Children's National

Is it Craniosynostosis or Plagiocephaly? What pediatricians need to know.

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Disclosures

- **No conflicts to disclose:**
- No financial or business interest, arrangement or affiliation that could be perceived as a real or apparent conflict of interest in the subject (content) of their presentation.
- No unapproved or investigational use of any drugs, commercial products or devices.

Objectives

- Identify normal and benign variations of head shape in the infant
- Describe clinical presentation of syndromic and nonsyndromic craniosynostosis
- Define indications for specialty referral for head shape concerns in the infant

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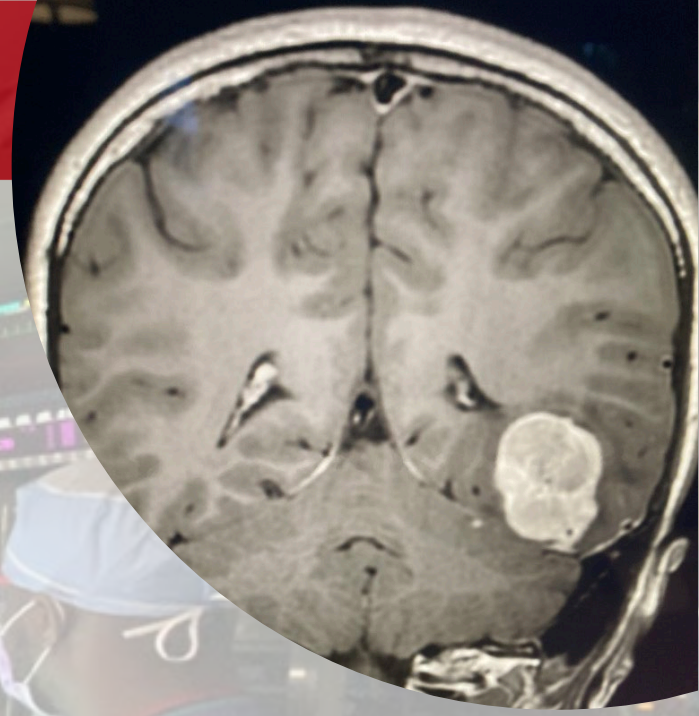
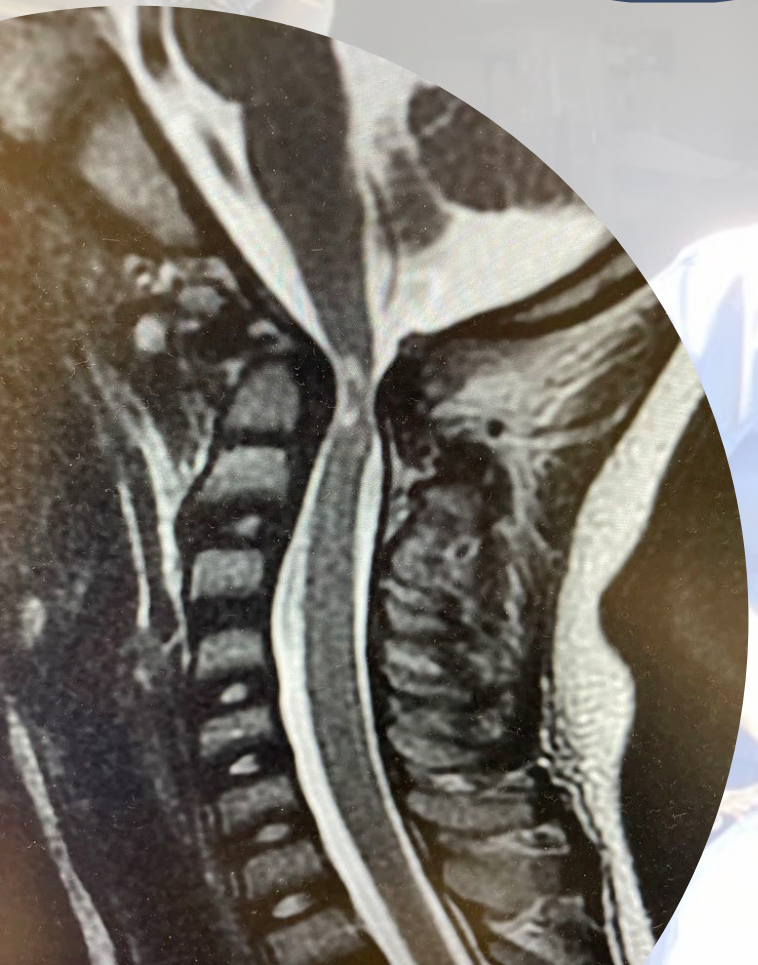
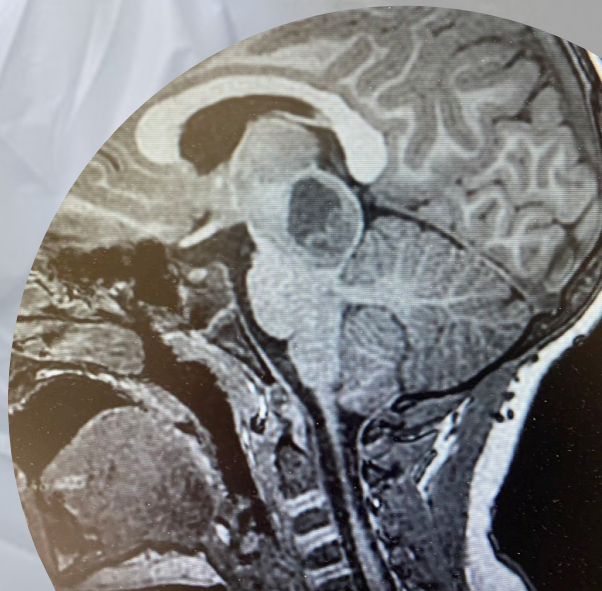




Complete Team Approach to Neurosurgical Care



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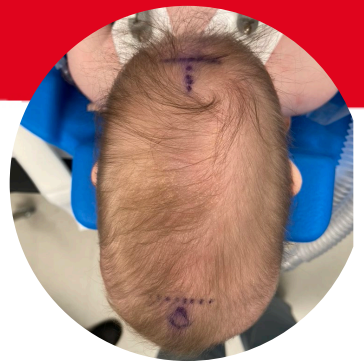
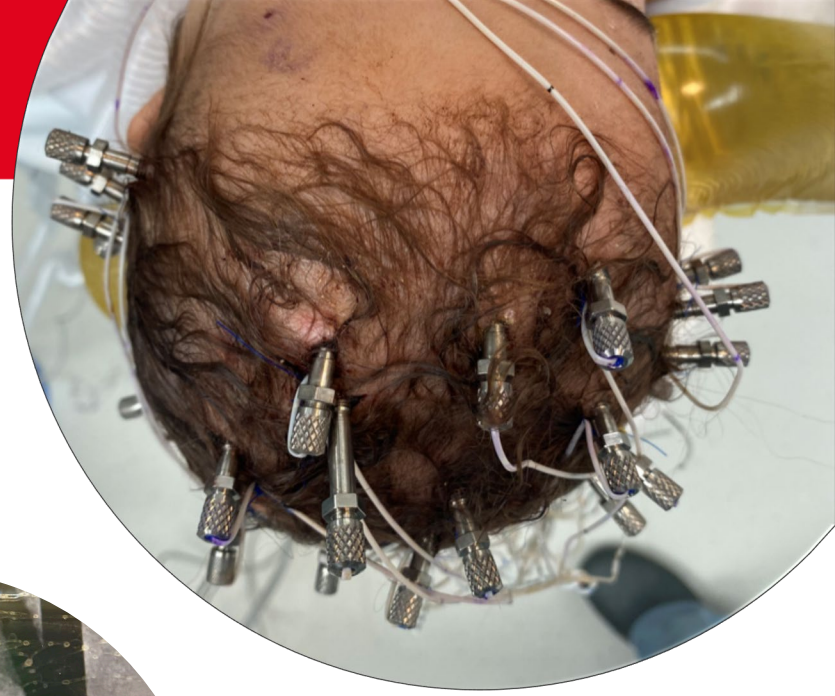
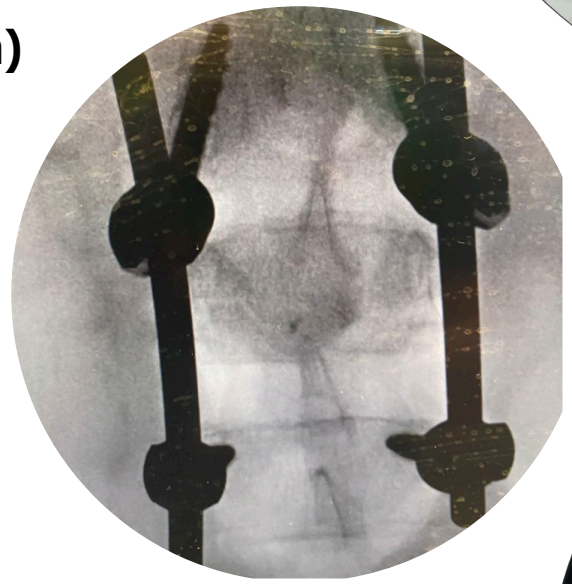
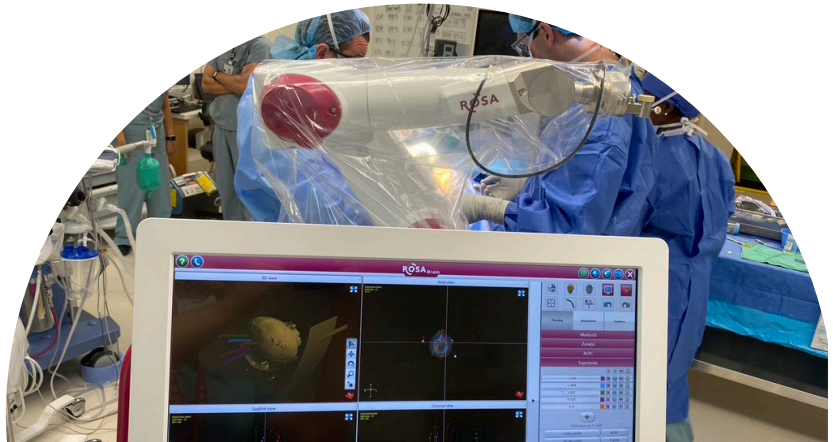


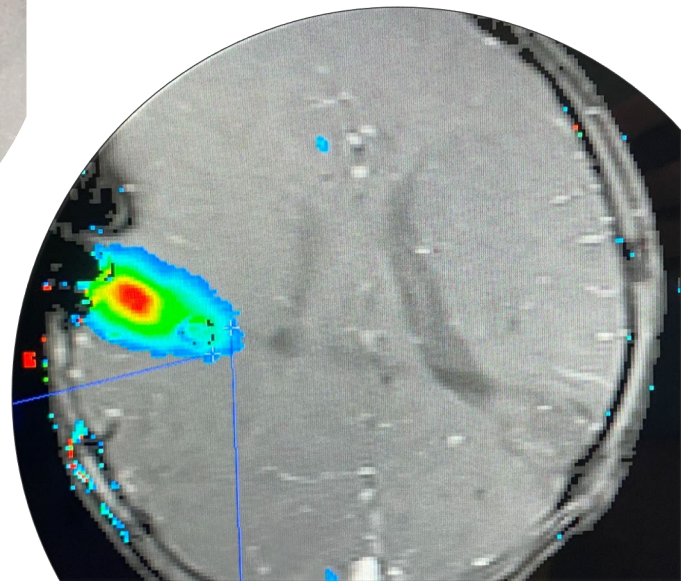


It's not (just) brain surgery

- **Epilepsy (laser, minimally disruptive, neurostimulation)**
- **Neuro-Oncology**
- **Neuromodulation & Movement Disorders**
- **Spine (tumor, trauma, congenital, Chiari)**
- **Craniofacial (craniosynostosis)**
- **Cerebrovascular (AVM, aneurysm, embolization)**
- **Brachial Plexus and peripheral nerve**

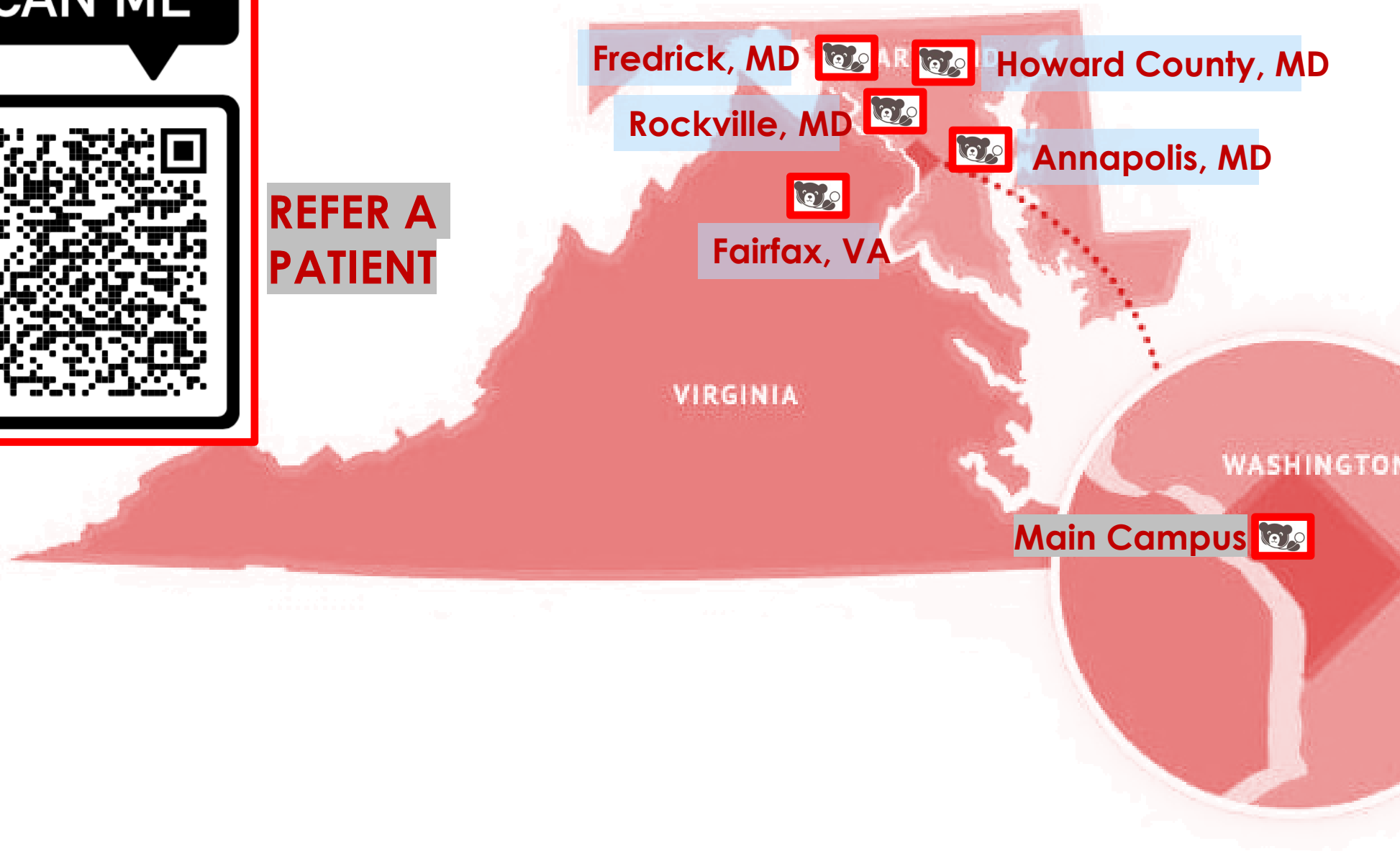






SCAN ME

REFER A PATIENT



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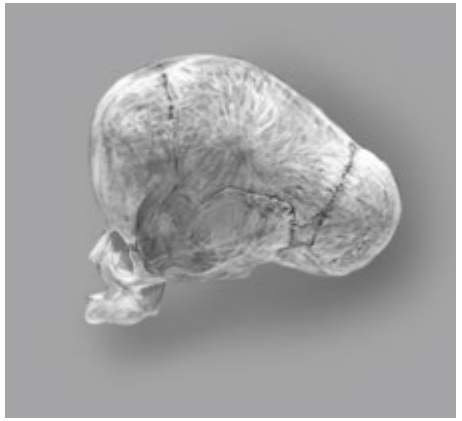
CRANIOSYNOSTOSIS

"Craniosynostosis intrigues me as a drama of nature in which the *Sturm und Drang* of a growing brain and its hydrodynamic forces compete against the rigidities and sometimes yielding barriers of a brain case derived from dermal placodes and primitive cartilage."

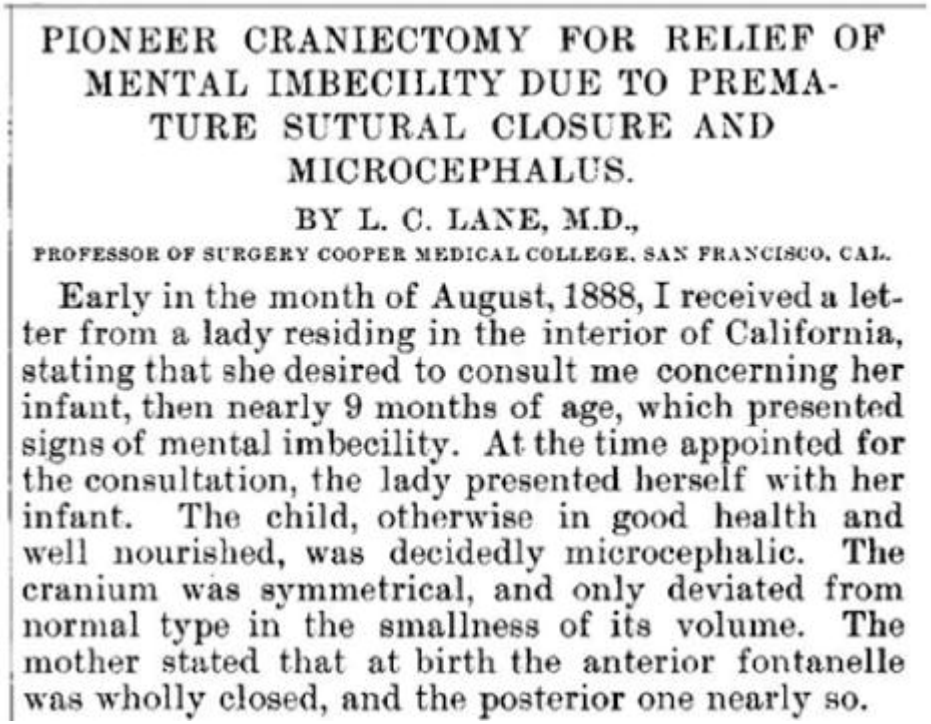
S. Pruzansky, 1978



HISTORY OF CRANIOSYNOSTOSIS

"Can you not unlock my poor child's brain and let it grow?"

- In response, Dr. L.C. Lane performs the first surgery for "premature sutural closure" in North America in 1892
- Lannelogue (Paris, 1890) contemporaneously described his own series – advocated release, not resection of fused suture



PIONEER CRANIECTOMY FOR RELIEF OF MENTAL IMBECILITY DUE TO PREMATURE SUTURAL CLOSURE AND MICROCEPHALUS.

BY L. C. LANE, M.D.,

PROFESSOR OF SURGERY COOPER MEDICAL COLLEGE, SAN FRANCISCO, CAL.

Early in the month of August, 1888, I received a letter from a lady residing in the interior of California, stating that she desired to consult me concerning her infant, then nearly 9 months of age, which presented signs of mental imbecility. At the time appointed for the consultation, the lady presented herself with her infant. The child, otherwise in good health and well nourished, was decidedly microcephalic. The cranium was symmetrical, and only deviated from normal type in the smallness of its volume. The mother stated that at birth the anterior fontanelle was wholly closed, and the posterior one nearly so.

