Pediatric Neurosurgical Disorders: from head to tail

Peter P. Sun, M.D. Pediatric Neurosurgery

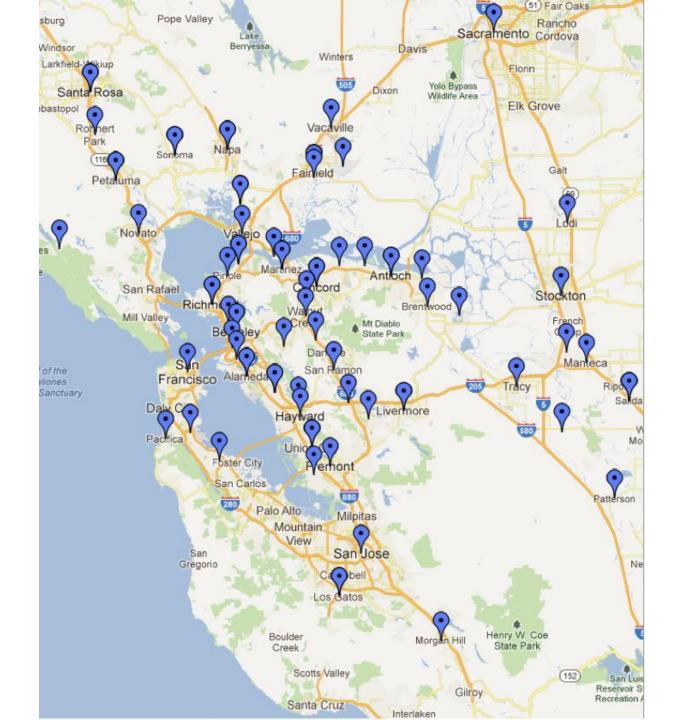


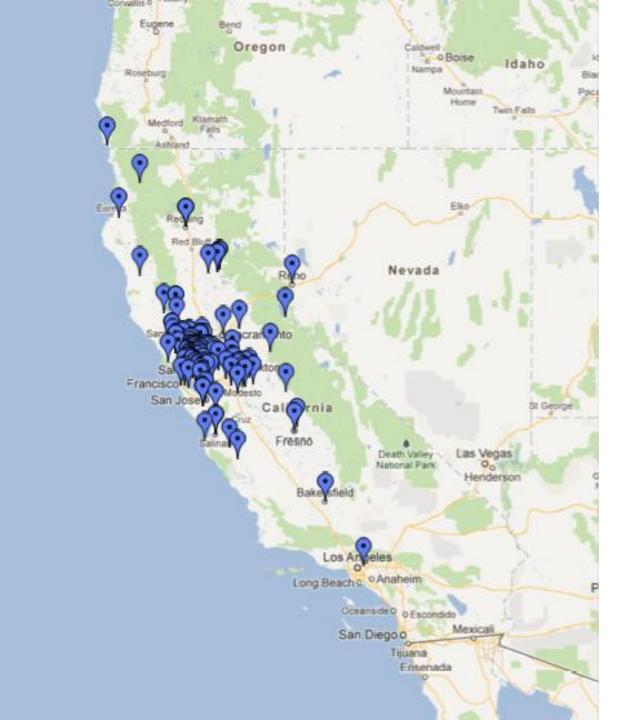












Cities of 2011 Surgery Patients



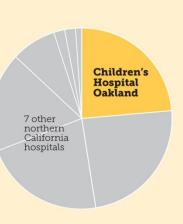




INPATIENT BRAIN TUMOR CASES, 2009

Children's performed nearly a quarter of all of northern California's pediatric inpatient brain tumor surgeries in 2009.

Primary & Secondary Service Areas (Alameda, Contra Costa, Marin, San Francisco, San Joaquin, Santa Clara, Solano, Sonoma, Stanislaus); Ages 0-17; ©2010 Children's Hospital & Research Center Oakland



INPATIENT CRANIOSYNOSTOSIS CASES, 2008-09

Children's treated over 39% of all of northern California's pediatric inpatient craniosynostsosis cases in 2008-09.

Primary & Secondary Service Areas (Alameda, Contra Costa, Marin, San Joaquin, Santa Clara, Solano, Sonoma, Stanislaus); Ages 0-17; ©2010 Children's Hospital & Research Center Oakland

8 other northern Califórnia hospitals

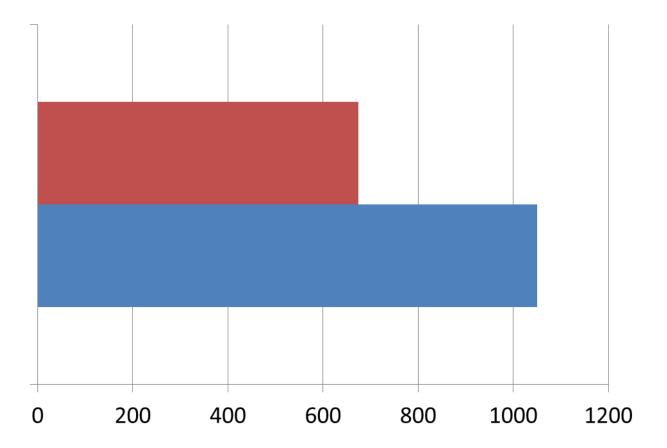
INPATIENT SPASTICITY CASES, 2008-09

Children's treated nearly half of all of northern California's pediatric spasticity cases in 2008-09.

Primary & Secondary Service Areas (Alameda, Contra Costa, Marin, San Francisco, San Joaquin, Santa Clara, Solano, Sonoma, Stanislaus);Ages 0-17; ©2010 Children's Hospital & Research Center Oakland

> 8 other northerm California hospitals

Surgical volume 2009-2011



OR Minutes, 2011

Neurosurgery

Top 5 Services Ranked by Minutes

Surgeon: Peter P. Sun, MD

Case Count: 307 Minutes: 74,591= 73% Minutes + Set Up/Tear Down: 83,291

Surgeon: Kurtis I. Auguste, MD

Case Count: 107 Minutes: 26,050 Minutes + Set Up/Tear Down: 29,800

Total Case Minutes: 101,592 (including Dr. Gupta and Dr. Sheinberg) Total Minutes + Set Up/Tear Down: 114,252

GPS Minutes: 166,653 = 32%

ORT Minutes: 120,626 = 23%

NES

Minutes: 101,592 = 19%

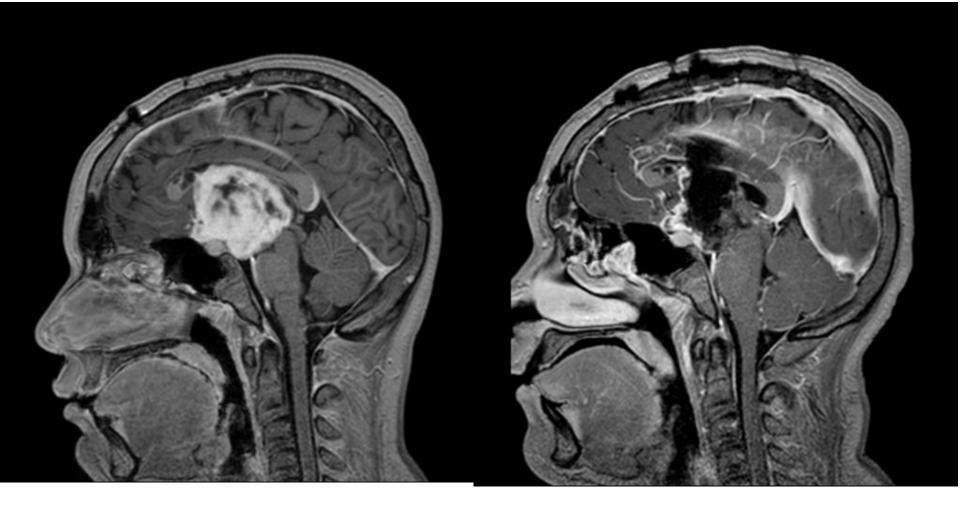
ΟΤΟ

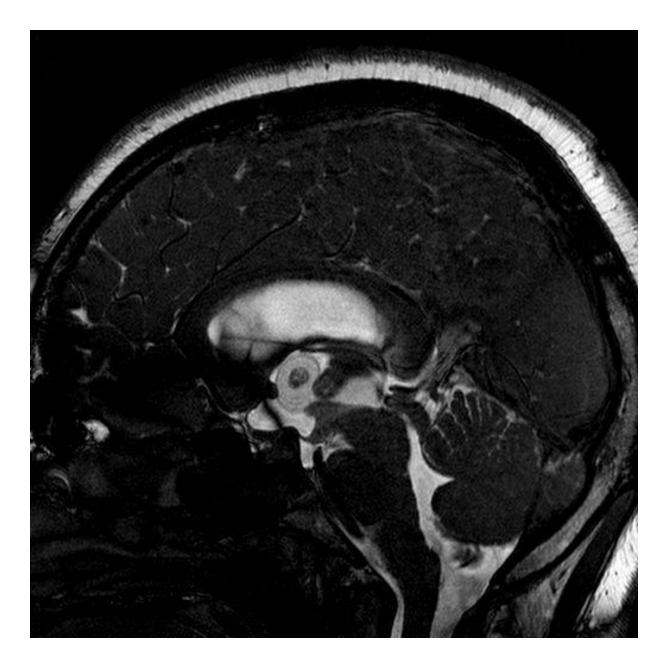
Minutes: 90,670 = 17%

PLS

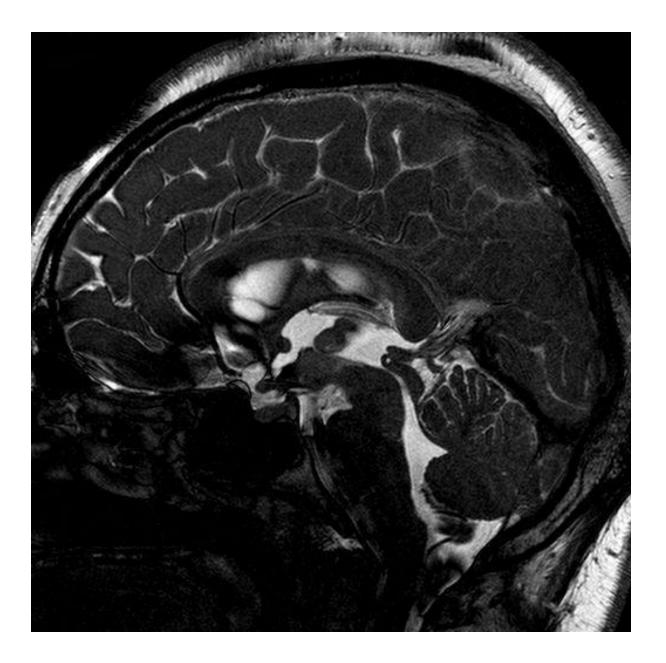
Minutes: 44,078 = 8%

Total OR Minutes (not including Set Up/Tear Down): 523,619

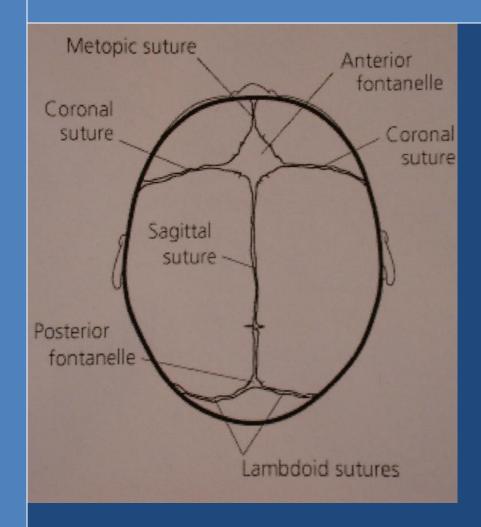


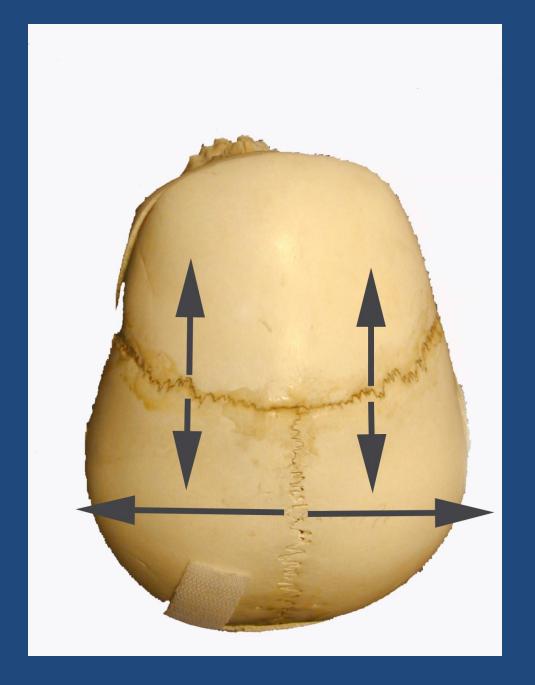


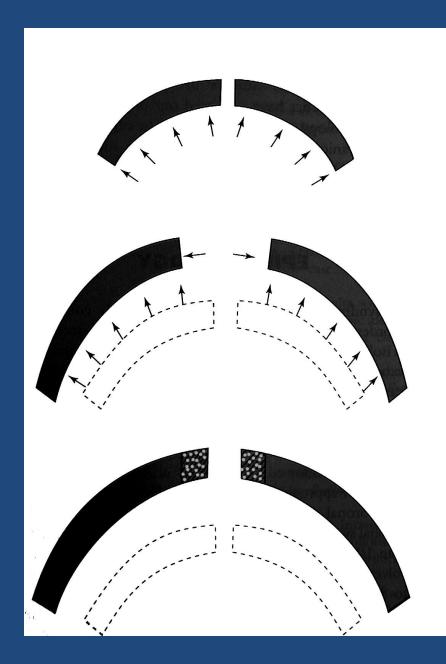












Skull Biology

- Sutures form at borders of advancing skull ossification centers with inductive signals from underlying dura
- New bone is laid down perpendicular to the suture line
- Bone deposition at the suture is driven by brain growth
- 87% of adult head size by age 2

Abnormal Head Shape

- Craniosynostosis: premature closure or absence of calvarial suture
 - creates characteristic skull deformities at birth
- External forces
- Compensatory changes from brain disorders

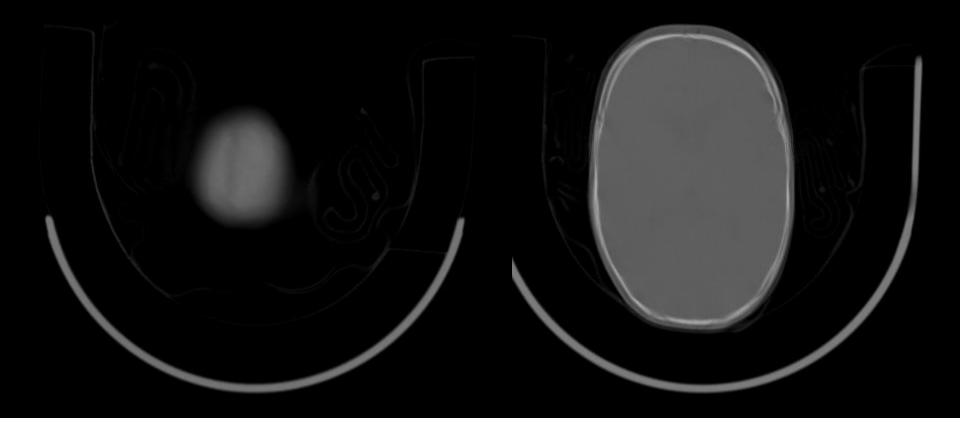
Abnormal Head Shape

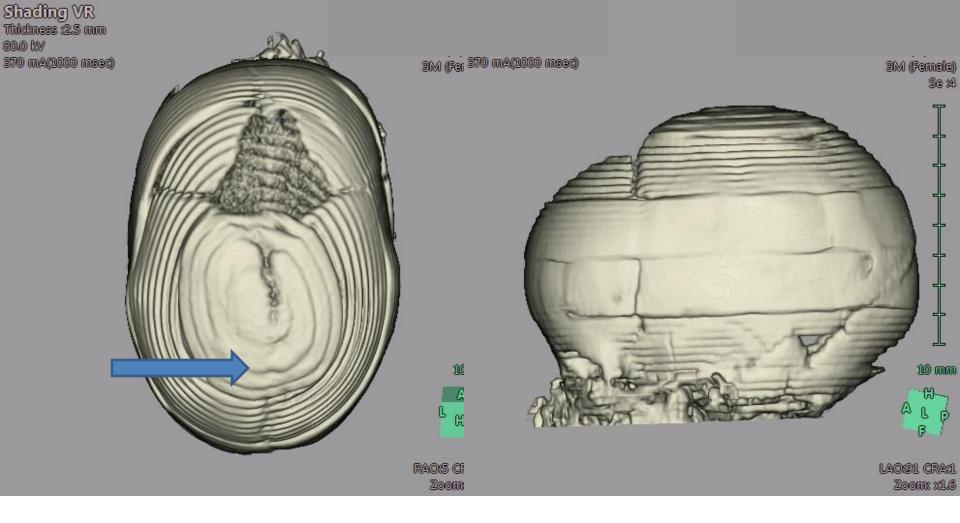
- Plagiocephaly
- Scaphocephaly/Dolichocephaly
- Turricephaly
- Brachycephaly
- Trigonocephaly

Diagnosis

History: present at or apparent shortly after birth Exam: shape palpate suture separation ridge over fused *midline* sutures Radiographs : confirmatory CT: for surgical planning







Computer reconstruction can give false positive !

Frontal plagiocephaly

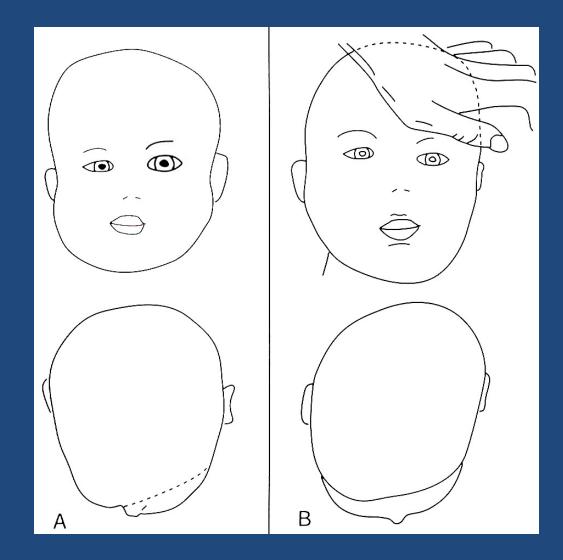
Positional frontal plagiocephaly

- Primary, or secondary to occipital positional plagiocephaly

Frontal plagiocephaly

Unilateral coronal synostosis:

- -1/10,000
- -frontal plagiocephaly, ipsilateral enlarged orbit, Contralateral nasal deviation, strabismus
- Not familial, 15% with FGR mutaitons





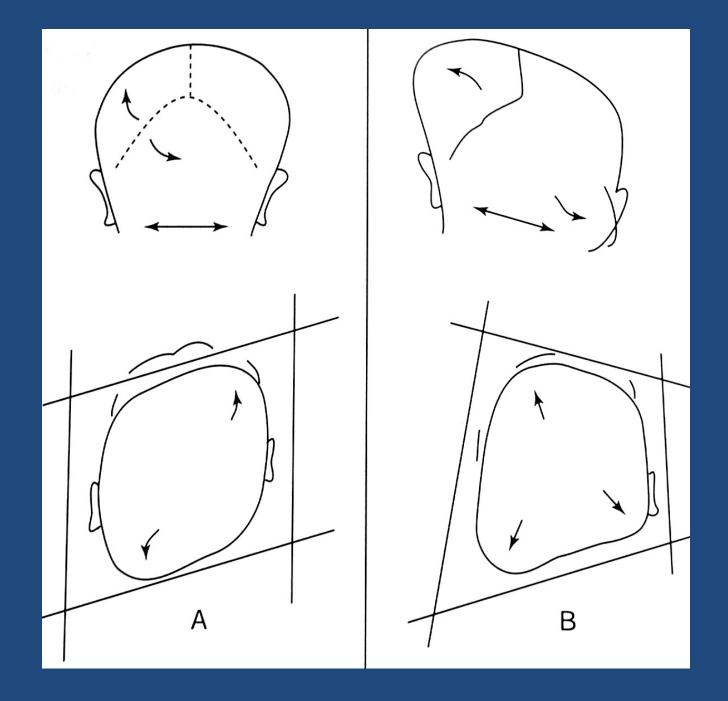


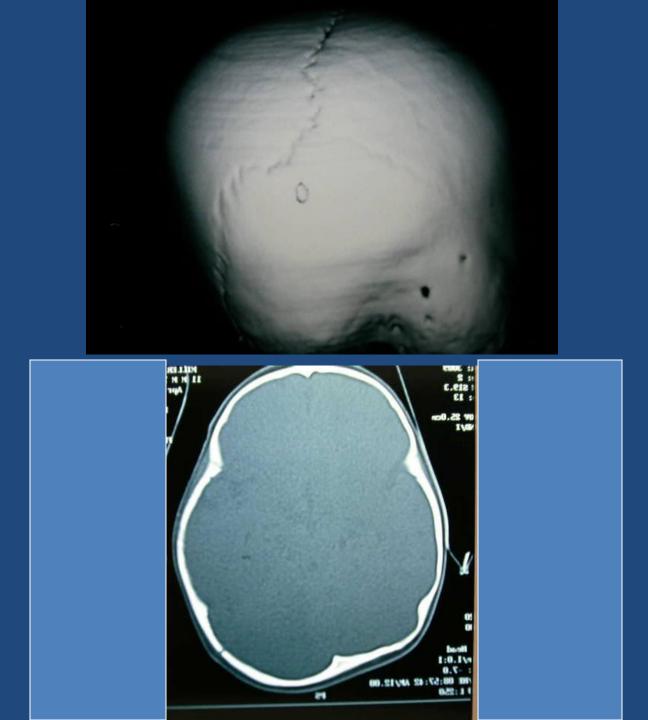


Occipital plagiocephaly

Positional plagiocephaly vs lambdoid synostisis

1/300 – 48% vs 3/100,000





SOMATOM PLUS 4 10928 spin +0 H-SP-CR 27-SE tilt +0 07-DE

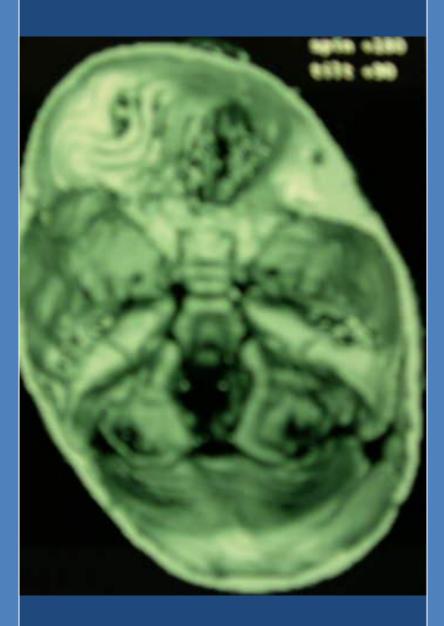
3000

W

1092864 27-SEP-1997 07-DEC-1998









Occipital plagiocephaly

Lambdoid synostosis

- -3/100,000
- -some ipsilateral occipital flattening compensatory parietal and mastoid bulges
 -trapazoid vertex view

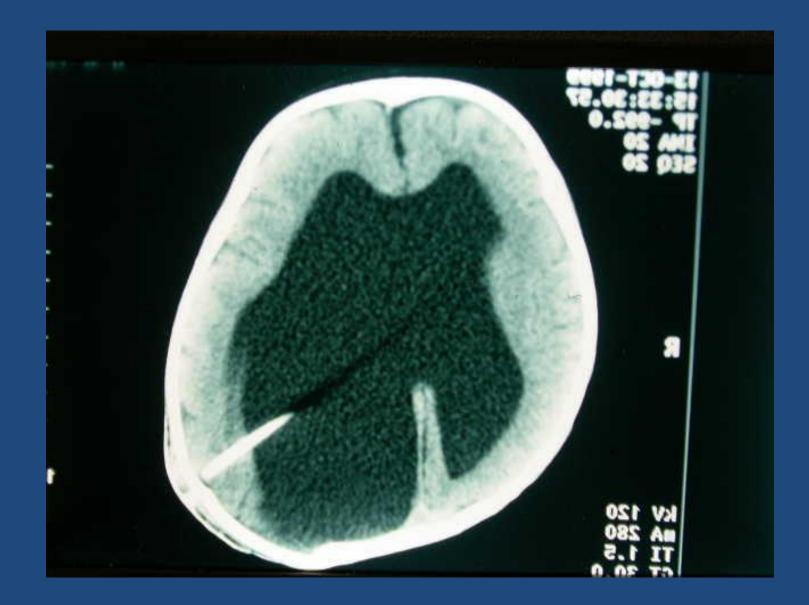
Occipital plagiocephaly

Positional plagiocephaly (deformational plagiocephaly, flat head)

