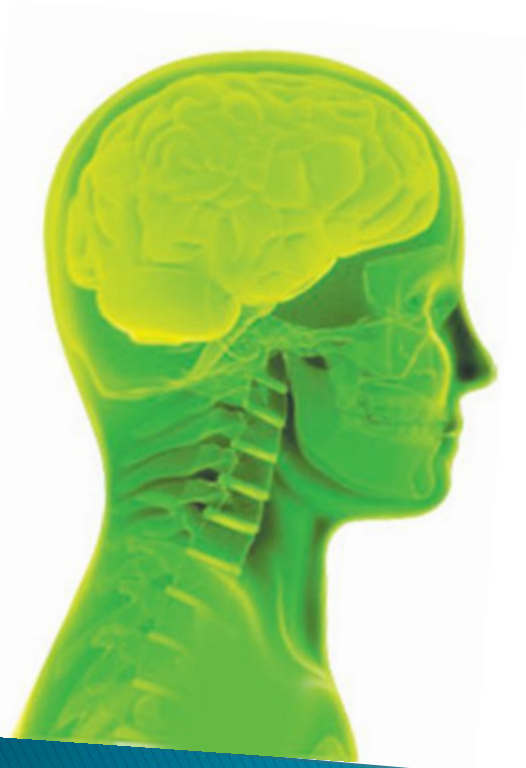
Weill Cornell Medical College

NewYork-Presbyterian
Phyllis and David Komansky
Center for Children's Health

Pediatric
Brain and Spine Center



The Misshapen Head in Infancy

Samuel Rhee, MD

Recent Director, Craniofacial Surgery Program

Department of Surgery

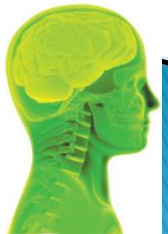
Weill Cornell Medical College

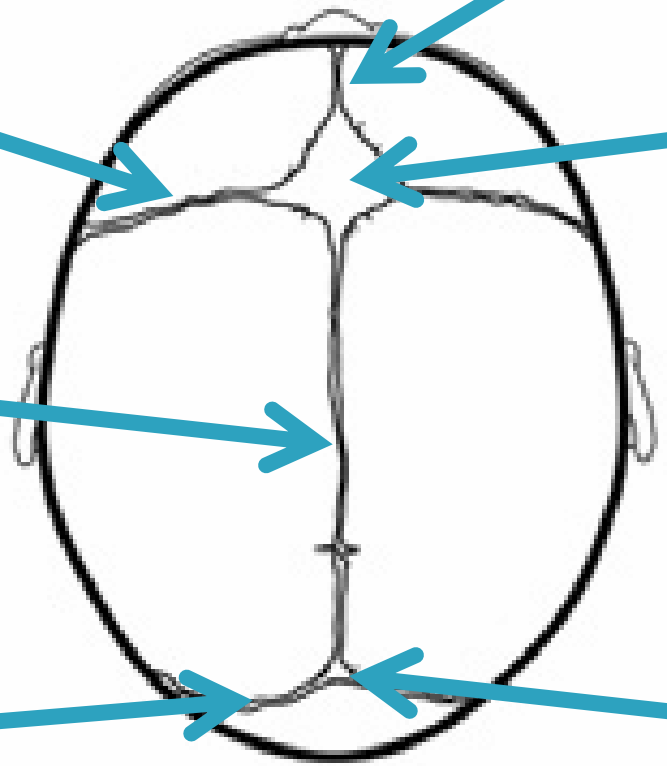
New York-Presbyterian Medical Center

Evaluation of head shape

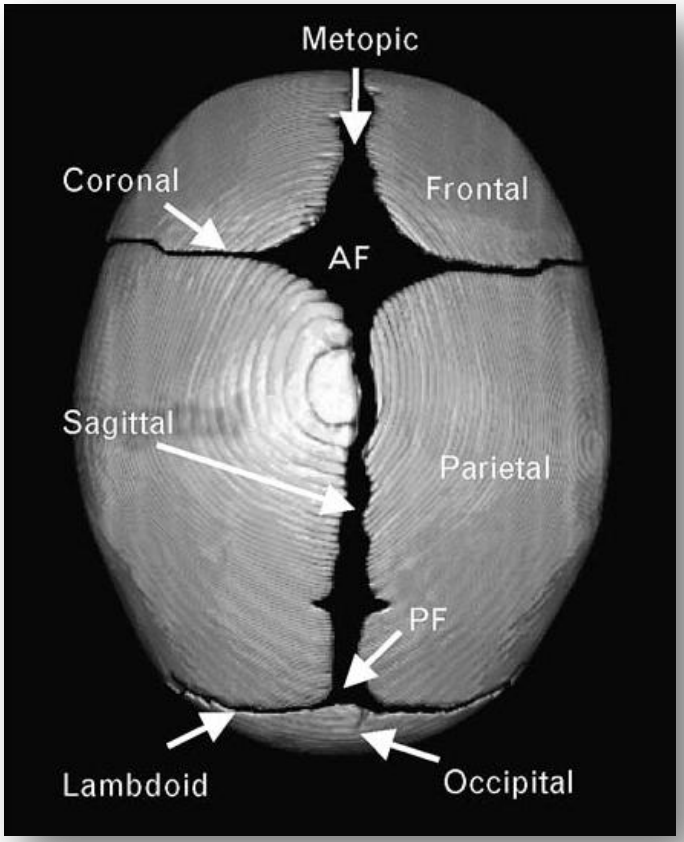
The "Outside" Perspective

- Initial assessment of patient
- Craniosynostosis vs. Positional head deformity
- Surgical correction of craniosynostosis
  - Minimally Invasive or Endoscopic surgery