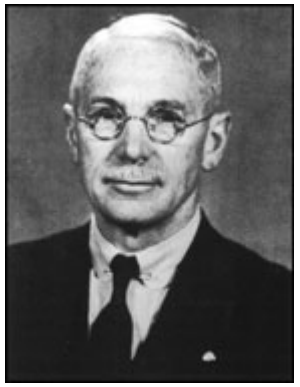
HISTORY OF CRANIOSYNOSTOSIS

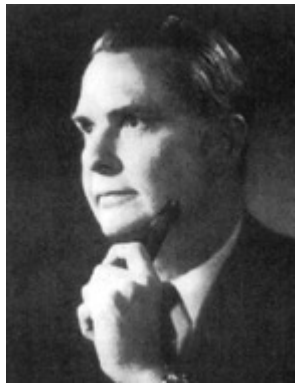
- This enthusiasm for the operation was halted by Jacobi, considered the Father of American Pediatrics, in an address entitled “Non Nocere” in Rome, 1894

“The hands take too frequently the place of brains…Such rash feats of indiscriminate surgery…are stains on your hands and sins on your souls…”

EVOLUTION OF SURGERY

- A resurgence of interest in surgery for craniosynostosis occurred in the 1930's
- Ingerham and Matson at the Children's Hospital in Boston popularized suturectomy
- Significant advances in anesthesia, blood transfusion, surgical technique
- 2 deaths in 394 patients





MODERN ENDOSCOPIC STRIP CRANIECTOMY

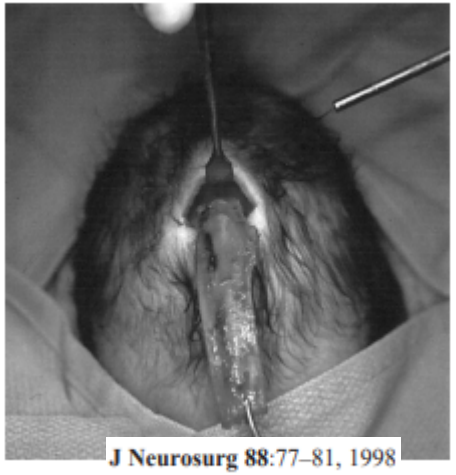
- Early 1990s: Jimenez and Barone recognized limitations of the approaches and proposed novel technique: simple suturectomy via an endoscopic approach

- 3 basic principles:
  - Faber and Towne, early surgery in life
  - Moss's functional matrix theory: rapidly growing brain would cause expansion of skull into a normal shape
  - Helmet remodeling (introduced by Pershing): to counteract tendency of cranial vault to revert to a prior shape

TABLE 1

*Patient characteristics and intraoperative data in four infants who underwent endoscopic strip craniectomy**

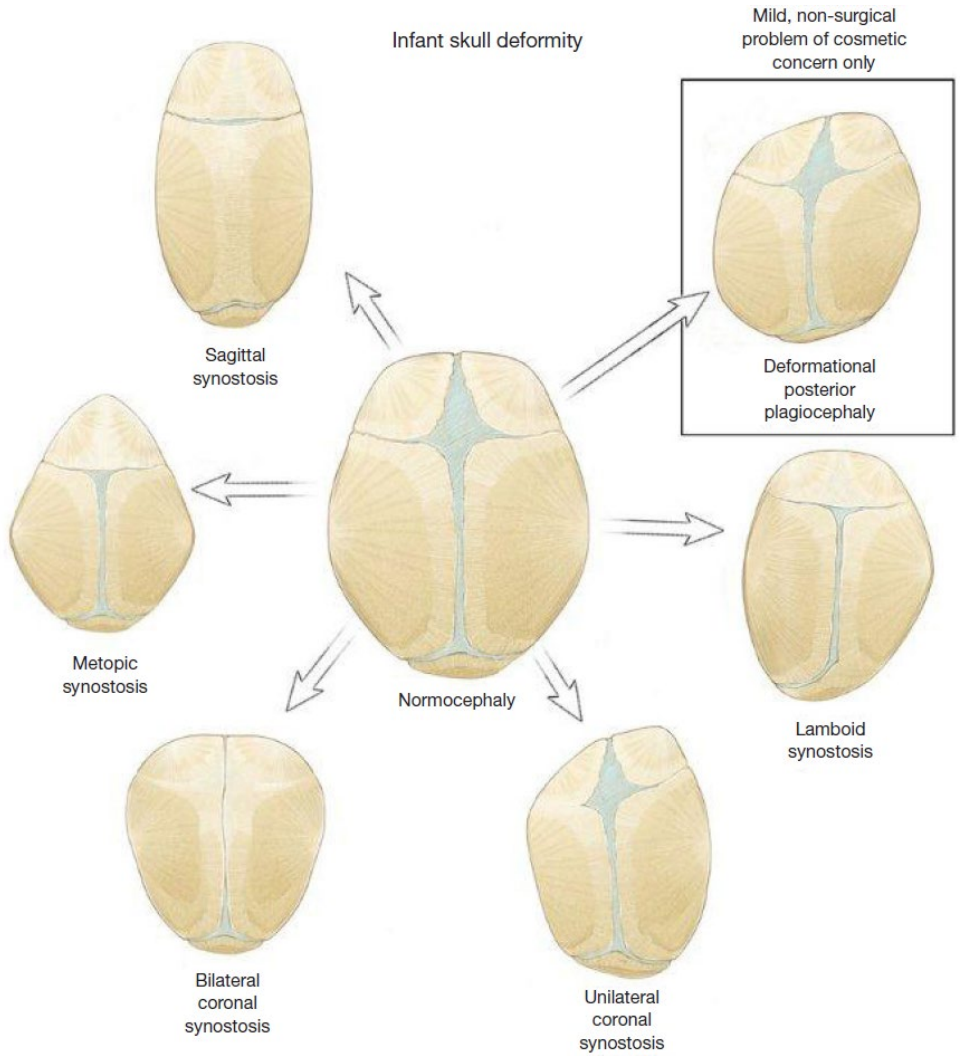
Factors	Case 1	Case 2	Case 3	Case 4
patient characteristics				
age (wks)	2	4	9	12
sex	M	F	M	M
weight (kg)	3.1	3.8	6.5	7.8
EBV (ml)	248	304	520	568
hematocrit (%)	32	37	33	34
intraop data				
length of surgery (hrs)	1.25	1.51	1.15	2.8
EBL (ml)	25	30	12	150
EBVL (%)	10	9.8	2.3	26.4
blood transfused (ml)	0	0	0	150
colloids (intake) (ml)	0	25	0	85
crystalloids (intake) (ml)	127	169	240	200



J Neurosurg 88:77–81, 1998

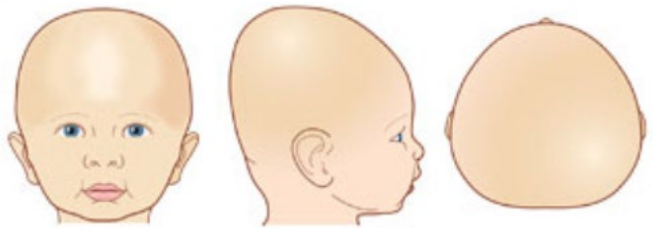
Evaluation of Head Shape

- History
  - Head shape at birth
  - Head turn preference
  - Torticollis
  - Family history of craniosynostosis
- Physical Exam Findings
  - Cranial Index
  - Position of ears, nose
  - Forehead asymmetry
  - Ridging along cranial suture
- Imaging: ? XR, CT, head US

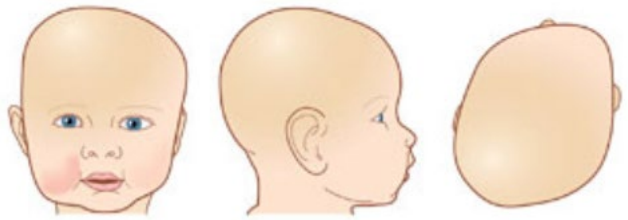


Cranial Index: biparietal diameter ÷ occipitofrontal diameter x 100

- **Cephalic index (CI)**
- Normal: 76 – 90%
- Normocephaly = CI 76% – 90%
- Brachycephaly = CI > 90%
- Dolichocephaly = CI < 76%



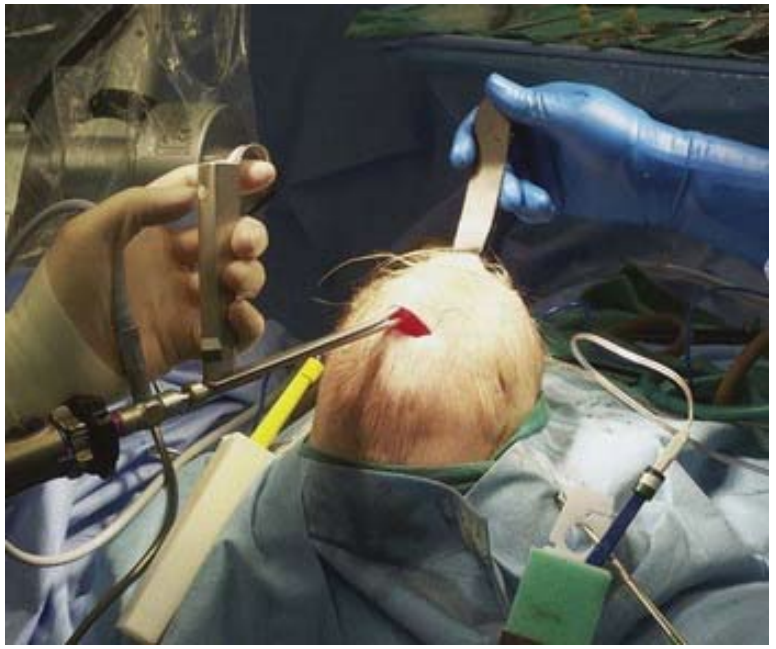
- **Diagonal difference (Plagiocephaly)**
- Normal head shape: 0 – 4 mm
- Mild: 5 – 9 mm
- Moderate: 10 – 15 mm
- Severe: >15 mm



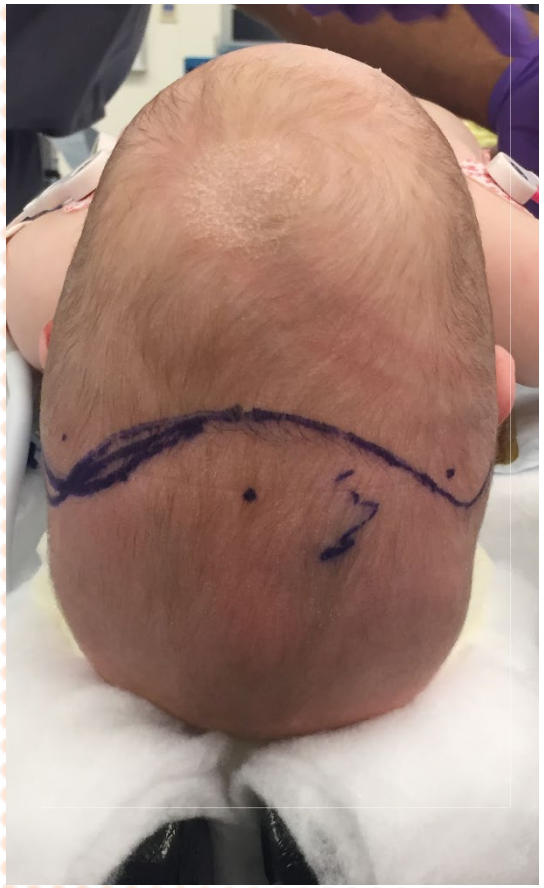
Surgical Management of Craniosynostosis

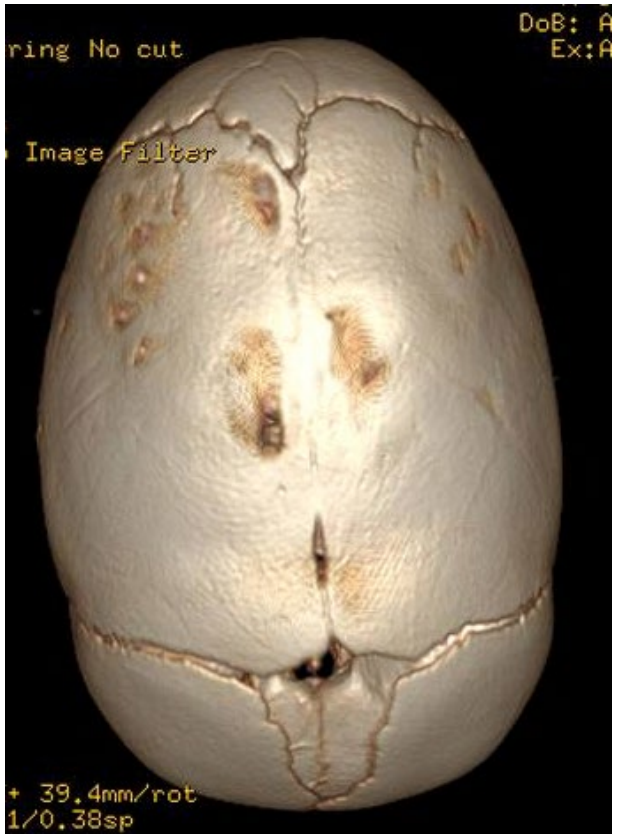
- Open Cranial Vault Remodeling

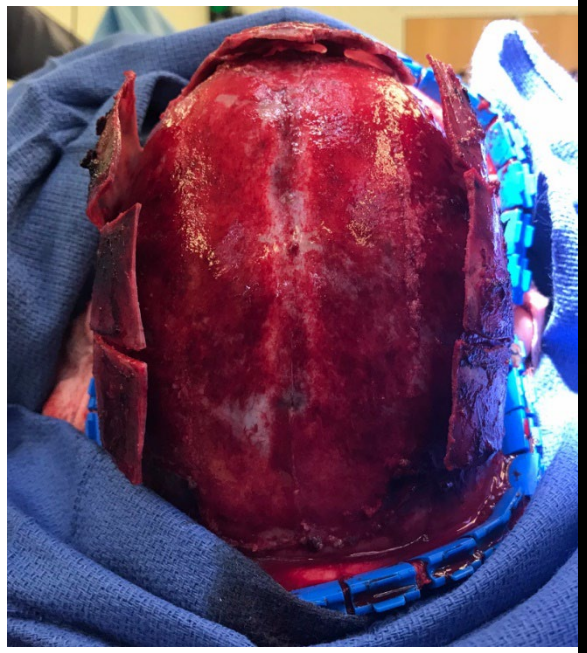
- Endoscopic Suturectomy

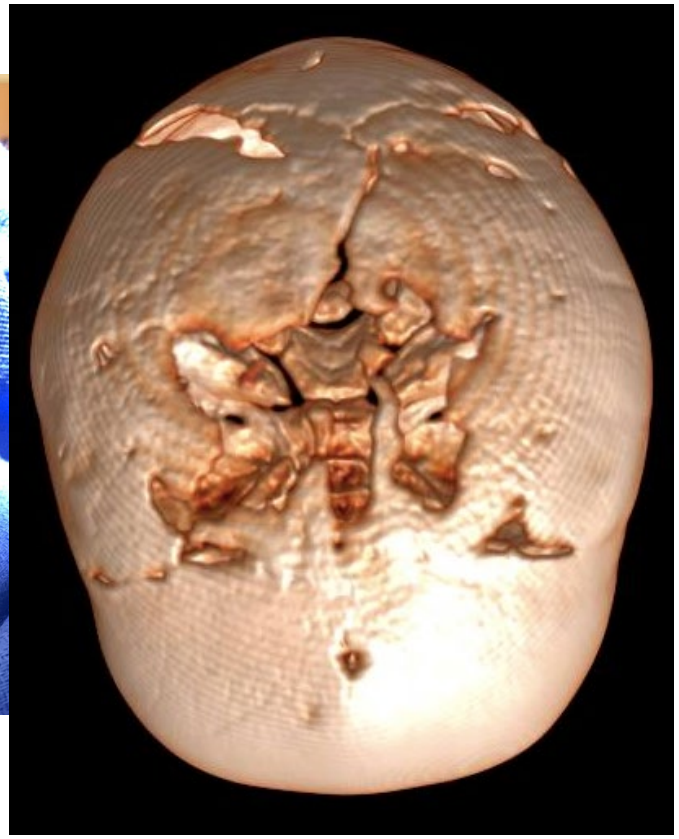


5 month old boy with scaphocephaly

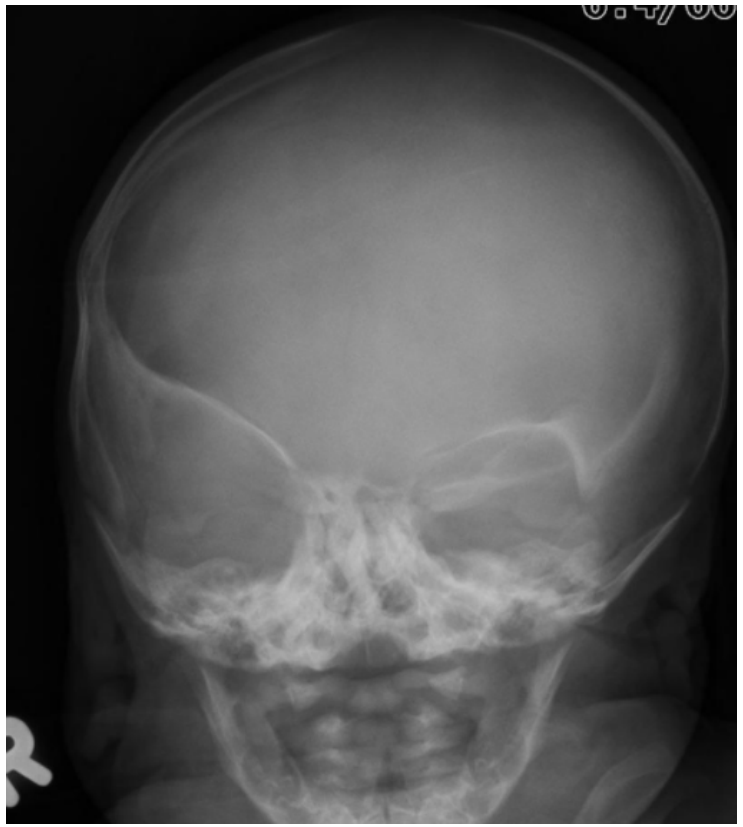


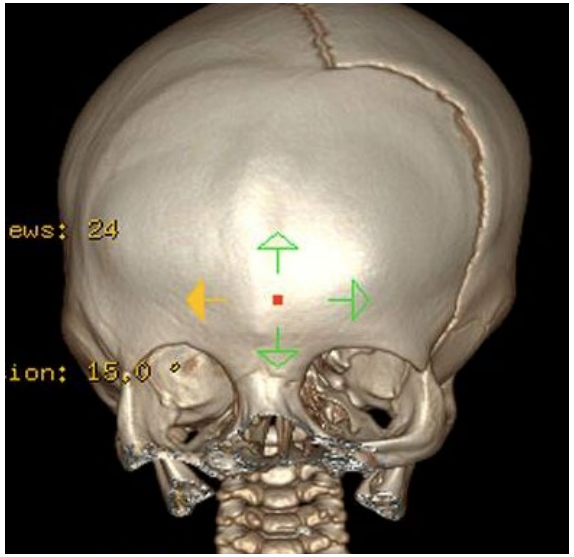


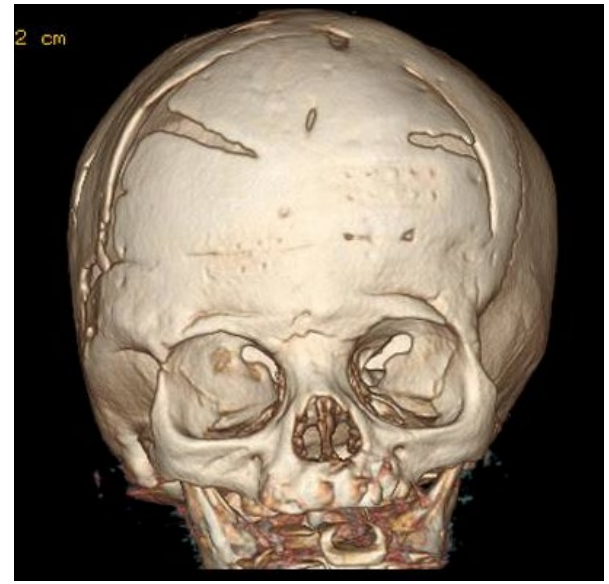




6 month old girl with unicoronal synostosis

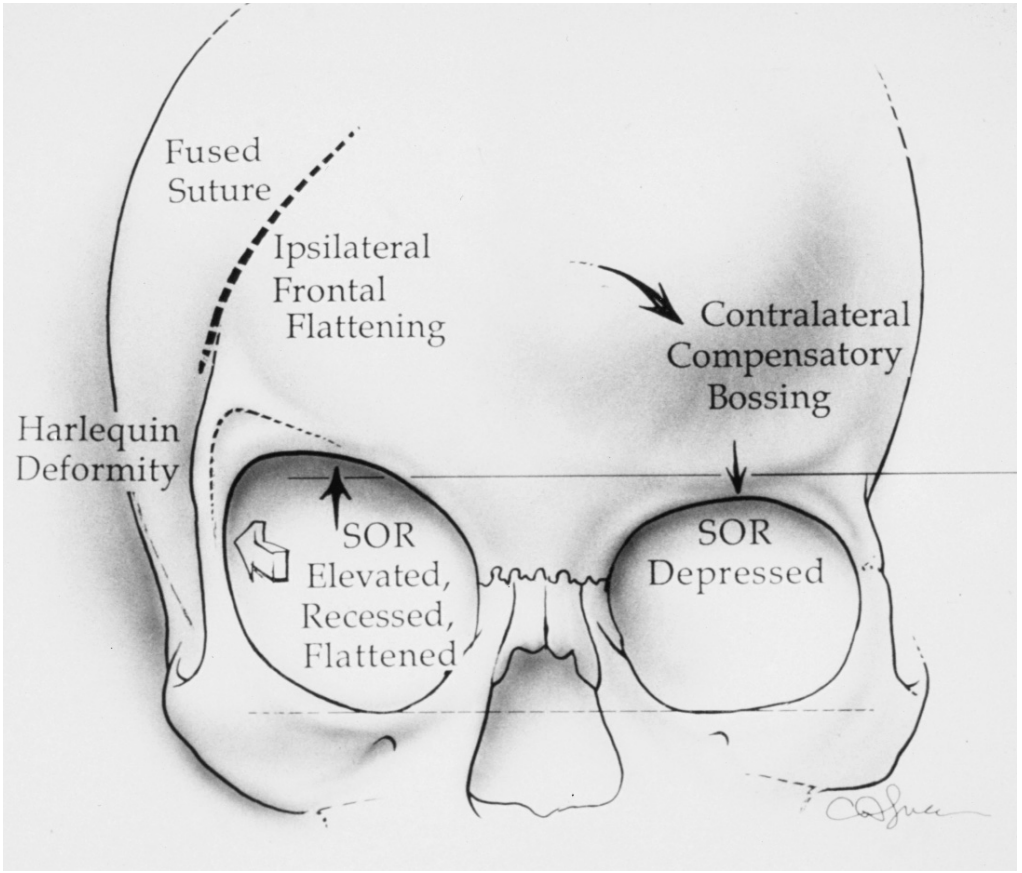




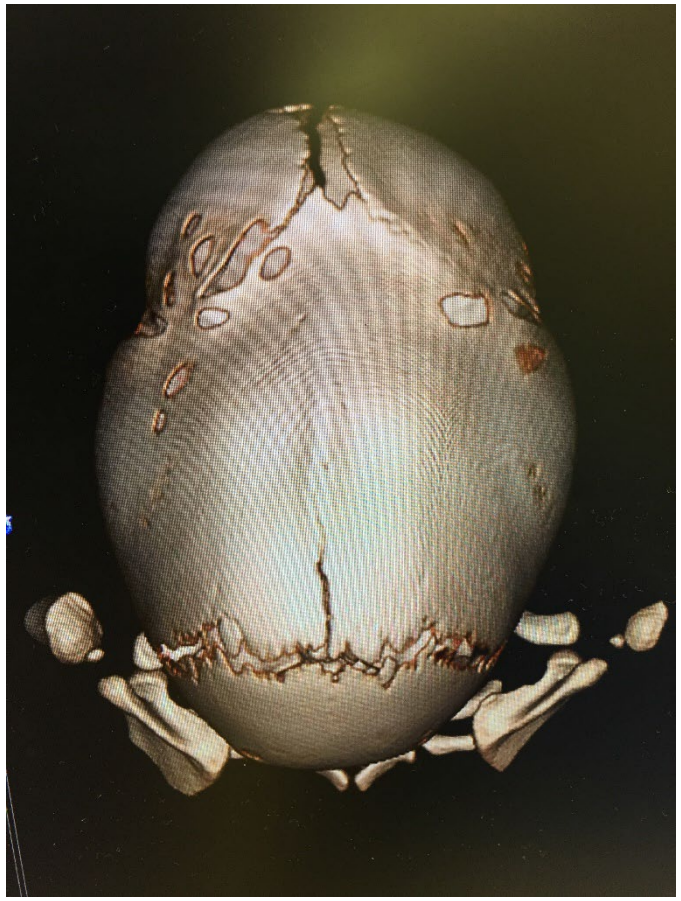


FACIAL DEFORMITY

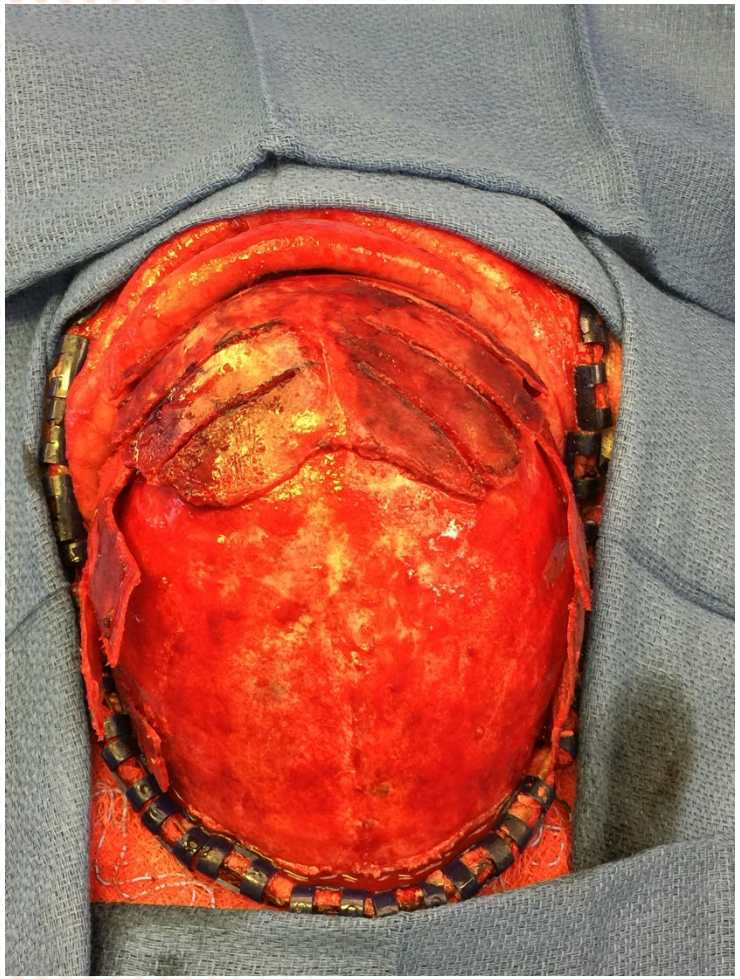
- Ispilateral
  - Eyebrow elevation
  - Opening of the palpebral fissure
  - Nasal bone deviation
  - Hemifacial expansion
- Contralateral
  - Eyebrow depression
  - Nasal tip, chin deviation
  - Anterior fossa expansion
  - Hemifacial compression