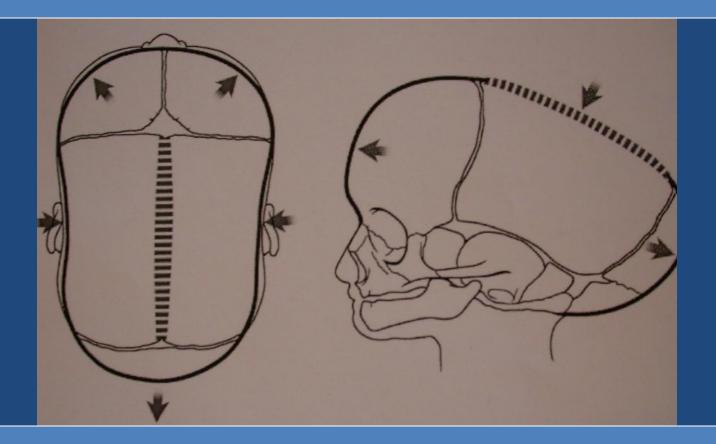
- -common
- -occipital only, parallelegram with compensatory frontal plagiocephaly, bi-occipital
- Helmet for severe cases







Scaphocephaly



Sagittal synostosis

Scaphocephaly

Sagittal synostosis

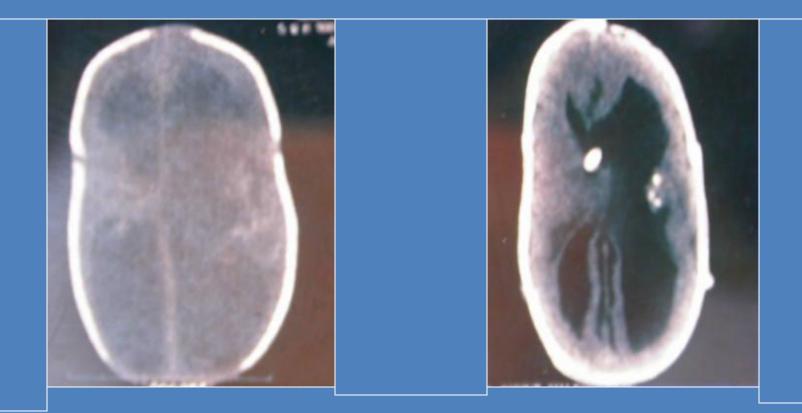
- 2/10,000
- -Frontal bossing, occipital keel, scaphocephaly
- -Early dx offers option of less invasive correction
- 15% of uncorrected pts develop elevated ICP (only single suture synostosis to have proven ICP complication)
- Possible higher incidence of developmental delays unrelated to the timing of correction
- 6% familial in an autosomal dominant pattern

16 M 18174822 Dec 24 2001	Se: 2 M 16 M 18174822 Surface No cut Dec 24 2001	Sur
	DFOV 23,0 cm BONE 154/2	DFO BON 154
	R	L





Shunted hydrocephalus Premi head shape



Trigonocephaly

Metopic synsotosis

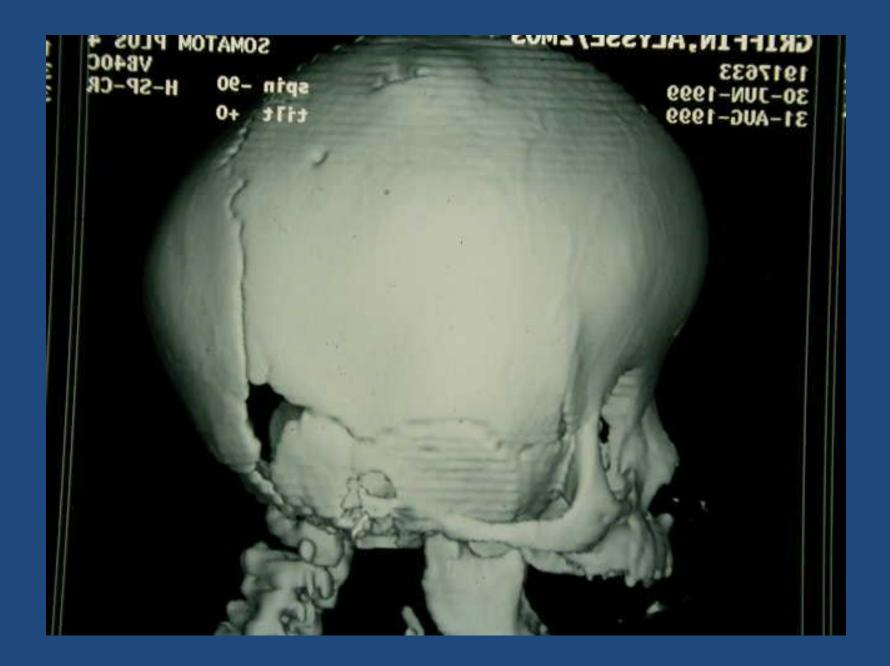
- 10-20% of synostosis
- Wide spectrum of manifestations: triangular forehead to minor midline ridge
- Surgery based on degree of deformity
- 15% associated with other anomalies: heart, GU, brain
- 5.6 % familial



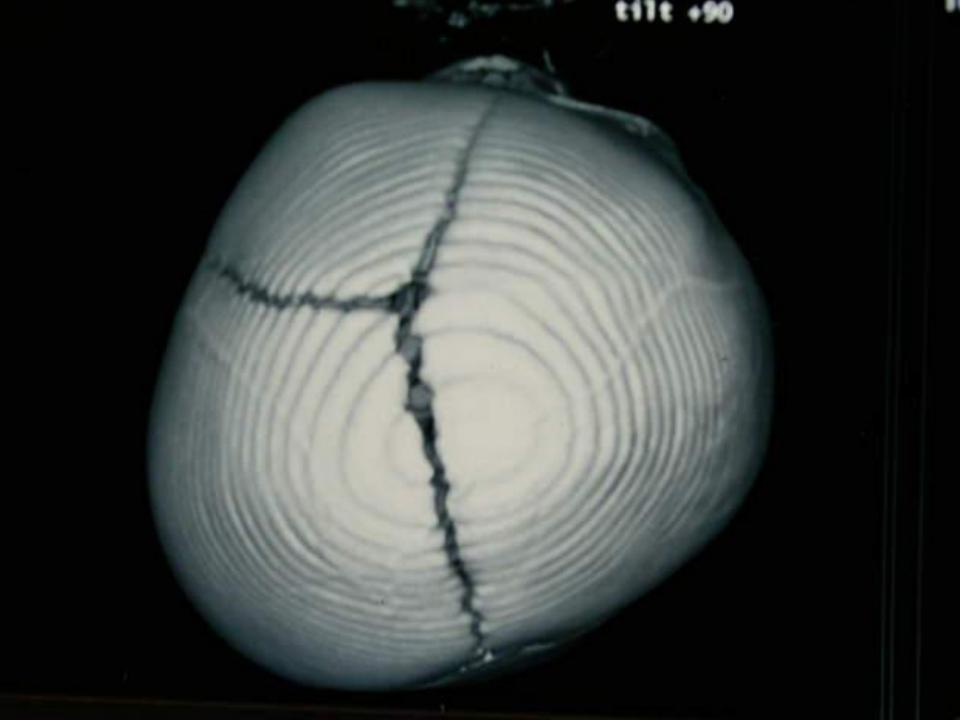
Turricephaly/Brachycephaly

Bicoronal synostosis

 Mostly with syndromic craniosynostosis: Apert's, Crouzon's, Pfeiffer's





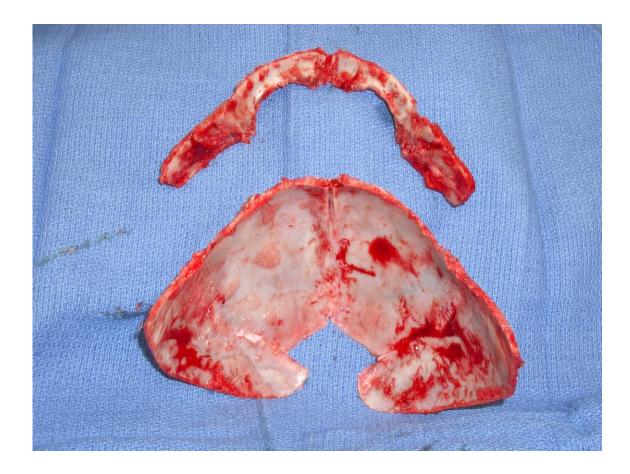


Craniosynostosis Treatment

- Correct deformity: psychosocial benefits of deformity correction outweighs risk of surgery
- Reduce the risk of elevated ICP in sagittal synostosis
- Traditional reconstruction 6-9 months of age
- Endoscopic assisted craniectomy + helmet < 3months of age
- Craniofacial Team approach for syndromic patients: neurosurgeon, plastic surgeon, ENT, orthodontist, oral surgeon, neuropsychologist, social worker etc...



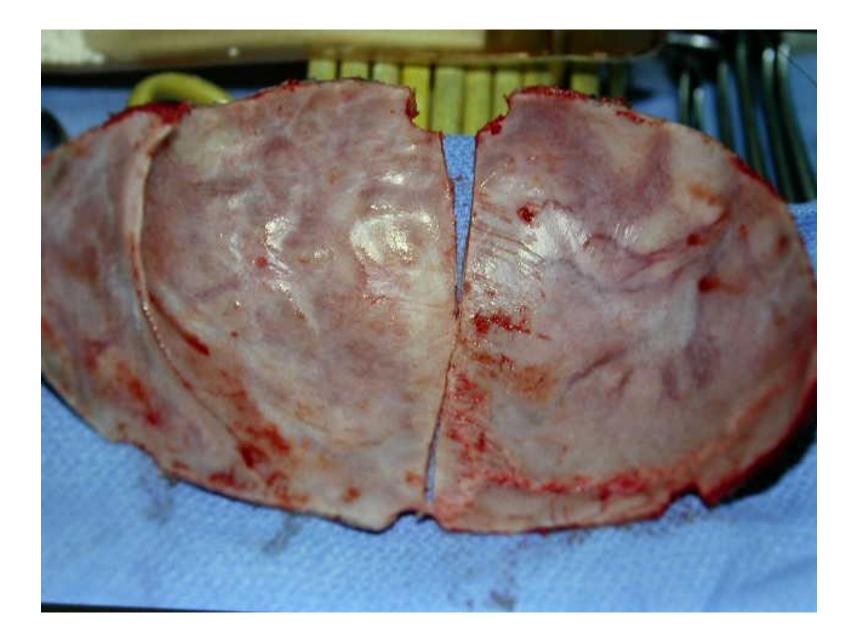












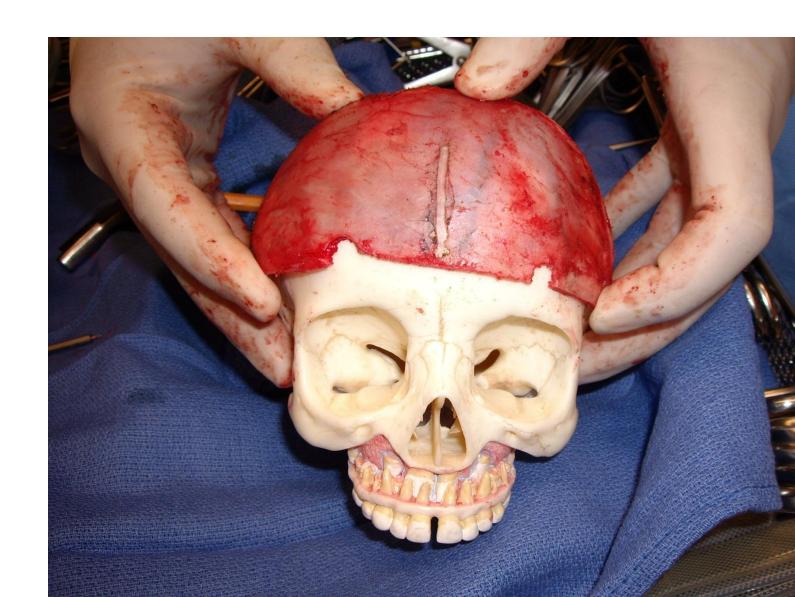






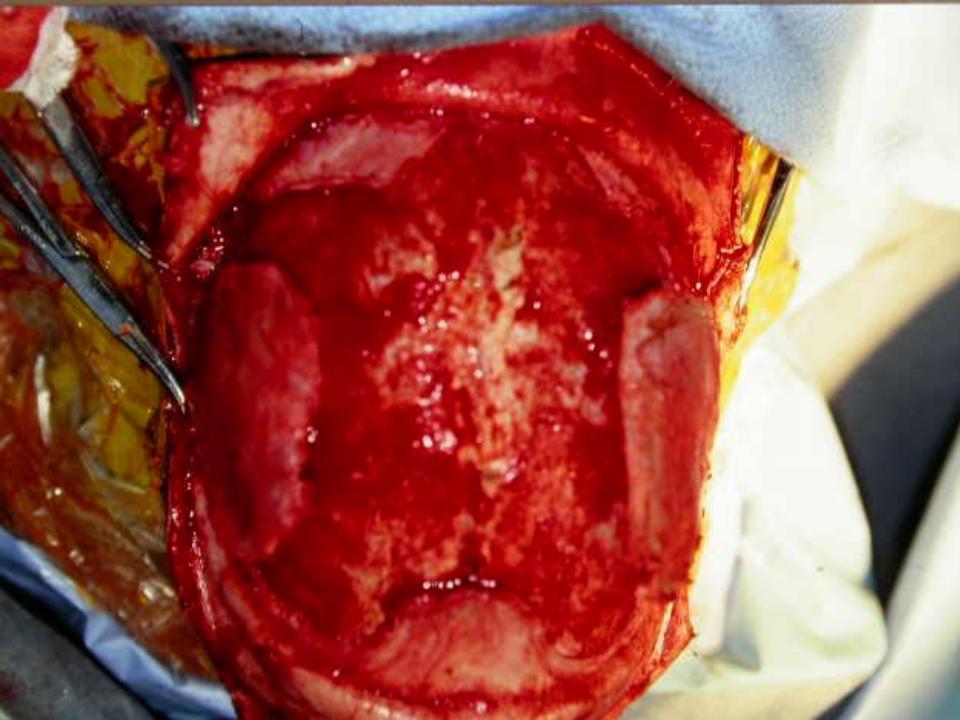






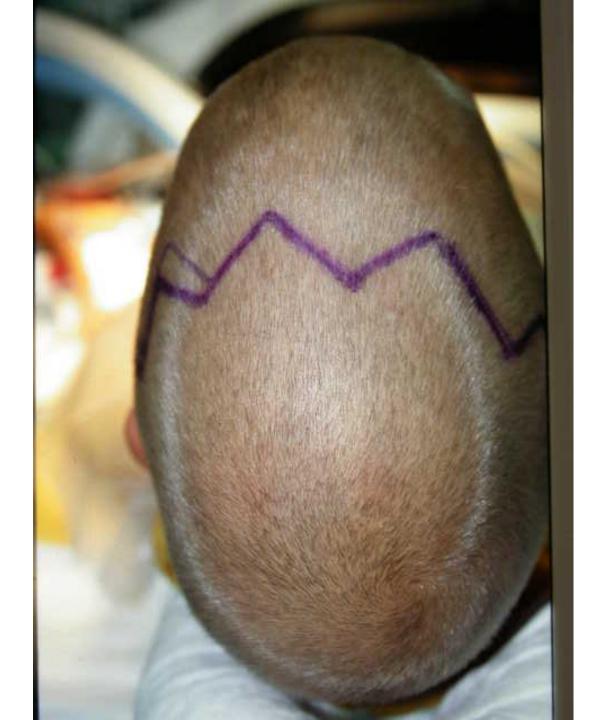


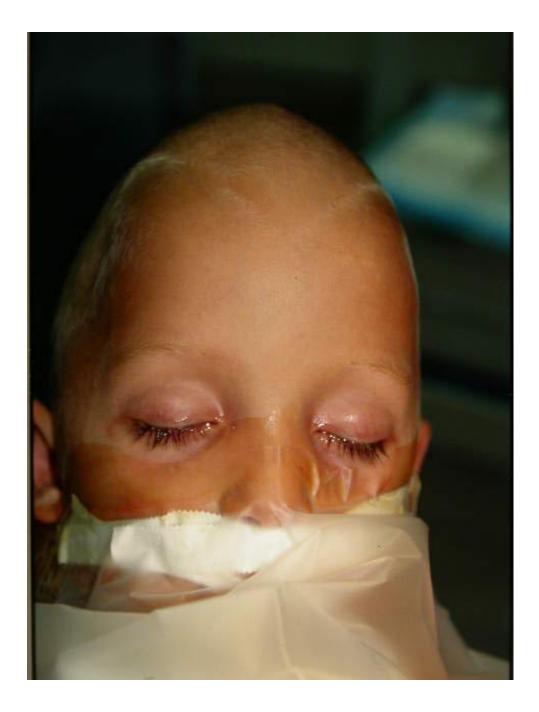




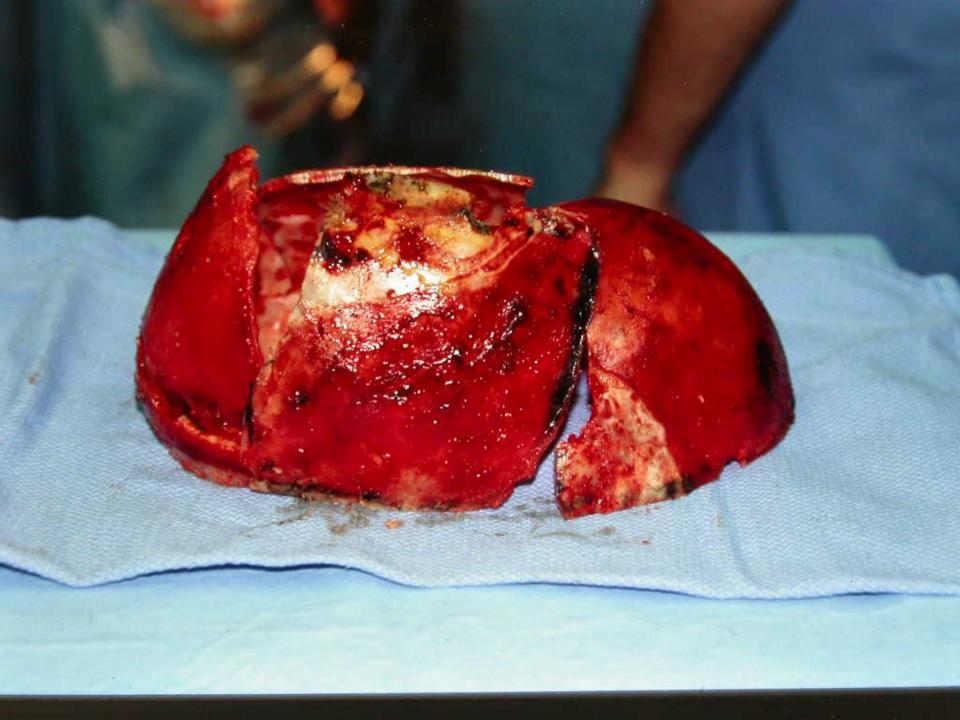






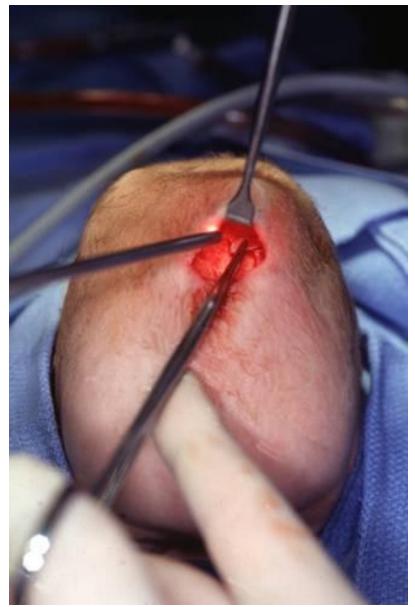








Endoscopically Assisted







Craniosynostosis Treatment

- Endoscopic repair:
- not yet widely accepted
- needs post op helmet for optimal correction
- 2 small scars = minimally invasive..
- EARLY DX!!