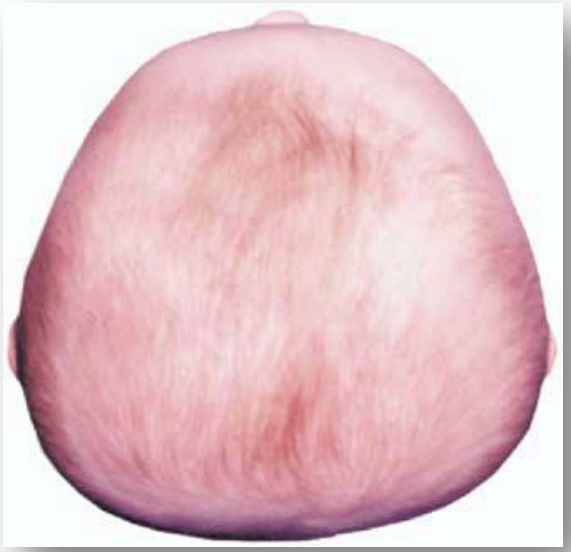



Metopic
Coronal
Anterior Fontanelle
Sagittal
Lambdoid
Posterior Fontanelle

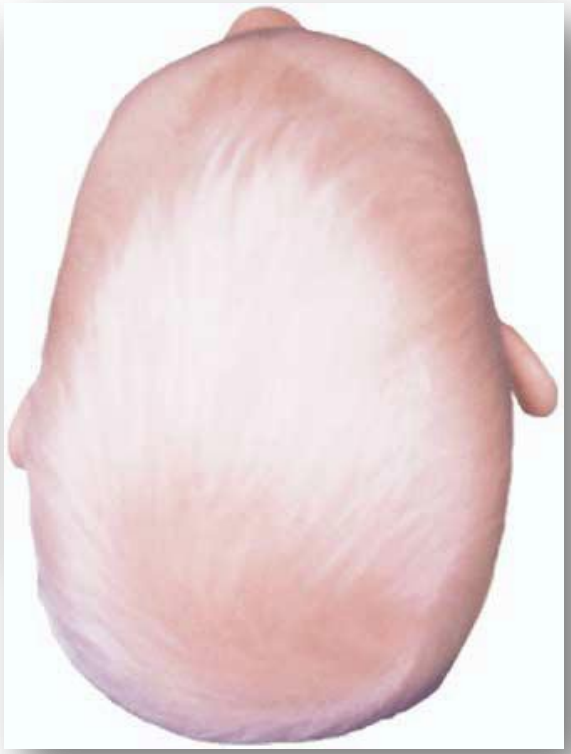
Timing of cranial suture closure



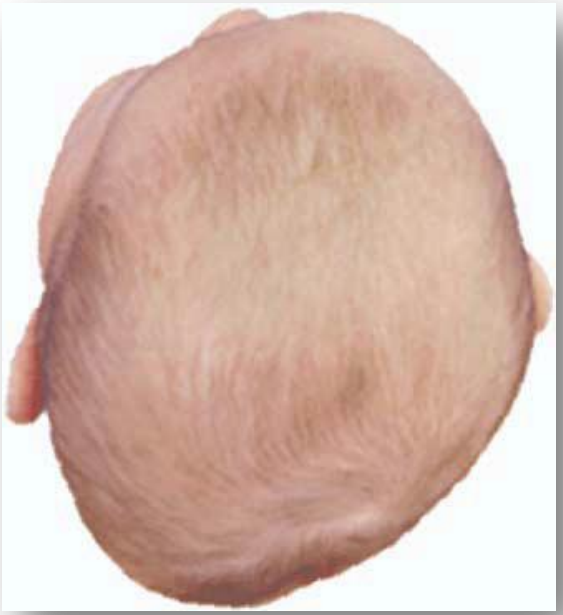