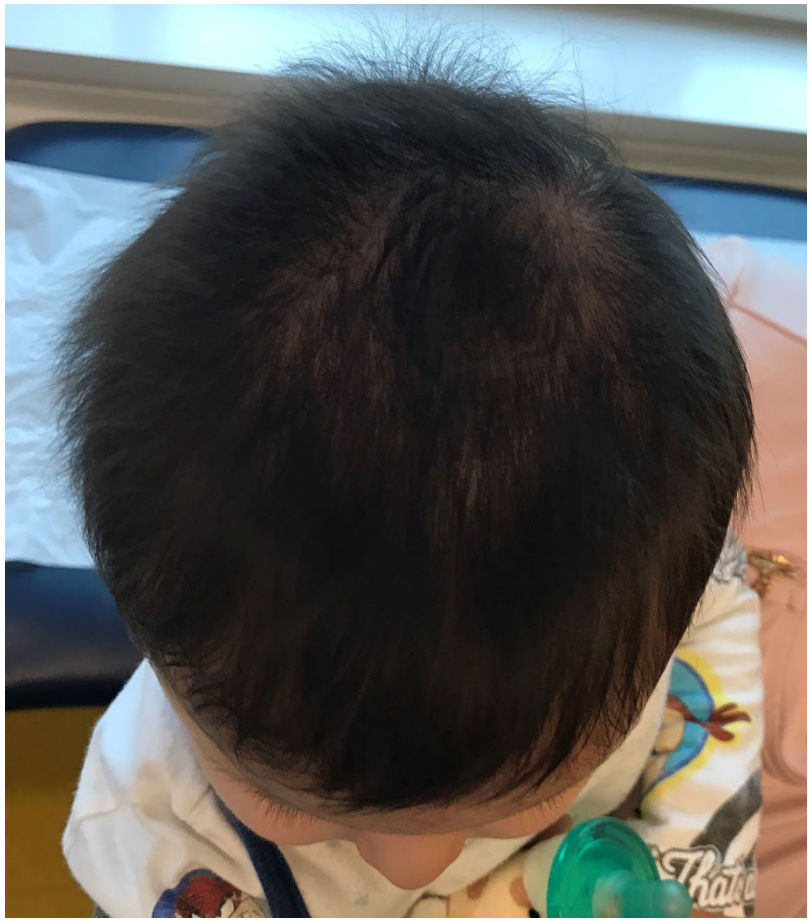


4 month old boy with bicoronal and sagittal synostosis

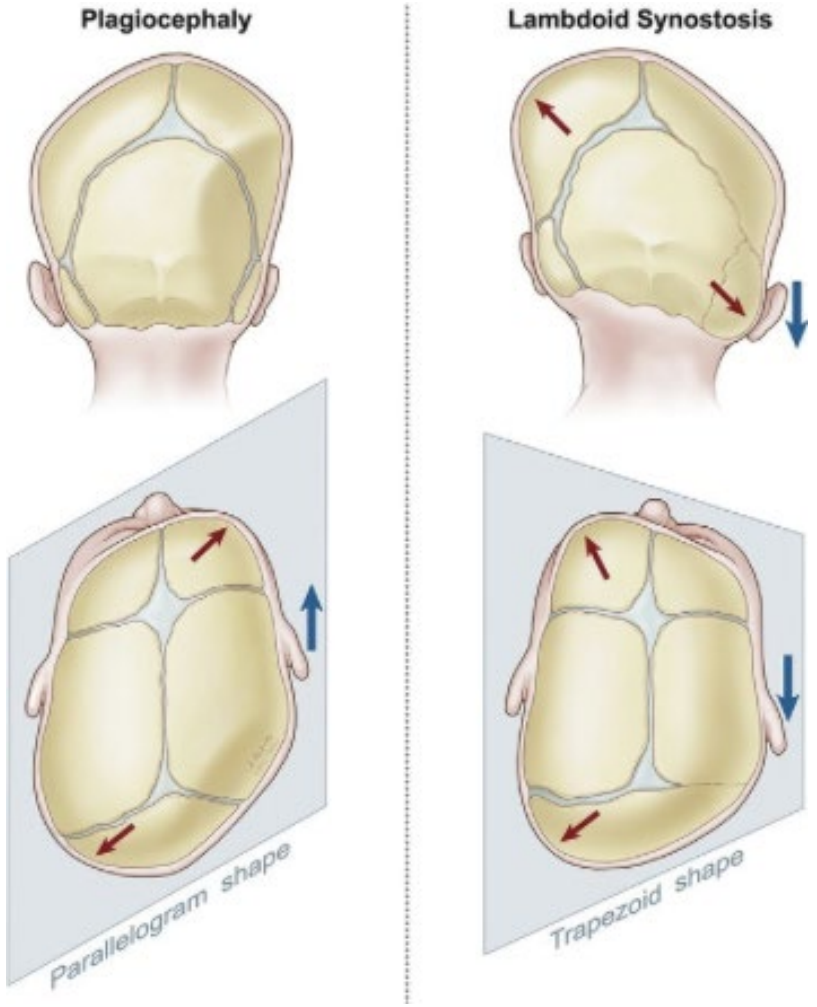


4 month old boy with bicoronal and sagittal synostosis





Positional Plagiocephaly vs. Lambdoid Synostosis



INDICATIONS FOR SURGERY

INDICATIONS FOR SURGERY

- Wide variation of management depending on center
- Treatment will vary based on:
  - Age of presentation
  - Location and number of synostoses
  - Severity of deformity
  - Preference of craniofacial team
- 2 main indications:
  - Correct skull shape for aesthetic and psychosocial considerations
  - Adequate space for brain growth

INDICATIONS FOR SURGERY

Endoscopic suturectomy

- 4-12 weeks of age
- During surgery, abnormal bone removed; helmet reshapes head
- Pros: 1-3 small incisions, less blood loss, shorter hospital stay
- Cons: helmet x 9-12 months, suture may re-fuse

CVR

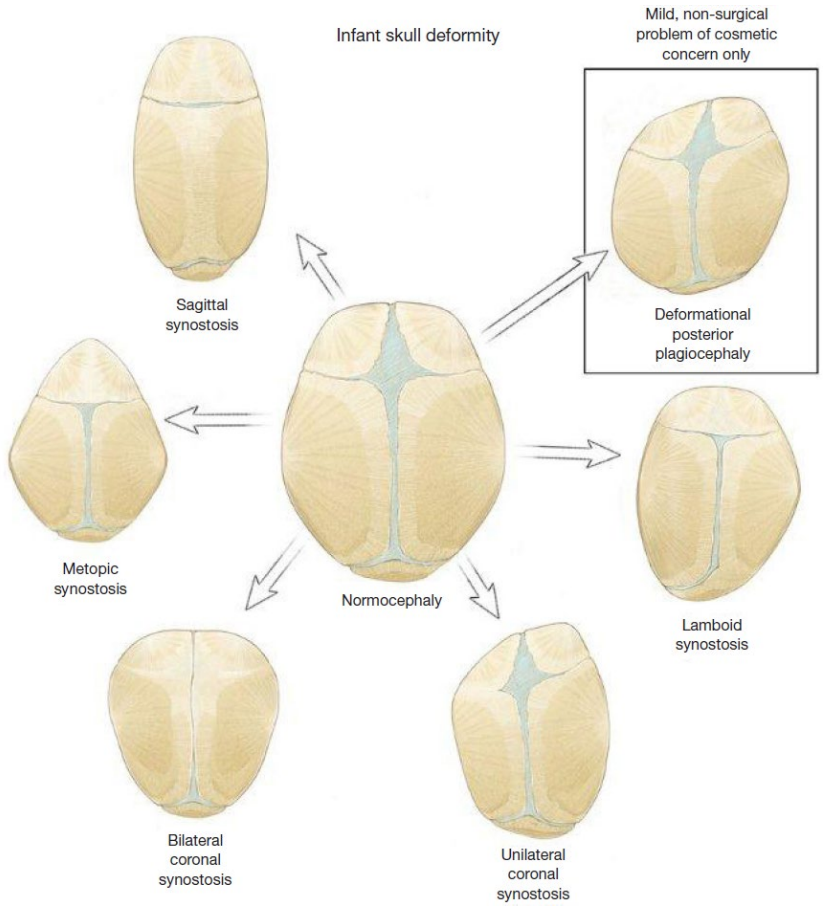
- 4-6 months of age for sagittal;
- 9 months of age for fronto-orbital advancement (FOA)
- During surgery, abnormal bone removed & head reshaped
- Pros: surgery “fixes” head shape, no helmet
- Cons: bicoronal incision, more blood loss, longer hospital stay

Cranial molding helmeting

- Baseline measurements prior to OR
- 3D scan 1 week post-op
- Helmet starts 2 weeks post-op
- Worn for 9 – 12 month
- *Must* be worn for 23 hours per day
- Adjusted Q 2 – 3 weeks

EPIDEMIOLOGY

- 1 in every 2000 to 2500 live births
- Single-suture or multi-suture
- Association with genetic conditions or syndromes
- Frequency:
  - Sagittal most common: 50-60%
  - Coronal: 17-29%
  - Metopic: 4-10%
  - Lambdoid: less than 2%
- Normal closure:
  - Metopic: 3-9 months
  - Sagittal: 22 years
  - Coronal: 24 years
  - Lambdoid: 26 years



ETIOLOGY

- Non-syndromic: incompletely understood
- Sporadic
- Genetic mutations
- Metabolic and hematologic syndromes
- Teratogens (valproic acid, retinoic acid)
- Maternal smoking
- Advanced paternal age

- More than 100 mutations have been identified
  - FGFR1-3, NELL1, MSX2, TWIST, GLI3 genes

TABLE V. Some Genes in Craniofacial Development

Neural crest
Pax3
Pax7
Pax9
Efnb1
Osteogenesis involving membrane and endochondral bone
Runx2
Mesenchymal condensations of skull
Alx4
Osteocalcin
BGLAP
Collagens
COL1A1 (bone)
COL2A1 (cartilage)[a]
Alkaline phosphatase
ALPL
Bony sutural edges
Fibroblast growth factors
Fgf2
Fgf4
Fibroblast growth factor receptors
Fgfr1
Fgfr2
Fgfr3
Muscle segment homeobox
Msx1
Msx2
Basic helix-loop-helix
Twist
Bone morphogenetic proteins
Bmp2
Bmp4
Bmp antagonist
Nog
Transforming growth factor β
Tgfb1
Tgfb2
Tgfb3
Nel-like 1
Nell1

Modified from Cohen [2005].

CRANIOFACIAL SYNDROMES

TABLE 1

Common Features of Craniosynostosis Syndromes

Feature	Apert Syndrome	Crouzon Syndrome	Saethre-Chotzen Syndrome	Pfeiffer Syndrome
Inheritance	Autosomal dominant	Autosomal dominant	Autosomal dominant	Autosomal dominant
Type of synostosis	Bicoronal	Bicoronal	Multiple suture	Bicoronal and lambdoid, occasionally sagittal
Hypertelorism and exorbitism	Present	Present	Absent	Present
Intelligence	Variable	Normal	Usually normal	Variable
Midface hypoplasia	Present	Present	Present	Present
Syndactyly	Present	Absent	Present	Present

TIMING OF SURGERY

TIMING OF SURGERY

- 3-9 months largely considered optimal
  - Passive postop endocranial remodeling
  - Reossification of calvariectomy defects
  - Malleability of calvarial bone
  - Minimize facial dysmorphisms

- >12 months age
  - Calvarial bone less easily molded
  - Unpredictable reossification
  - Endocranial base does not change
  - Facial dysmorphisms persist or progress

TIMING OF SURGERY

- Less than 4 months: Endoscopic techniques in which postoperative helmets are used to direct skull growth
- 6-8 months or more: Spring or distractor mediated techniques can be used in older infants
- 6-12 months: Open cranial vault remodeling procedures

WHAT ABOUT OLDER CHILDREN?

- Clinical symptoms
  - Headache, lethargy, developmental delays
- Clinical signs
  - Signs of elevated intracranial pressure
  - Ophthalmology evaluation: Papilledema
- Intracranial pressure monitoring

HOW DO YOU COUNSEL PARENTS ON SURGERY?

Risks

- Bleeding
- Infection
- Durotomy
- CSF Leak
- Need for reoperation

Preoperative Considerations

- A-line
- Central line?
- Precordial dopplers
- Blood in OR

WHAT ARE THE COMPLICATIONS?

- Acute
  - Blood loss – nearly continuous
    - Avoid dilutional coagulopathy
    - Persists 12-24 hours postop -> ICU
  - Dural tears – CSF leak
    - Immediate repair if noted
    - Loss of continuity of osteogenic dura may lead to cranial defect in long term
    - Infection via communication to sinuses

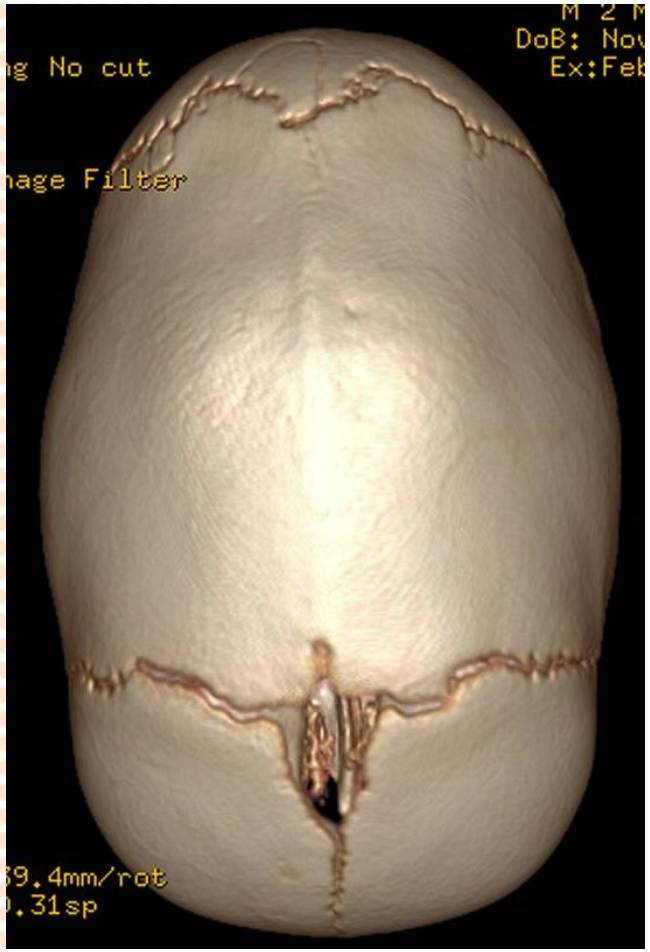
WHAT ARE THE COMPLICATIONS?

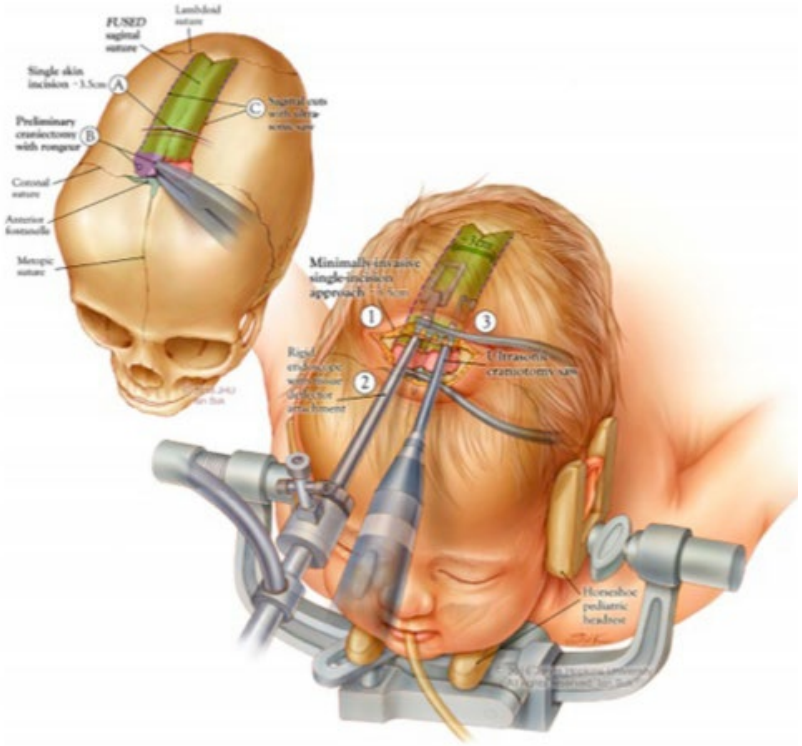
- Late – abnormal bone healing
  - Age over 1 – decreased ability to heal defects
  - General rule: defect >2cm in age > 1, should be filled with split calvarial graft
  - Infection may lead to resorption

- Reports of transcranial plate migration (Persing, 1996)
  - No harmful sequaelae reported
  - Use resorbable

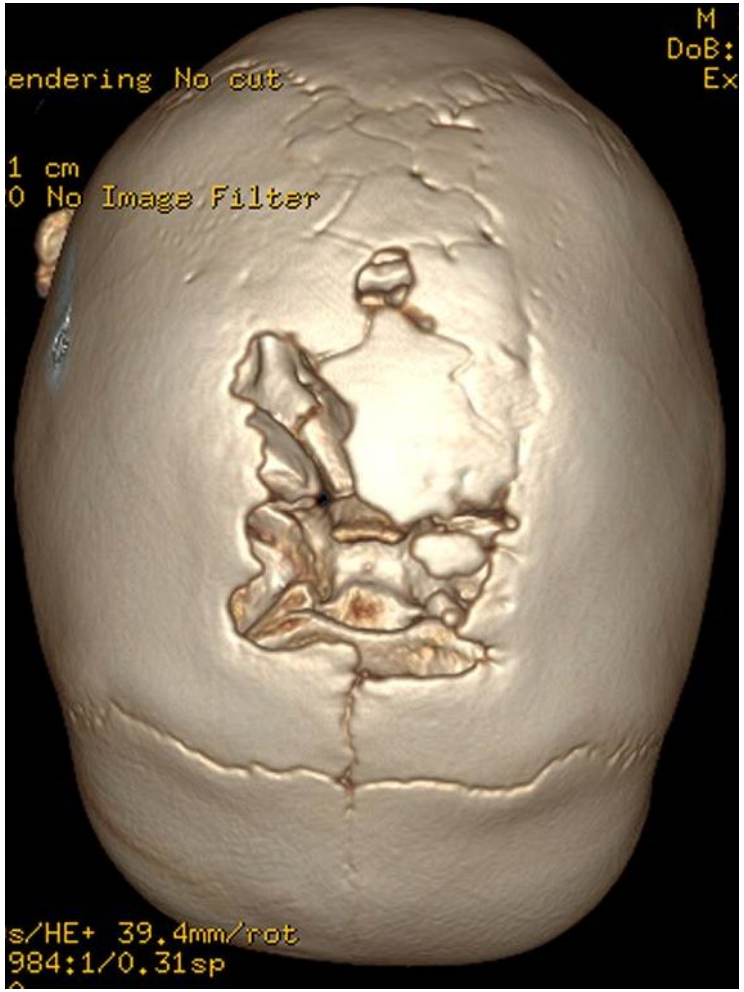
- Mortality – 1.5-2%
  - Six center combined experience reported 1.6% (Whitaker, 1979)

3 MONTH OLD BOY WITH SCAPHOCEPHALY





Childs Nerv Syst (2017) 33:1–5



ENDOSCOPIC VERSUS CVR

Analysis of clinical outcomes for treatment of sagittal craniosynostosis: a comparison of endoscopic suturectomy and cranial vault remodeling

- N=207 patients (187 endoscopic sutrectomy and 20 CVR)
- Operative time: 45 vs. 195 minutes
- LOS: 1 vs. 3 days
- Transfusion rate: 2% vs. 85%
- CI Z-scores were initially more favorable for ES; at 3 years equal
- 4 *syndromic* patients treated by ES required secondary expansion for raised ICP

- ES is an effective treatment for nonsyndromic sagittal synostosis

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TABLE 2. Patient characteristics of the study population

Characteristic	CVR (n = 20)	ES (n = 187)
Males, n (%)	10 (50)	137 (73)
Age at presentation, mos		
Median (IQR)	13.5 (8.0–20.3)	2.0 (1.3–3.0)
Range	6.0–47.0	0.5–6.8
Age at operation, mos		
Median (IQR)	14.0 (11.8–23.8)	3.0 (2.5–4.0)
Range	8.0–48.0	1.5–7.0
Duration of helmeting, mos		
Median (IQR)	Not applicable	8.0 (7.0–9.0)
Range		2.0–14.0

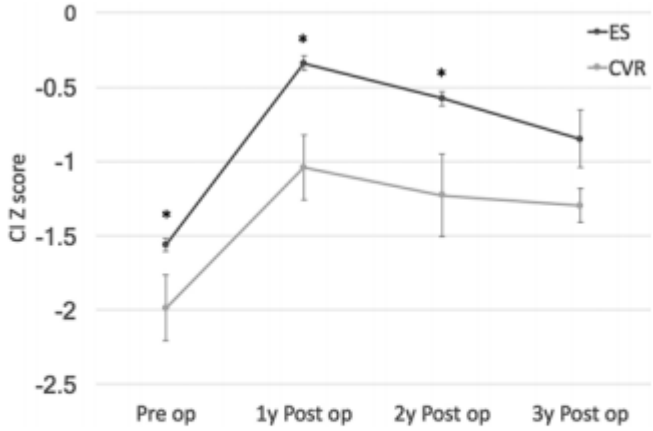


FIG. 2. Line graph of mean CI Z-score by treatment group measured at defined time points: preoperatively, and postoperatively at 1, 2, and 3 years. A CI Z-score = 0 represents a CI equal to the population mean. Error bars represent standard error. *Significant difference between the ES and CVR groups ($p < 0.05$).