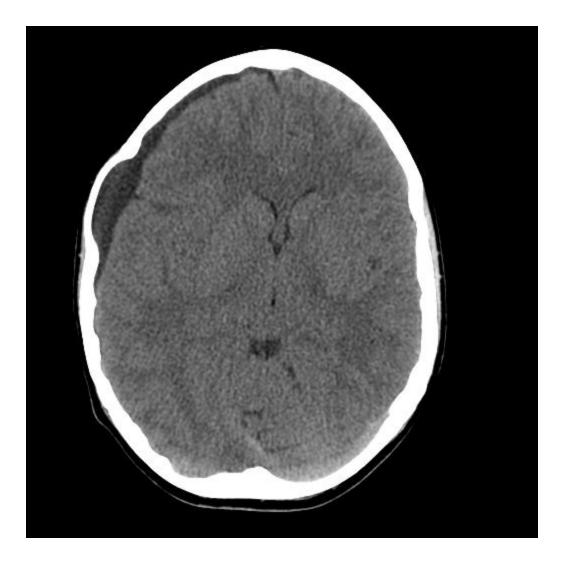


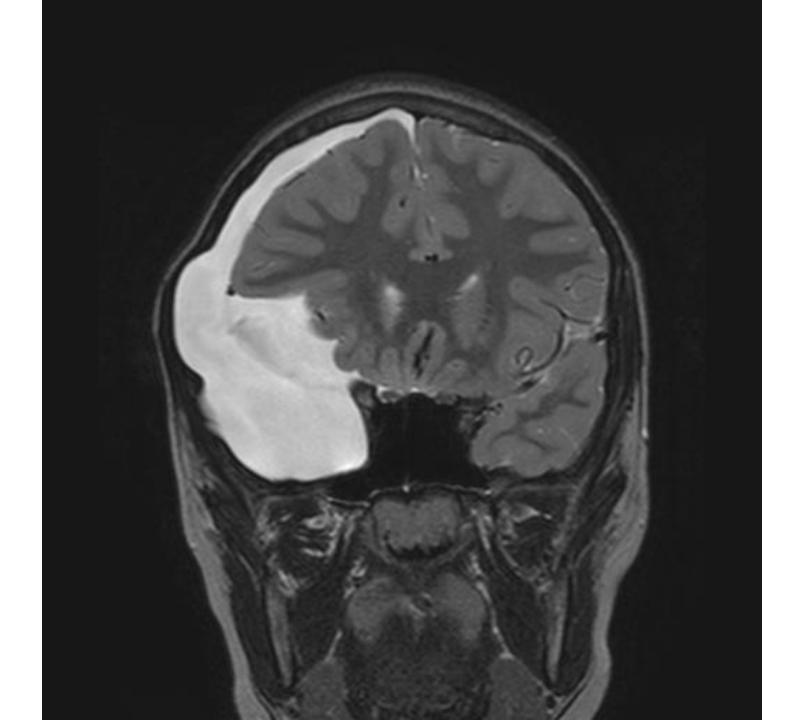


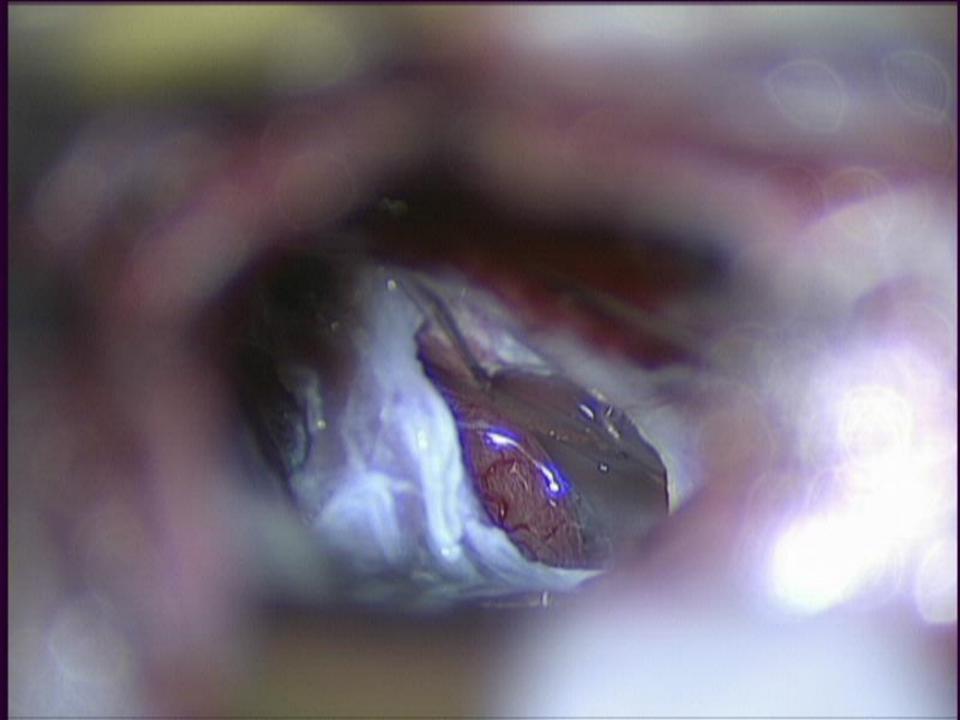




20:52:37 ANTERIOR Can 11 P -857 PAGE 11 26-JAN-91 26-J Scan TP -1 062 RIGHT







#:1/1 h:8/15 #g_R0.0

1:32

SE R:5050 E:100/Ef C:1/1 62.5kHz

IEAD

05/25/99 13:50 MF: 1.2



BRAIN/U / -5.0mm 12.0 OVER 02:16:35.2 PM

P





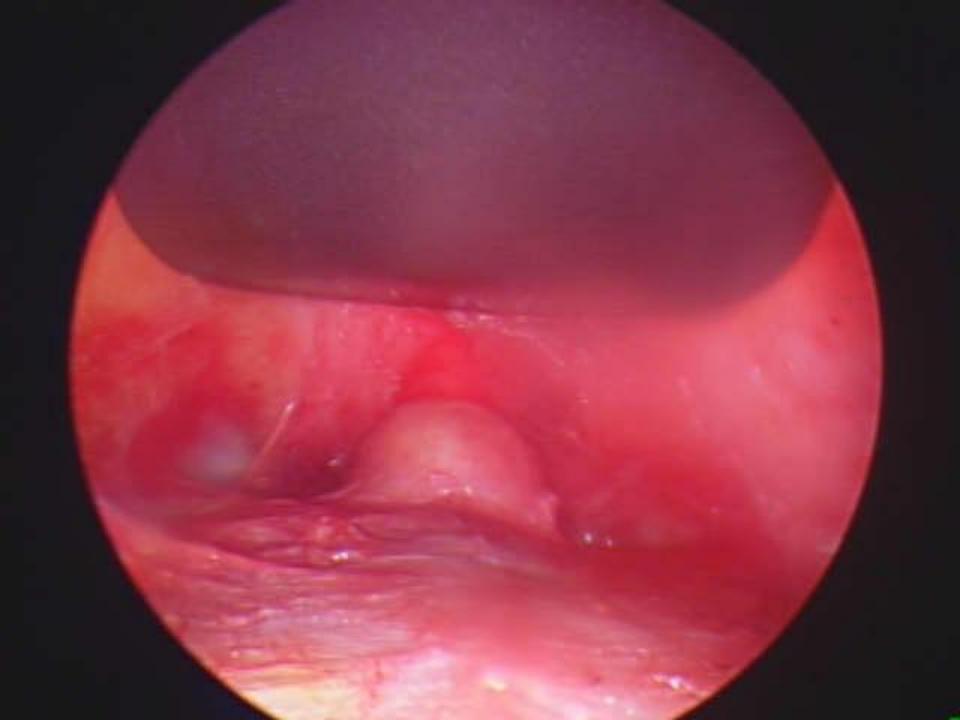


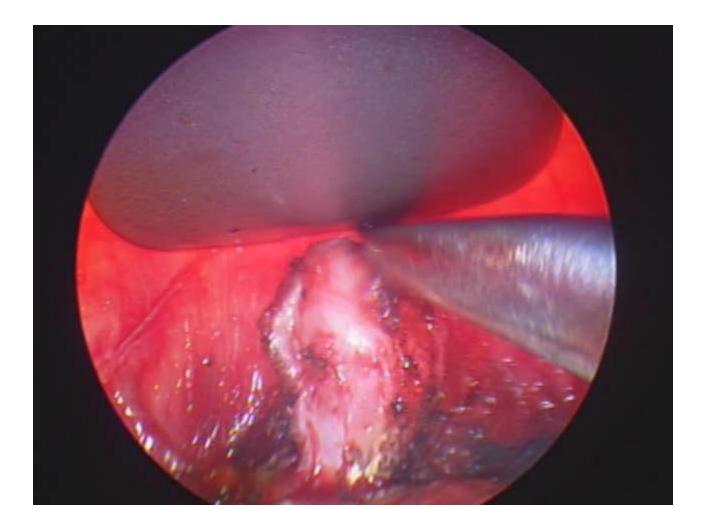
ET:12

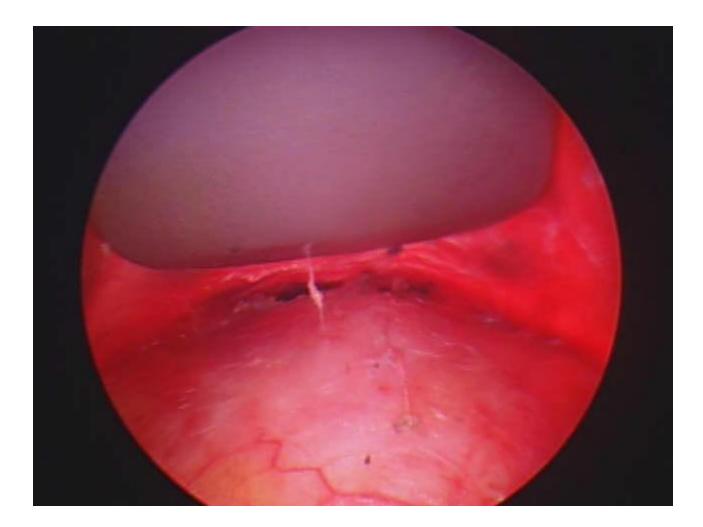
FSE TR:3300 TE:96/Ef EC:1/1 31.2kHz

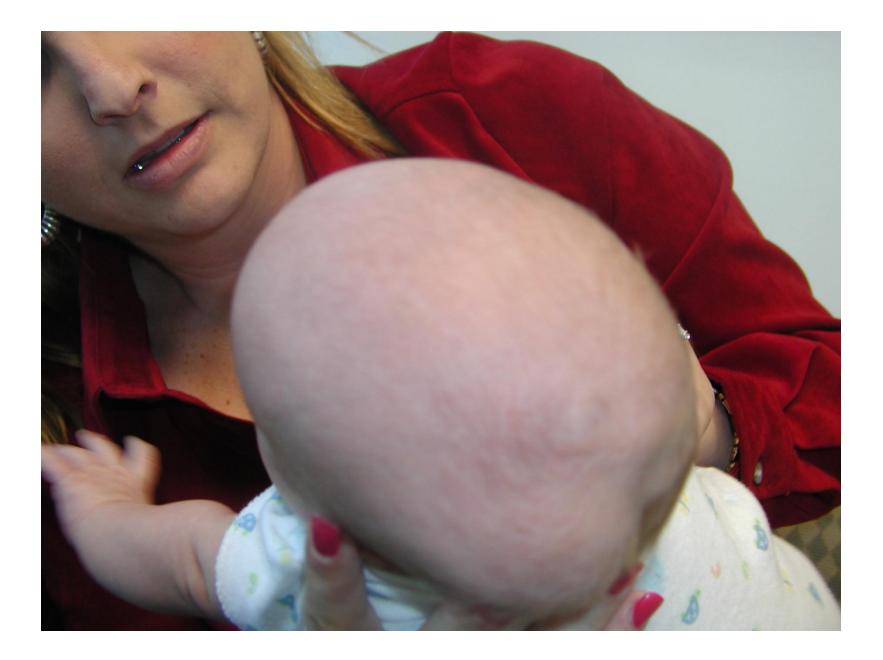
HEAD FOV:20x20 3.0thk/0.5sp

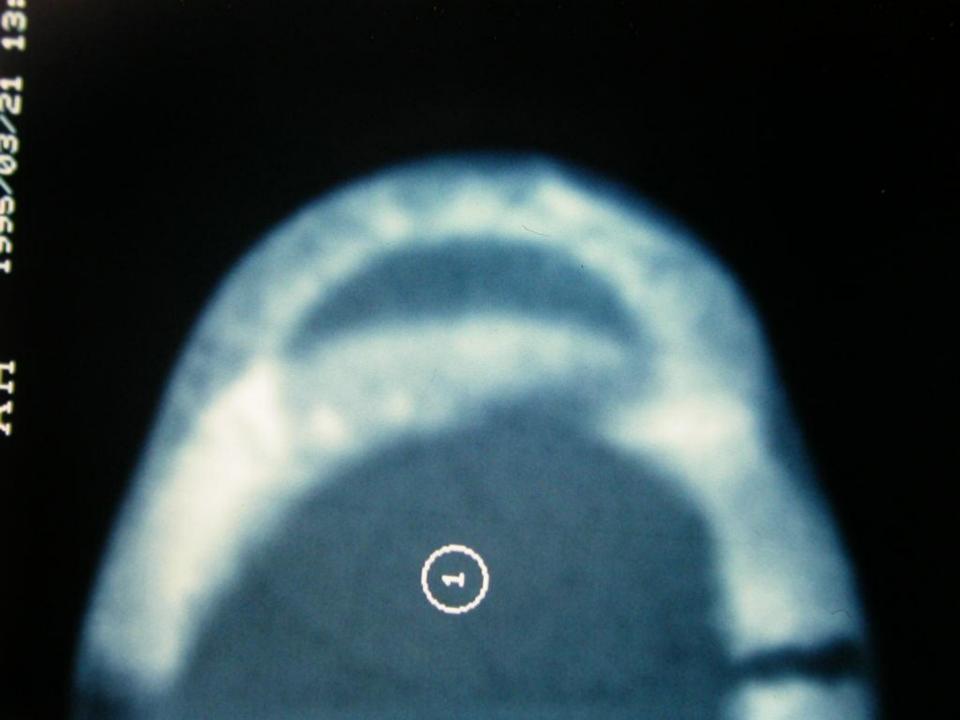




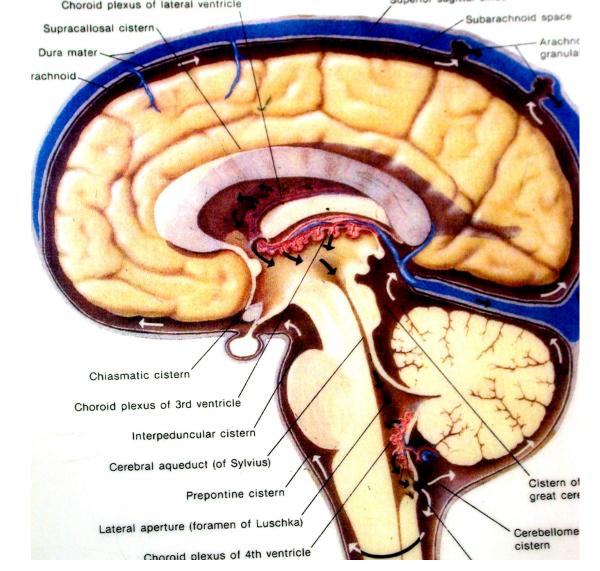












Flow: Lat vent - foramen of Monroe - IIIrd ventaqueduct of Sylvius - IV vent - Luschka & Magendie subarachnoid space

Hydrocephalus: Mismatch of CSF production and absorption

Ventriculomegaly: Large ventricles

Etiology of Hydrocephalus

Congenital

Chiari II (myelomingocele) 85% **Aqueductal Stenosis** Dandy Walker malformation Acquired infectious post-hemorrhagic - IVH: 20-50% tumor post-operative

1.0

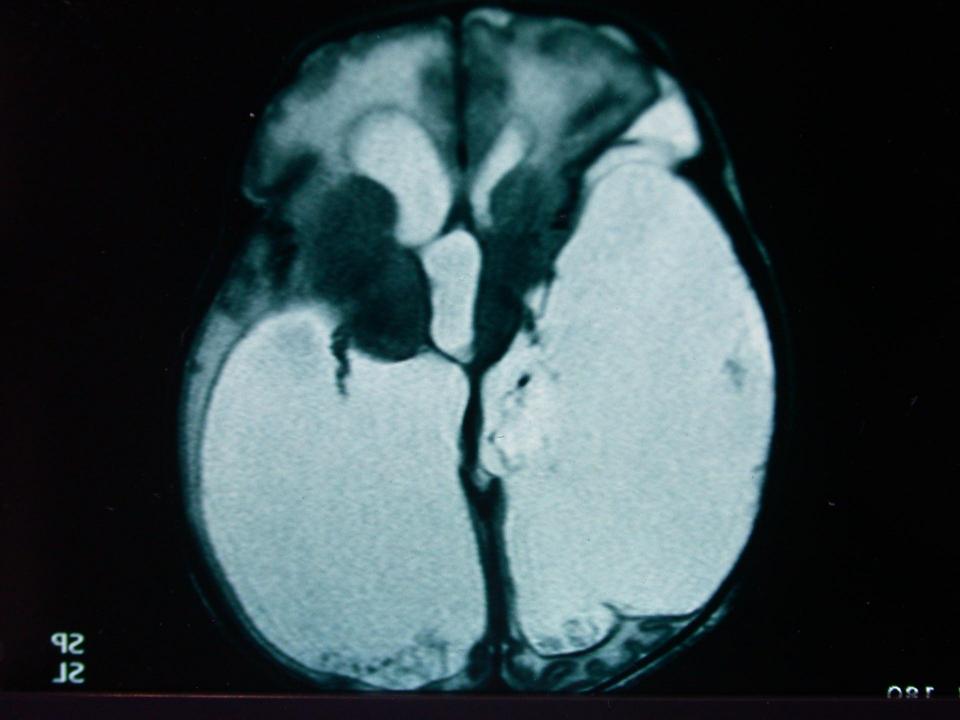
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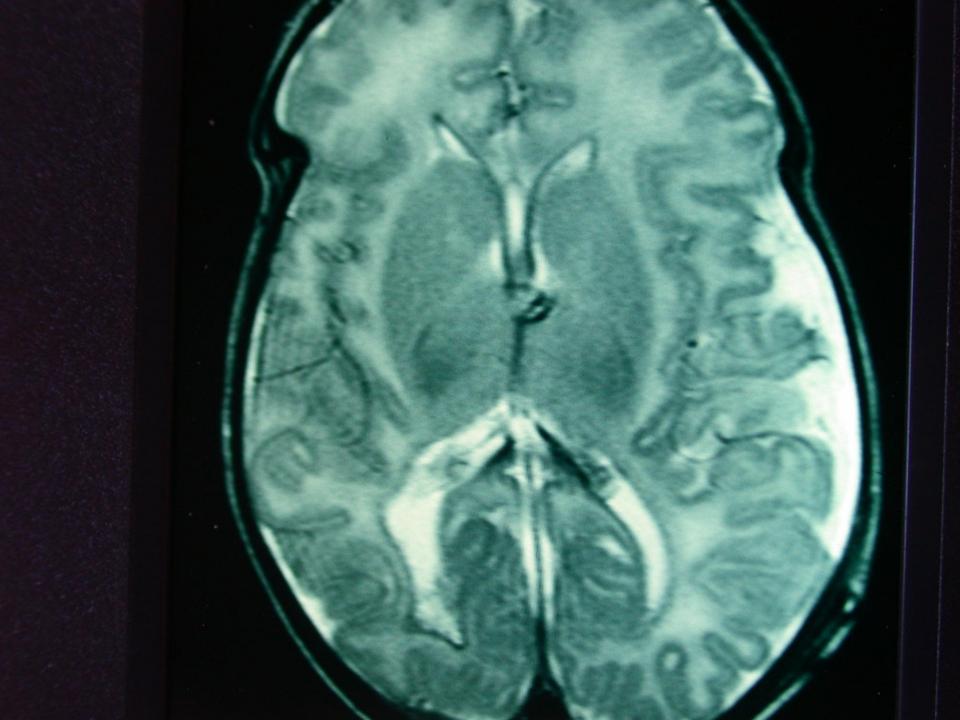
> SE TR:500 TE:20 EC:1/1 16kHz

HEAD FOV:20x20 5.0thk/1.0sp 16/03:16 04:14:52 Mag = FL: ROT:

Clinical Characteristics

Newborns	Infants
Full Fontanelle	Enlarging HC
Sunsetting eyes	IV nerve palsy
Split Sutures	Papilledema
A&Bs	Irritability
Macrocephaly	Emesis





Outcome with shunts

Natural history of untreated hydrocephalusMortality70%Disability75%Mental Retardation>50%

Jansen J, Acta Neurol Scand 1985 Foltz EL, J Neurosurg 1963 Laurence KH, Arch Dis Chil 1965

Outcome of shunted hydrocephalusMortality<15%</td>IQ>8050%(verbal IQ > performance IQ)Palliative

Hirsch JF, Child's Nerv Syst 1994

Prognostic variables of shunted hydrocephalus

•Etiology

- •Age at Diagnosis
- Degree of Ventriculomegaly

•Age at Treatment

Outcome of shunted hydrocephalus

Etiology

Chiari II Aqueductal Stenosis Dandy-Walker IVH

70%¹⁻³ 50-65%^{4,5} 30-50%^{6,7} ~Grade & lesion

Hemmer R, Arch Psych Nerv (German) 1981
 McCullough DC, J Neurosurg 1982
 Raimondi AJ, Am J Dis Child 1974
 Villani R, Child's Nerv Syst 1995
 Guiffre R, J Neurosurg Sci 1986
 Hirsch JF, J Neurosurg 1984
 Sawaya R, J Neurosurg 1981

IQ > 80

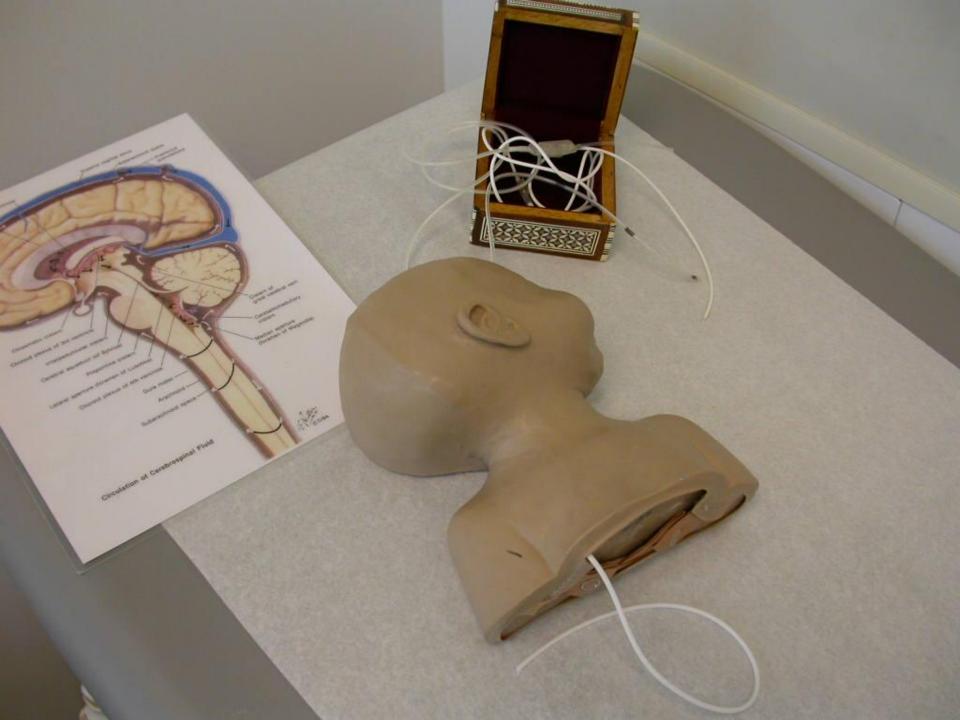
Fetal hydrocephalus: AW> 20mm for greater than 2 months leads to uniformly poor outcome

-Rapidly progressing fetal hydrocephalus, early delivery and shunting may be considered after 32 weeks (C-section)



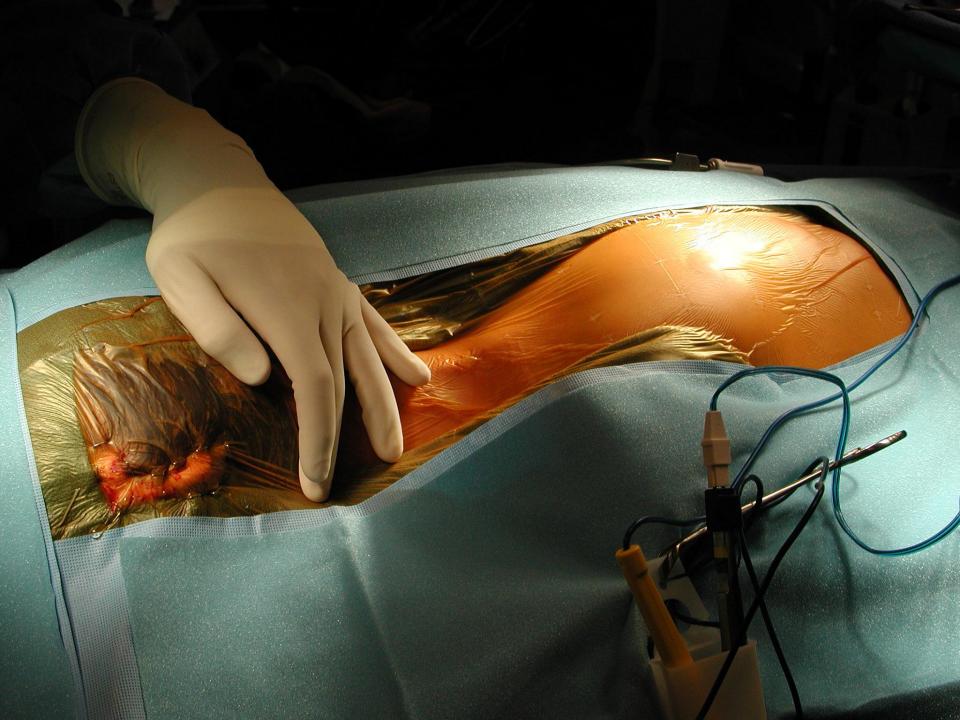
• Age at Treatment

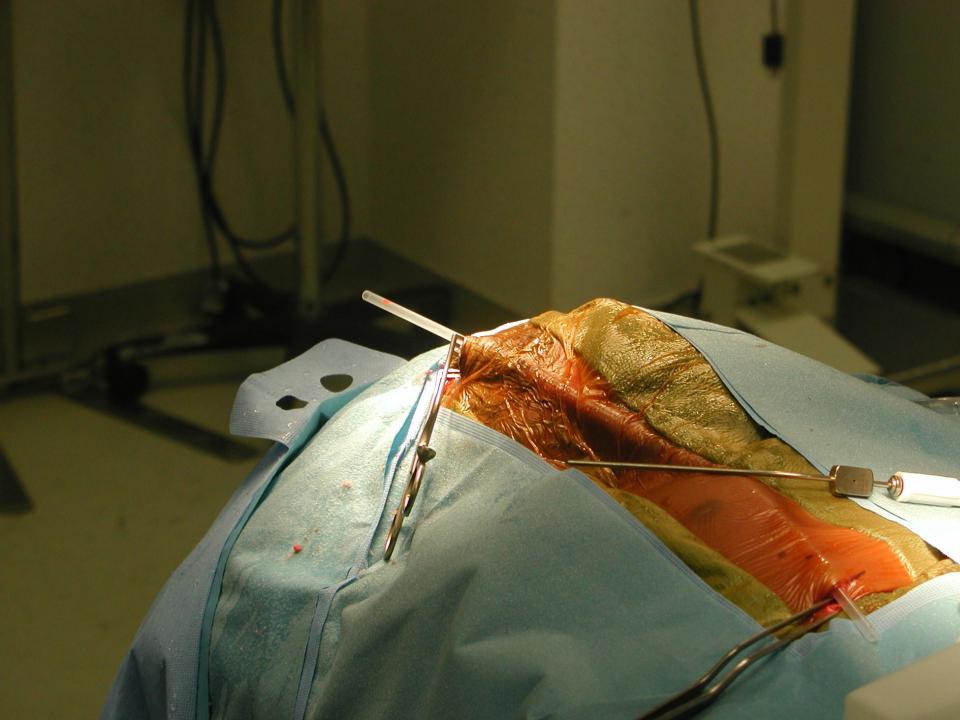
For infants with overt hydrocephalus, clinical experience points to a critical limit of age 2-5 month for shunting to achieve a good outcome













- 1. No traffic
- 2. IV abx
- 3. IT Vanco
- 4. Abx irrgation (Baci/Kefzol)
- 5. Triple prep

2010: Infection rate 1.3%!