Examination of the infant skull



Brachycephaly



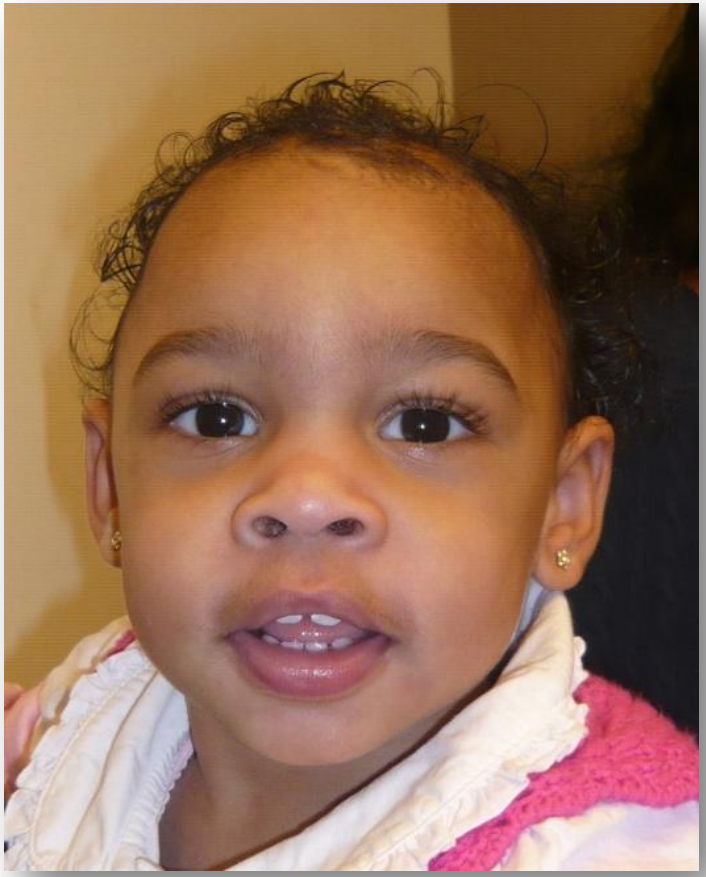
Dolichocephaly

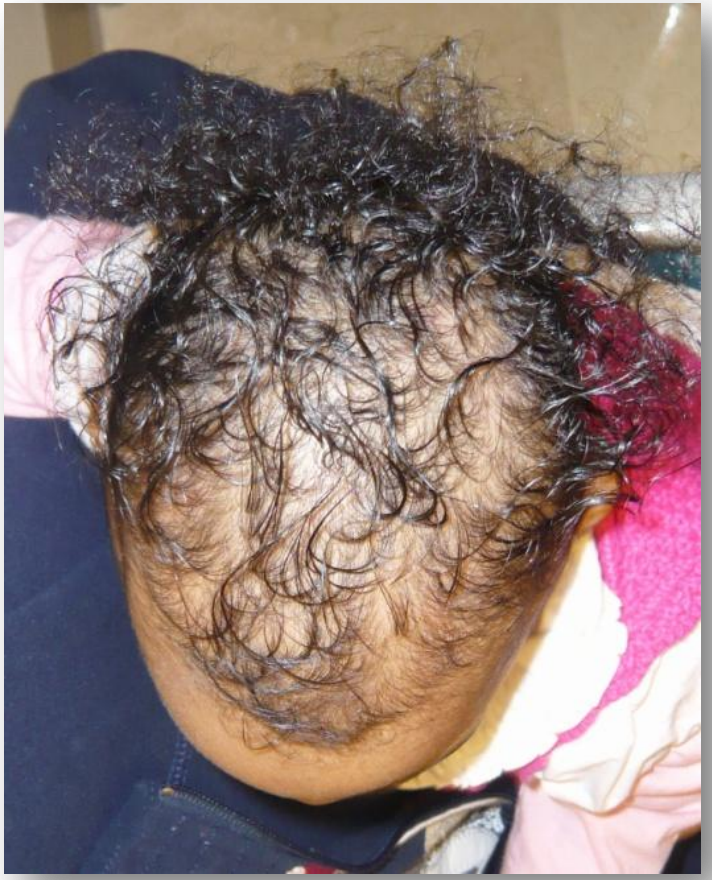


Plagiocephaly

Hummel P, Fortado D. Impacting infant head shapes. Adv Neonatal Care. 2005 Dec;5(6):329-40.