ENDOSCOPIC VERSUS CVR

Analysis of clinical outcomes for treatment of sagittal craniosynostosis: a comparison of endoscopic suturectomy and cranial vault remodeling

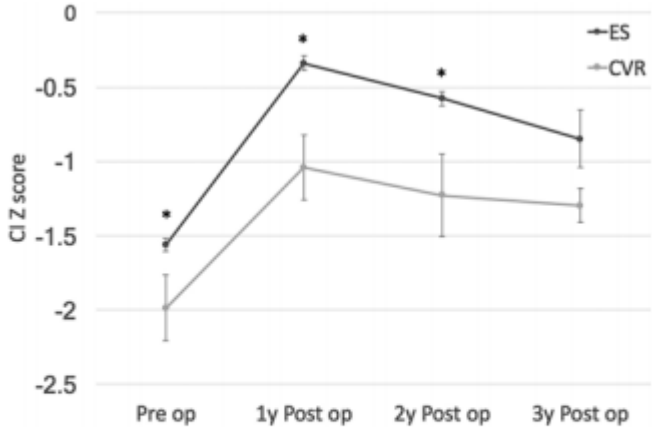
- N=207 patients (187 endoscopic sutrectomy and 20 CVR)
- Operative time: 45 vs. 195 minutes
- LOS: 1 vs. 3 days
- Transfusion rate: 2% vs. 85%
- CI Z-scores were initially more favorable for ES; at 3 years equal
- 4 *syndromic* patients treated by ES required secondary expansion for raised ICP

- ES is an effective treatment for nonsyndromic sagittal synostosis

J Neurosurg Pediatr Volume 22 • November 2018

TABLE 2. Patient characteristics of the study population

Characteristic	CVR (n = 20)	ES (n = 187)
Males, n (%)	10 (50)	137 (73)
Age at presentation, mos		
Median (IQR)	13.5 (8.0–20.3)	2.0 (1.3–3.0)
Range	6.0–47.0	0.5–6.8
Age at operation, mos		
Median (IQR)	14.0 (11.8–23.8)	3.0 (2.5–4.0)
Range	8.0–48.0	1.5–7.0
Duration of helmeting, mos		
Median (IQR)	Not applicable	8.0 (7.0–9.0)
Range		2.0–14.0



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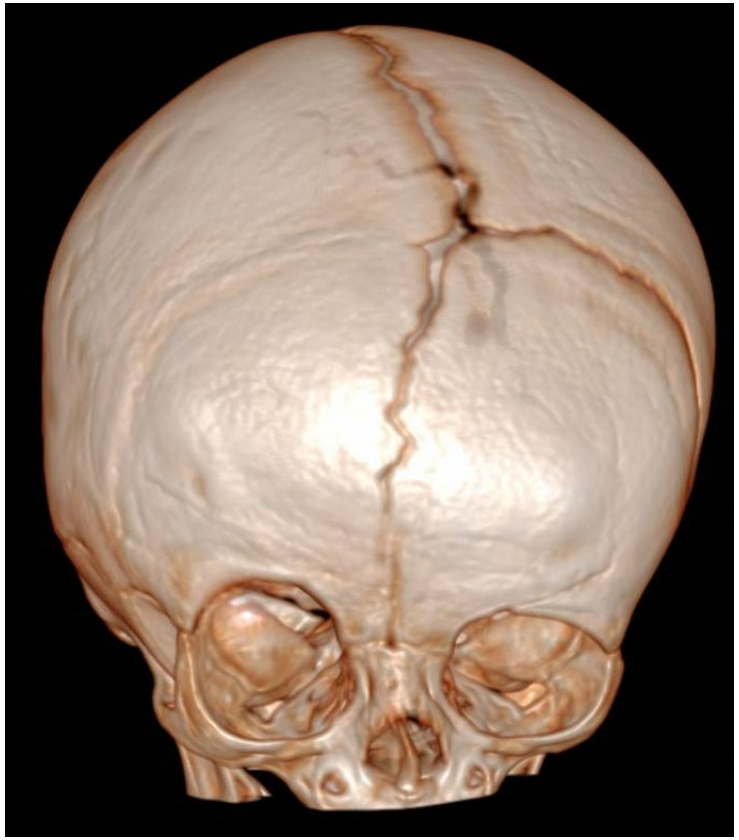
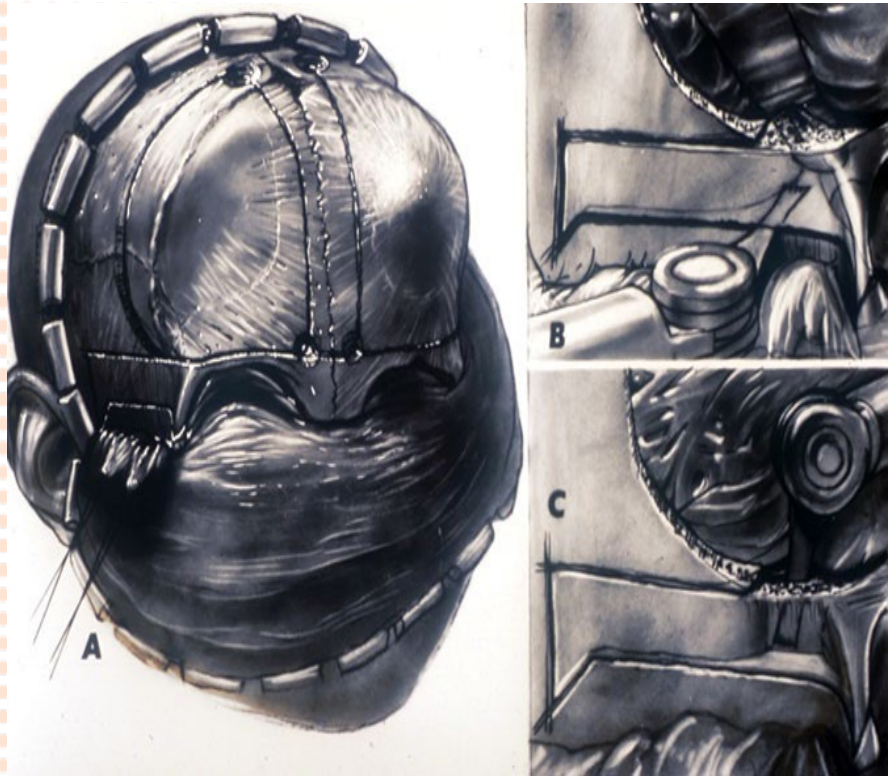
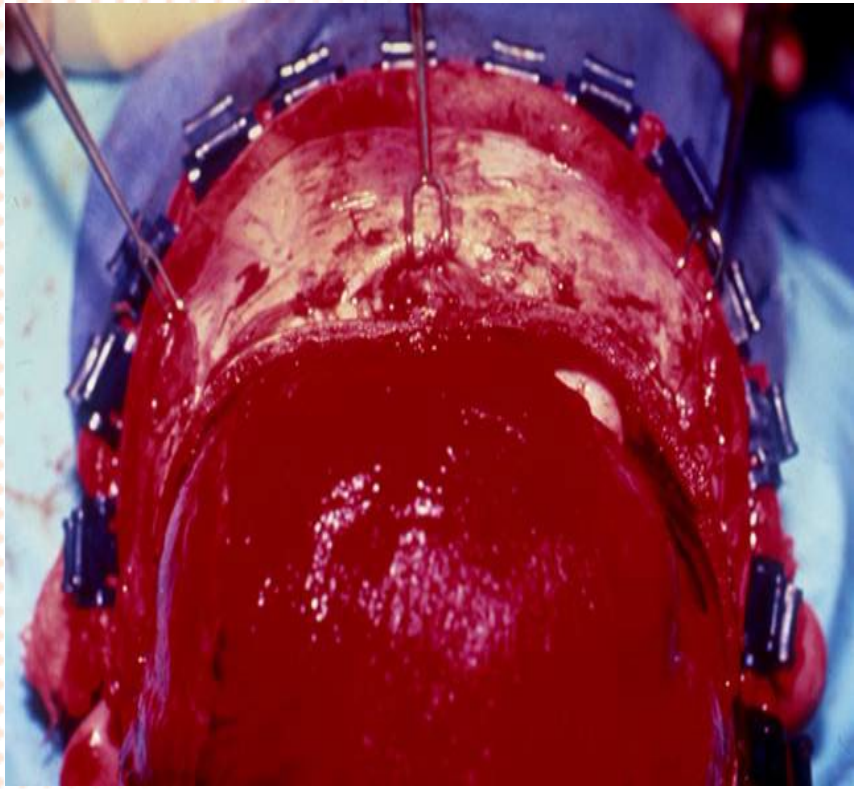
10 MONTH OLD GIRL WITH RIGHT UNICORONAL SYNOSTOSIS

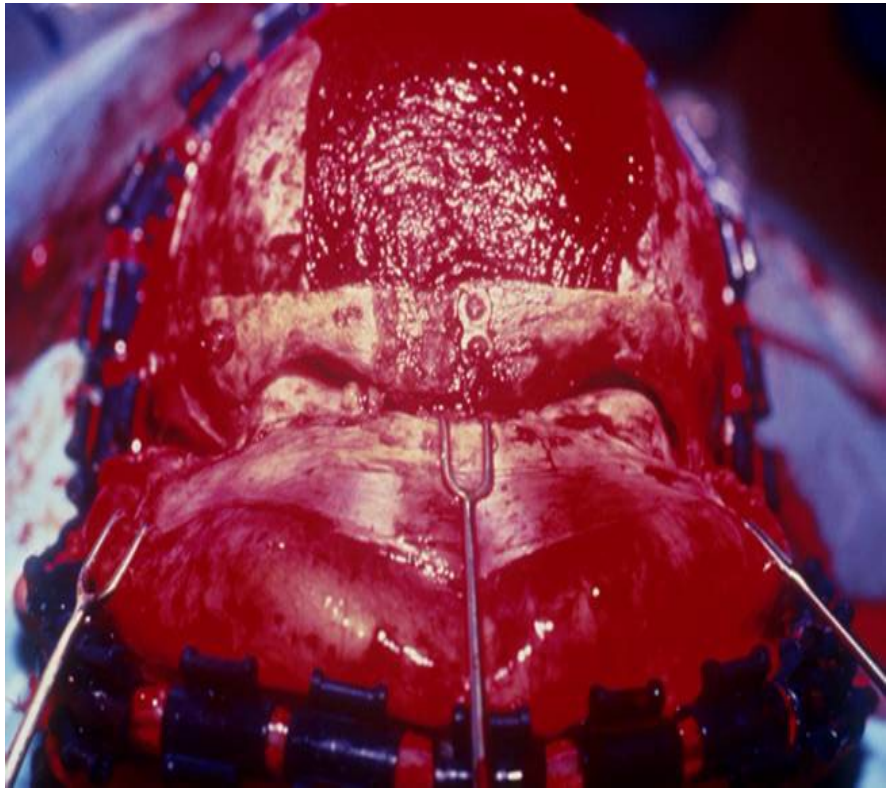


SURGICAL TECHNIQUE



SURGICAL TECHNIQUE





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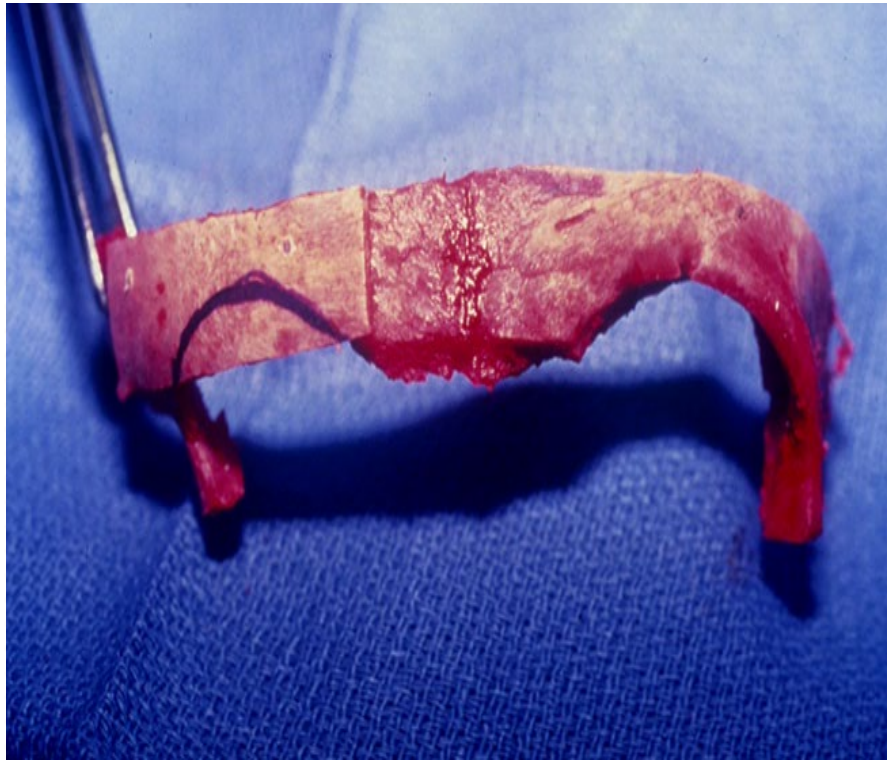

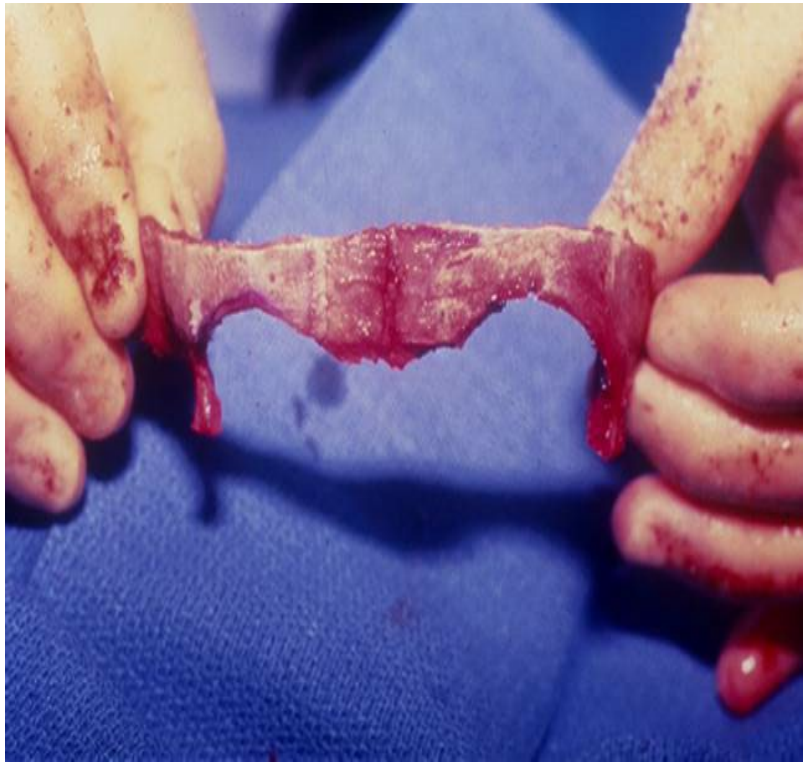

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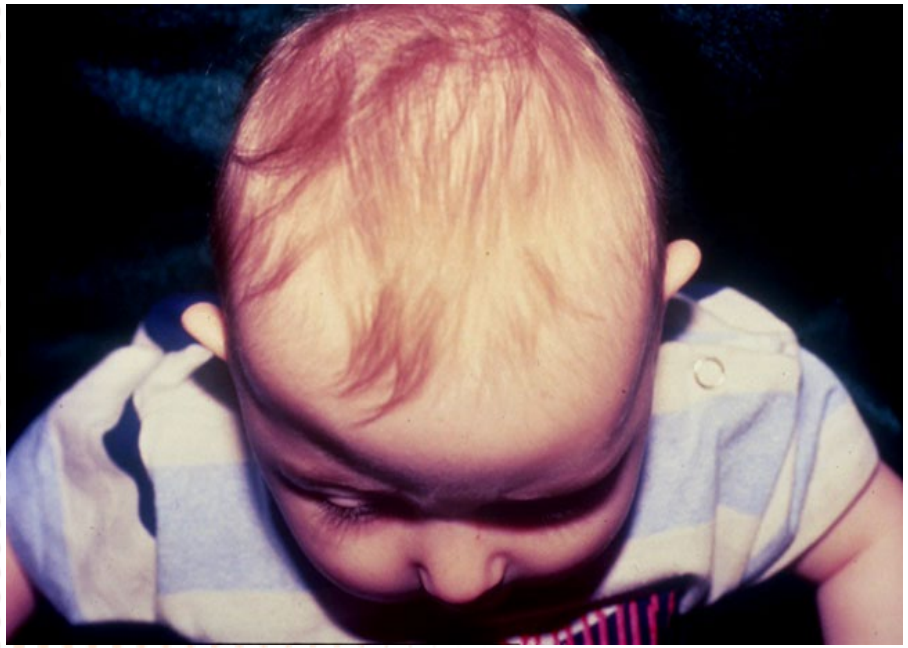


3 MONTH OLD BOY WITH RIGHT UNICORONAL SYNOSTOSIS

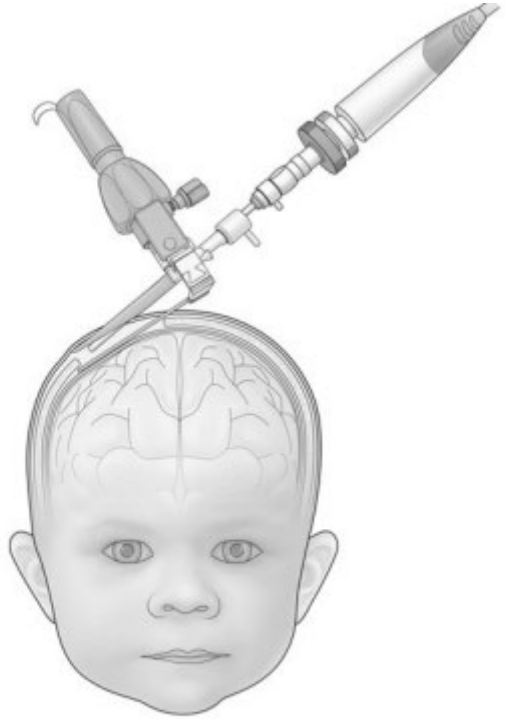
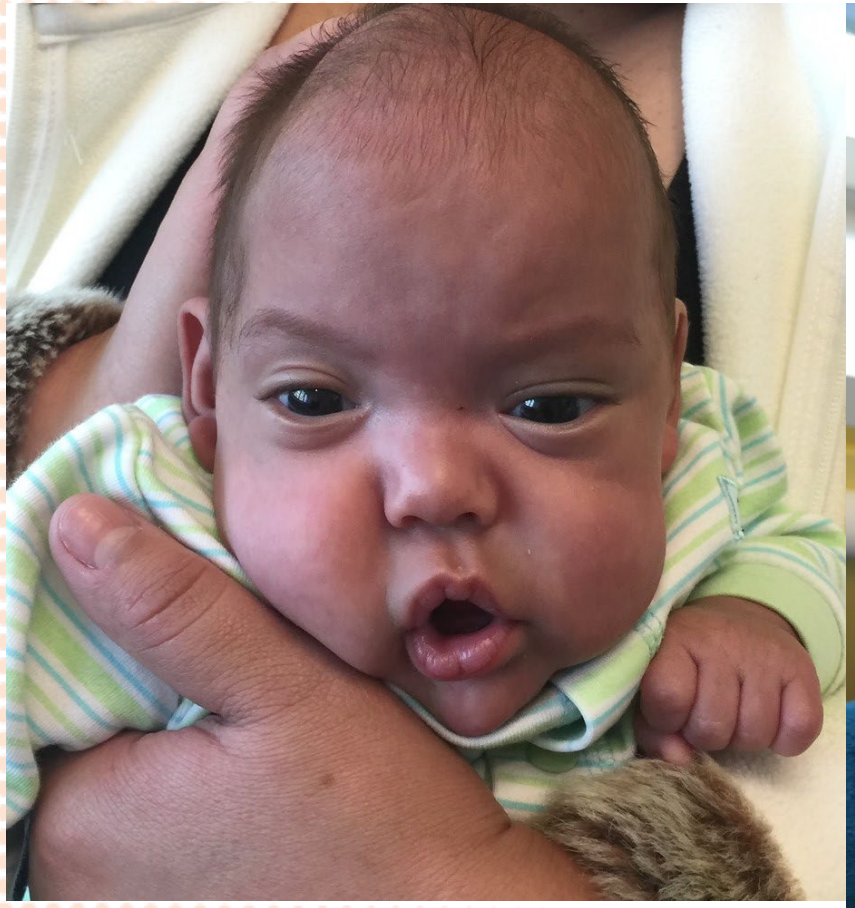
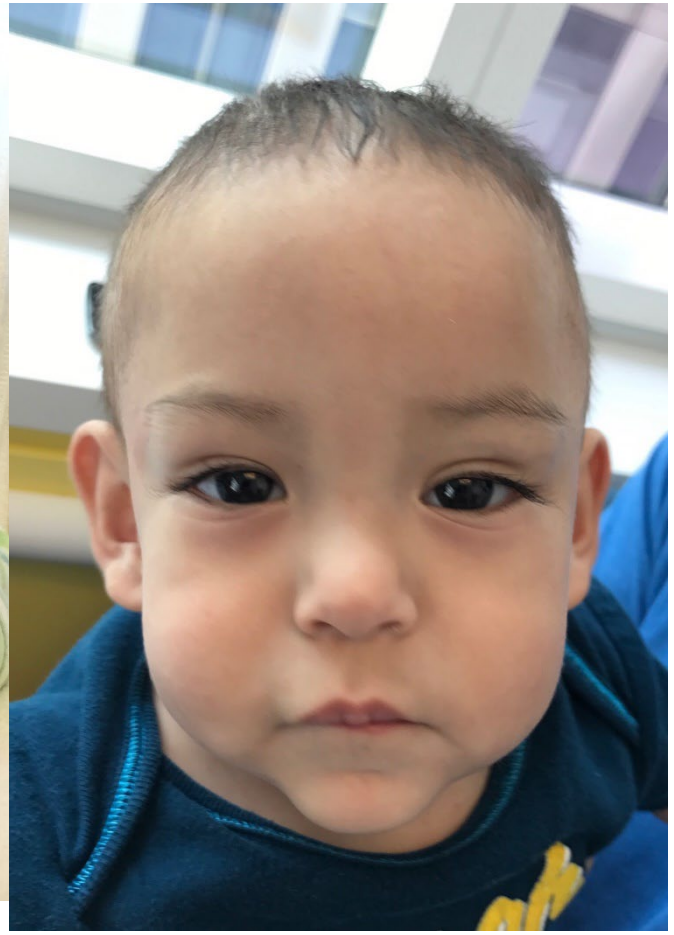