Implications of a shunt

• Lifetime dependence

Shunt Malfunction

- 30-40% failure rate in 1st year¹
- @ 4-6 revisions / 20yrs¹
- independent of shunt type, 50% in 2 years²

Proximal Shunt Malfunction

- choroid plexus / ventricular surface:
 <u>OVERDRAINAGE</u> siphoning,
 one way valve
- too short
- poor positioning



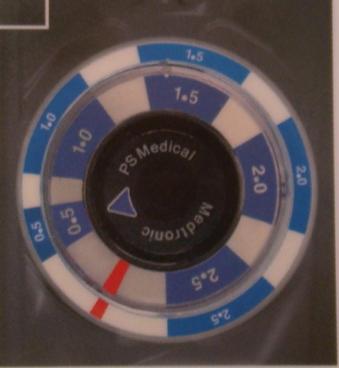
ett ustabl



(Right) Keeping the Locator Tool in position, set the Indicator Tool into the Locator Tool while aligning the red bands on the tools and rotating the Indicator Tool as necessary until it settles into place. Record current performance level setting.

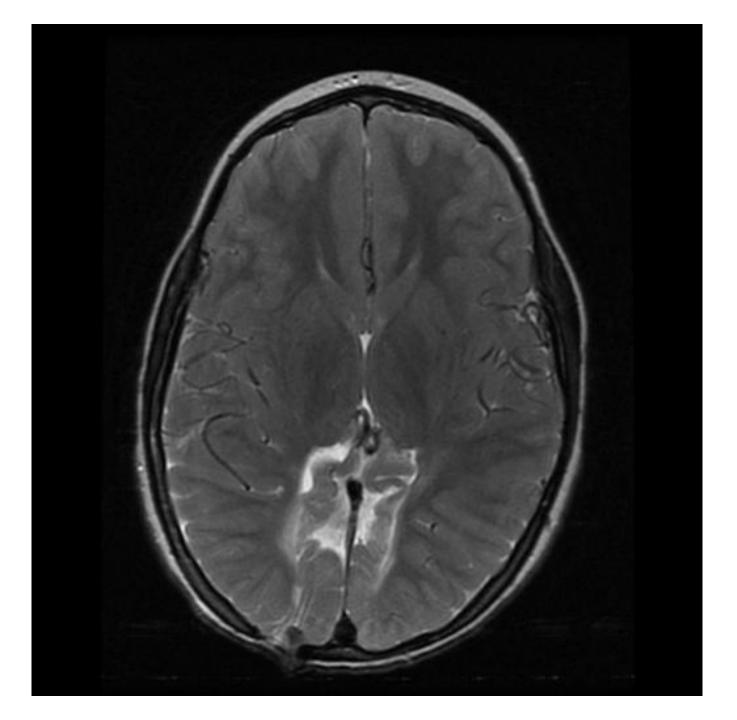
NOTE: All valves are preset to

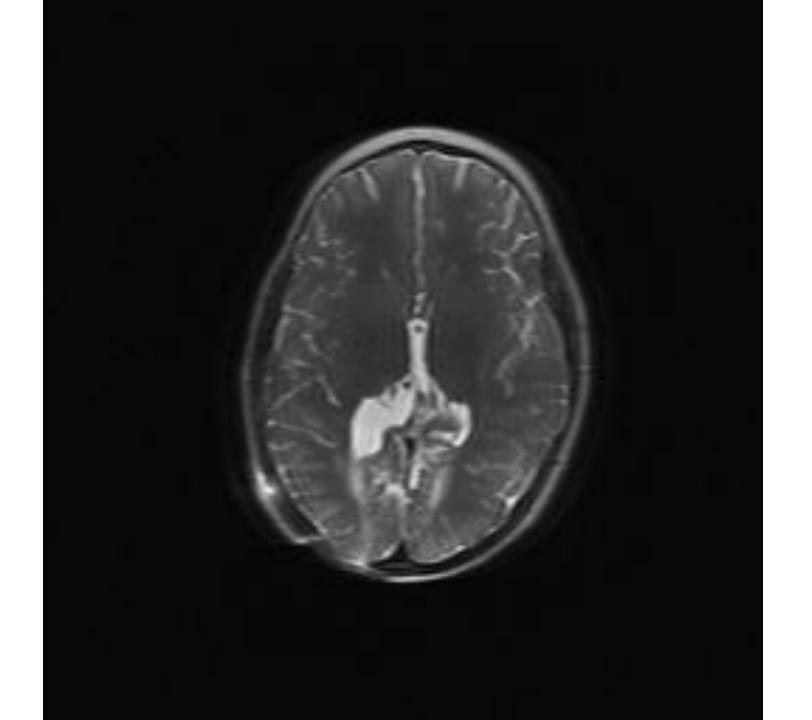
(Left) Do not remove the valve from the sterile packaging. Turn sterile package face down so that the translucent tray and valve are facing up. Position the Locator Tool above the valve so that the Locator Tool's blue flow direction arrow is aligned and centered with the direction of CSF flow through the valve.

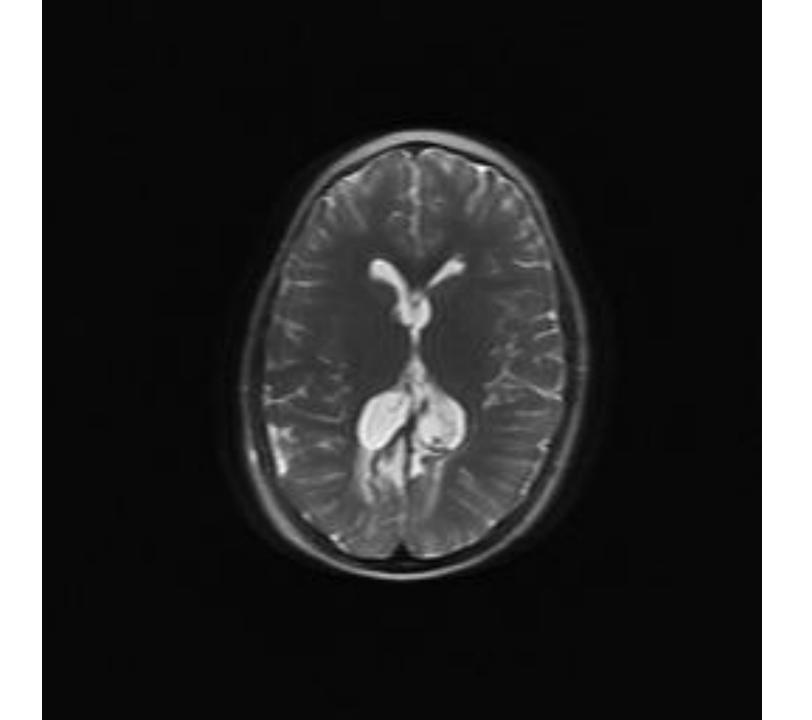












Adjustable Shunts + Intraoperative Imaging+ Computer Navigation

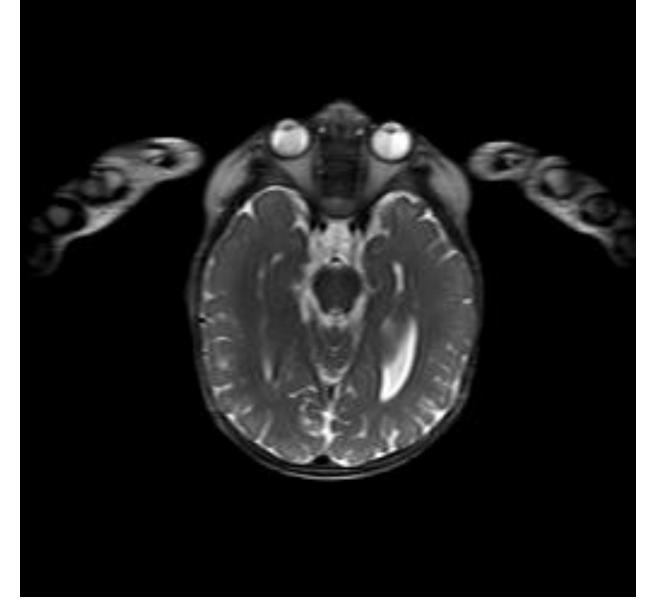
Shunt Revision: 2001: N=128 (240)

2010: N=47 (400+)

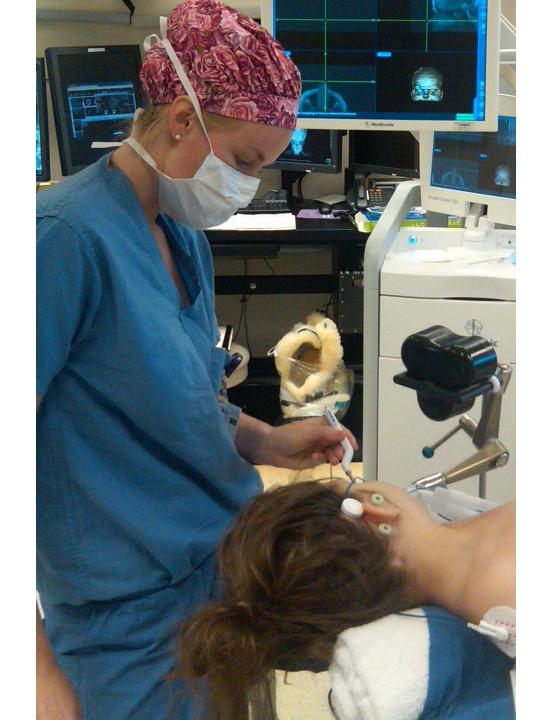








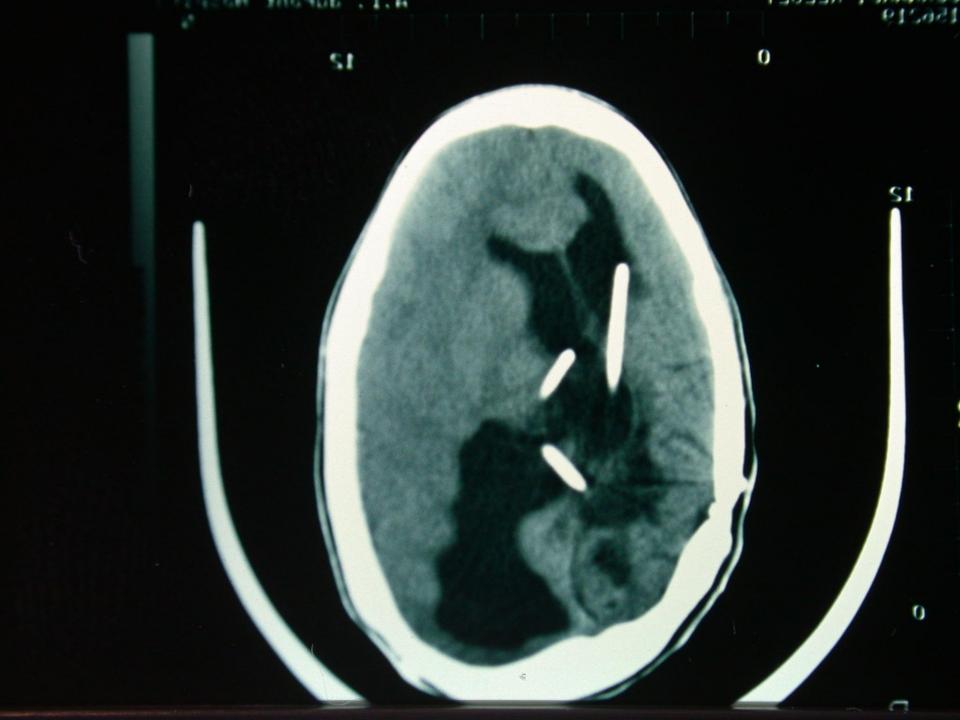
No more CT scans!!! "quick MRI": no sedation, no radiation!!



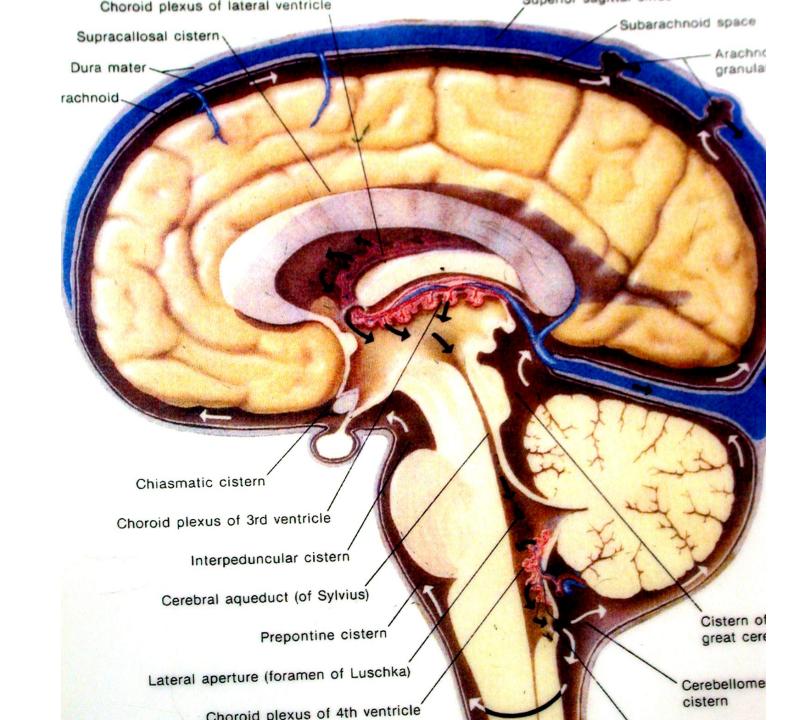
• Endoscopic fenestration:

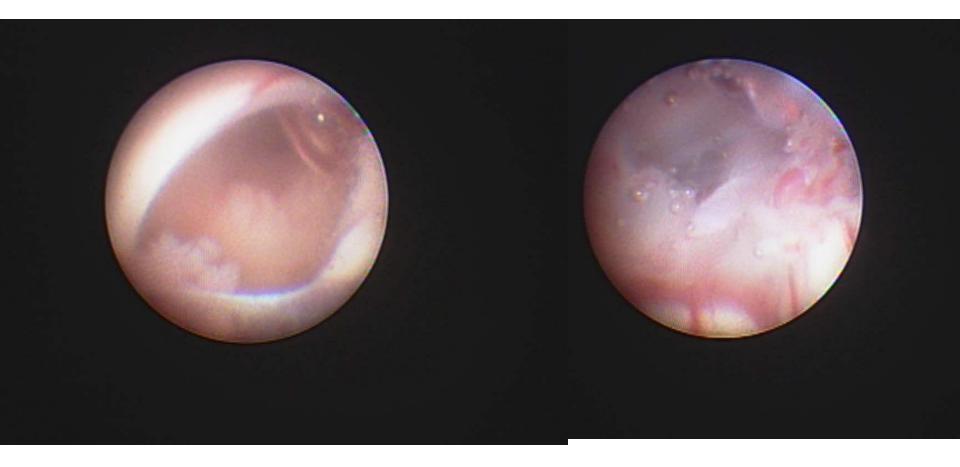
- large instrumentation not ideally suited for neonatal brain
- Endoscopic 3rd ventriculostomy
- obstructive hydrocephalus
- relies on well developed CSF circulation, high failure rate <2yo

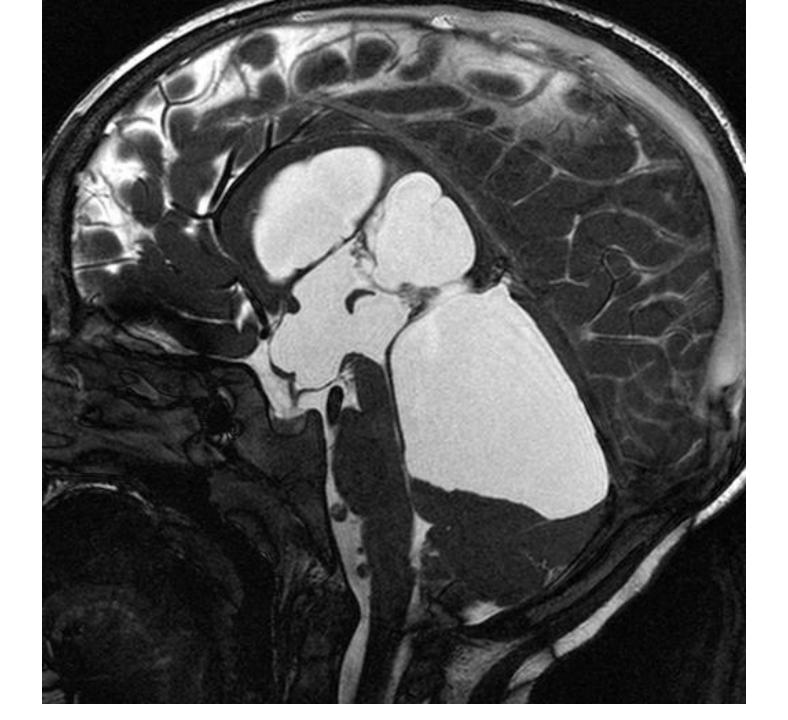




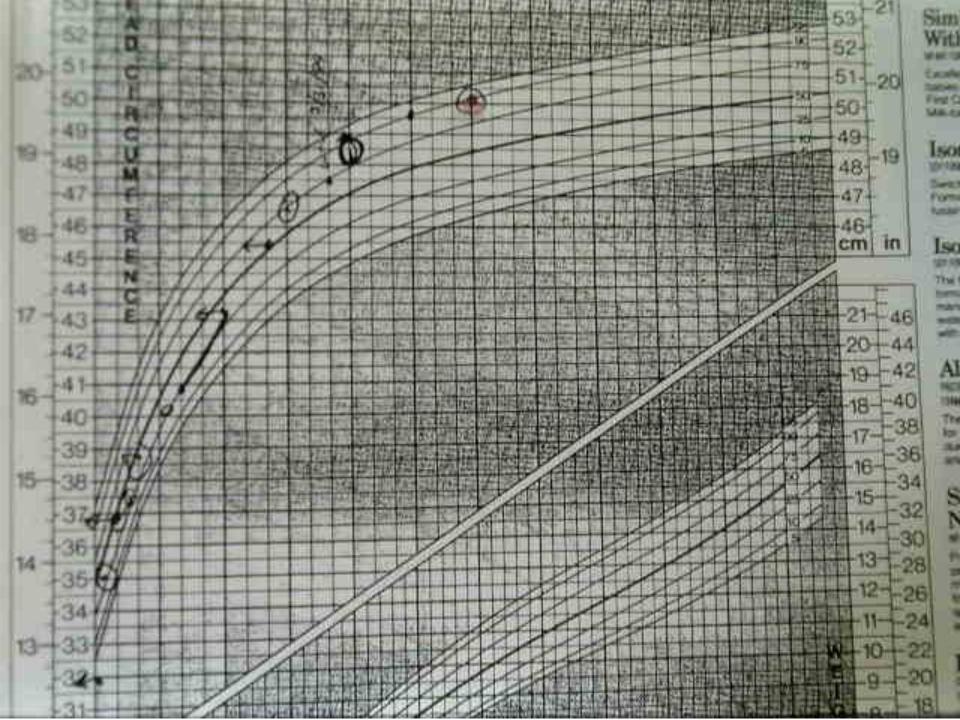










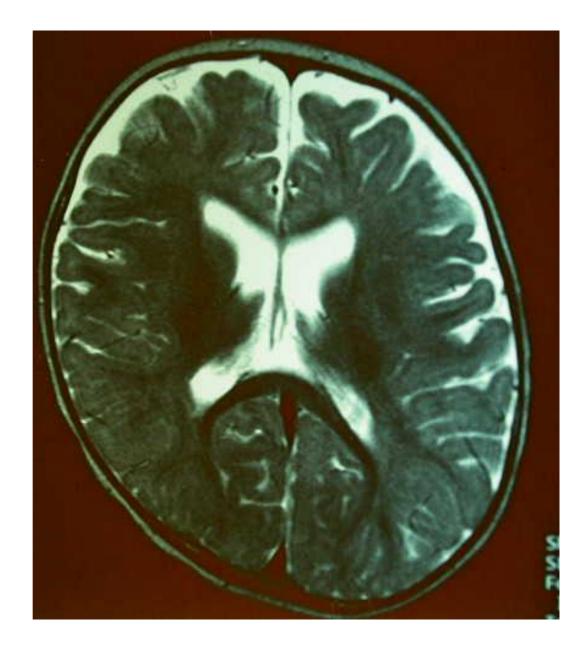


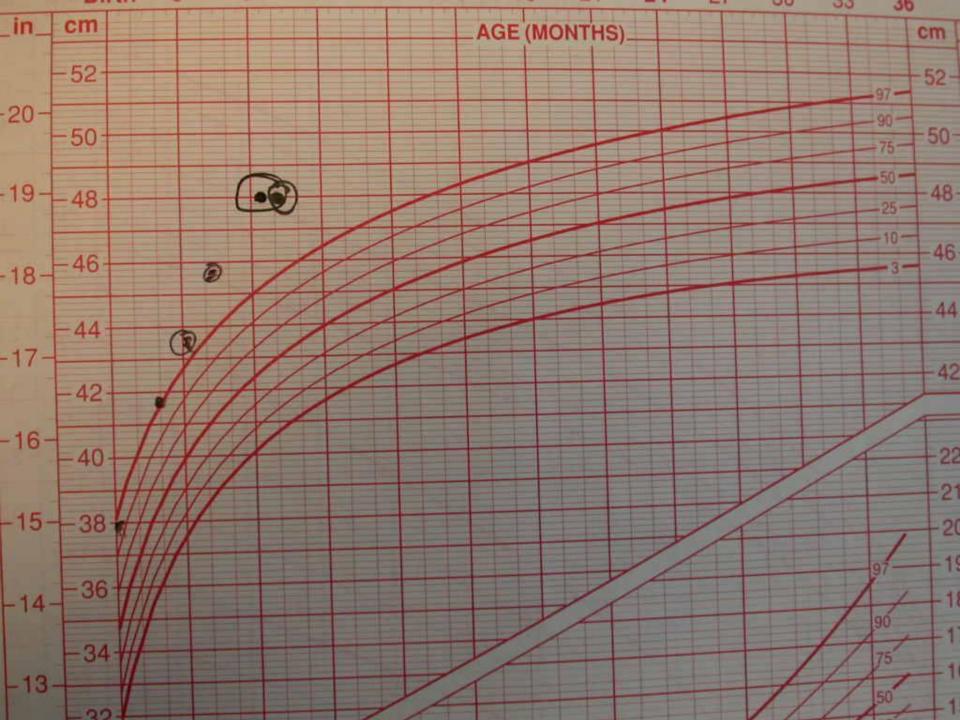
Benign External Hydrocephalus*

Presentation: Macrocephaly Imaging: Prominent SA spaces with mild ventriculomegaly Resolves by 12-24 mo Developmentally normal (40% interim motor delay) Associated with positional plagiocephaly 88% had family history of macrocephaly

Do not Shunt

*Alvarez L, Pediatrics 1986



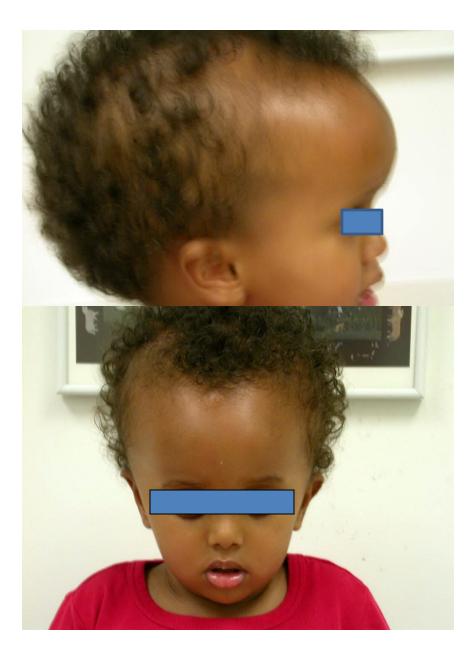


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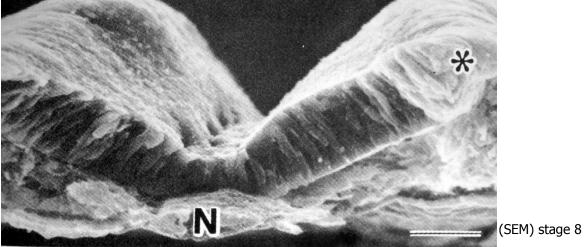
> SE TR:500 TE:20 EC:1/1 16kHz