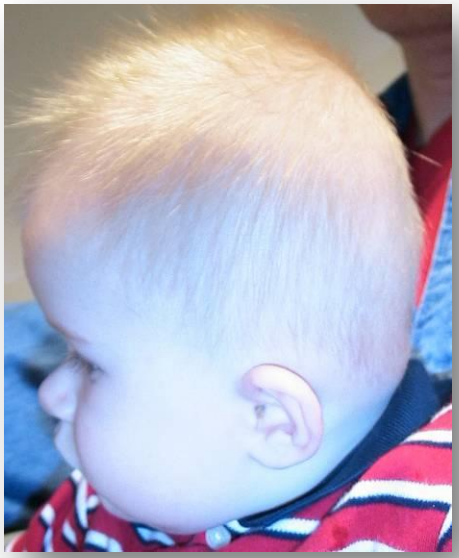
Conditions affecting sutures

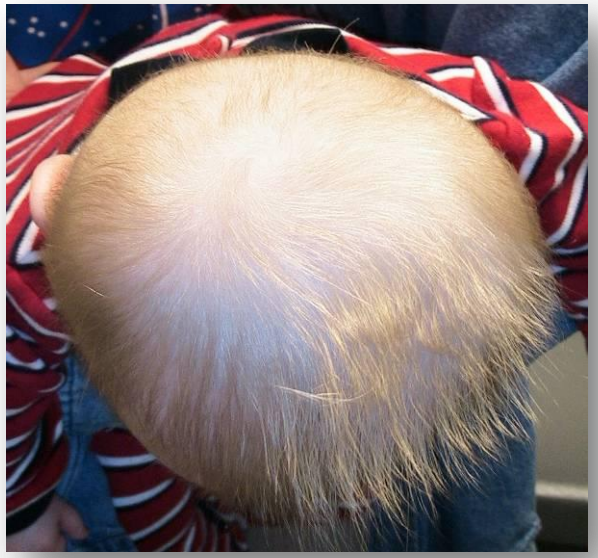


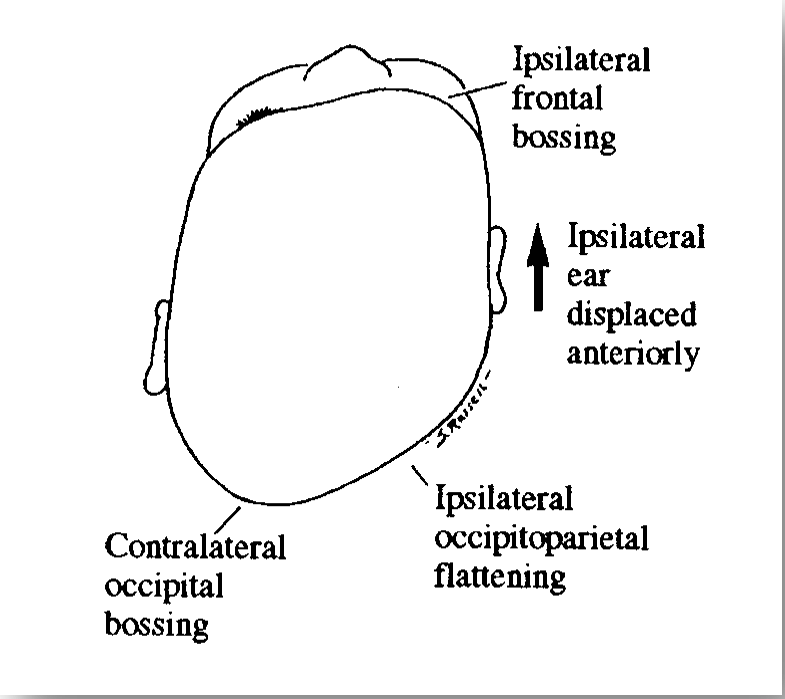


Craniosynostosis vs. PHD

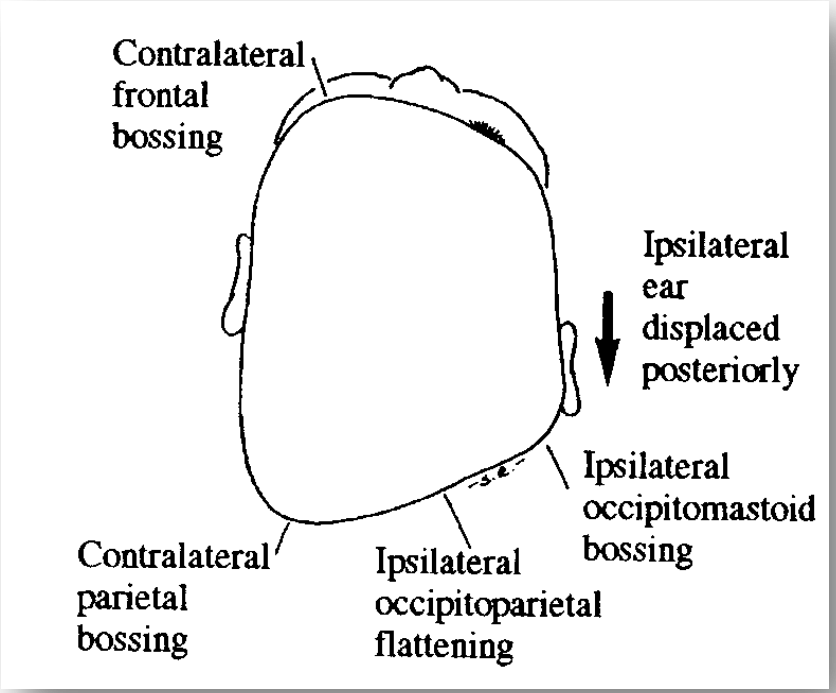
- Clinical diagnosis
  - Can be difficult even for experienced clinicians
- Imaging studies
  - Plain radiographs unless neurologic concerns