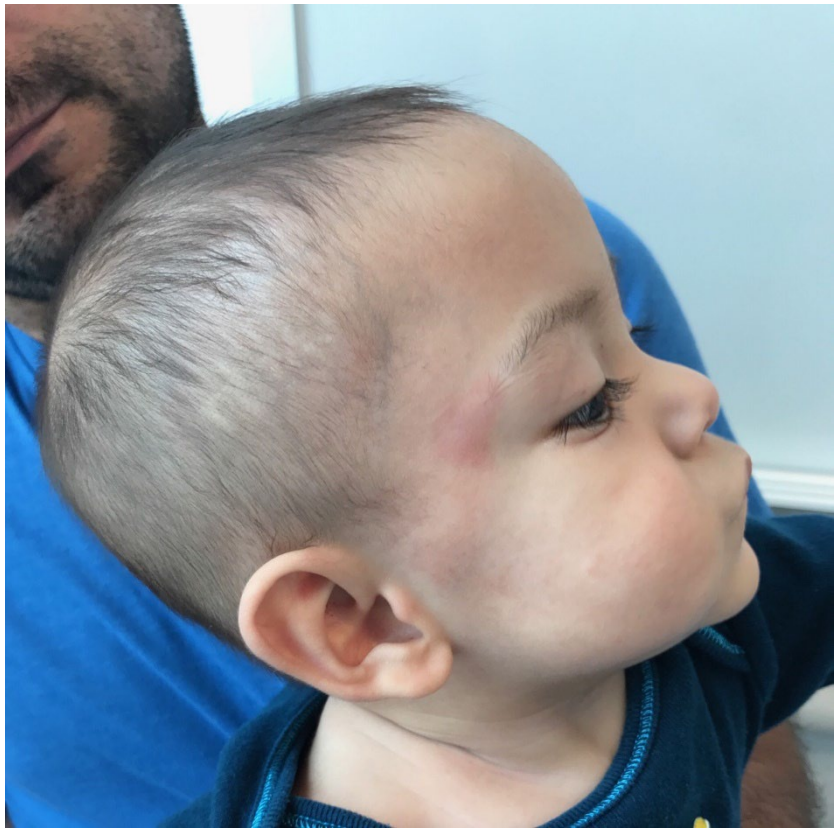
Fig. 1 Artist's rendition of a patient with coronal synostosis undergoing endoscopic surgery. A small incision is used to place the J&B Dural Retractor. The insulated blades retract the scalp and protect the dura as a zero degree endoscope provides adequate and direct visualization

Childs Nerv Syst (2012) 28:1429–1432

3 MONTH OLD BOY WITH RIGHT UNICORONAL SYNOSTOSIS

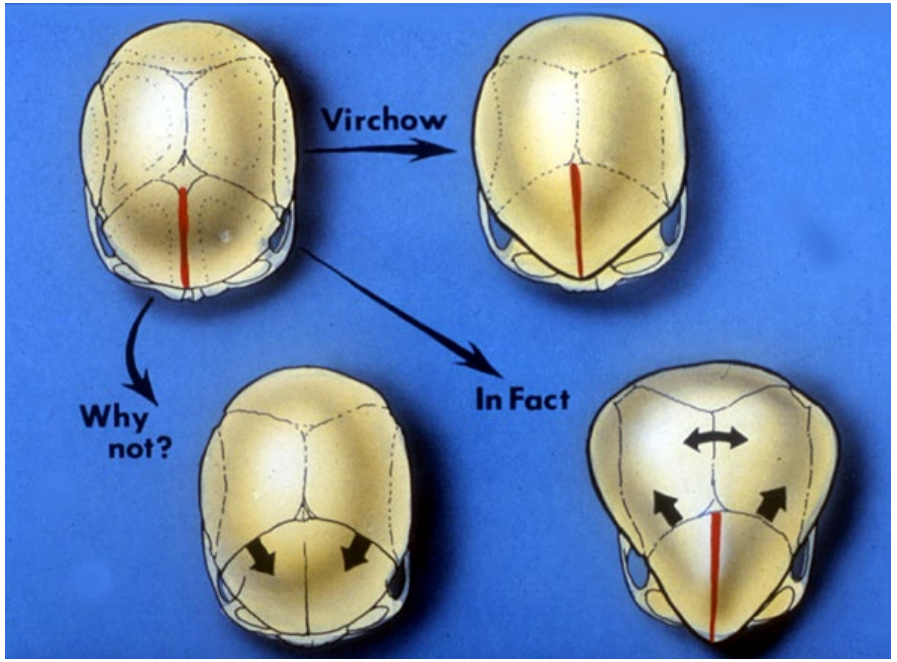






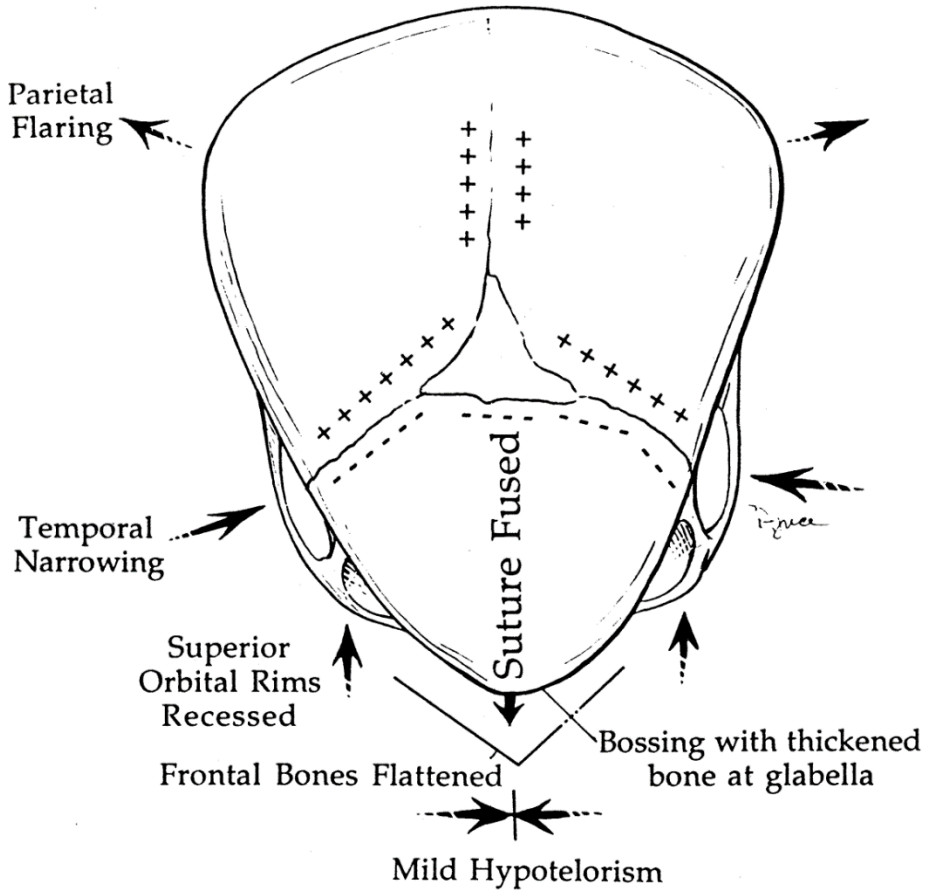
METOPIC SYNOSTOSIS

- 3rd most common; M:F 7:3
  - Only suture that normally fuses
- Trigonocephaly (triangular shape)
  - Midfrontal keel
  - Bifrontotemporal narrowing
  - Parieto-occipital protrusion
- Considered most at risk for cognitive or behavioral impairment
  - Higher CNS anomalies, chromosome defects than other non-synodromic synostosis



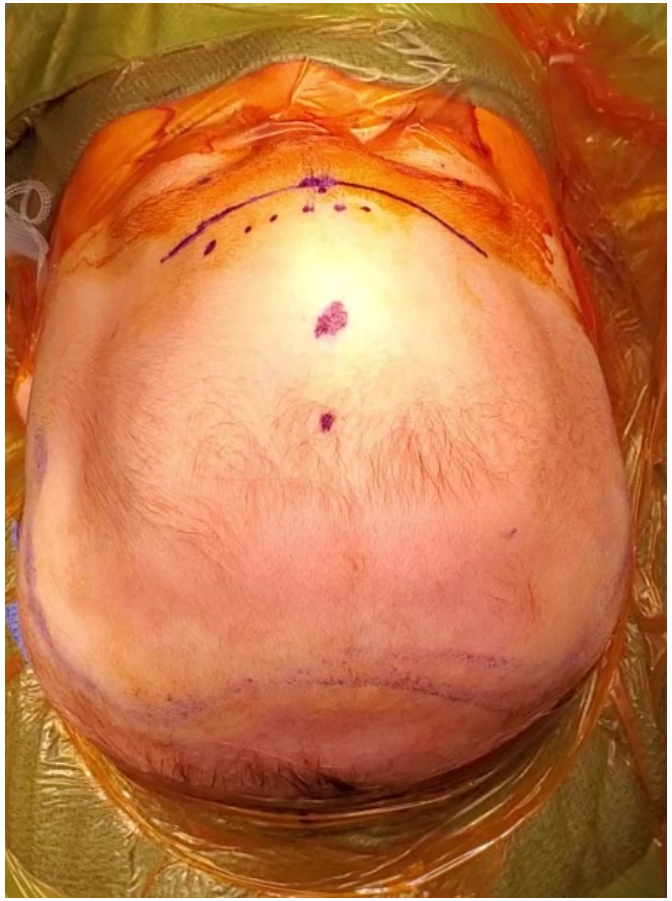
FACIAL DEFORMITY

- Excessive narrowing of the interorbital space
  - Orbital hypotelorism
  - Epicanthal folds
  - Superolateral orbital rim retrusion
  - Low nasal dorsum



SURGICAL TECHNIQUE

- Bifrontal craniotomy with bilateral recontouring, fronto-orbital advancement
  - Achieve symmetry of forehead
  - Similar dissection initially to bilateral coronal synostosis
  - Bandeau divided at midline
  - Contoured with Tessier forceps
  - Interposition graft connecting bandeau
  - Frontal bones divided through keel and contoured



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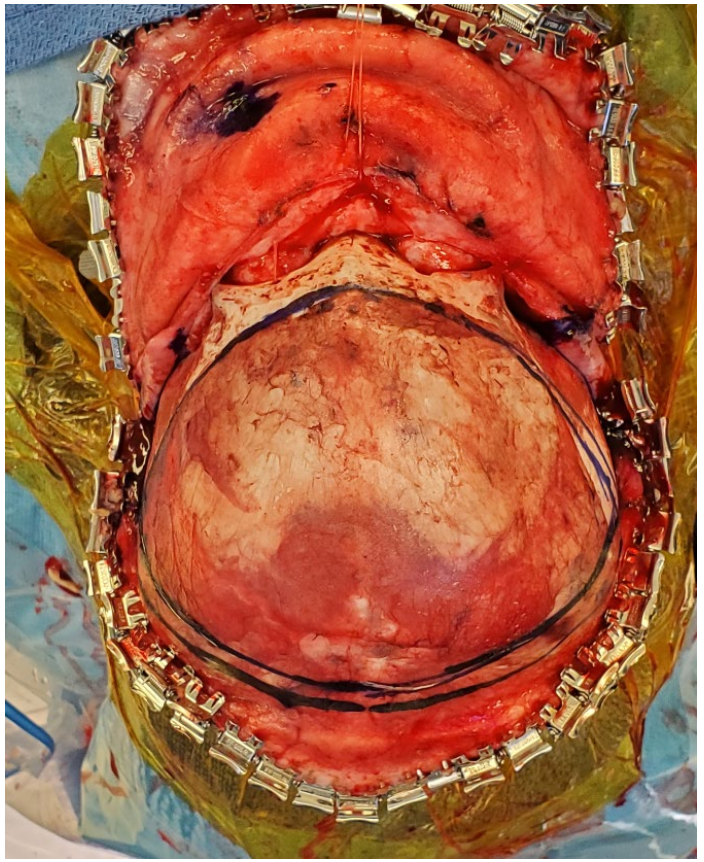

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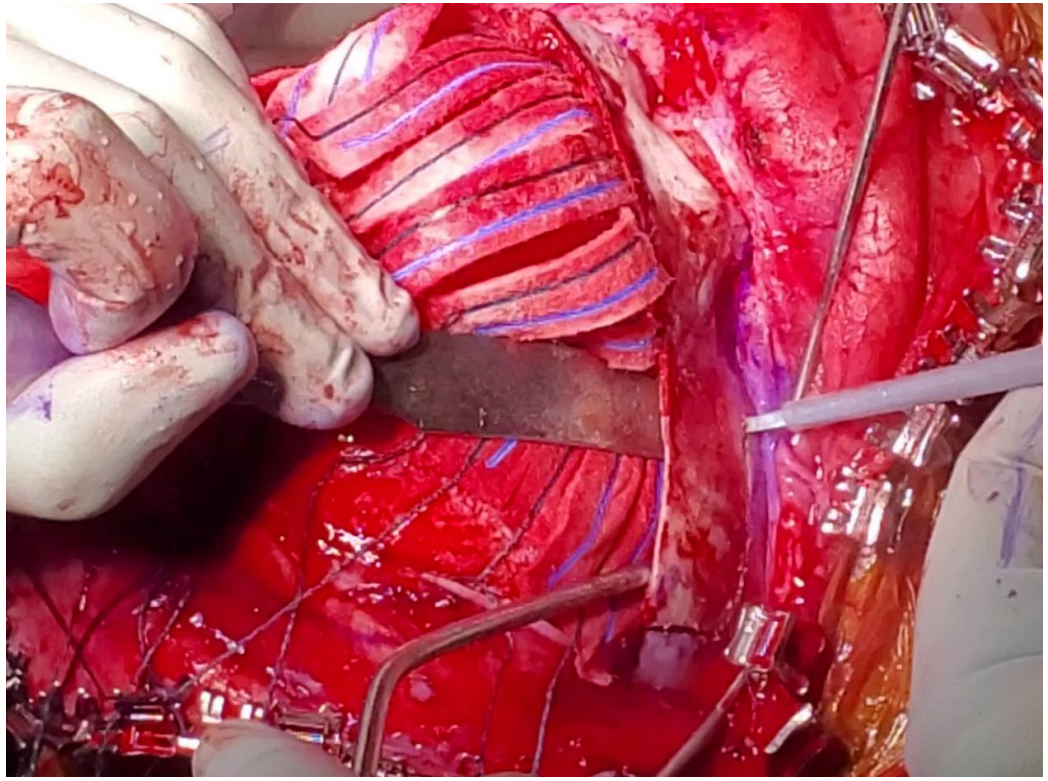

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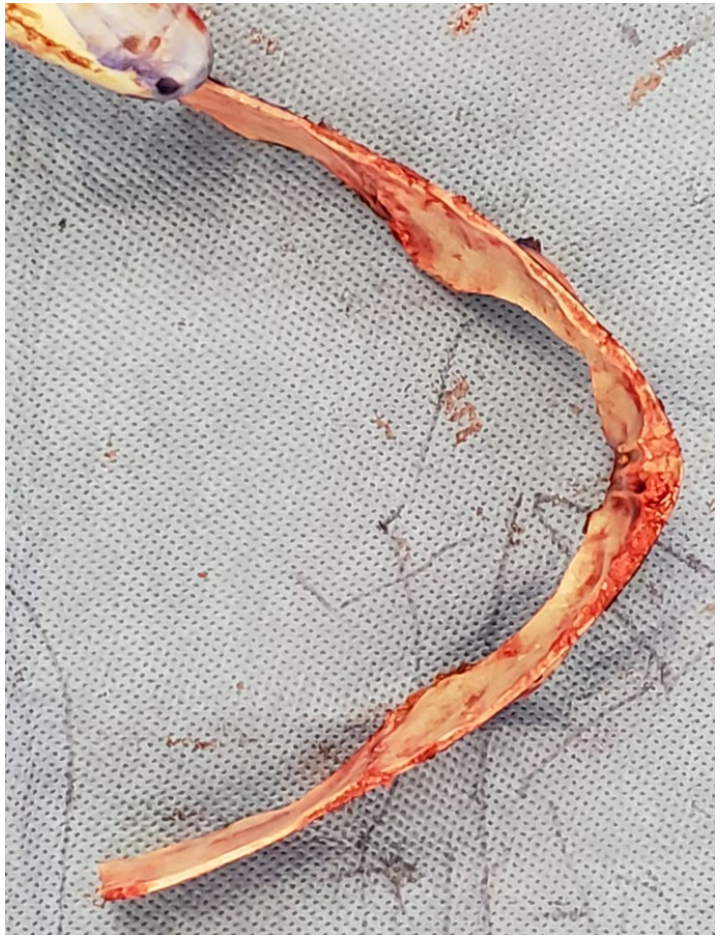

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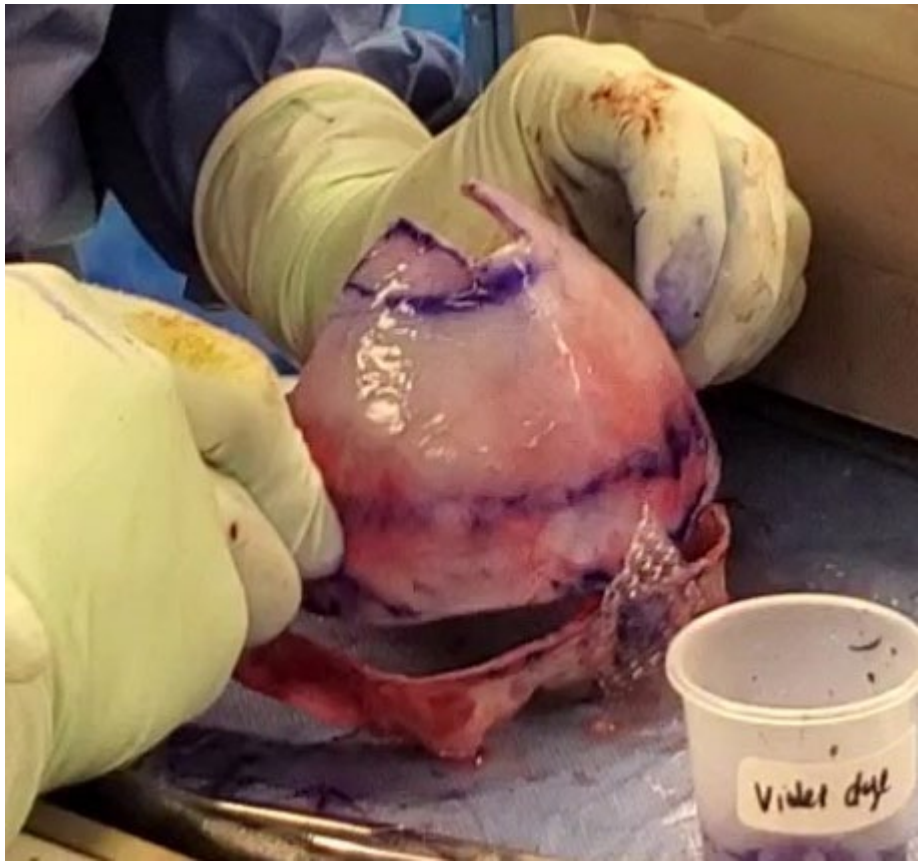

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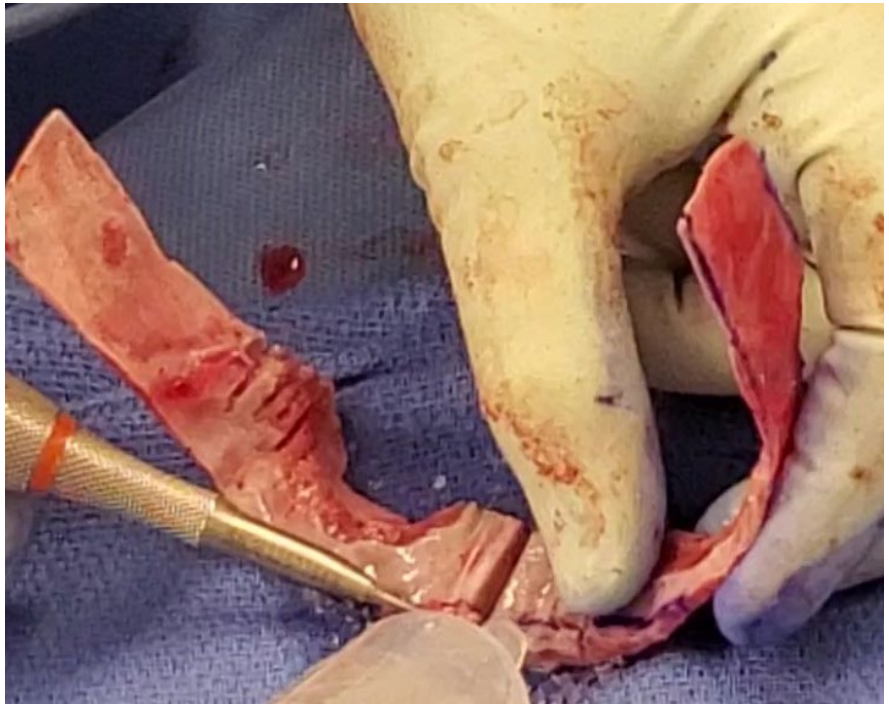

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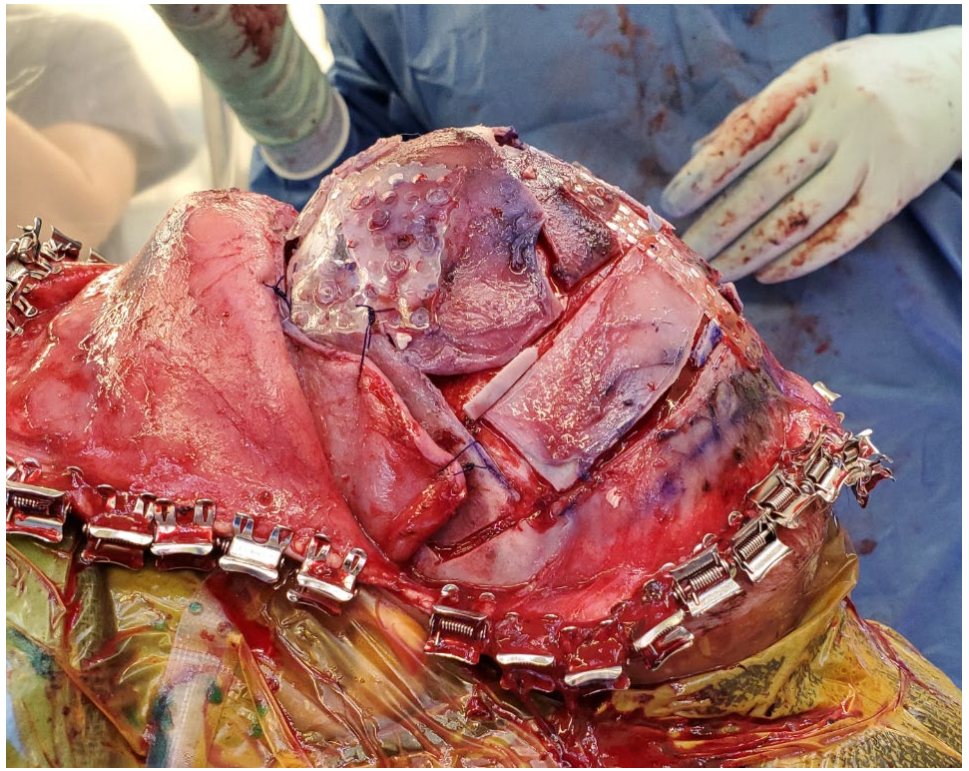