HEAD FOV:20x20 5.0thk/1.0sp 16/03:16 04:14:52 Mag = FL: ROT:

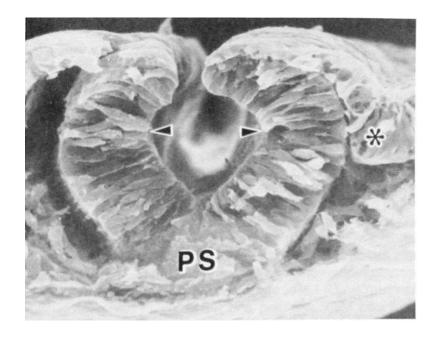


Parietal bossing

Primary Neurolation - (C1-S2)



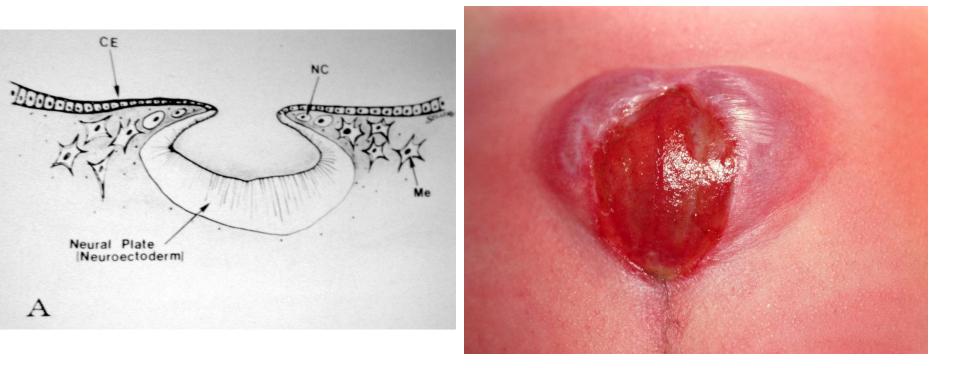
(SEM) stage 8 chick embryo



Abnormal Dorsal Midline Neural Development: conceptual classification

Abnormal Primary Neurulation

Failure of Neural Tube Closure - Neural Tube Defects: spina bifida aperta / myelomeningocle



Tethered spinal cord

• Defective dorsal midline formation (*spina bifida, myelodysplasia, spinal dysraphism*)

• Allows inelastic mesenchymal elements to fuse with the spinal cord at the caudal level of malformation

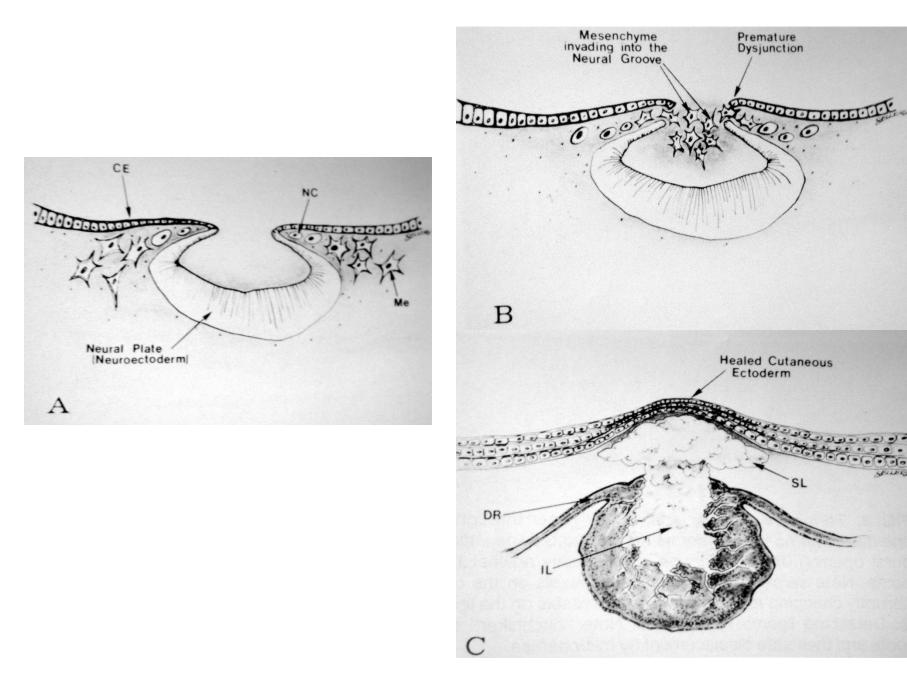
• Tethers the spinal cord during rapid spinal column growth

Tethered spinal cord

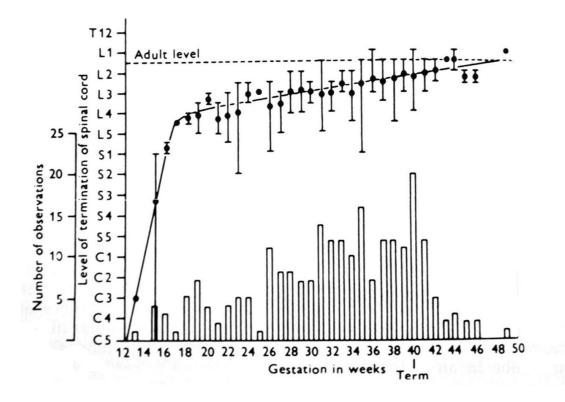
Stretched spinal cord, conus below L2

Altered blood flow, stretched neuronal membrane, neuronal and interneuron degeneration

Progressive neurological deficits



Ascent of the Conus Medullaris

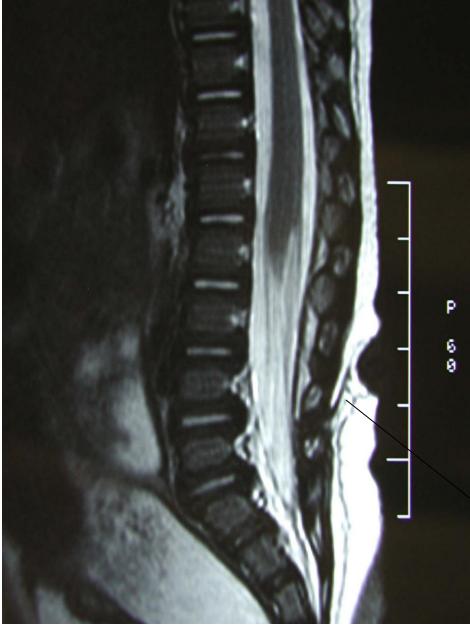


Tip of conus at birth: L2-3 interspace Tip of conus >2months of age: **L1-2** interspace

• Tip of the conus at or below L2-3 suggests spinal cord is tethered

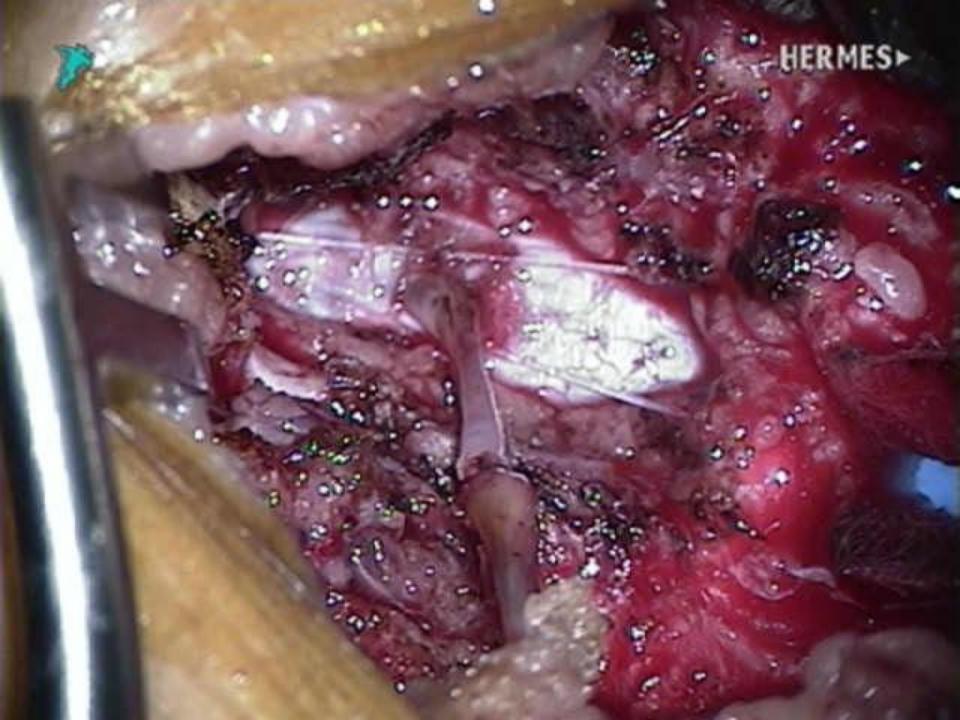


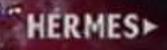




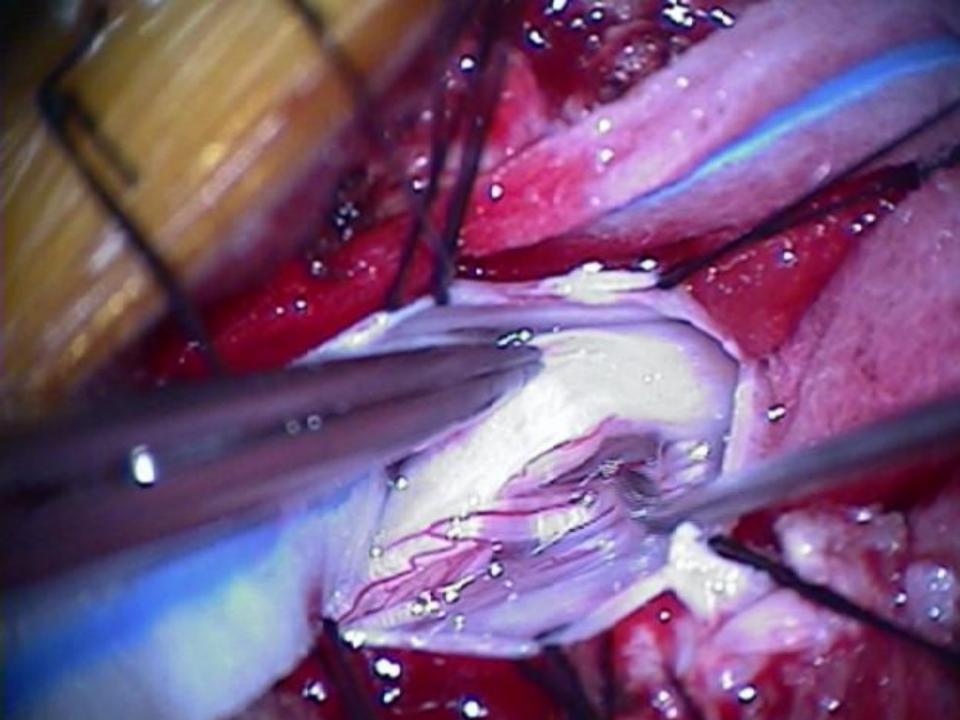


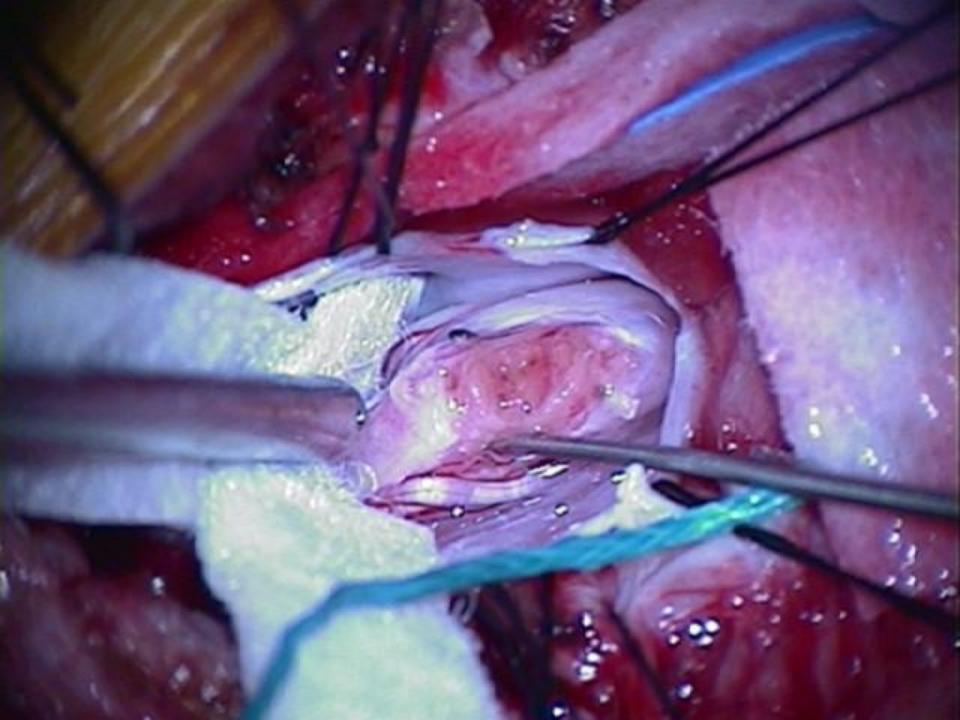
Subcutaneous tract

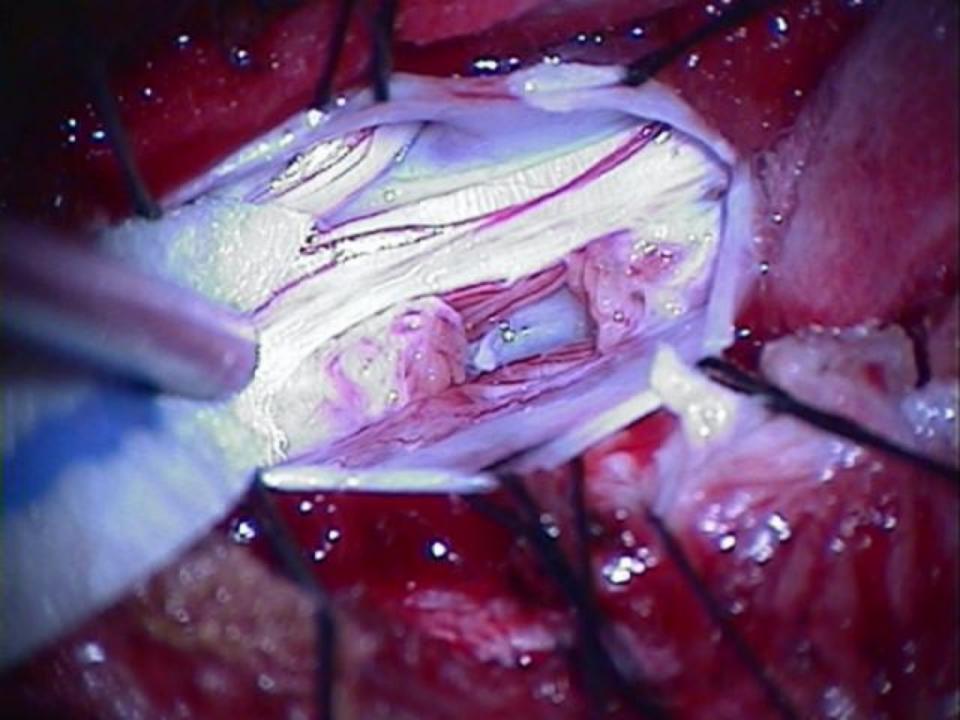












Tethered spinal cord Diagnosis

80-90% have cutaneous manifestations

MRI screening for high risk patients with cutaneous manifestations and those patients with associated caudal abnormalities

Tethered spinal cord Cutaneous Manifestations

- 1. Dimple above the crease
- 2. Capillary Hemangioma / dystrophic skin
- 3 Subcutaneous Lipoma
- 4. Hair Tuft
- 5. Asymmetrical Crease
- 6. Midline Appendage

Tethered spinal cord Cutaneous Manifestations

1. Dimples that represent tethered spinal cord from a dermal sinus tract is above the intergluteal cleft (S2)





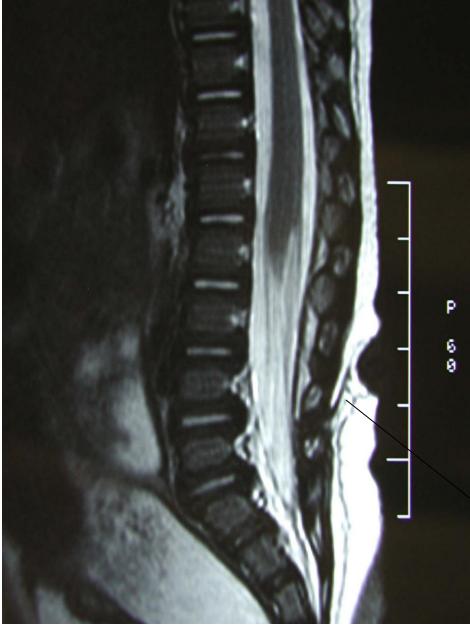




Dimples

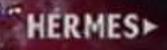
•Shallow or deep

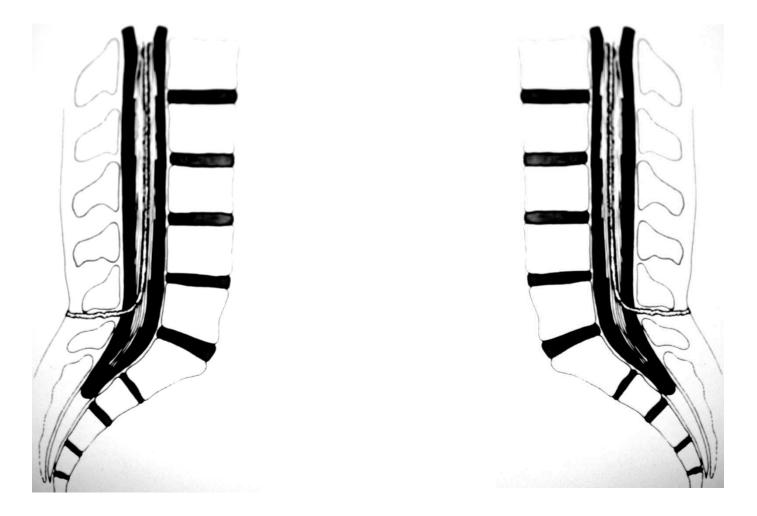
•Frequently associated with dystrophic skin, hemangioma



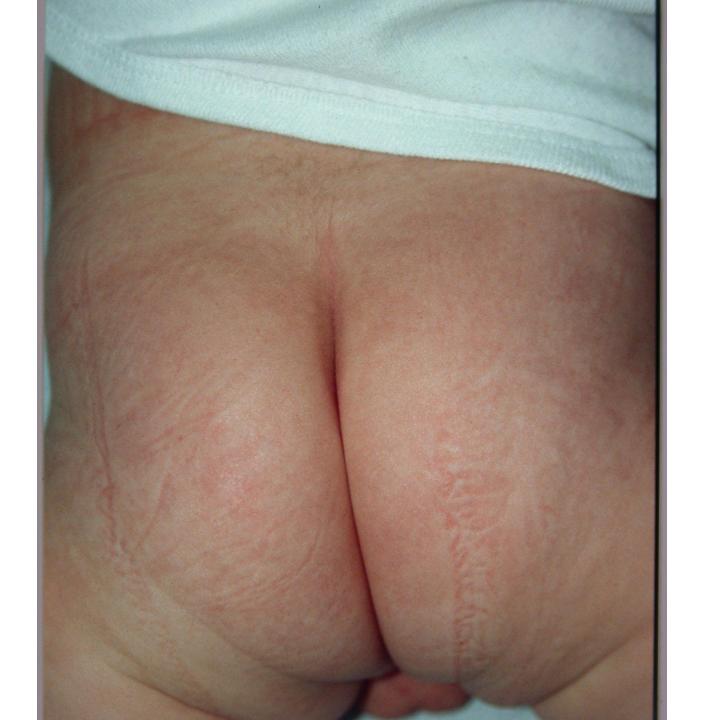


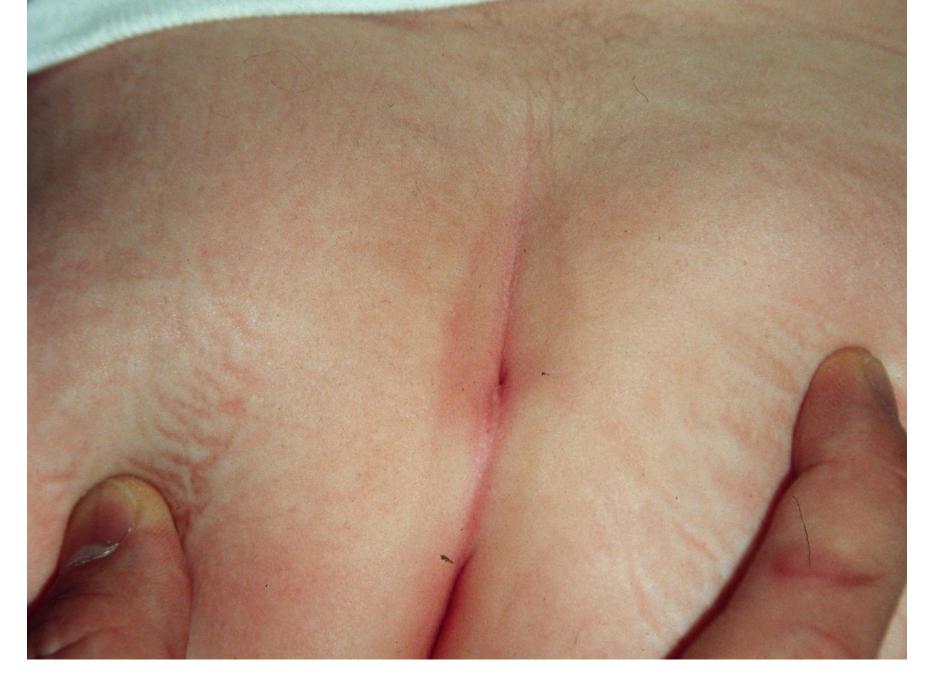
Subcutaneous tract



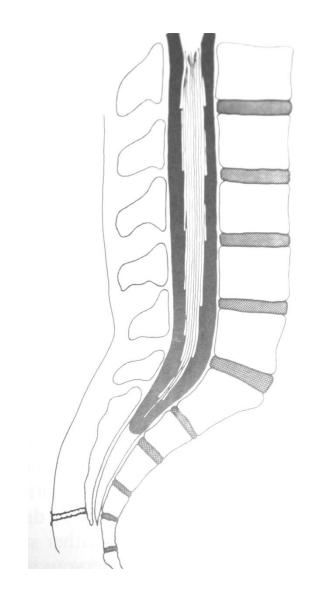


Dermal sinus tract: tethering lesion mass lesion source of infection (epidural abscess/meningitis)

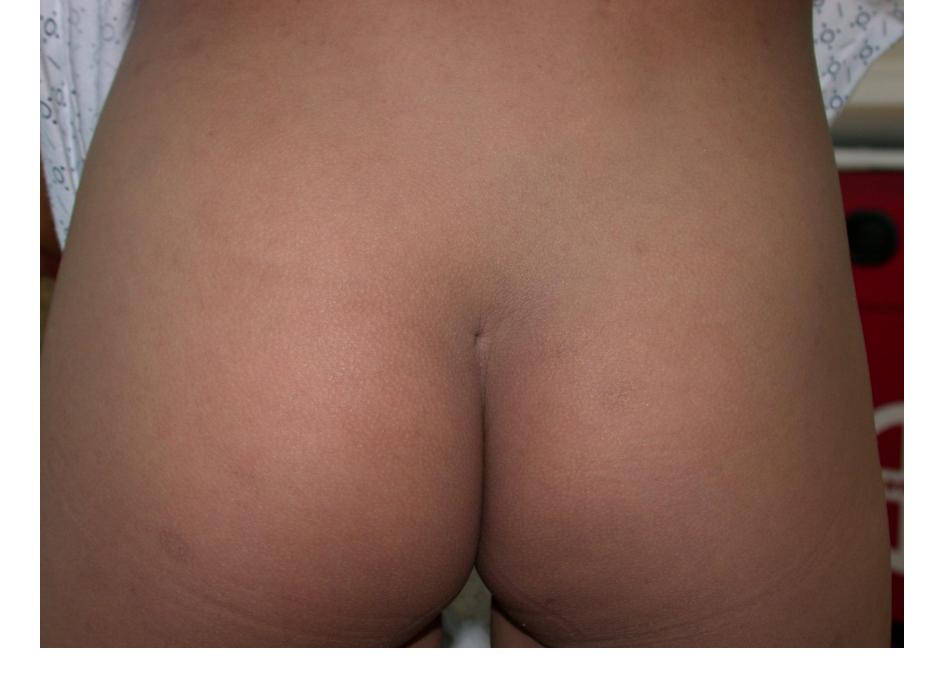




Dimple in a straight crease= normal



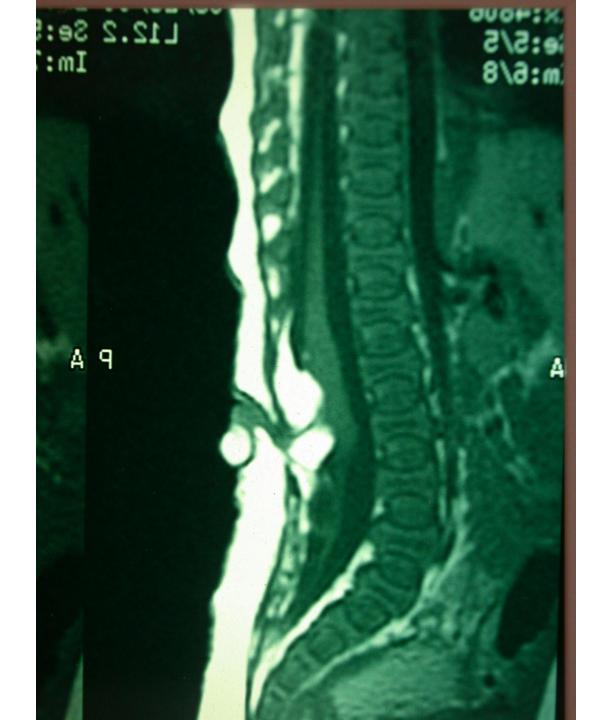
Sacral dimple: -in the crease -near the tip of the coccyx -does not extend into the spine -2% of infants