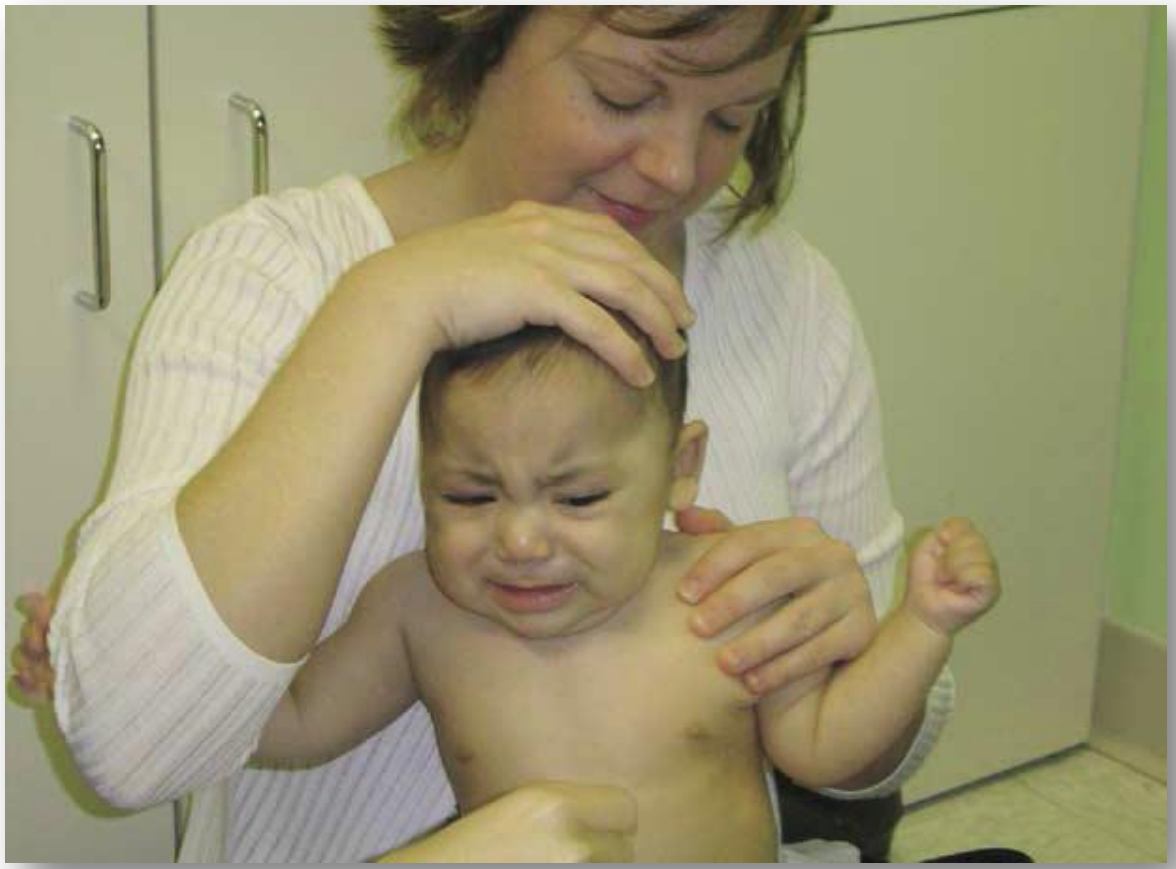
Positional head deformity



True lambdoid synostosis

Anatomic Feature	Synostotic	Deformational
Ipsilateral superior orbital rim	Up	Down
Ipsilateral ear	Anterior and high	Posterior and low
Nasal root	Ipsilateral	Midline
Ipsilateral cheek	Forward	Backward