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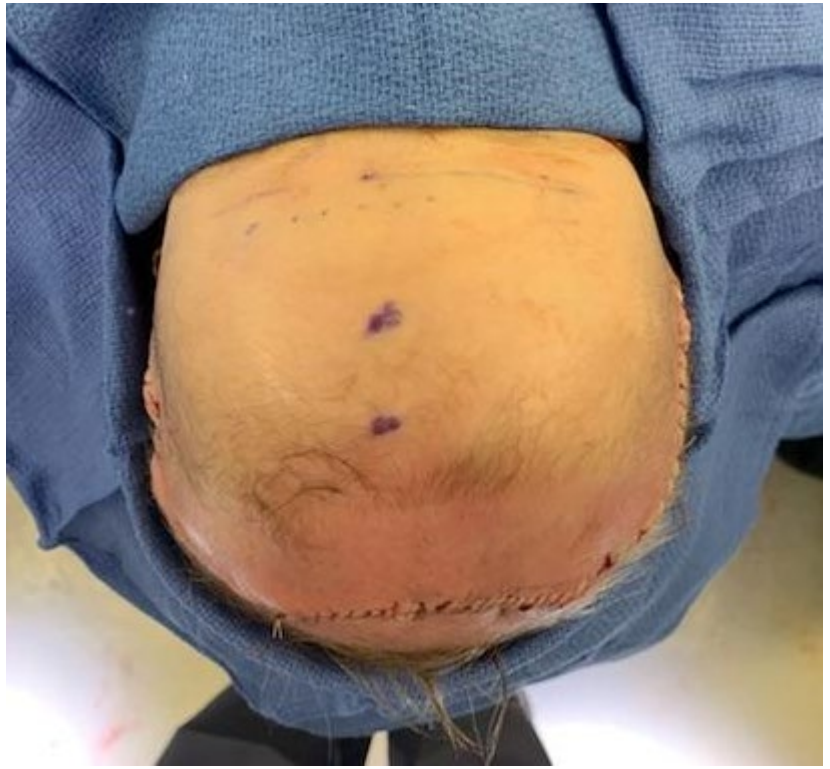
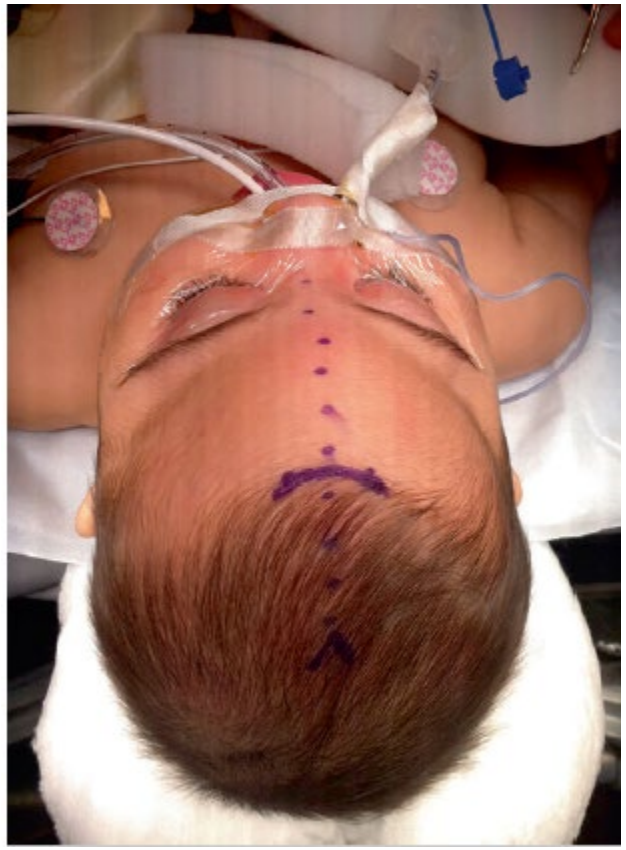


ENDOSCOPIC TECHNIQUE

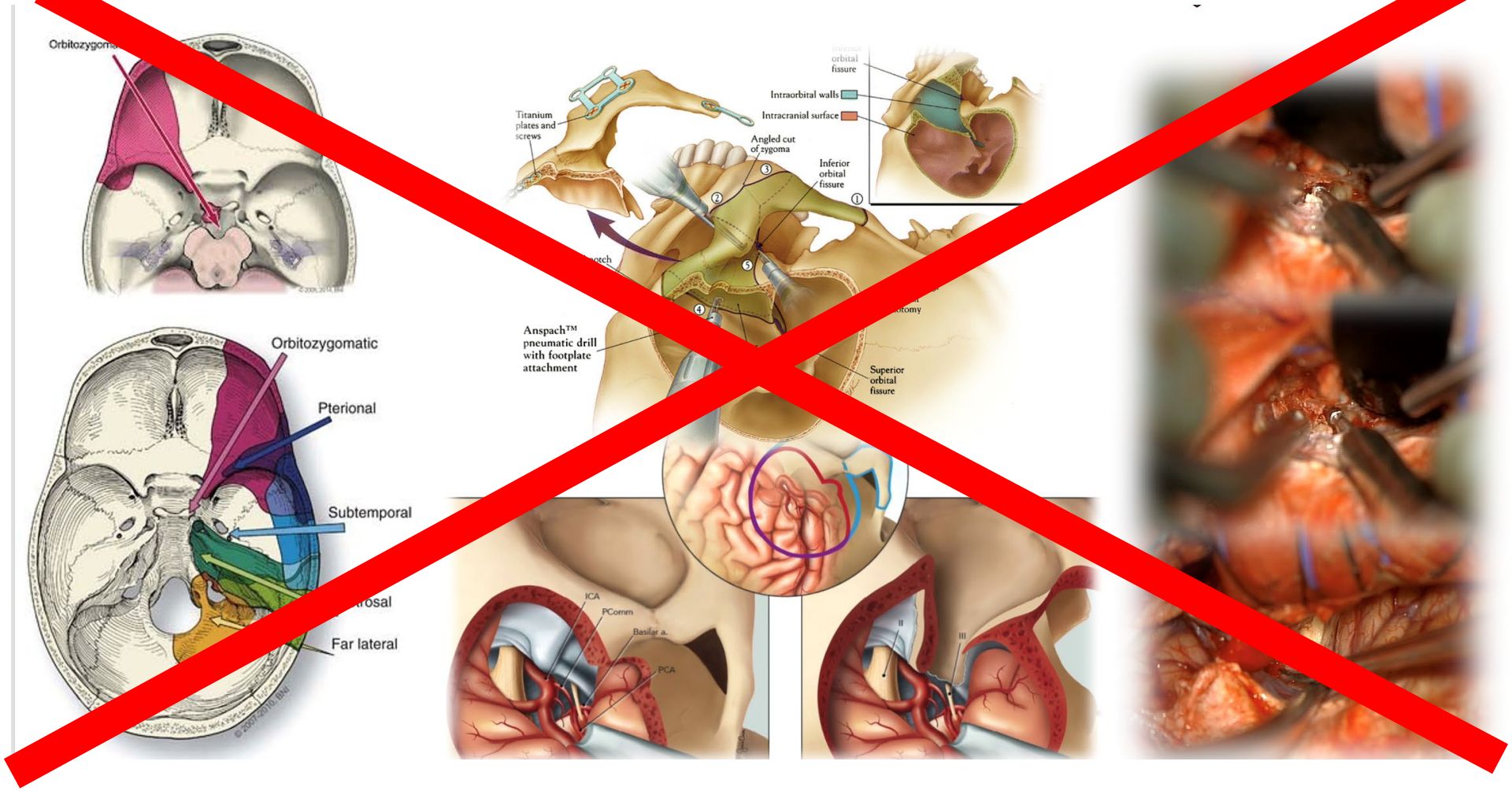
- Infant younger than 4-6 months
- Ultimate frontal bone projection often falls slightly short of that achieved with traditional surgery
  - Correction of superlateral orbital rim retrusion is often not as significant as what can be achieved in open procedure

- Paced supine
- 1.5 to 2.5 cm incision made just posterior to midline
- Often two emissary veins encountered when dura stripped and can be coagulated
- Ultrasonic bone aspirator to reach to nasofrontal juncture

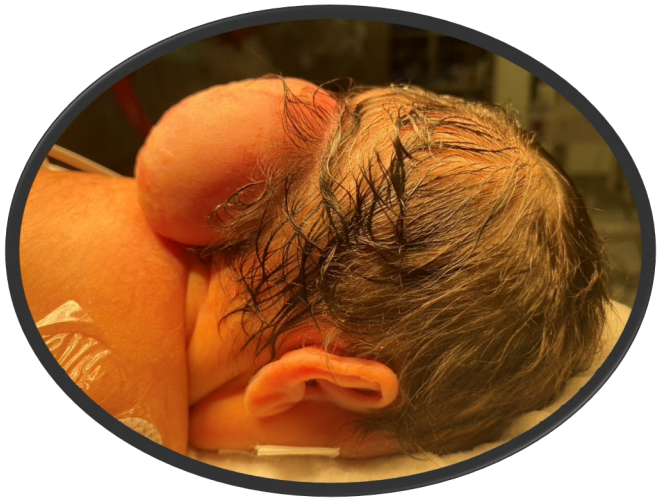


BUILDING A CRANIOSYNOSTOSIS CENTER

- Community pediatricis team and timing of surgery
- Encouraging referral as soon as there is suspicion of abnormal head shape
- Early diagnosis and referral gives family option to consider techniques
- Establishing multi-disciplinary care of craniosynostosis
  - Craniofacial surgeons
  - Audiologists
  - Dentist
  - Ophthalmology
  - Pediatrician
  - PT/OT



Children's National



Today

1.00

0.01

p(neurosurgery)

"A 1 month old with a sacral dimple …"

"A 3 month old with abnormal head shape …"

"A 9 month old with large head …"

"A 3 year old with ataxia …"

"An 8 year old with headaches …"

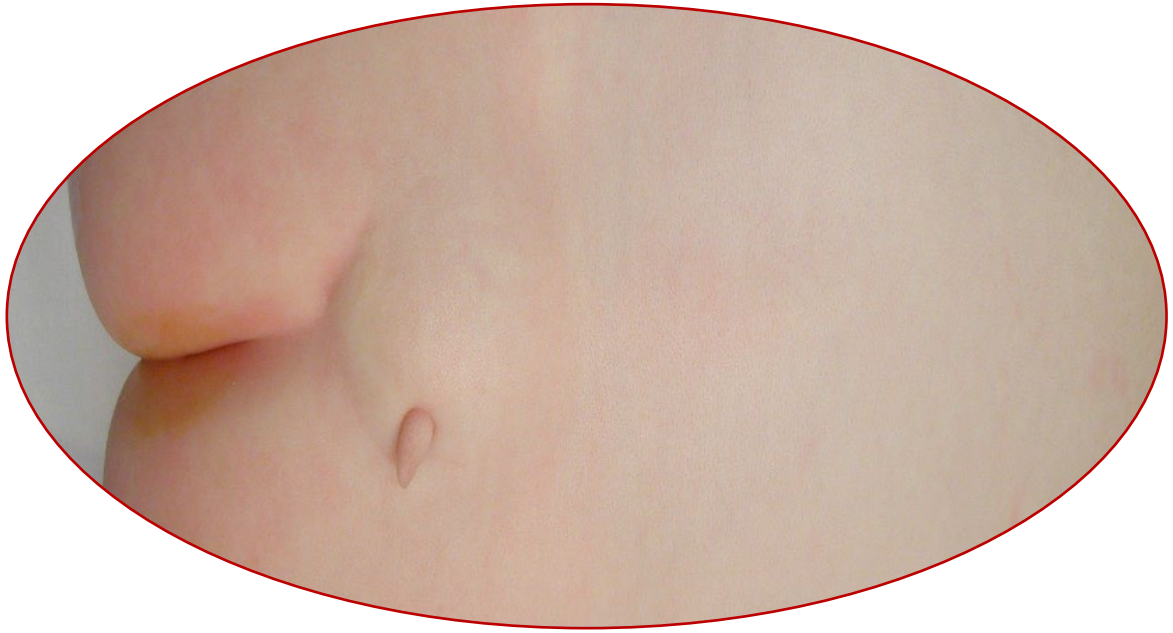
"A 4 year old with head trauma…"

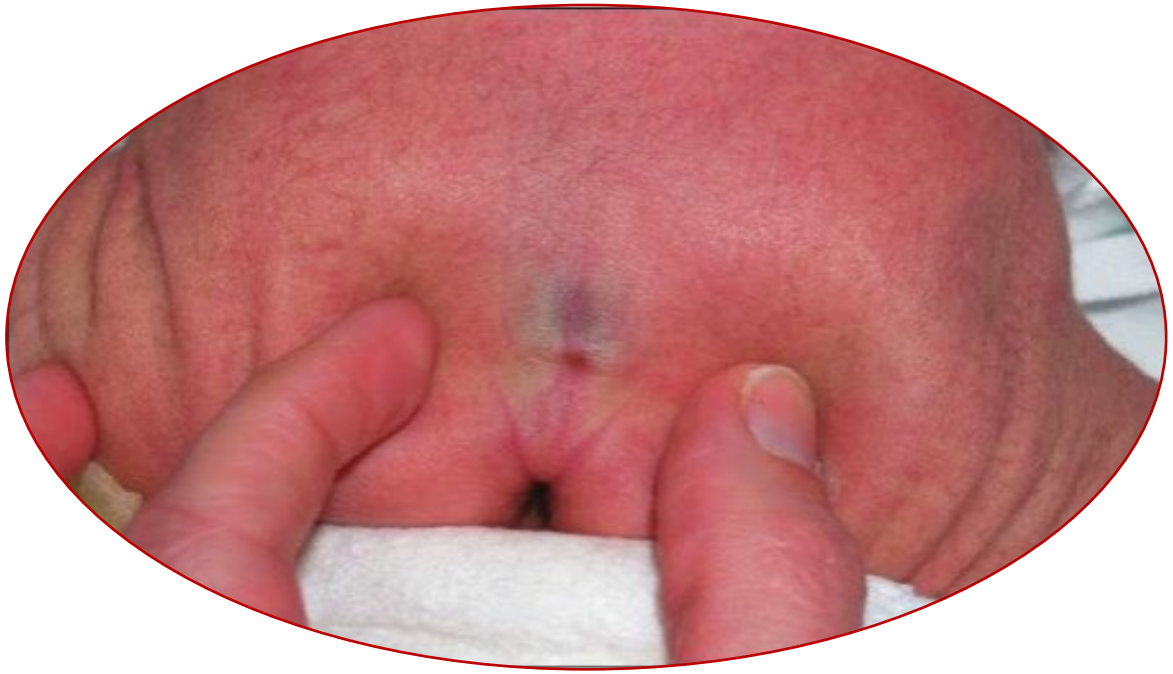
1.00

0.01

p(neurosurgery)

Newborn with a sacral dimple





1.00 p(neurosurgery) 0.01

Newborn with a sacral dimple – what's at stake?

Clinical examination (looking for…)
MRI spine with and without contrast
Strongly consider sedation if > 1 month (dx early…)
91% of <1 yr need anaesthesia

Refer to neurosurgery (ok to refer prior to ordering imaging)

We found no significant association between the number of sedated MRI scans and cognitive outcome at 4.6 y in our cohort after adjustment for confounding variables, including the number and timing of surgeries. Our institutional prac-

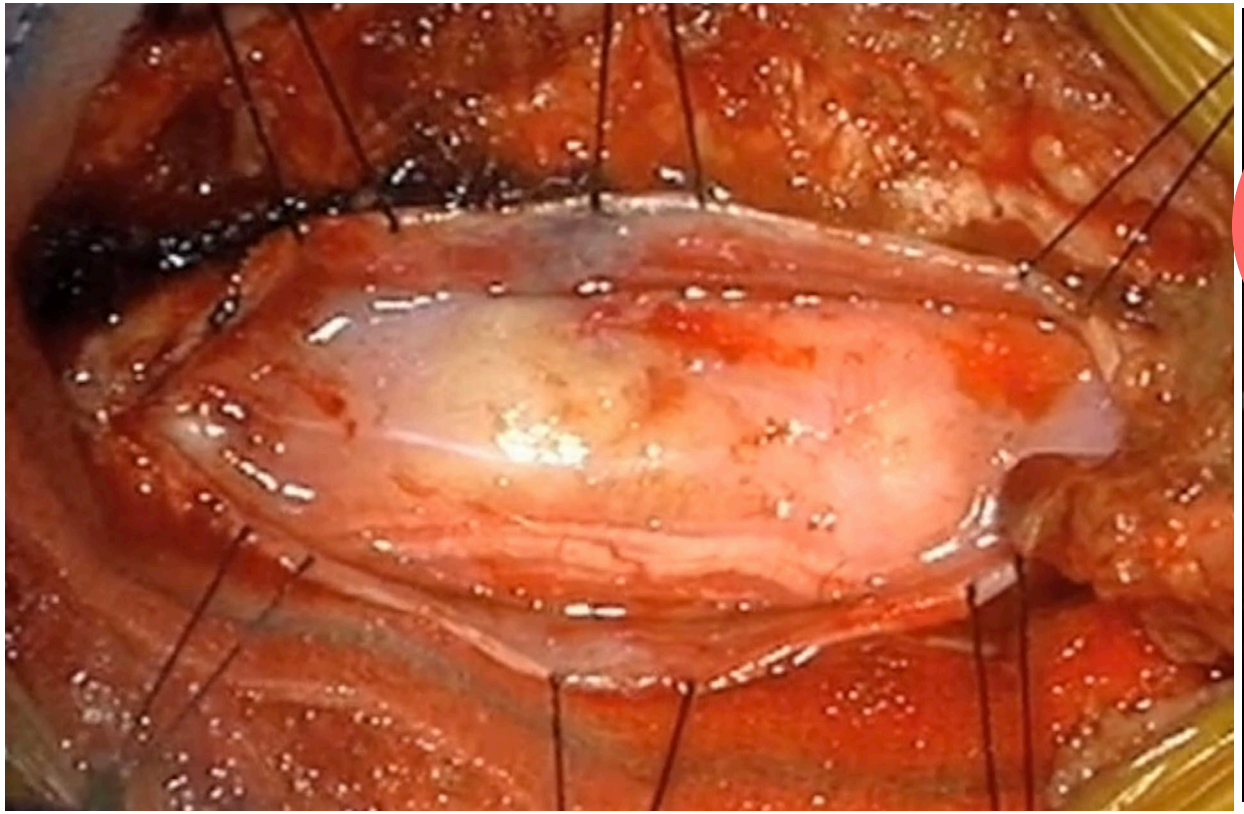
Impaired cognitive performance in premature newborns with two or more surgeries prior to term-equivalent age

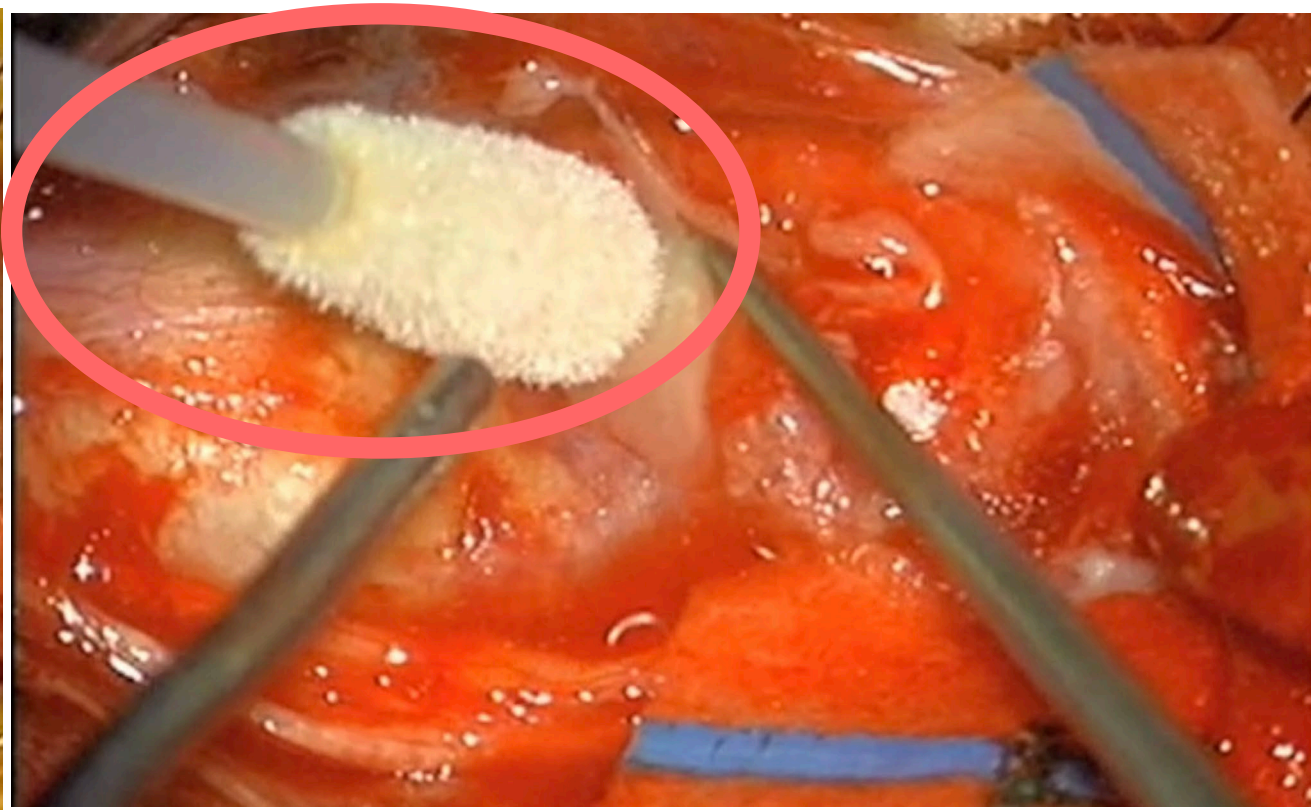
Dawn Gano[1], Sarah K. Andersen[2], Hannah C. Glass[1,3], Elizabeth E. Rogers[1], David V. Glidden[4], A. James Barkovich[1,3,5] and Donna M. Ferriero[1,3]

Pediatric RESEARCH Volume 78 | Number 3 | September 2015

Children's National

Newborn with a sacral dimple – what's at stake?