Dimples above the crease= not normal



Not normal





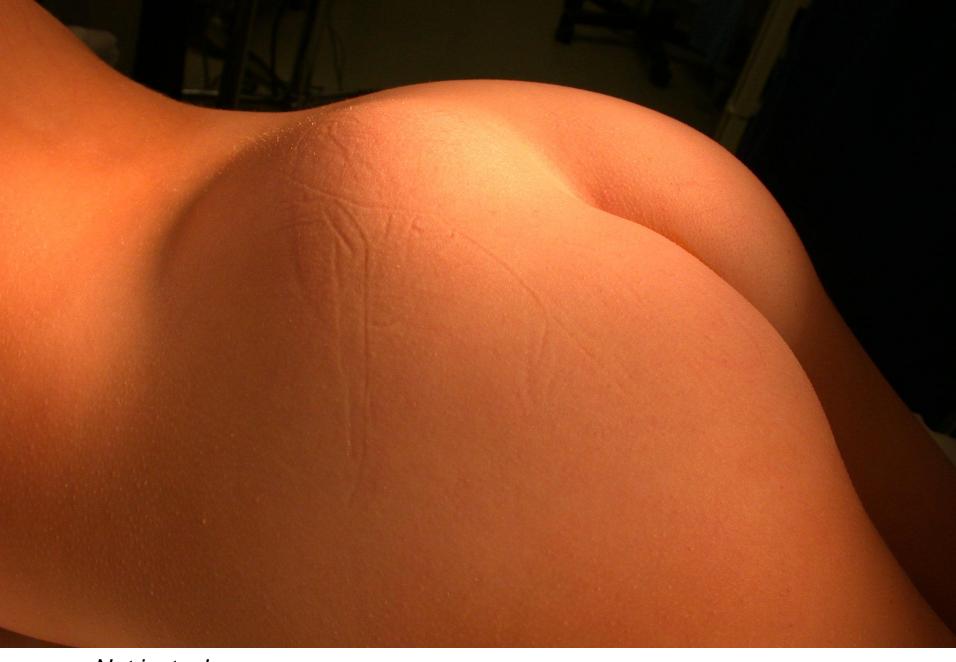
Lumbar lesions=not normal







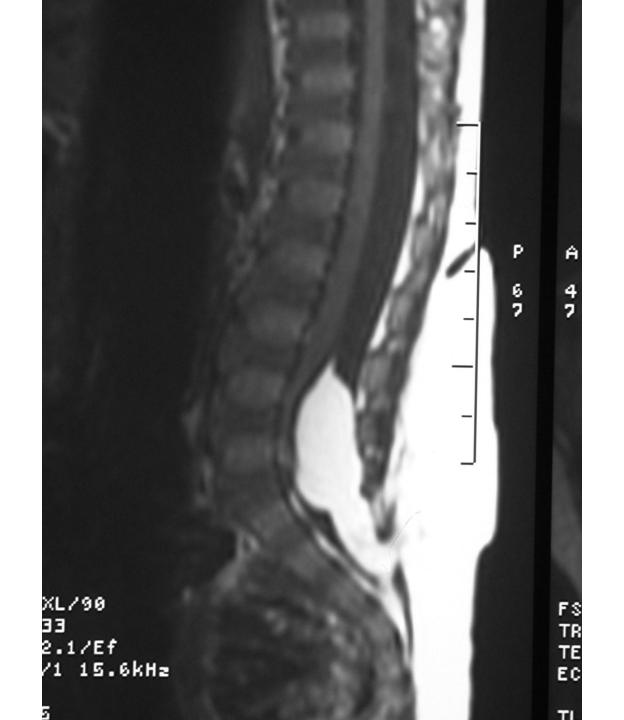
Smooth capillary hemangiomas may be normal

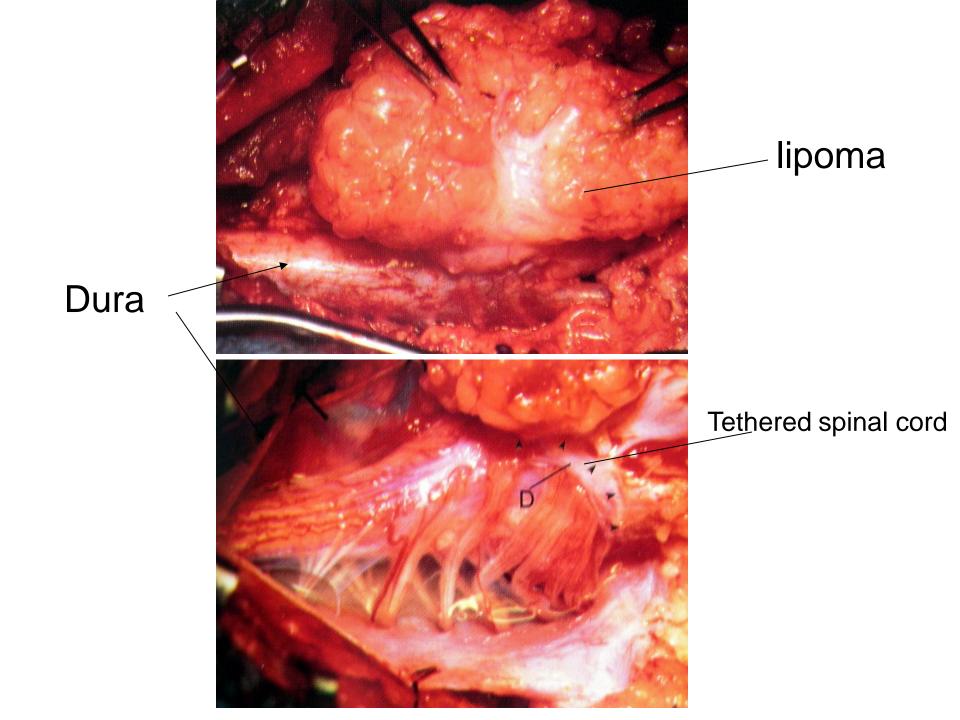


Not just a bump



Like wise.....



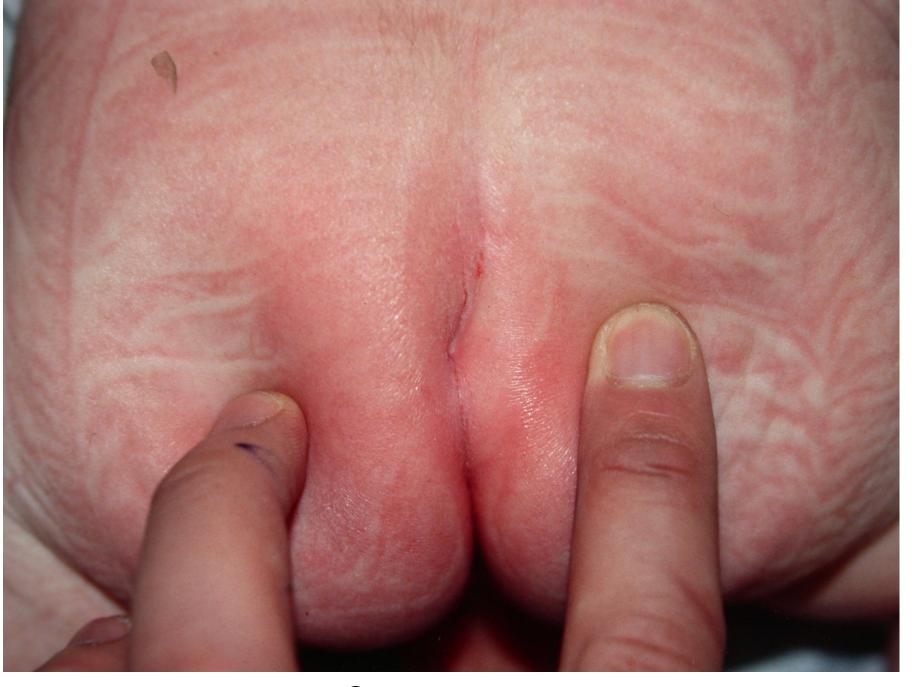




This hair is too long







Minor crease deviation @15% tethered cords, needs screening





Not just a skin tag...

Tethered spinal cord Associated caudal lesions

MRI recommended

- Sacral agenesis
- Imperforate anus (54%)
- Cloacal exstrophy, VATER
- Urinary tract anomalies
- Genital duct anomalies

Tethered spinal cord Management

MRI: 3 months of age

Surgery:

-Generally, tethered cords should be untethered
6-12 months of age: relatively low risk,
no transfusions, 3 days hospitalization
- Routine neurophysiological monitoring

Pediatric Brain Tumors:

-Almost never present with headaches alone
-Tend to be midline, and large
-Biology determined by both histology *and* location developing molecular characterization
-Most are curable; in many others long term control achieved through combination of surgery, chemotherapy and radiation
-Long term sequelae requiring requiring comprehensive

pediatric subspecialty care

Pediatric Brain Tumors:

Incidence rate: 3.9 cases per 100,000 children (0-19)

@2500 new cases in the United States 2000¹

1. Central Brain Tumor Registry of the United States (CBTRUS)

<u>Site of Origin</u> (1038 Pediatric CNS Tumors)

Cerebrum (30%) Posterior Fossa (37%) Ventricular (10%) Optic Nerve/ Chiasm (9%) Sellar/Parasellar/Pituitary (7%) Spinal Cord (4%) Pineal (2%)

Presentation: Symptoms*

(supratentorial / infratentorial)

Headache +1: 99.1% / 99.4% +2: 87.7% / 93.1% +3: 64.8% / 71.3%

```
nausea/vomiting (71% / 86%)
seizures
speech/personality/academic performance change <6 (87% / 80%)
visual symptoms/diplopia (90% / 80%)
difficulty walking/balance (65% / 92%)
motor weakness
bladder symptoms
neck pain
back pain
failure to thrive
```

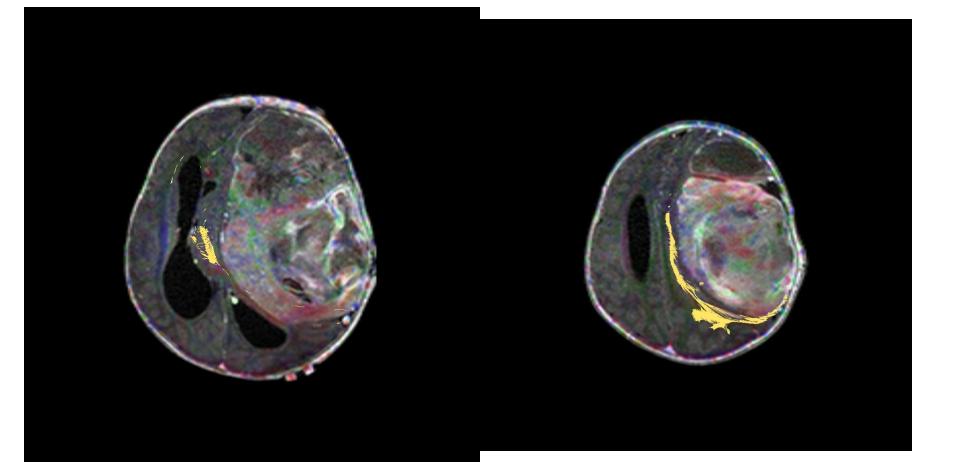
Presentation: Symptoms* (supratentorial / infratentorial)

Headache +1: 97.7 / 99.0% +2: 90.8 / 97.2% +3: 82.6 / 92.5 %

confusion/stupor head tilt lethargy paresis ataxia papilledema (64%/80%) visual field defect >4 (55%/9%) tense fontanel coma irritability stiff neck abnormal reflexes hypesthesia >3 decreased visual acuity CN palsy: VI, VII

*The epidemiology of headache among children with brain tumor, *J Neuro-oncology* 10: 31-46, 1991



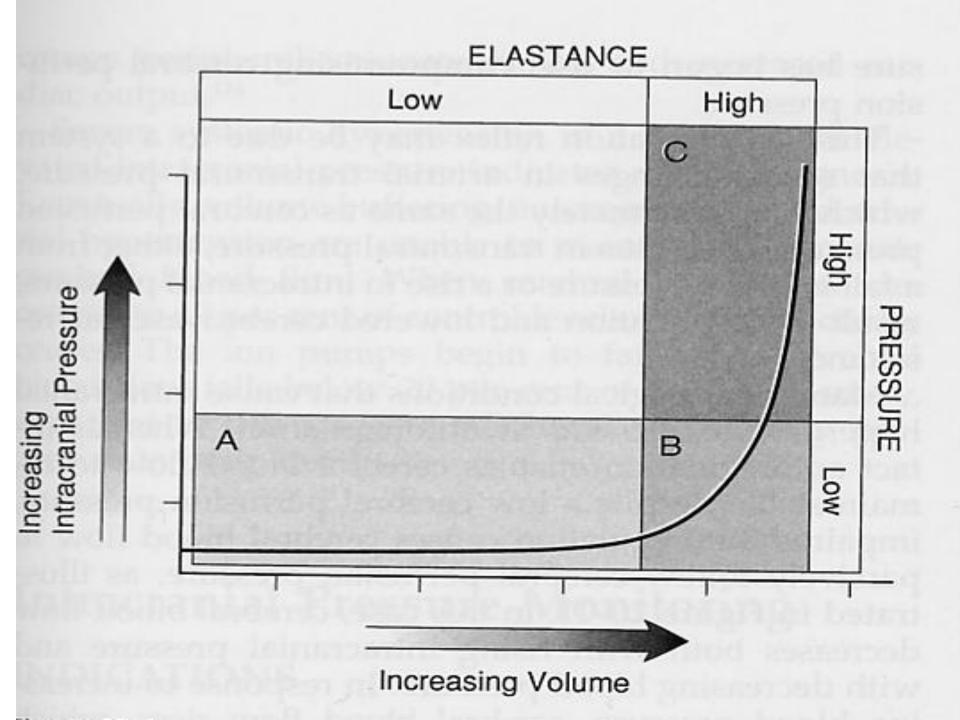


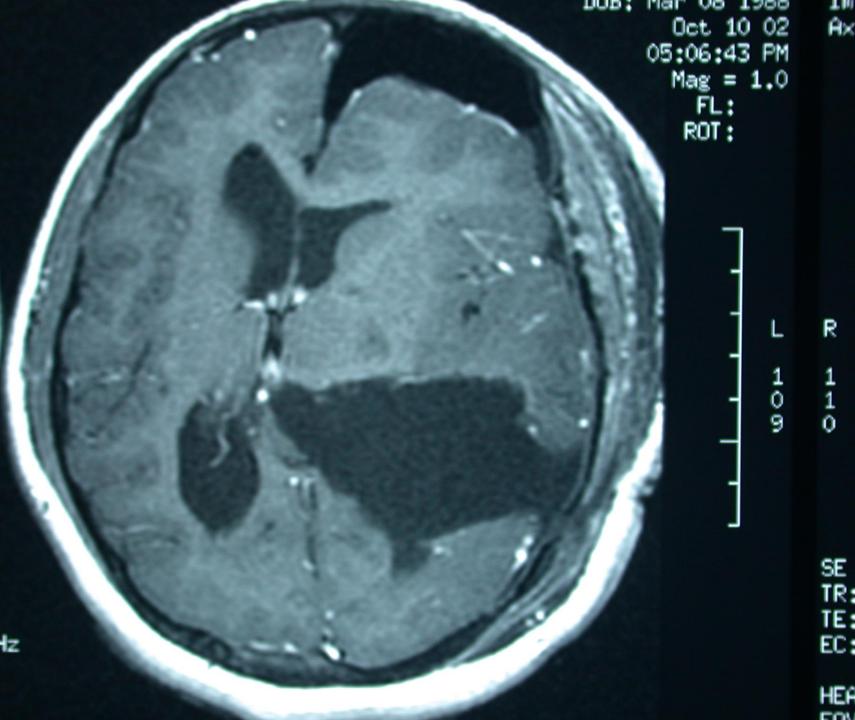
R

 $1 \\ 1 \\ 0$

SE TR:250 TE:8/Fr EC:1/1 16kHz Mag = FL: ROT:

HEAD FOV:22×16 5.0thk/2.5sp





:233 :14 :1/1 16kHz

AD 1+22-16

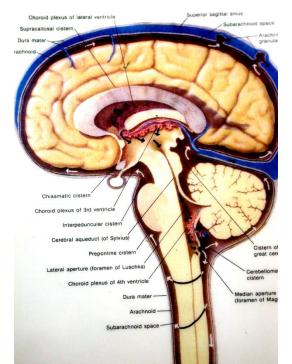
\$6.6+C

Low Grade Gliomas

Good

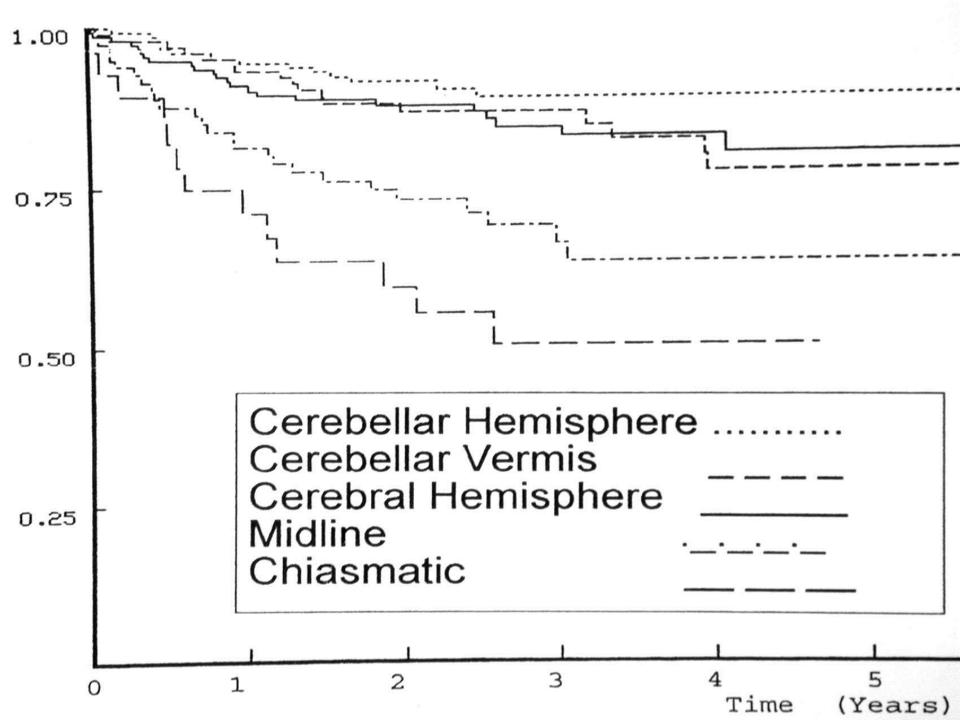
JPA (may have well defined margins)

cerebellum cerebral hemisphere tectum midbrain spinal cord



Bad

Fibrillary (infiltrative) thalamus hypothalamus basal ganglion pons optic pathway/chiasmatic



Low Grade Gliomas

Treatment:

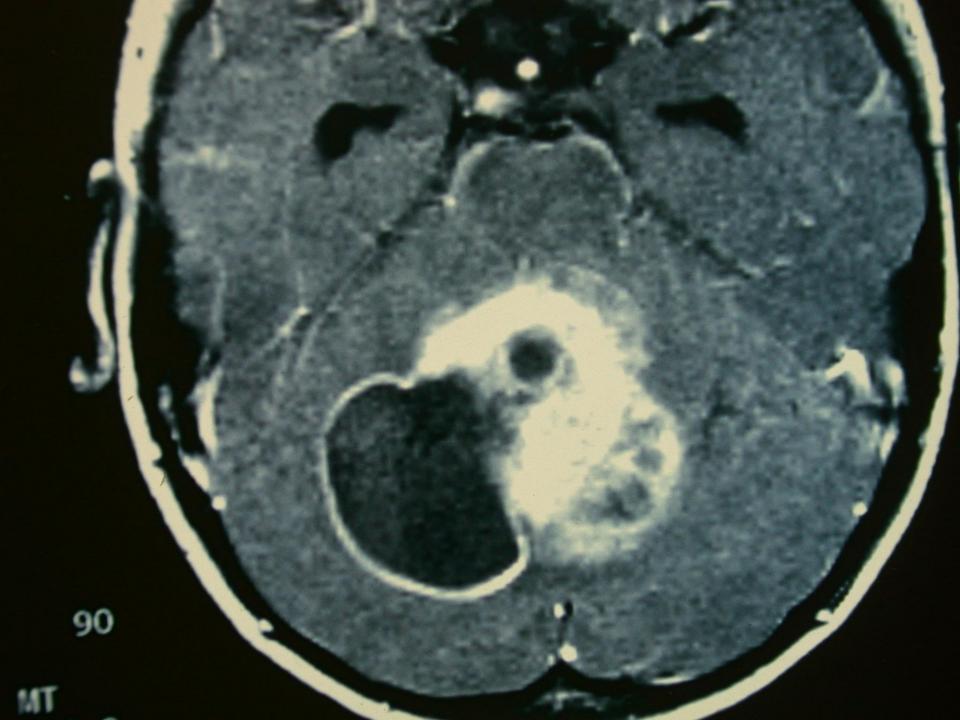
Surgery for tissue diagnosis and remove as much tumor safely as possible.