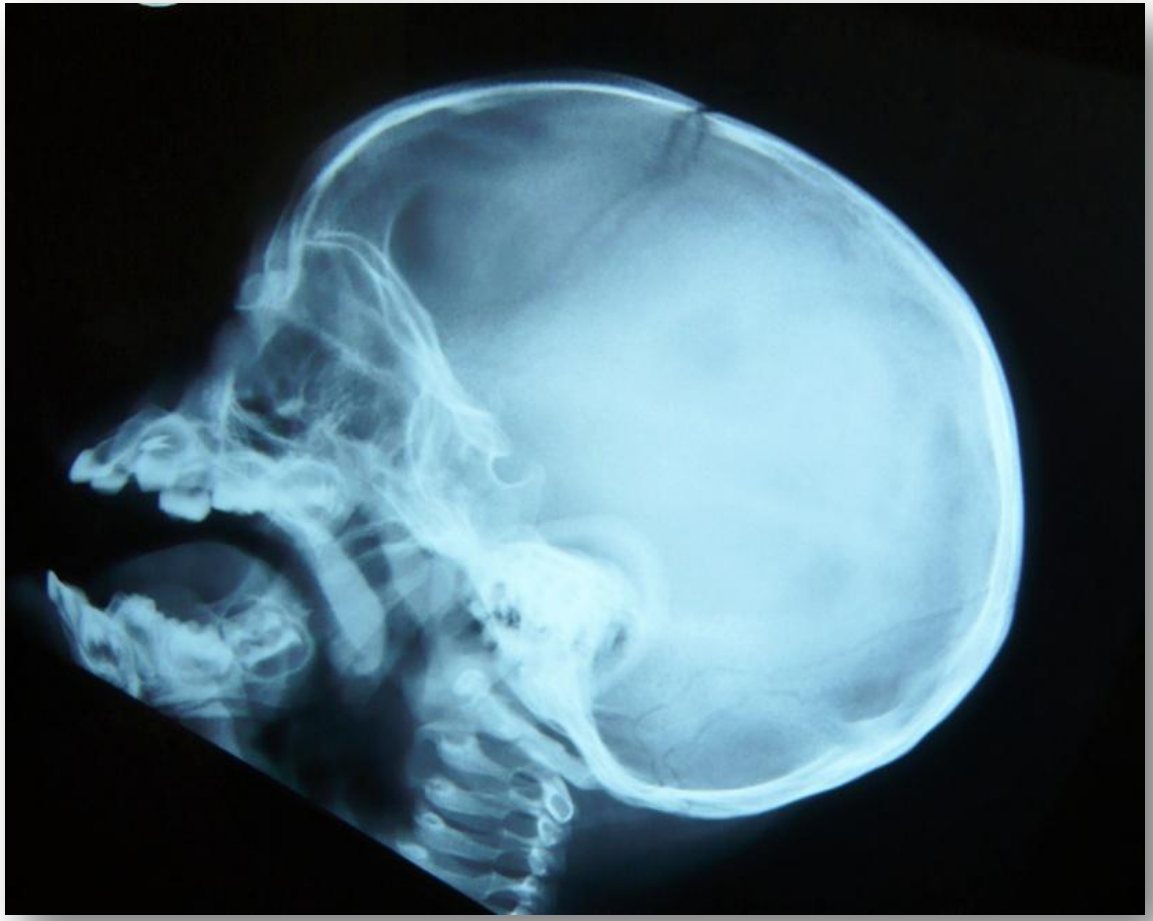
Treatment of torticollis



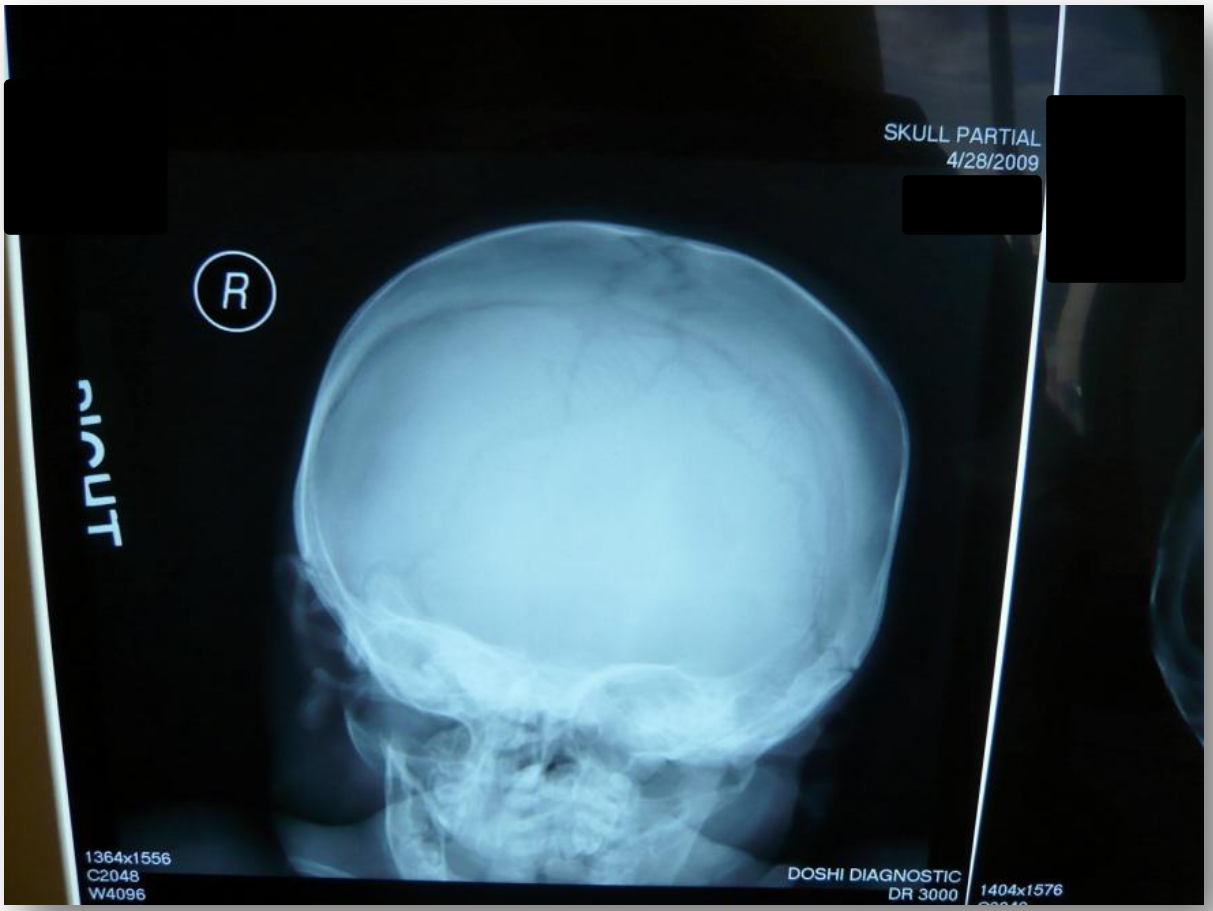
Hummel P, Fortado D. Impacting infant head shapes. Adv Neonatal Care. 2005 Dec;5(6):329–40.