Which Sacral Dimples are Dangerous?

Generations of physicians have been taught that a dimple is innocent if its base can be visualized and abnormal if its bottom cannot be seen; *this teaching is incorrect*. The presence or absence of a "bottom" to the dimple has little to do with its pathologic nature. Rather, *it is the location of the dimple along the craniocaudal axis* that is the most important feature. As the name implies, the innocent *coccygeal* dimple is more caudally located than the pathologic *lumbosacral* DST. It

CLINICAL REPORT Guidance for the Clinician in Rendering Pediatric Care

American Academy of Pediatrics

DEDICATED TO THE HEALTH OF ALL CHILDREN™

Congenital Brain and Spinal Cord Malformations and Their Associated Cutaneous Markers

Mark Dias, MD, FAANS, FAAP, Michael Partington, MD, FAANS, FAAP, the SECTION ON NEUROLOGIC SURGERY

"SIMPLE DIMPLE RULES" FOR SACRAL DIMPLES[6]

The following parameters define which sacral dimples are high risk:[6,7]

- Larger than 0.5 cm in size.
- Located more than 2.5 cm cephalad to the anal verge.
- Associated with overlying cutaneous markers:
  - True hypertrichosis, or hairs within the dimple (distinctly different than the mild hairiness seen in **Figure 6**).
  - Skin tags.
  - Telangiectasia or hemangioma (**Figure 7**).
  - Subcutaneous mass or lump.
  - Apparent aplasia cutis.
  - Abnormal pigmentation.
- Bifurcation (fork) or asymmetry of the superior gluteal crease (**Figure 8**).

ANY of these

The Enigmatic Sacro-Coccygeal Dimple: To Ignore or Explore?

Stan L. Block, MD, FAAP

PEDIATRIC ANNALS 43:3 | **MARCH 2014**

Spinal DSTs may be investigated using spinal ultrasonography and/or MRI, although it is important to point out that the decision to treat is made *solely on the presence of the pathologic dimple*, regardless of imaging findings. The DST may not be visualized, and the spinal cord is not always radiographically tethered (ie, below the mid-body of L2); even high-resolution MRI may miss as many as 50% of DSTs.[21] The value of neuroimaging is, therefore, largely to look for associated anomalies or the presence of dermoid or epidermoid cyst(s) as part of surgical planning.

All spinal DSTs should be repaired regardless of imaging studies,

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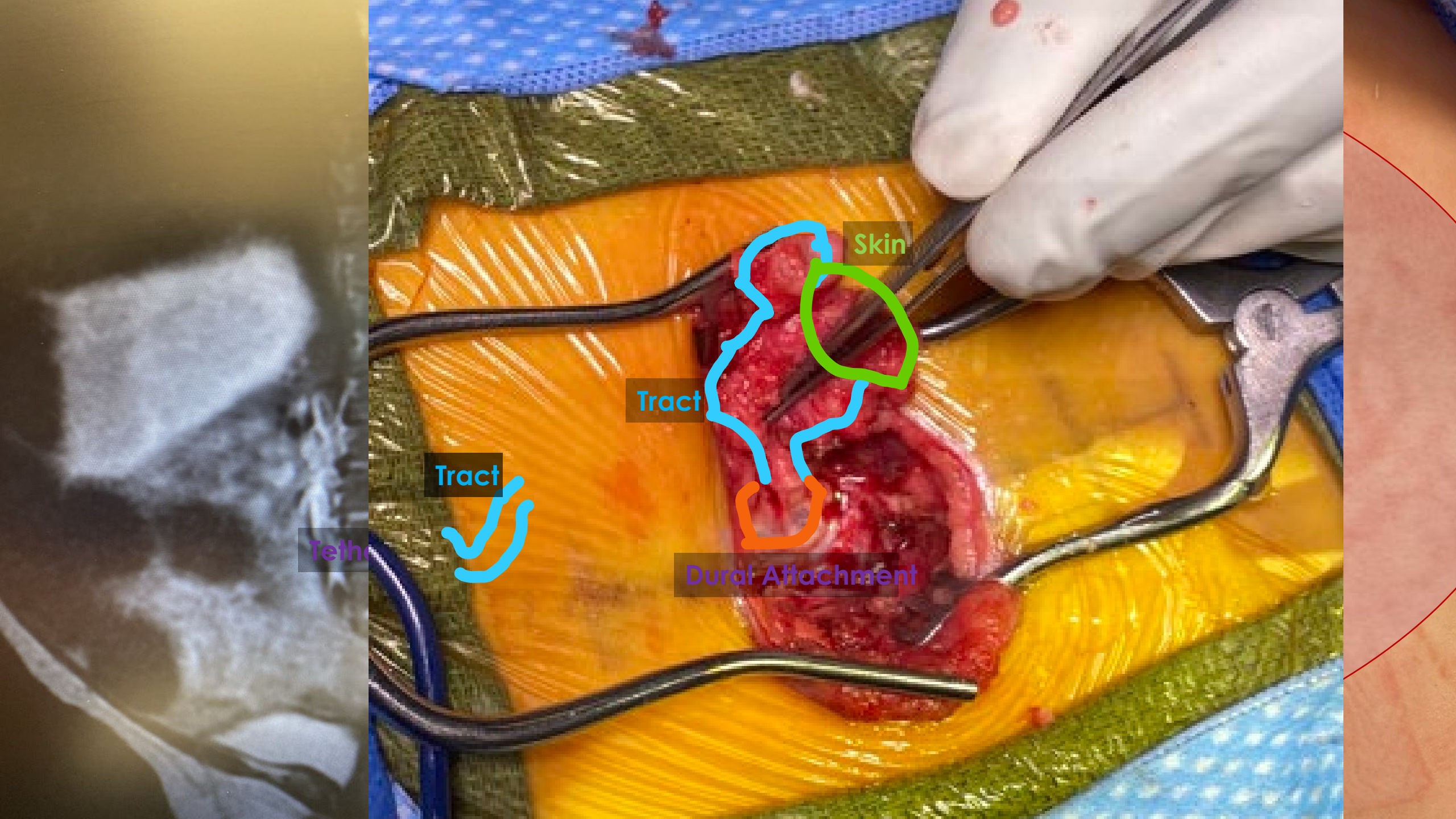
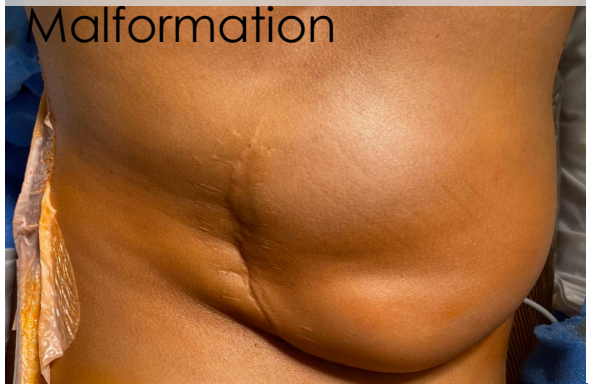
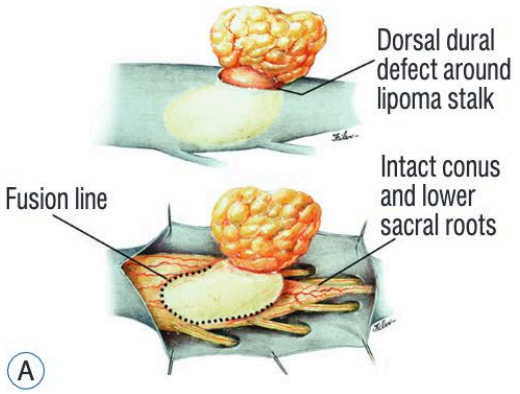
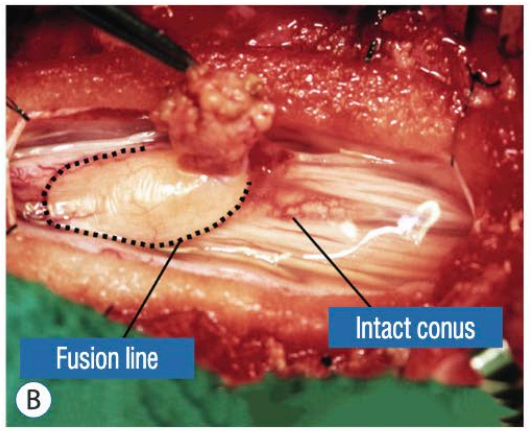

Skin
Tract
Tract
Dural Attachment

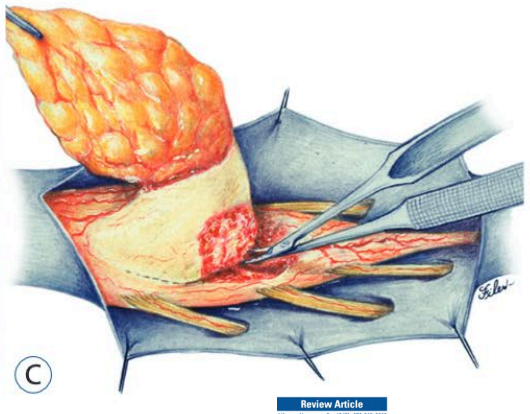
Many Spinal Malformations Follow These Ideas

Lipomatous Malformation







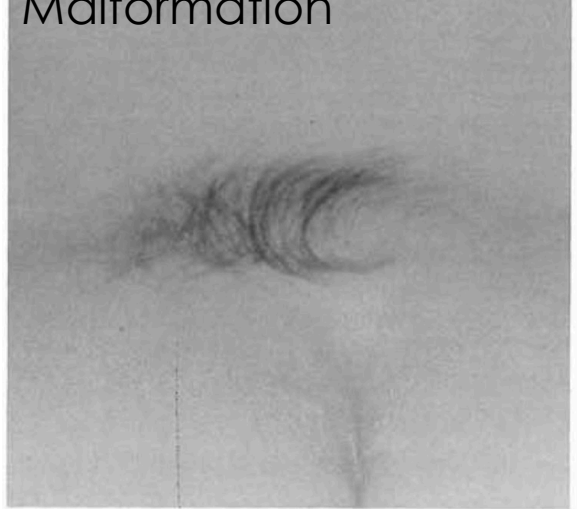


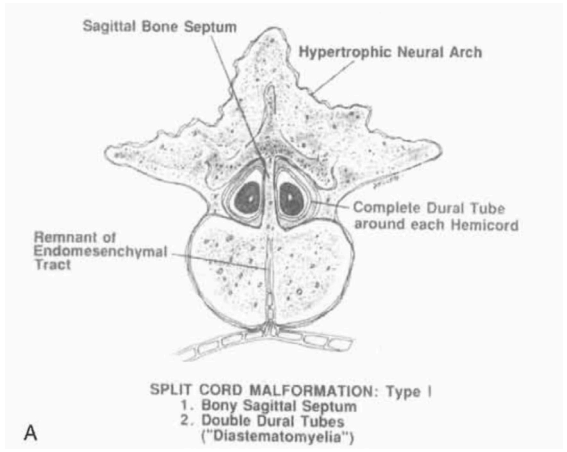
Review Article

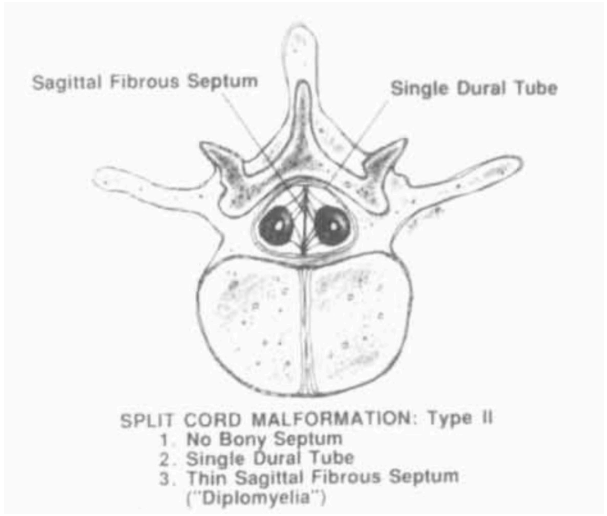
Surgical Management of Complex Spinal Cord Lipomas : A New Perspective

Dachling Pang

Split Cord Malformation







SPINAL DYSRAPHISM 1042-3680/95 $0.00 + .20

SPLIT CORD MALFORMATIONS

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Children's National

Big head … which one is hydrocephalus?

Primary CSF disorders in Infancy

Benign Enlargement of the Subarachnoid Spaces

Fontanelle flat or sunken, sutures opposed

Large head, more notably brachycephalic

≥50% famililal (**measure the parents esp. dad**)

Progressive, peaks from 4-12 months of age

Obtain HUS to confirm diagnosis, self limited

Subdural hematomas are common (5%) **not diagnostic** of NAT

May have mild motor delays (macrocephaly) but should normalize in function and HC by age 2-3

Hydrocephalus

Fontanelle bulges, sutures splay (≥2mm)

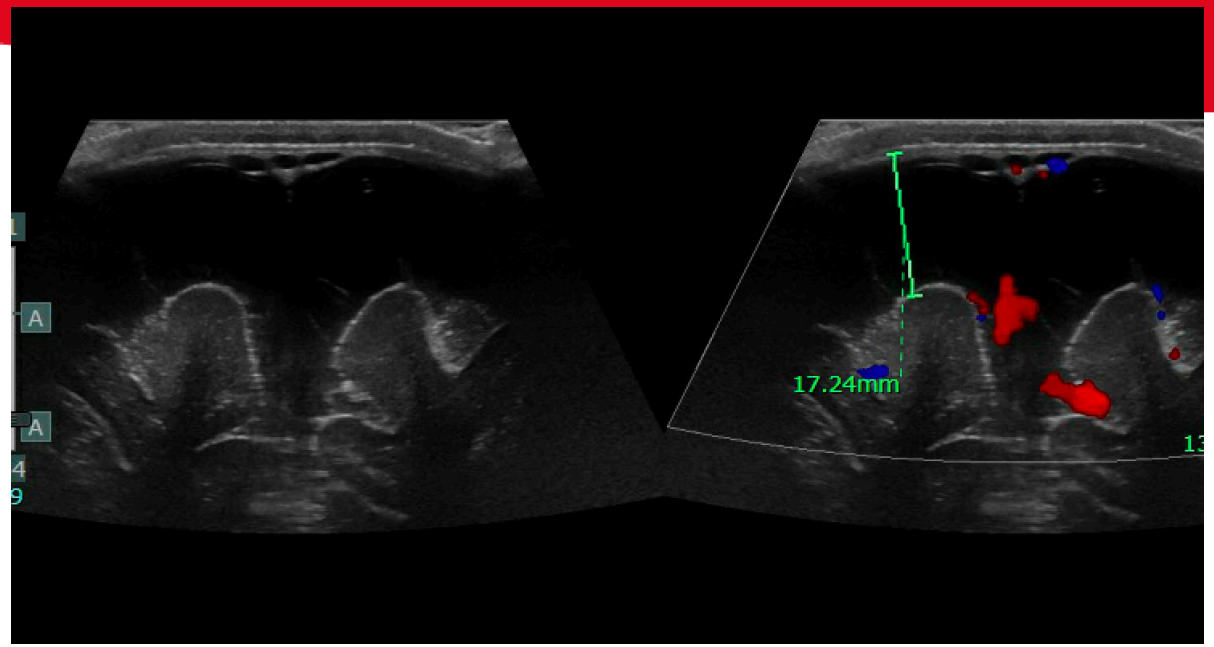
Frontal bossing, scalp veins dilated

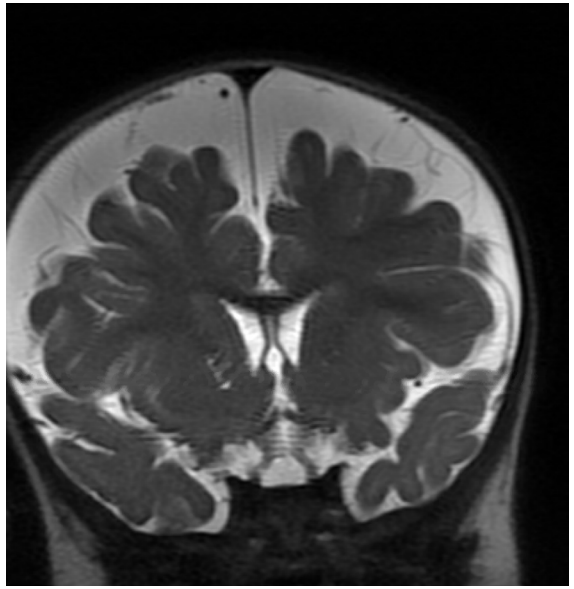
Usually sporadic (except X-linked)

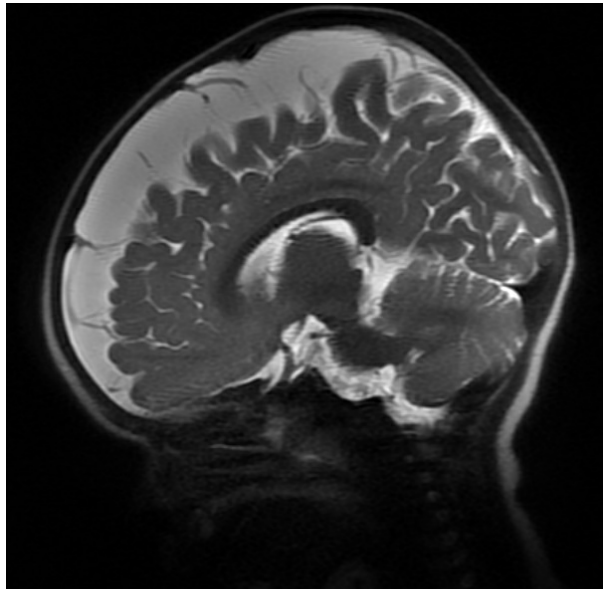
Recognized at any age, often ≤6 months

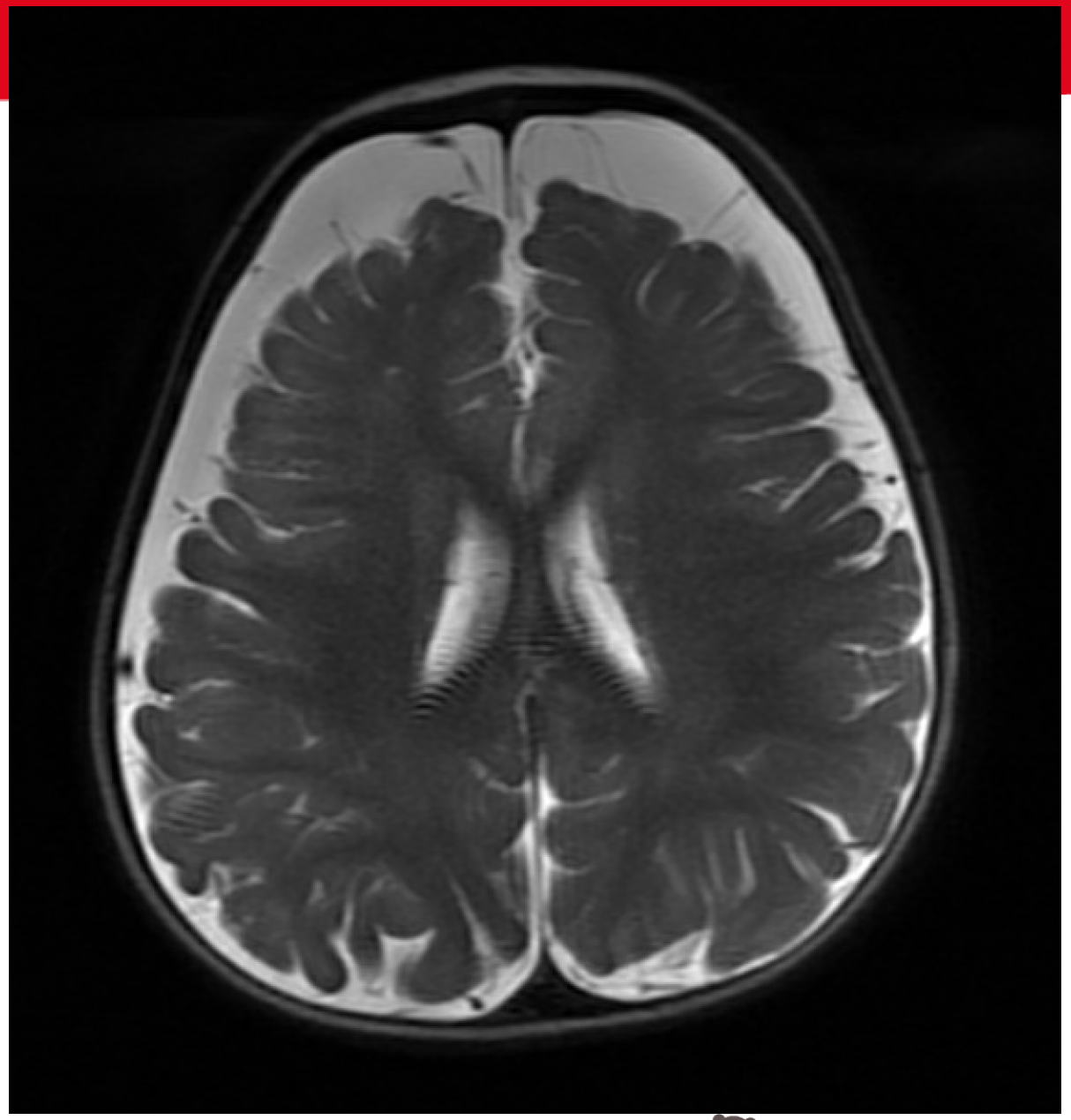
Danger Signs: Send to ED

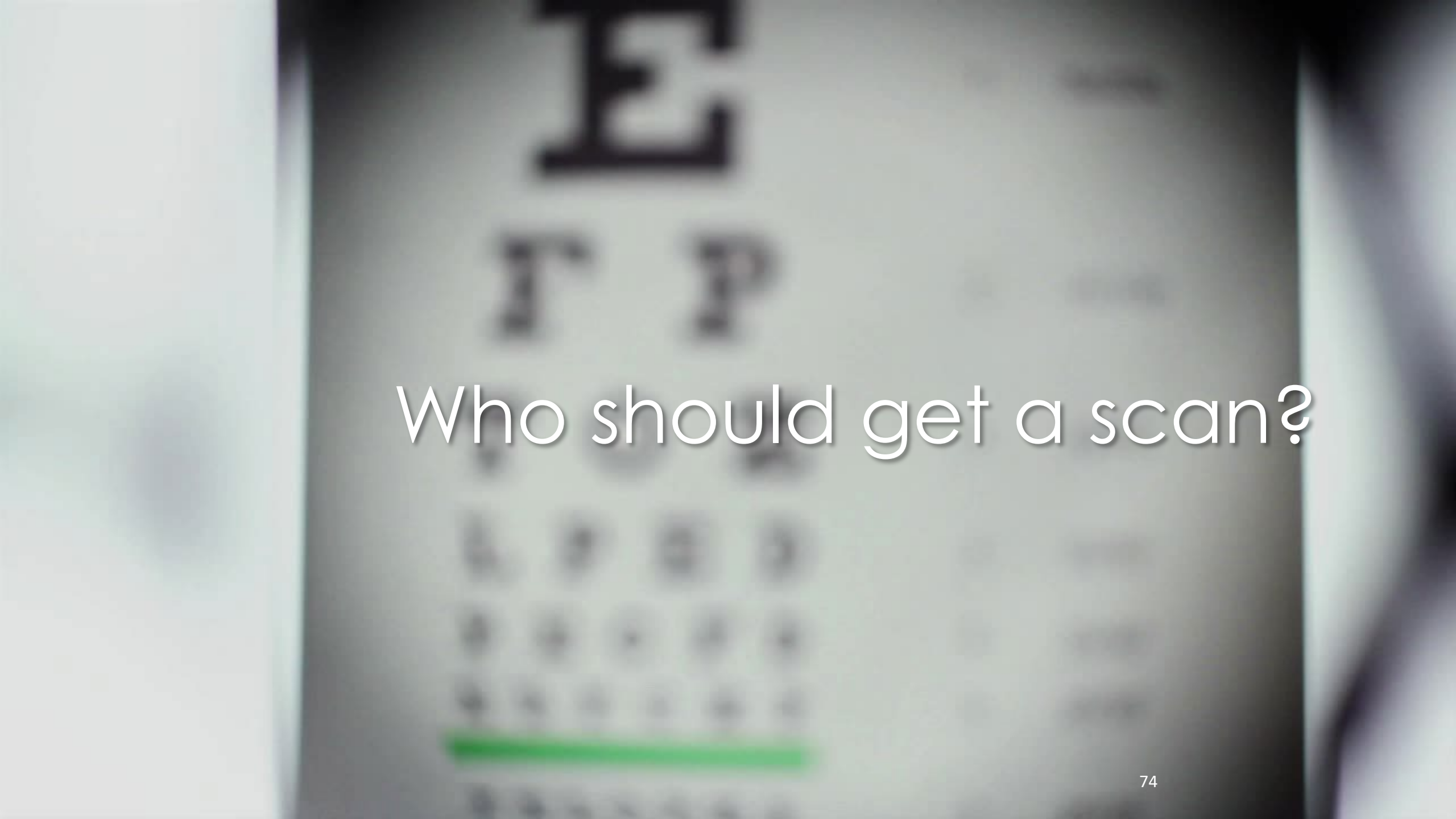
Bradycardia, vomiting, sundowning eyes (sclera persistently visible above iris), tense fontanelle, seizure, lethargy or extreme irritability


17.24mm









Who should get a scan?