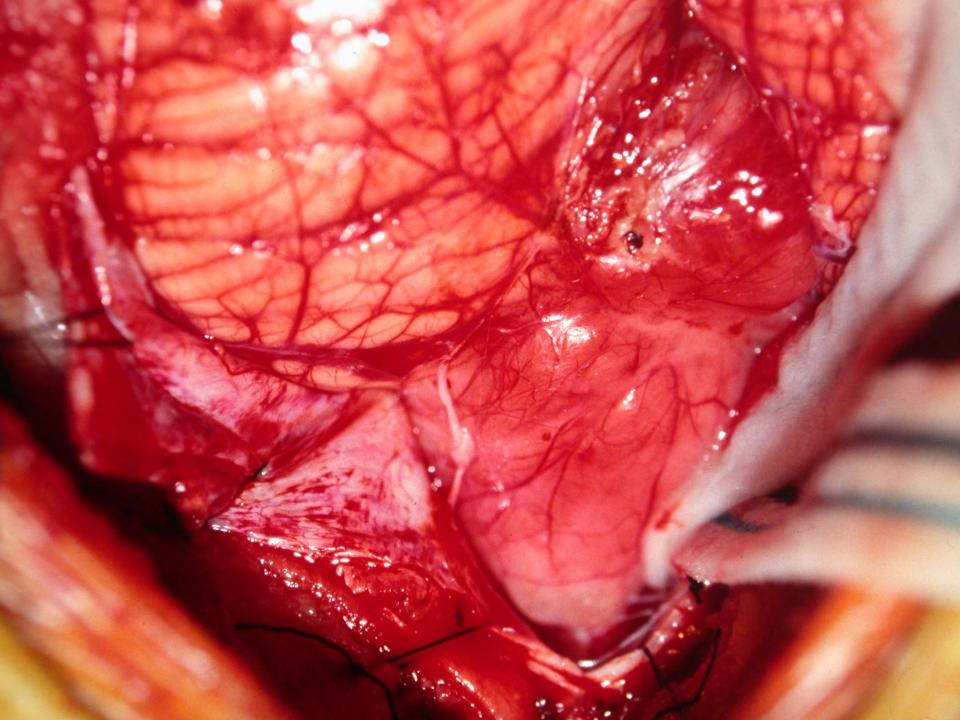
-Total resection offers long term DFS and is potentially curative (95-100% 10yr DFS for cerebellar astrocytomas)

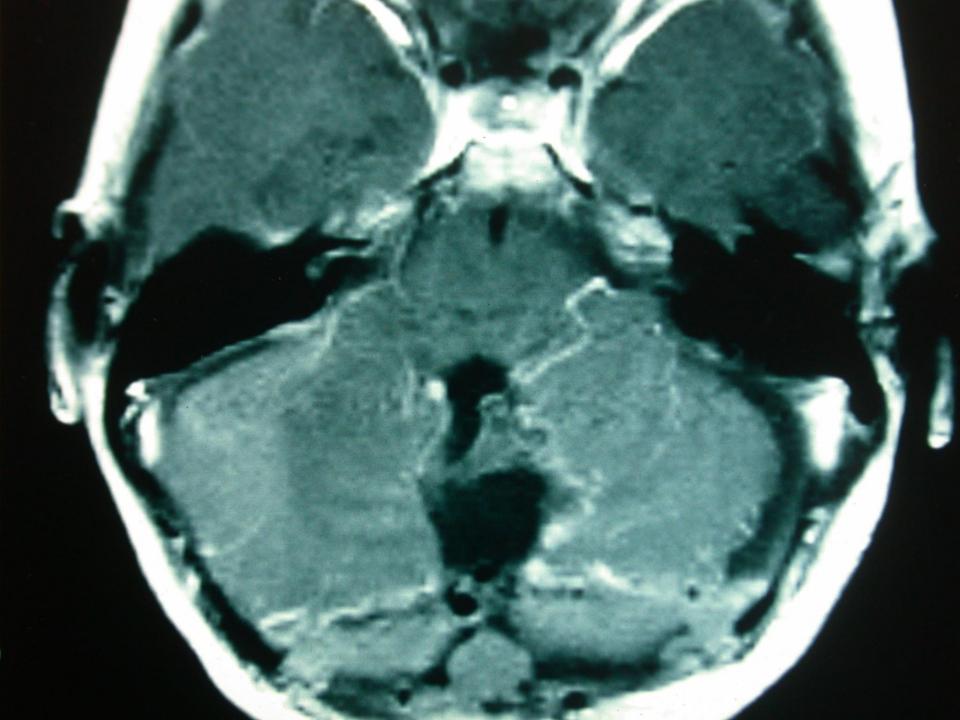
- -Resection limited by certain locations and infiltrative margins
- -Radical resection >90-95% improves EFS (84% vs. 65% 2yr EFS Wisoff,

CCG9891, 726 pts 1991-1996)

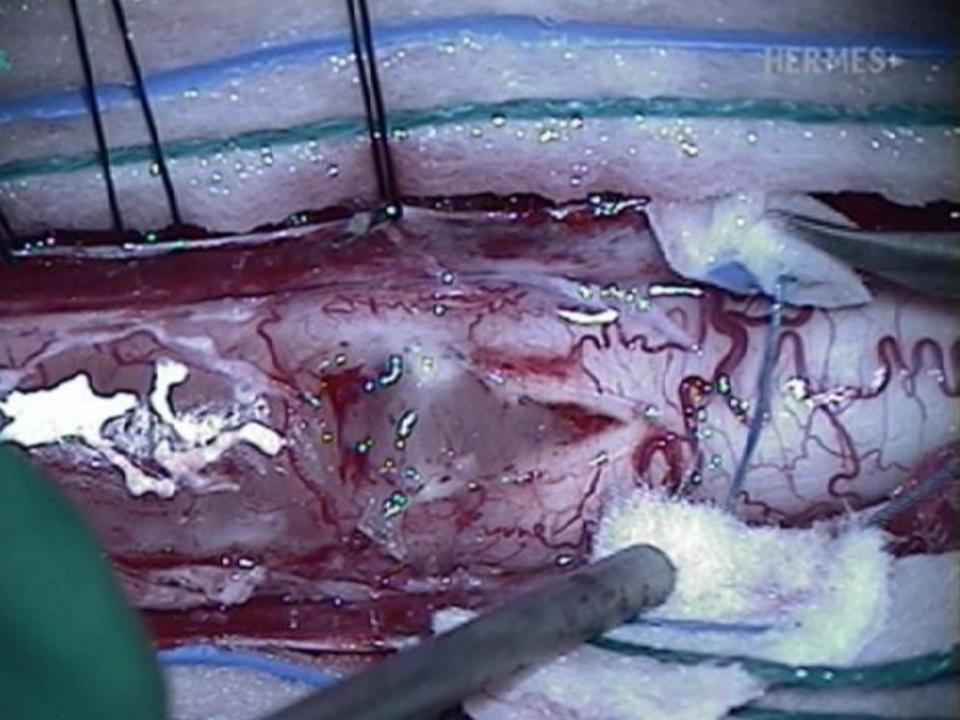
- -small residual disease may be followed
- -mindful that adjuvant therapy can offer long term control

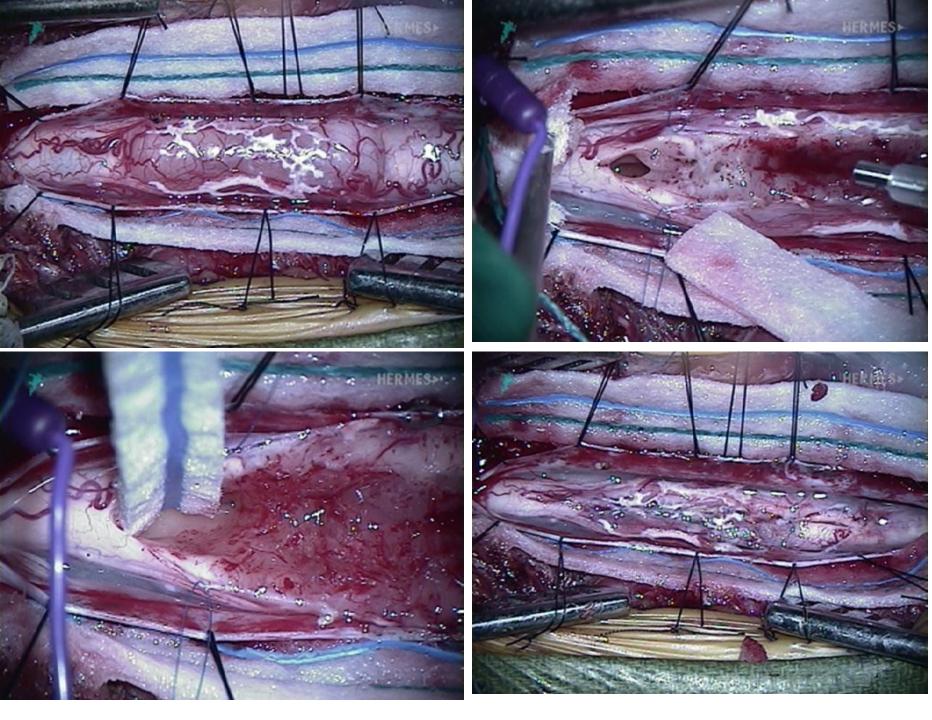






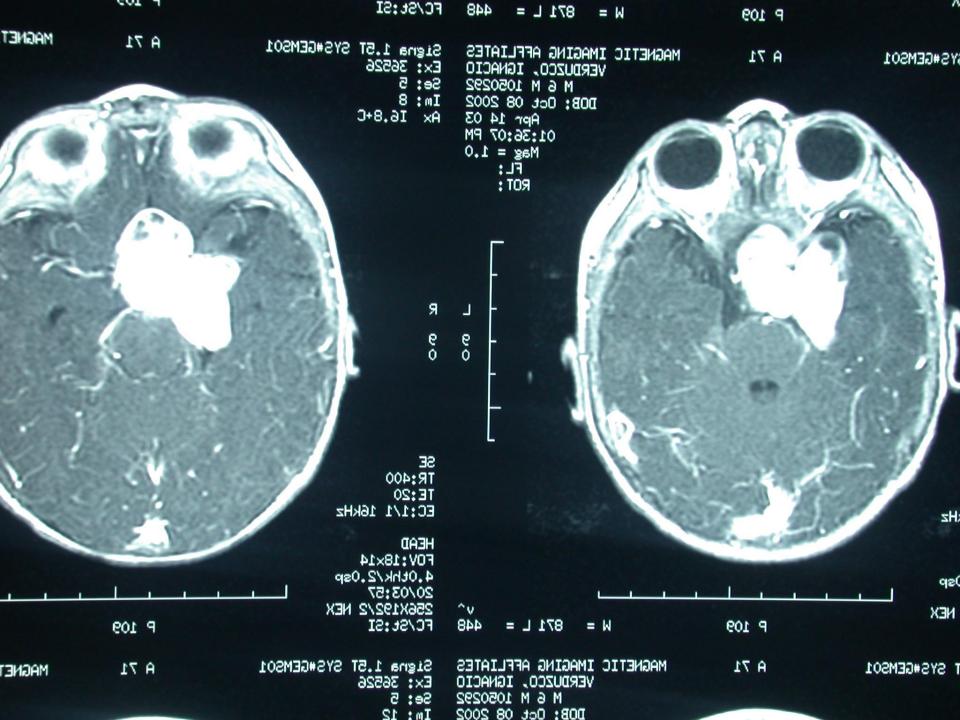


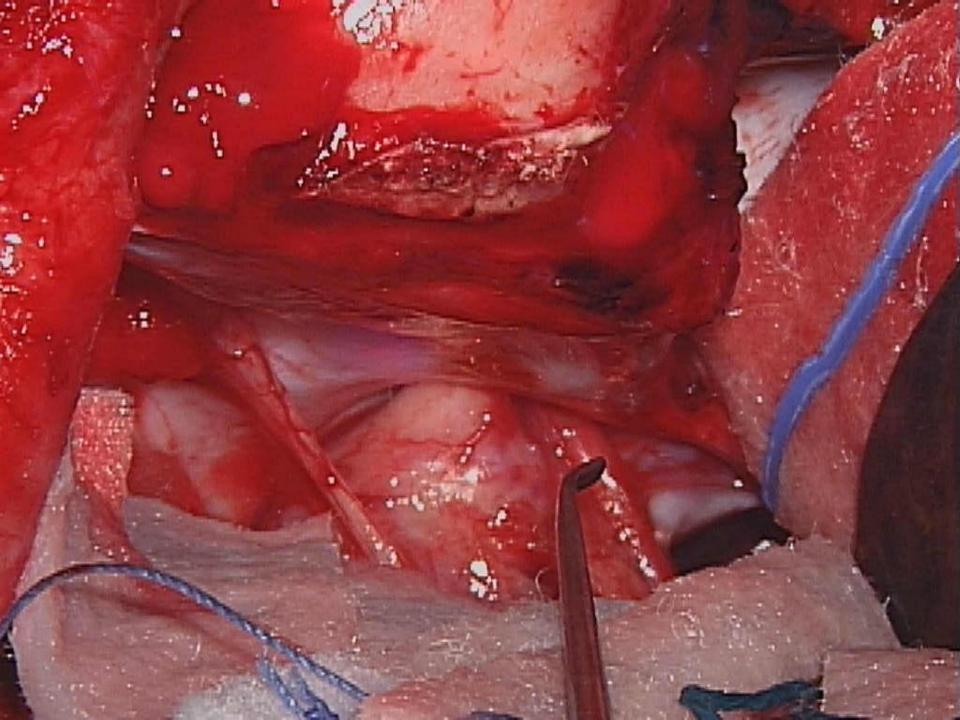


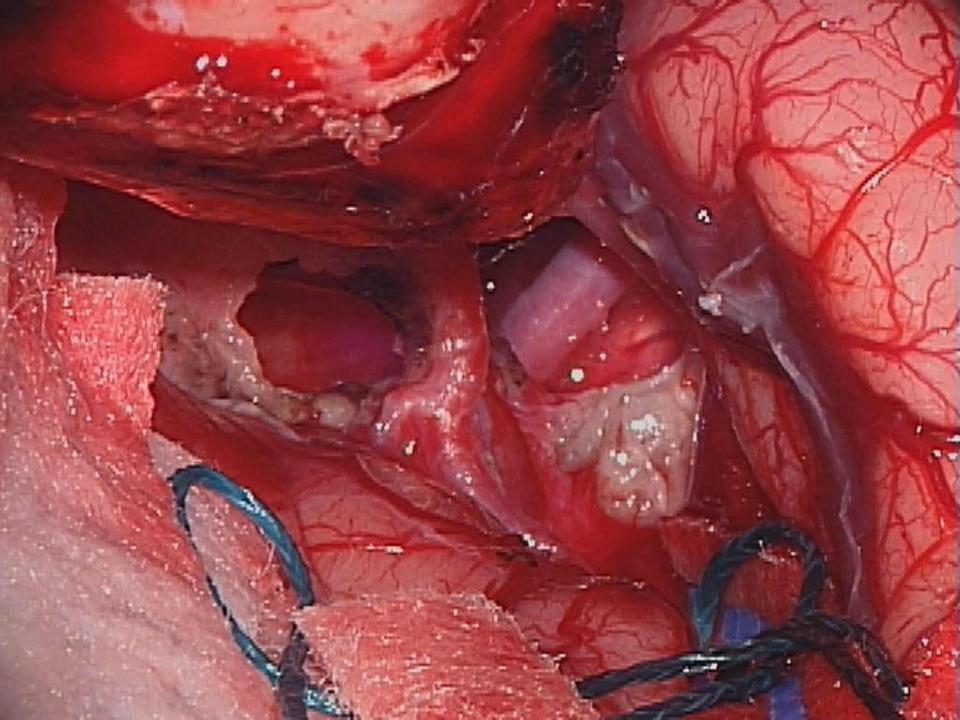




All gone

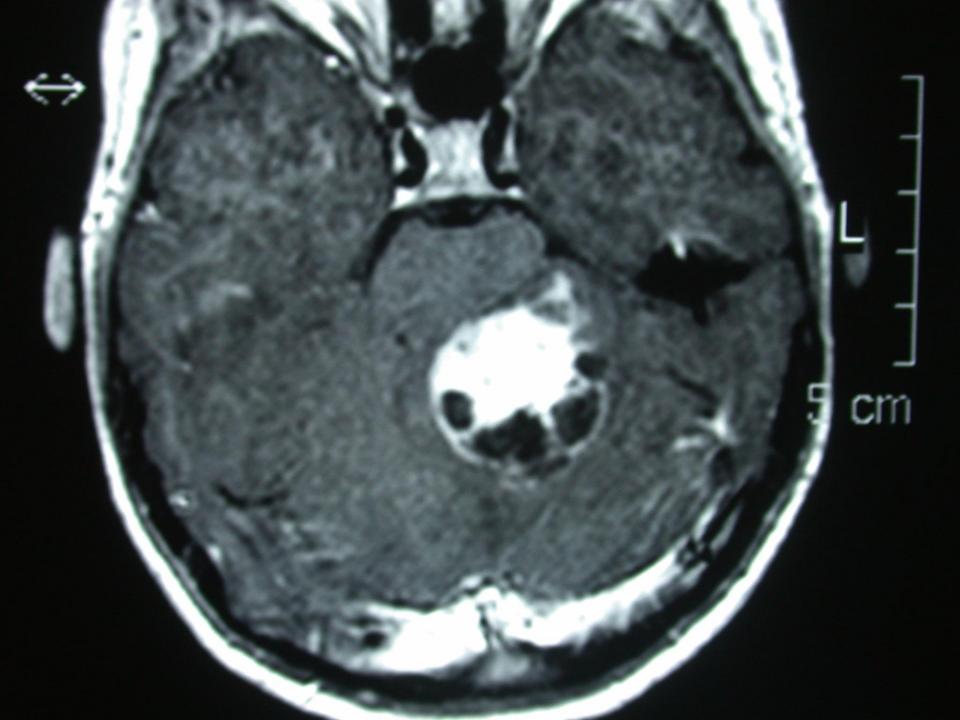


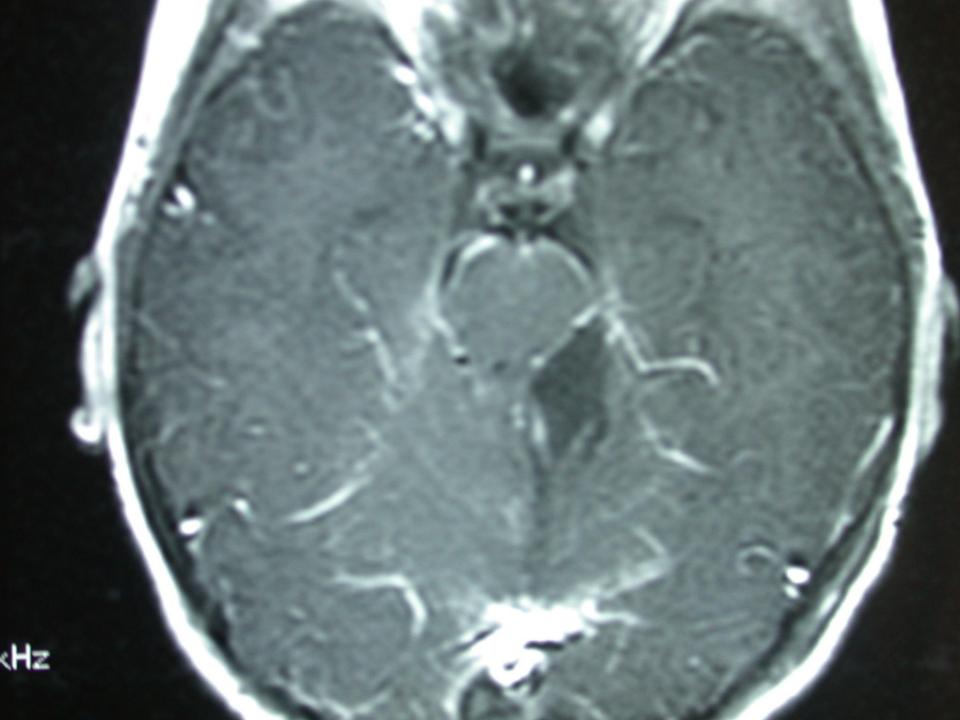


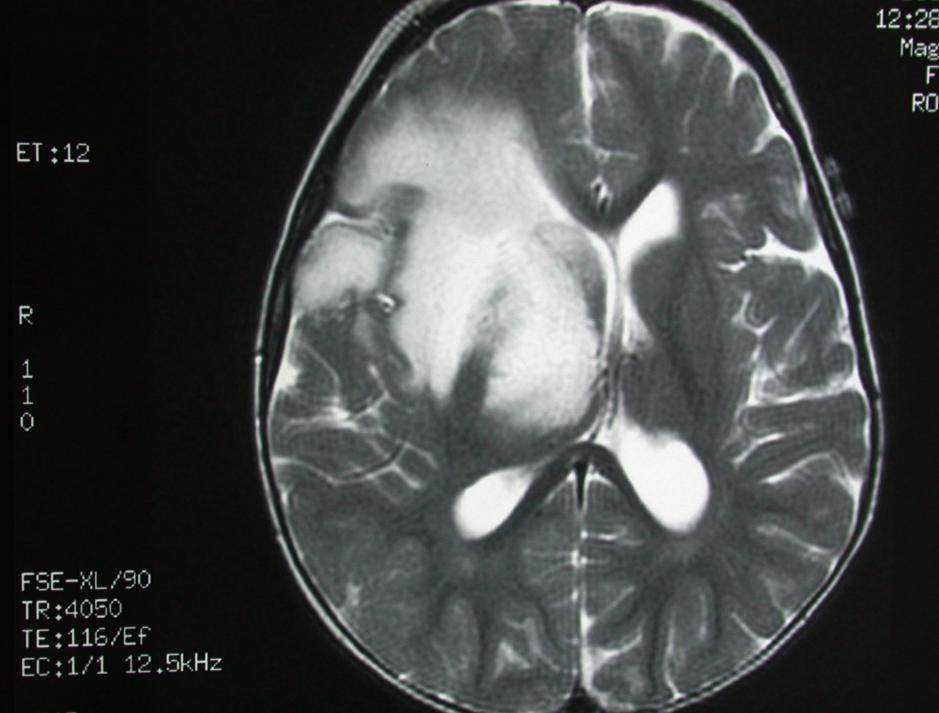


00 NEX	P 91	W = 704 L = 3	v^ 256X192/1.00 NEX 74 FC/St:SI	P 91	М
F SYS#GEMS01	A 88	MACUETTO THOOTHE OFFI TOT	C Store 1 57 SYS#GEMS01	A 88	MAGNETIC
HC C	ANR C	DOB: Oct 08 20 Apr 24 04:00:17 Mag = 1 FL: ROT:	03 Ax I20.0+C		
			L R 9 8 5 5		
16kHz			SE TR:400 TE:20 EC:1/1 16kHz		
x14 /1.7sp 08 2/1.00 NEX :SI	P 91	W = 704 L = 3	HEAD FOV:18×14 4.3thk/1.7sp 20/02:08 v^ 256X192/1.00 NEX 374 FC/St:SI	P 91	
1.5T SYS#GEM501 5758 3 2.0+C	A 88	MAGNETTO THOOTHS OFFI TO Apr 24 04:00:17	10 Ex: 36758 32 Se: 4 32 Im: 14 03 Ax S4.0+C	A 88	MAGNETIC IM VE D

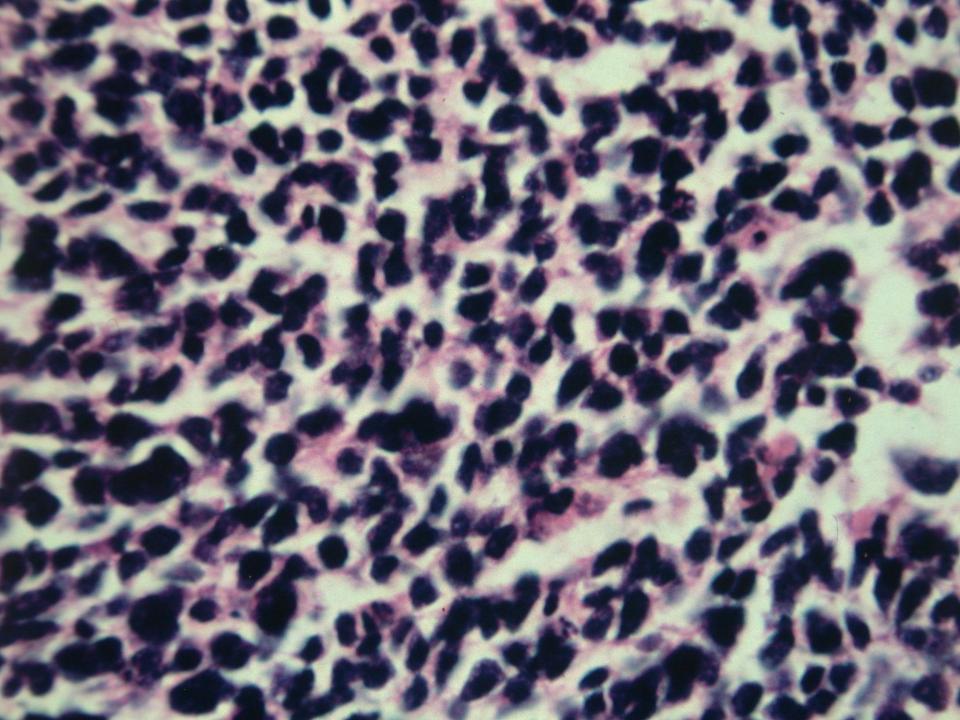
Not all gone, rest intertwined with optic nerve treated with chemo