Treatment of PHD

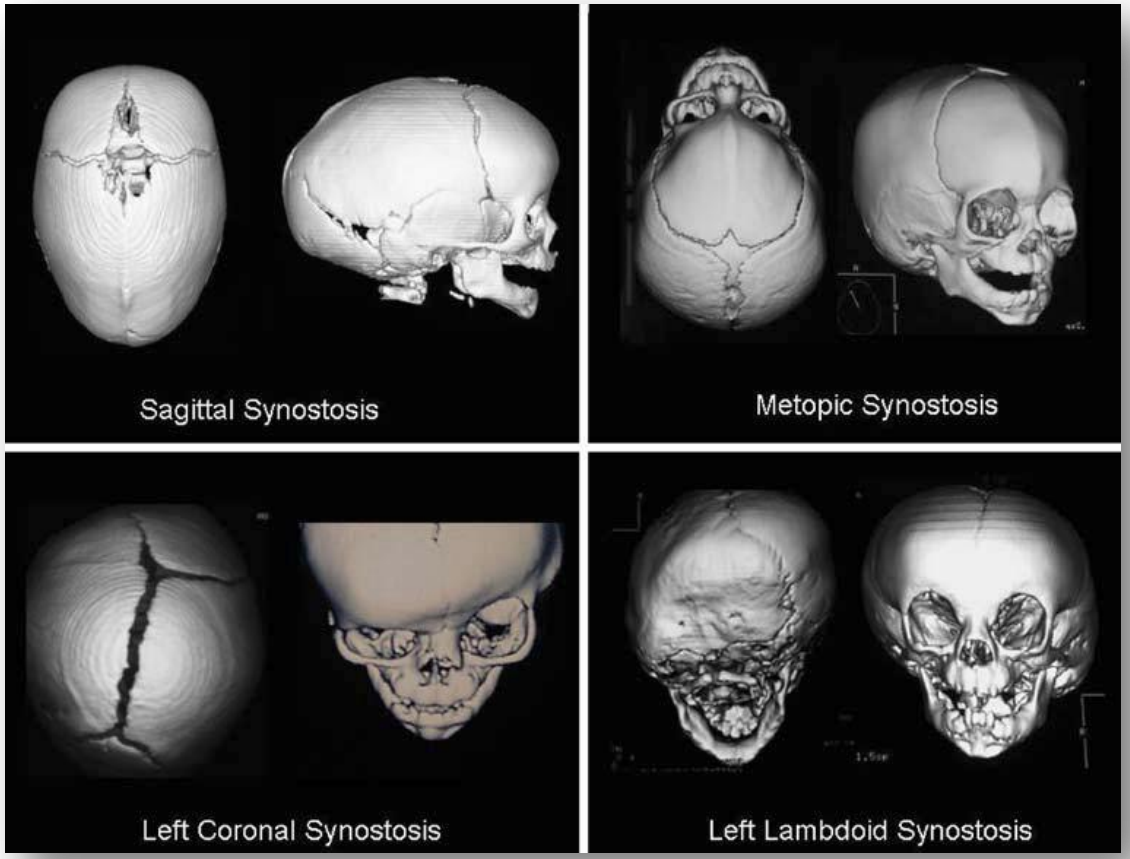
Assessment with imaging



Assessment with imaging



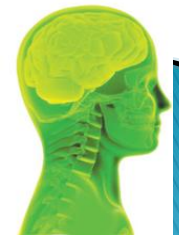
CT scans



Neurodevelopment of children with single suture craniosynostosis: a review. Kapp-Simon KA, Speltz ML, Cunningham ML, Patel PK, Tomita T. Childs Nerv Syst. 2007 Mar;23(3):269-81. Epub 2006 Dec 21

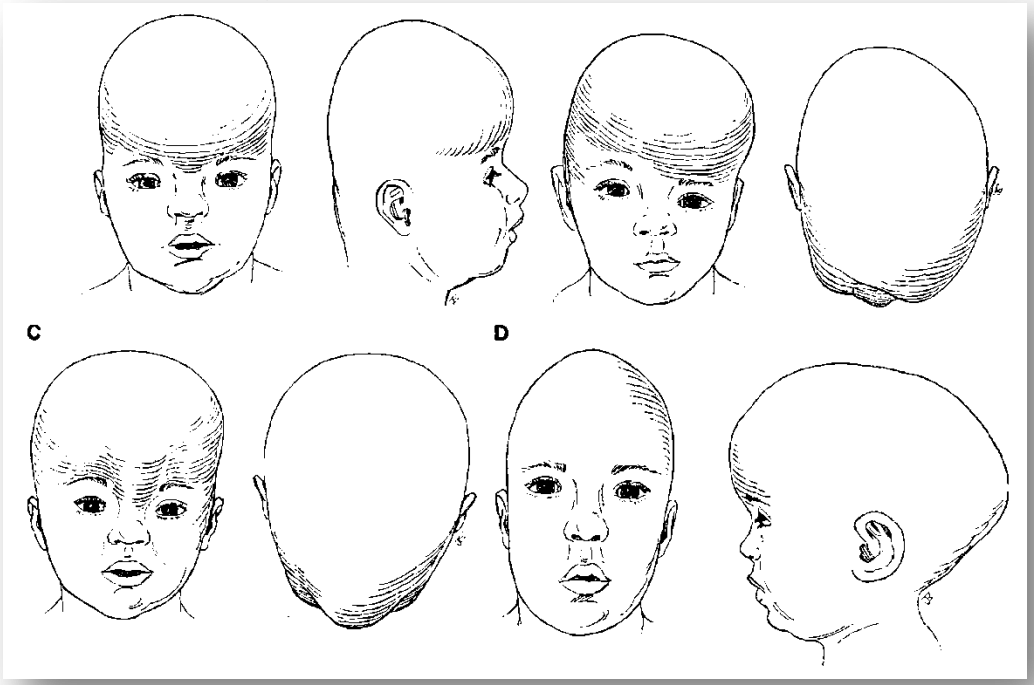
Radiation exposure

EXAM TYPE	RELEVANT ORGAN	DOSE (mSv)
Pediatric Head CT Unadjusted Settings (200 mAs, neonate)	Brain	60
Pediatric Head CT Adjusted Settings (100 mAs, neonate)	Brain	30
Pediatric Abdominal CT Unadjusted Settings (200 mAs, neonate)	Stomach	25
Pediatric Abdominal CT Adjusted Settings (50 mAs, neonate)	Stomach	6
Chest X-ray (PA/lateral)	Lung	0.01/0.15
Screening Mammogram	Breast	3

