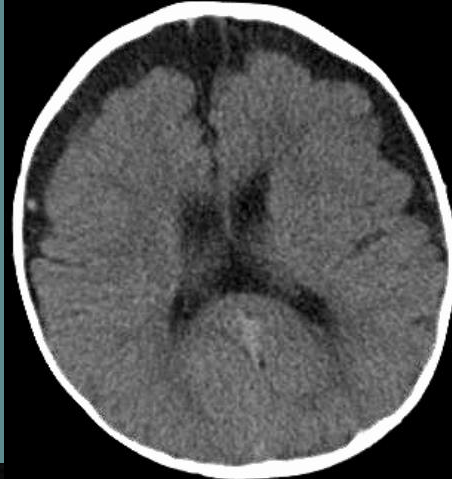
BEAFI & SUBDURAL HEMATOMA

- Bleeding can be either post-traumatic or spontaneous
- Leads to concerns for non-accidental trauma



BEAFI & SUBDURAL HEMATOMA

- Can also occur post chemotherapy with thrombocytopenia



BEAFI & SUBDURAL HEMATOMA

- Typically a significant enough trauma to cause the child to have a skull fracture or mild concussion
- Fall off the swing, table, grocery cart, down stairs
- Usually not ground level fall or playing with child such as tossing in the air

BEAFI & SUBDURAL HEMATOMA

- Educating the parents is paramount
- Risk period is until the BEAFI resolves, typically until age 3 or 4 years
- Unfortunately this is also the at risk group for falls as toddlers learn to walk and develop coordination
- The possible spontaneous occurrence really scares parents
- No helmet required, just makes them more top heavy

BEAFI & SUBDURAL HEMATOMA

- Call the office during regular hours & discuss with us, probably will get scanned
- After hours we request they come to the ER and get a “OneShot” brain MRI or CT of the head, even if acting OK after the initial event
- Blood can “silently” accumulate in the subdural space in these children
- Blood in subdural space gets admitted for observation
- A negative scan can go home if clinically well

BEAFI & SUBDURAL HEMATOMA

- In our practice, we see numerous children each year with BEAFI
- This trauma scenario occurs 2-3x per year





Lunar Eclipse/Blood Moon - Rick King