ULTOD

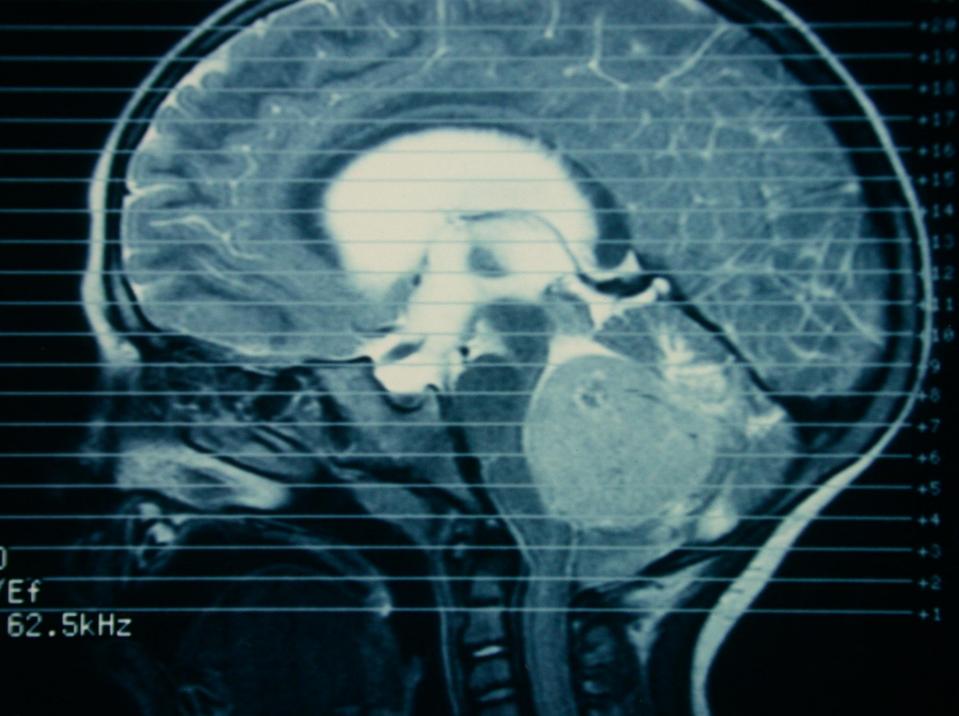


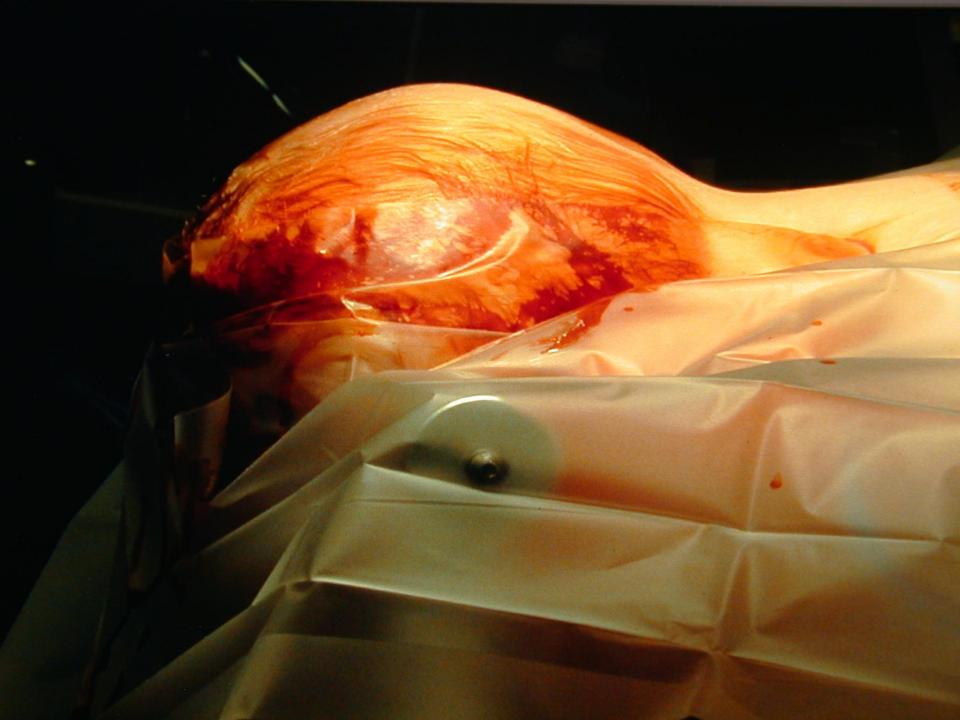
PNET (medulloblastoma)

Treatment: Surgery (<1.5cm²), chemotherapy, craniospinal XRt +local Xrt

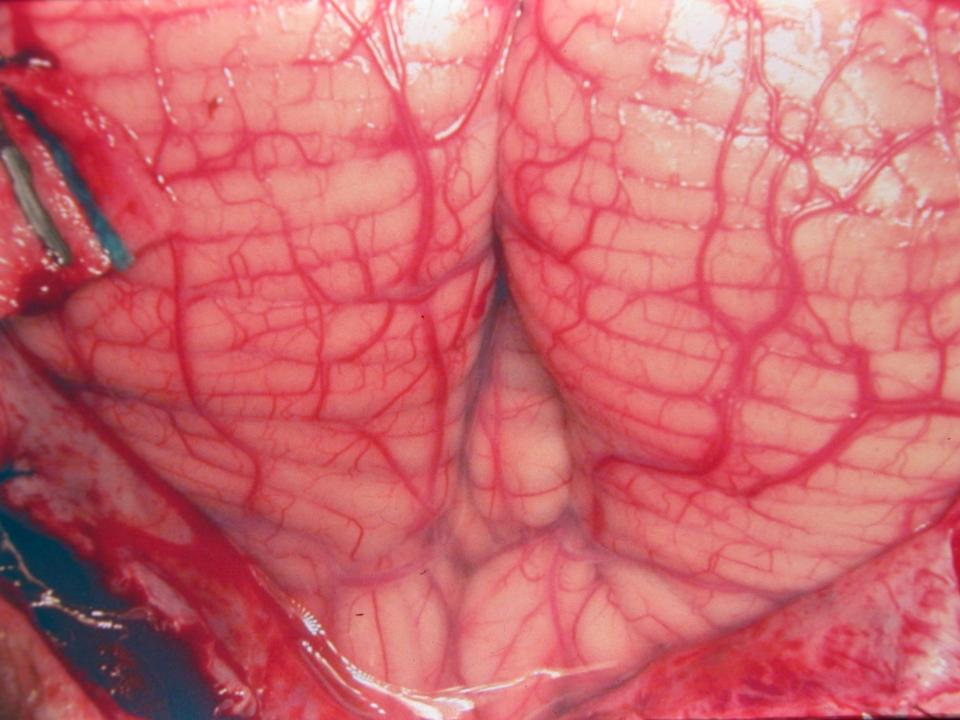
Prognosis

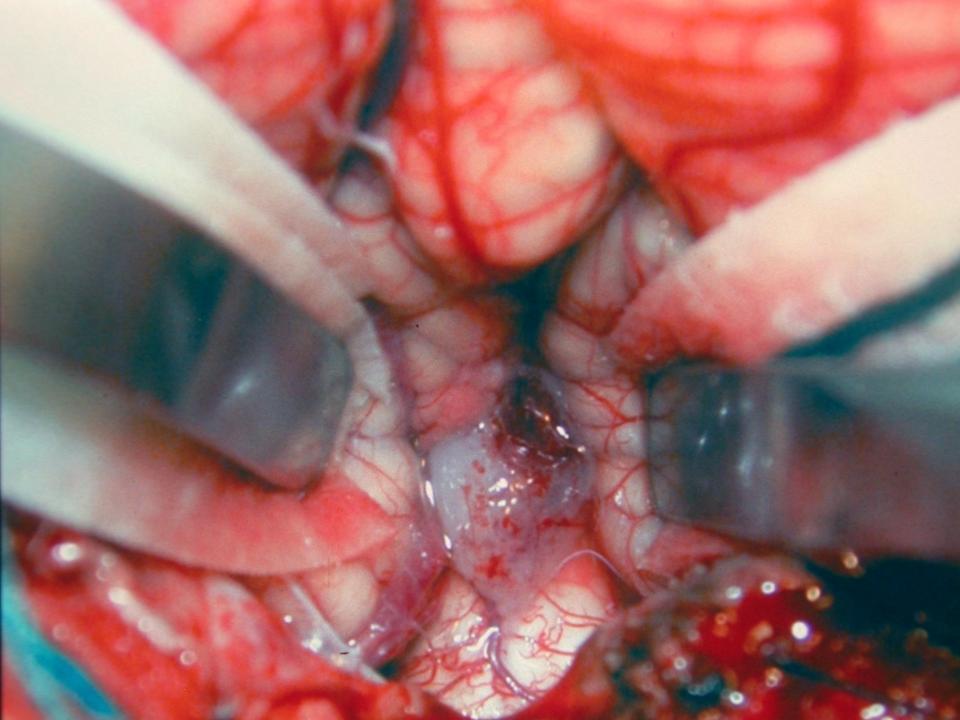
- 1. Staging, amount of post operative residual tumor
- 2. Age < 3yo
- 3. Emerging evidence for histology, molecular markers

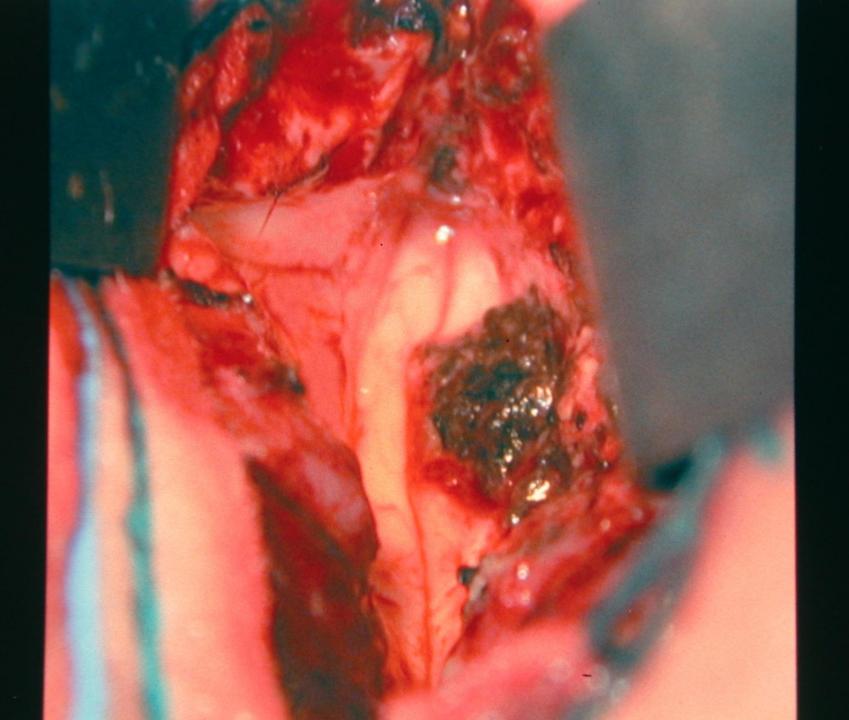




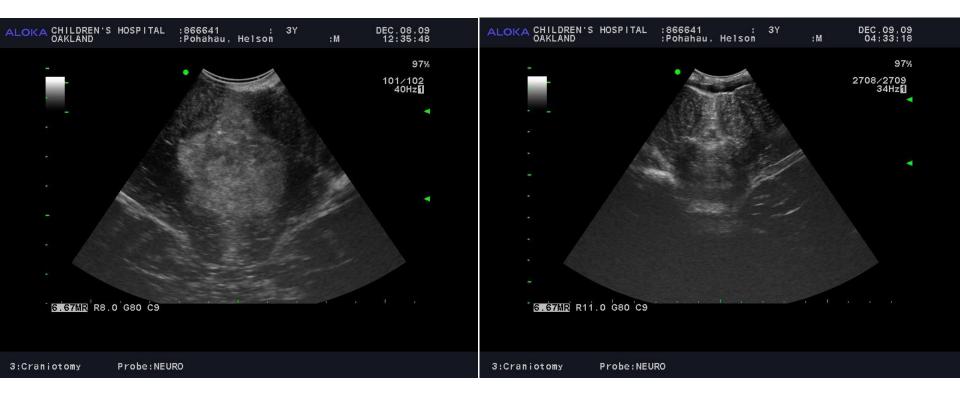








Intraoperative ultrasound:





PNET (medulloblastoma)

Survival: 65-85 % standard risk 40-50% high risk

Relapses (2-4 yrs, locally and CSF pathways) are very difficult to cure

Current directions:

HEAD START 3 and 4- chemo for high risk pts, less Xrt (1800cGy) for standard risk pts to reduce neurocognitive sequelae, high dose chemo and bone marrow transplant with no Xrt for young children CHO is the only Head Start institution in Northern California

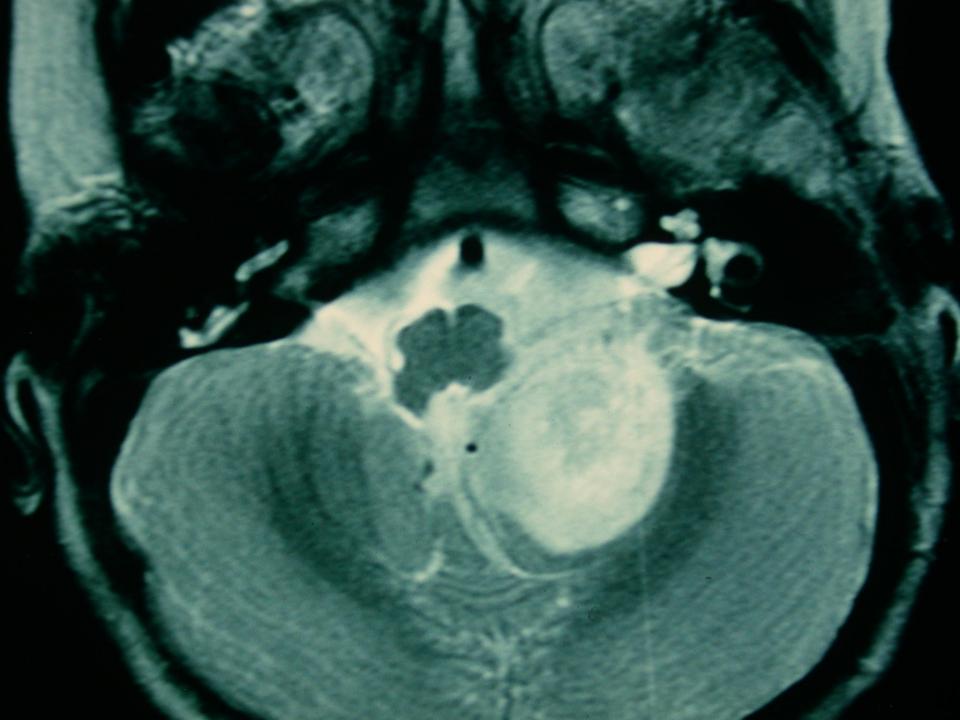
Ependymoma

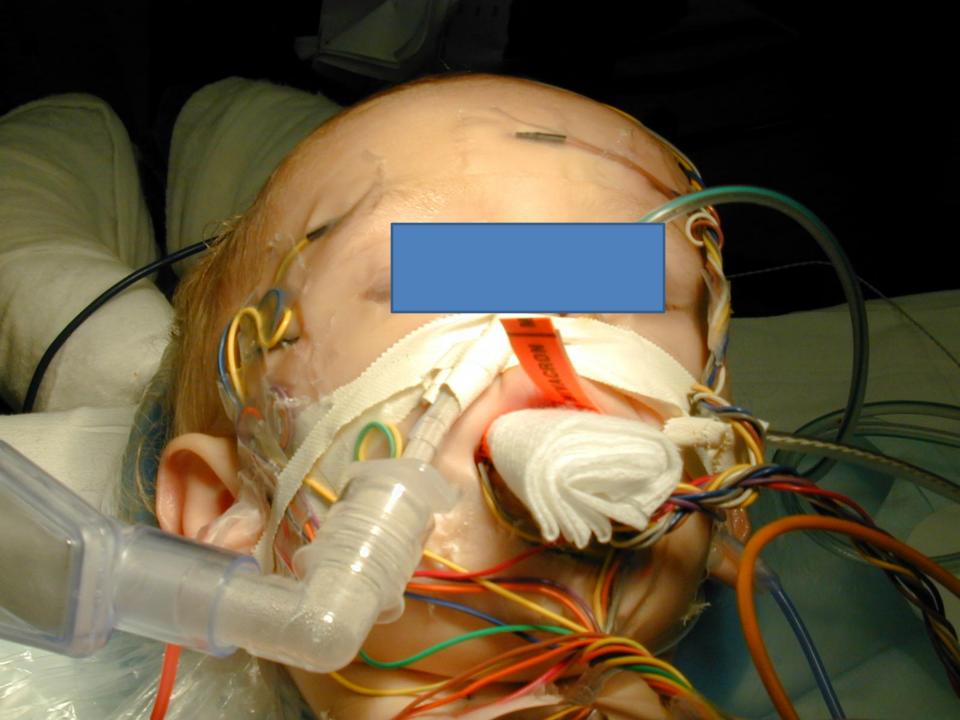
4th ventricle, fingers projecting out of foramens

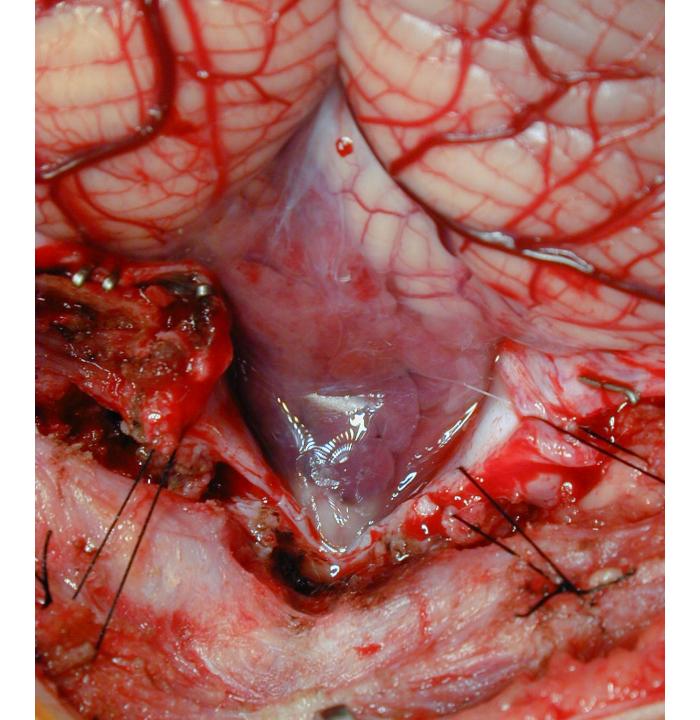
Treatment: Surgical resection major determinant of outcome Complete Resection + Xrt: 60-70% 5 yr EFS

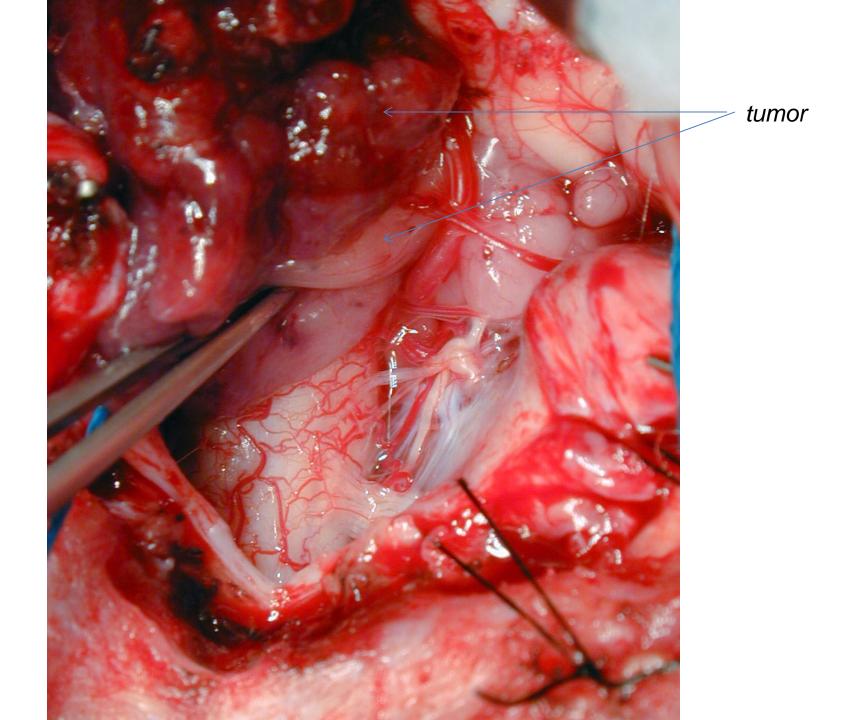
Disseminated Disease (11%), subtotal resection: 20-30%













CN 11

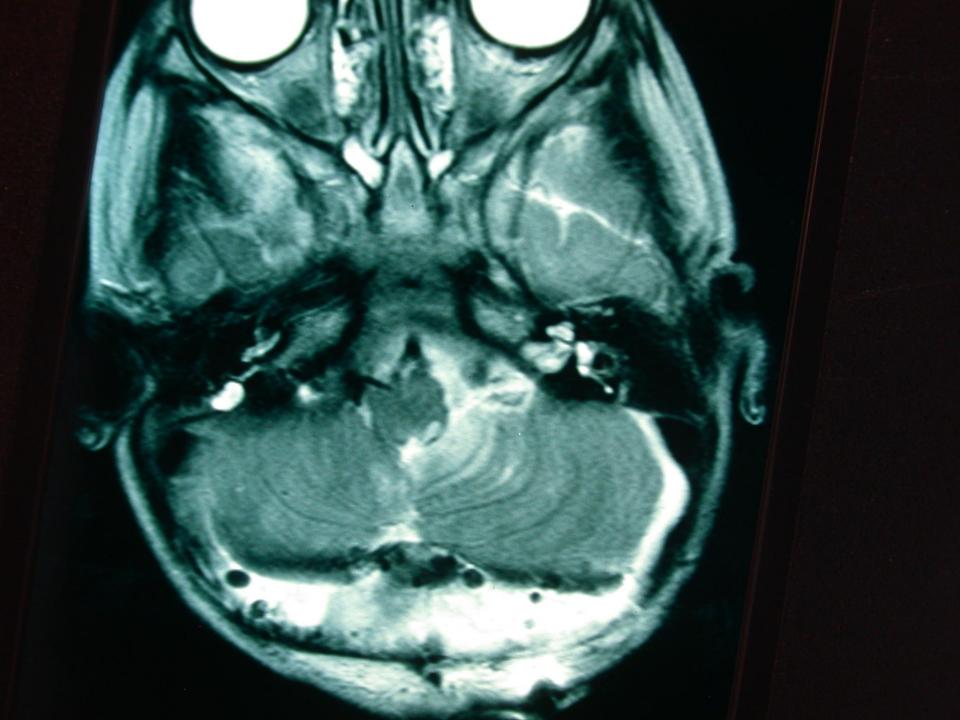
CN 9,10

C1

PICA

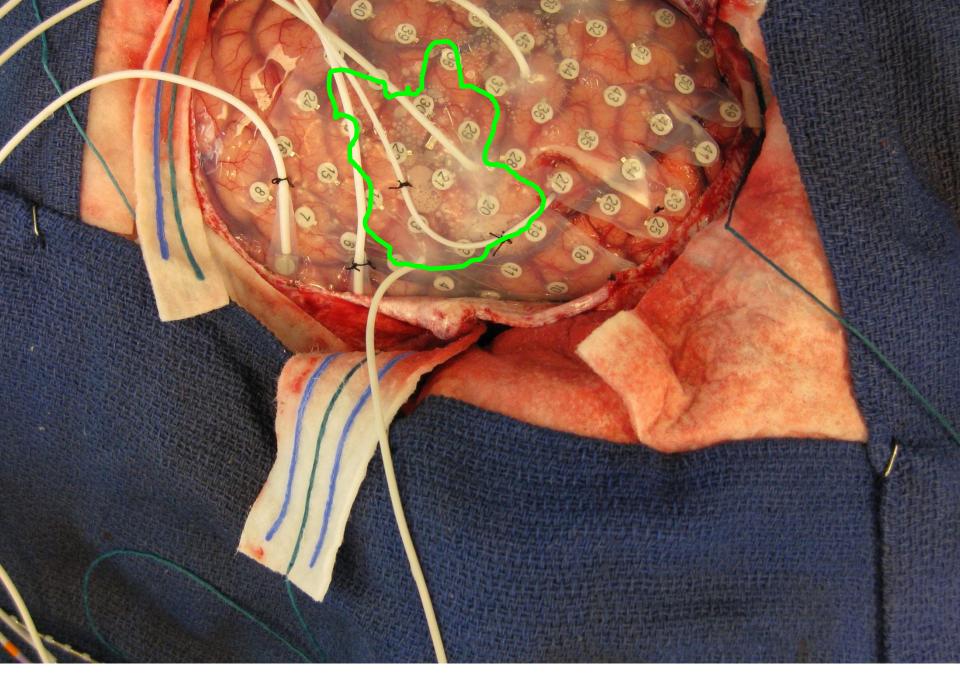
CN 12

Vertebral Artery



K. A. MEG 5/4/07 MRI 11/28/05





Surgical removal of seizure focus