What is the role of head circumference screening?

N=75,412 in an integrated HCN
BESS: 233
Hydrocephalus: 24
CSDH: 15
Cyst/tumor: 17
Assoc. Conditions: 29

BESS 34:10,000

BESS:everything else is 4:1

HC measurements are not sensitive

Large relative (≥ 4 major %ile)

and absolute (>95-97 %ile) increases in HC

are ≥90% specific

Most patients with an intracranial process are not detected by screening using HC

Daymont *et al. BMC Pediatrics* 2012, **12**:9
http://www.biomedcentral.com/1471-2431/12/9

BMC Pediatrics

RESEARCH ARTICLE Open Access

The test characteristics of head circumference measurements for pathology associated with head enlargement: a retrospective cohort study

Carrie Daymont[1,2,3,4*], Moira Zabel[3,4], Chris Feudtner[3,5,6] and David M Rubin[3,5,6]

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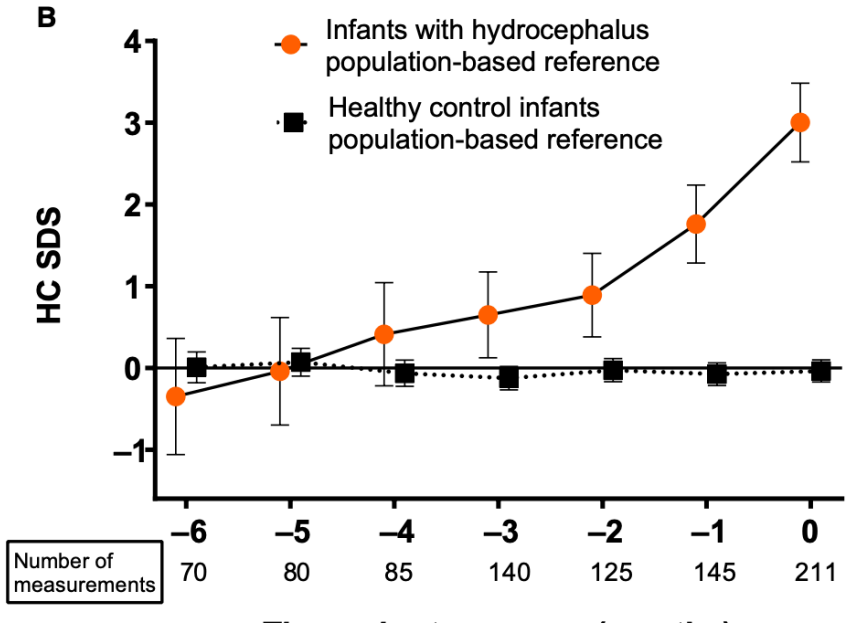
What is the trajectory of head circumference in acquired hydrocephalus?



Standard deviation score (SDS)	WHO HC standard		Population-based HC reference	
	Specificity (%) (95% CI)	Sensitivity (%) (95% CI)	Specificity (%) (95% CI)	Sensitivity (%) (95% CI)
1.5	46 (44.8-46.4)	85 (73.8-93.0)	86 (85.1-86.3)	70 (57.4-81.5)
2.0	69 (68.3-69.7)	75 (62.7-85.5)	94 (93.8-94.5)	61 (47.3-72.9)
2.5	85 (84.3-85.4)	72 (59.2-82.9)	98 (97.5-98.0)	51 (37.7-63.9)
3.0	94 (93.7-94.5)	52 (39.3-65.4)	99 (99.2-99.5)	41 (28.6-54.3)

Abbreviations: CI, confidence interval.

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REGULAR ARTICLE

ACTA PÆDIATRICA WILEY

Screening of hydrocephalus in infants using either WHO or population-based head circumference reference charts

Marjo Karvonen[1,2] | Antti Saari[1,2] | Marja-Leena Lamidi[1] | Tuomas Selander[3] | Tuija Löppönen[1] | Tuula Lönnqvist[4] | Leo Dunkel[5] | Ulla Sankilampi[1,2]

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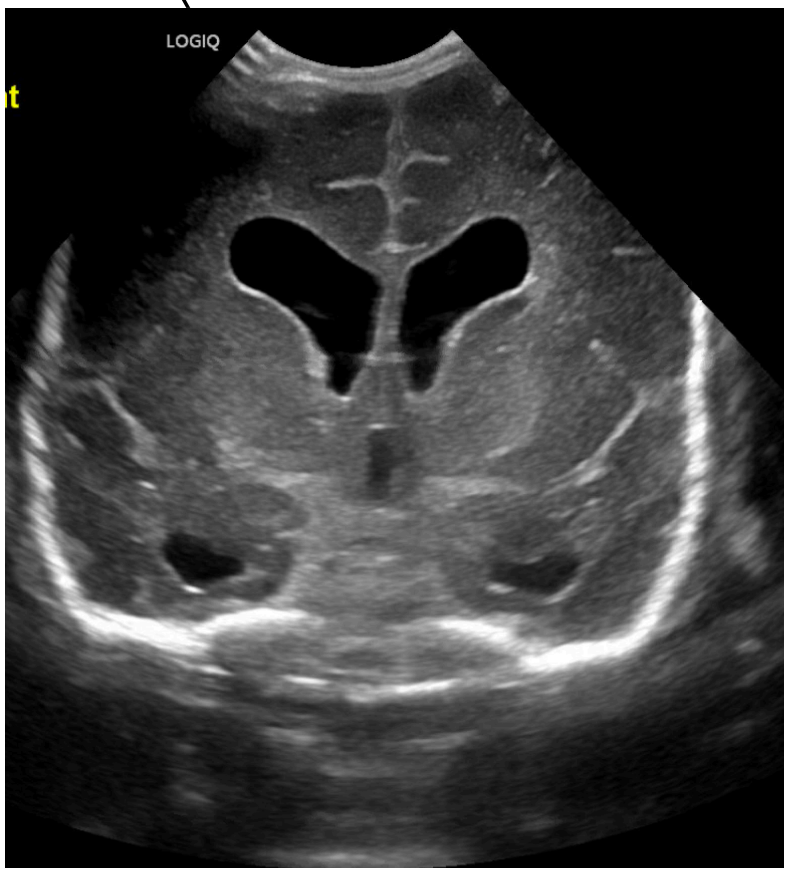
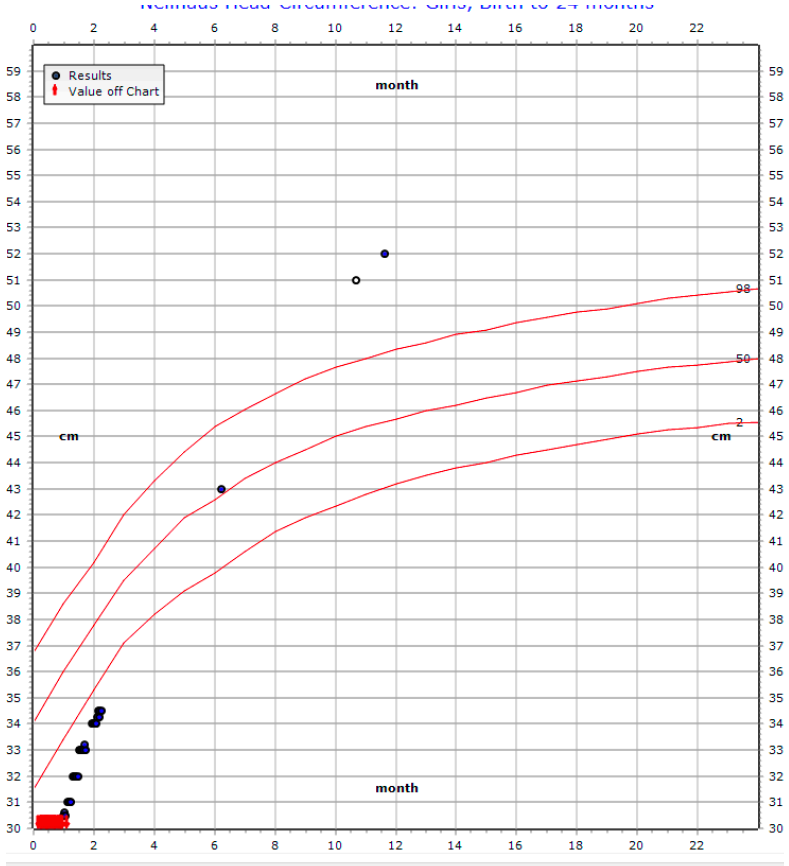
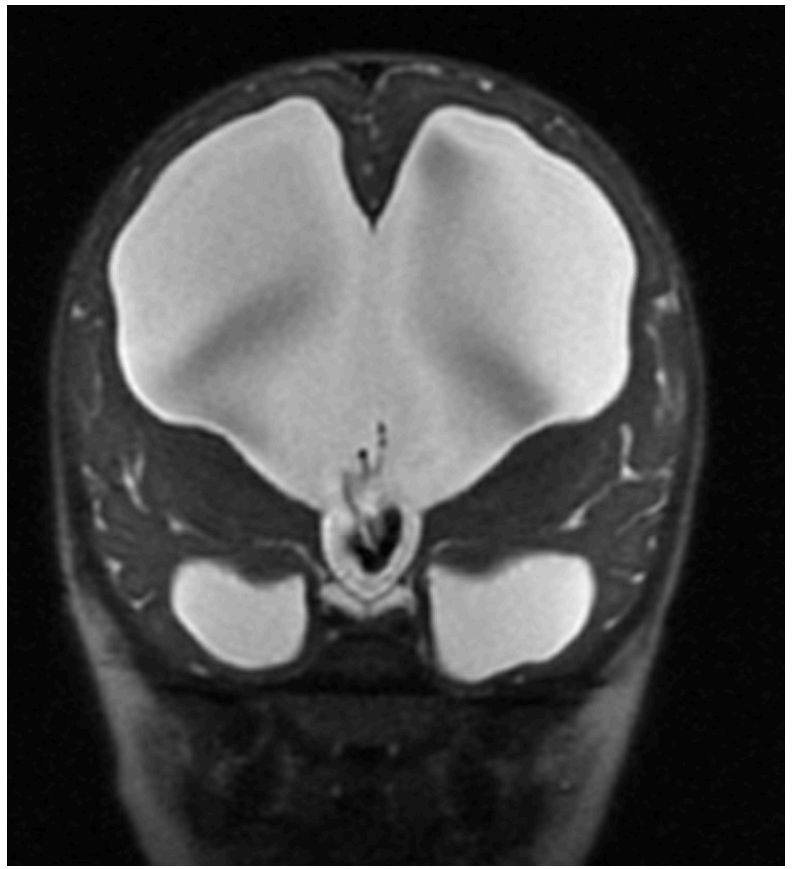
When is a “normal” head circumference not normal?

Ex-30 weeker with bl Gr IV IVH and progressive macrocephaly







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“Does this child have hydrocephalus?”

Head US is the image of choice in infants
Beware early macrocephaly before 4 months
Measure the parents (esp. dad) and siblings
Finding BESS is worthwhile (SDH vs. NAT)
MRI is used for surgical planning or unusual cases (can do limited MRI without sedation)
Watchful of clinical signs and symptoms (danger signs)
Imaging:
Absolute HC ≥ 2 SD or ≥ 95 %ile
Relative HC ≥ 2 SD
Disproportionate HC ≥ 2 SD

Hydrocephalus Treatment

Hydrocephalus is a surgical disease

Two options:
shunt placement
endoscopic third ventriculostomy

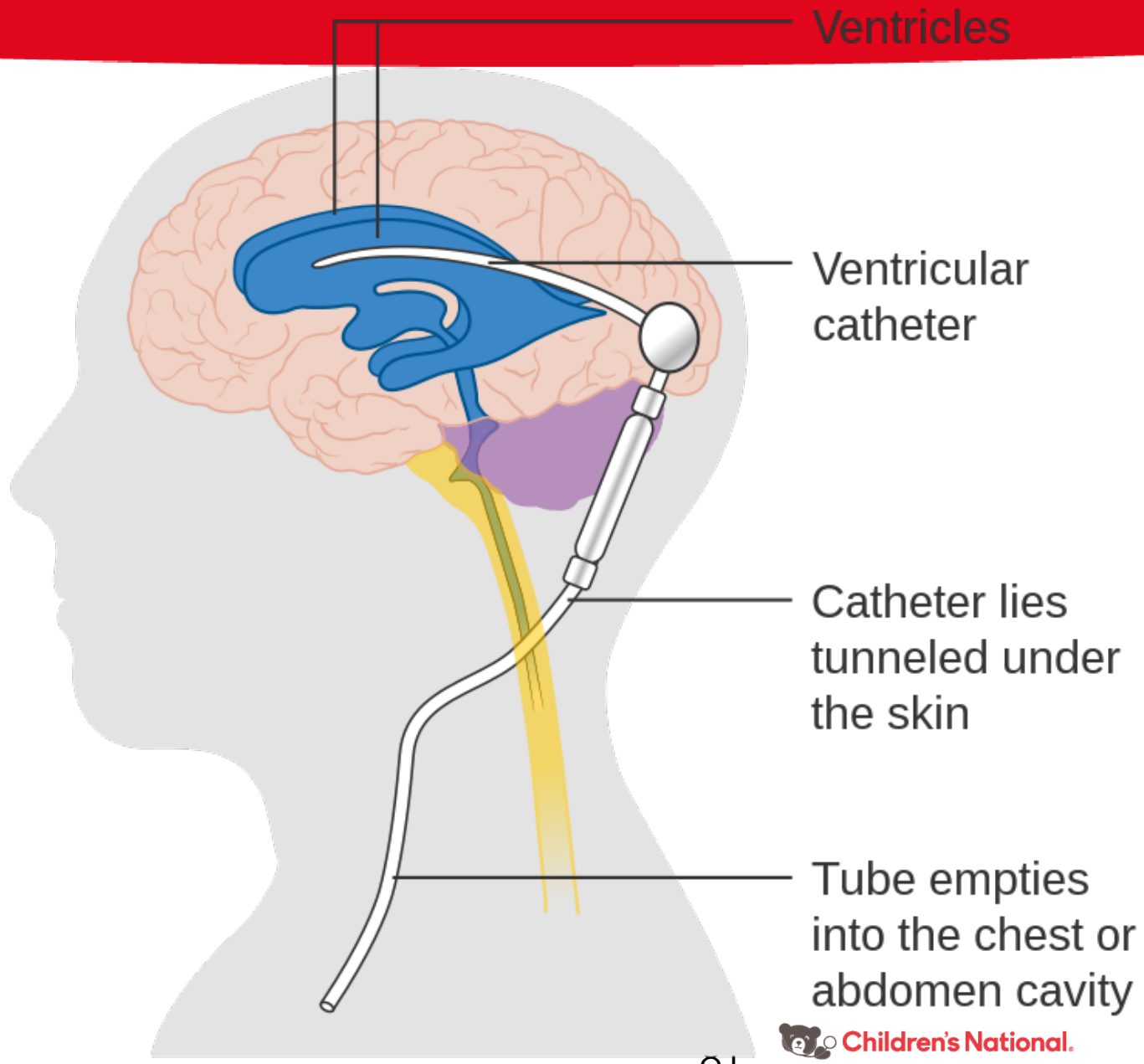
One is not better than the other (ESTHI trial, ongoing).

Almost any child with hydrocephalus can receive a shunt; some children have unfavorable anatomy or pathology for ETV.

Ventricular Shunts

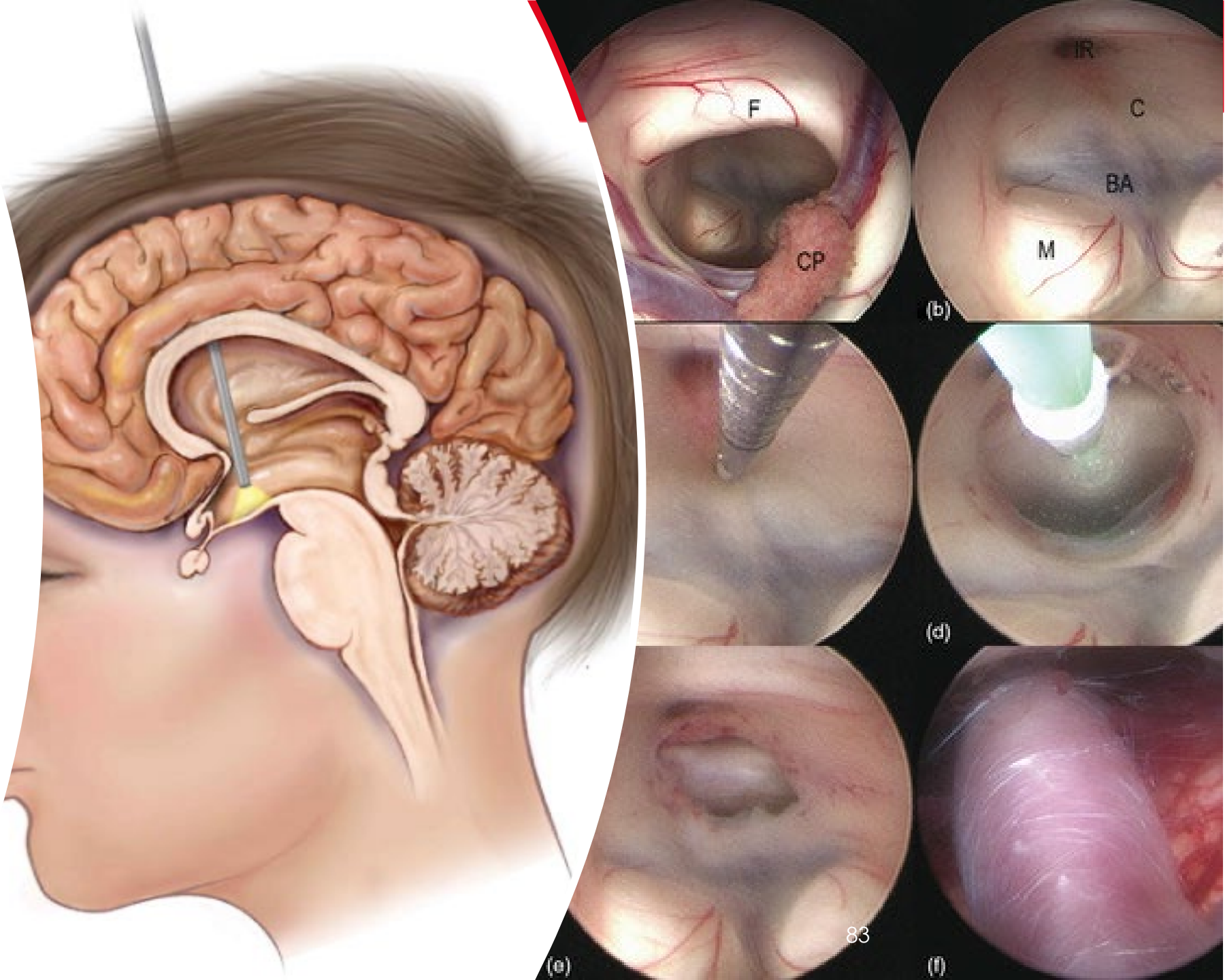
Shunts are a CSF diversion system with at least one proximal catheter, reservoir +/- valve, and distal catheter. The proximal catheter can have an anterior or posterior entry site

Valves can be **fixed** or **programmable**
Programmable valves that are not MR-resistant require reprogramming after MRI (don't get an MRI in outpatient radiology)
The distal catheter commonly ends in the peritoneum, but right atrial, pleural, other termini are also used.
50% of shunts fail within two years of placement



There is no shunt like no shunt

Endoscopic Third Ventriculostomy



SCAN ME

CLINICAL REPORT Guidance for the Clinician in Rendering Pediatric Care

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Incidental Findings on Brain and Spine Imaging in Children

Cormac O. Maher, MD, FAAP, Joseph H. Piatt Jr, MD, FAAP, SECTION ON NEUROLOGIC SURGERY

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Congenital Brain and Spinal Cord Malformations and Their Associated Cutaneous Markers

Mark Dias, MD, FAANS, FAAP, Michael Partington, MD, FAANS, FAAP, the SECTION ON NEUROLOGIC SURGERY

Thank you!

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Thank You!