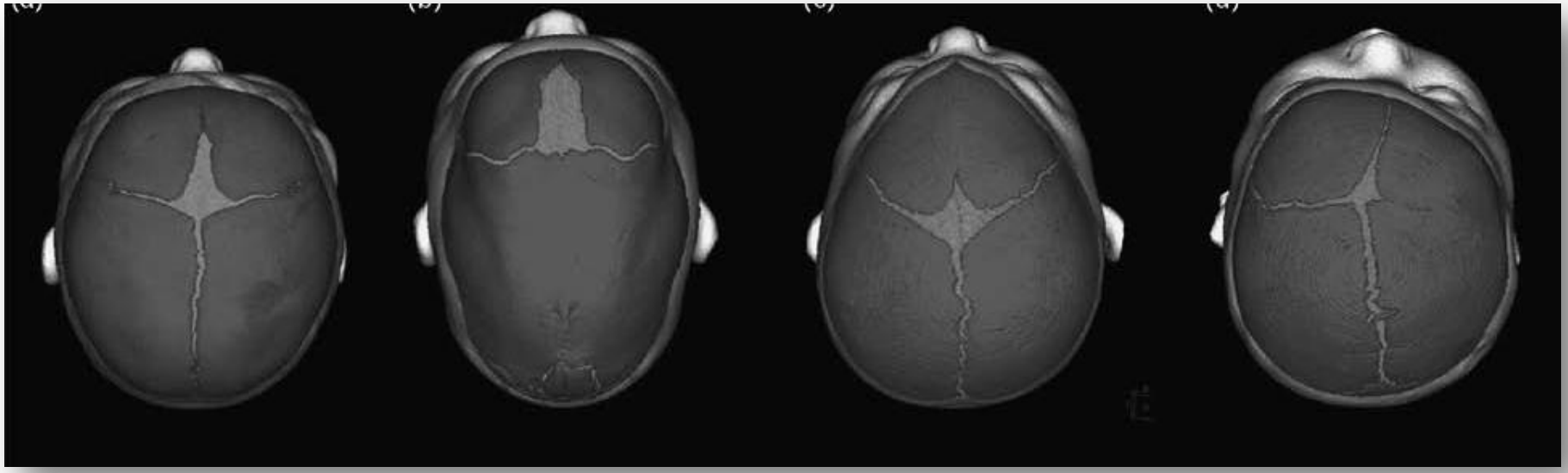


Craniosynostosis

- Premature fusion of cranial sutures
- Compensatory growth in other sutures (Virchow's Law)



Craniosynostosis



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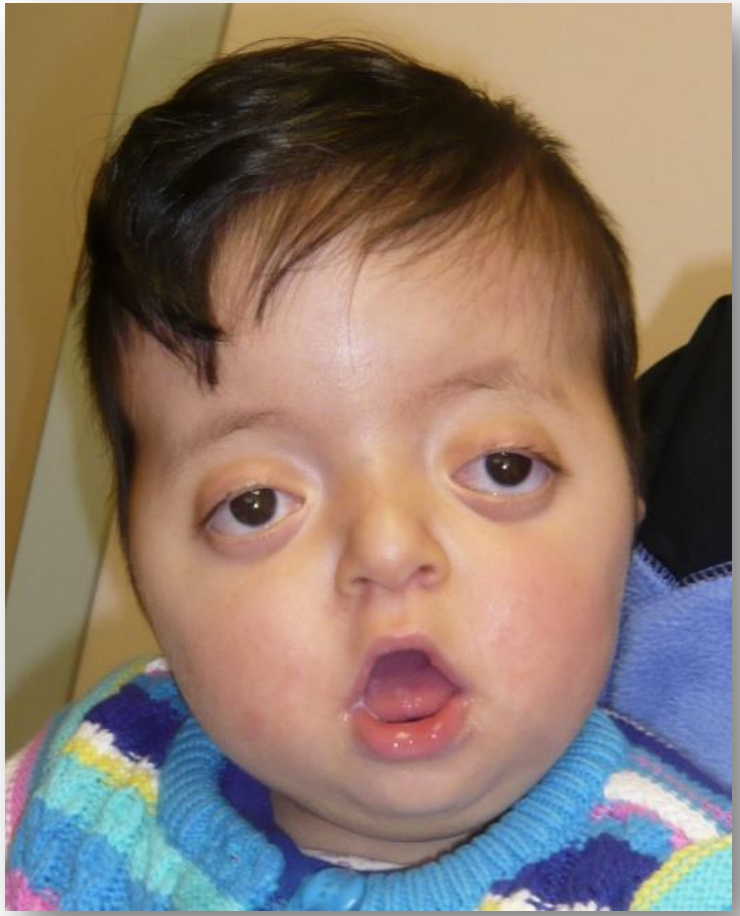
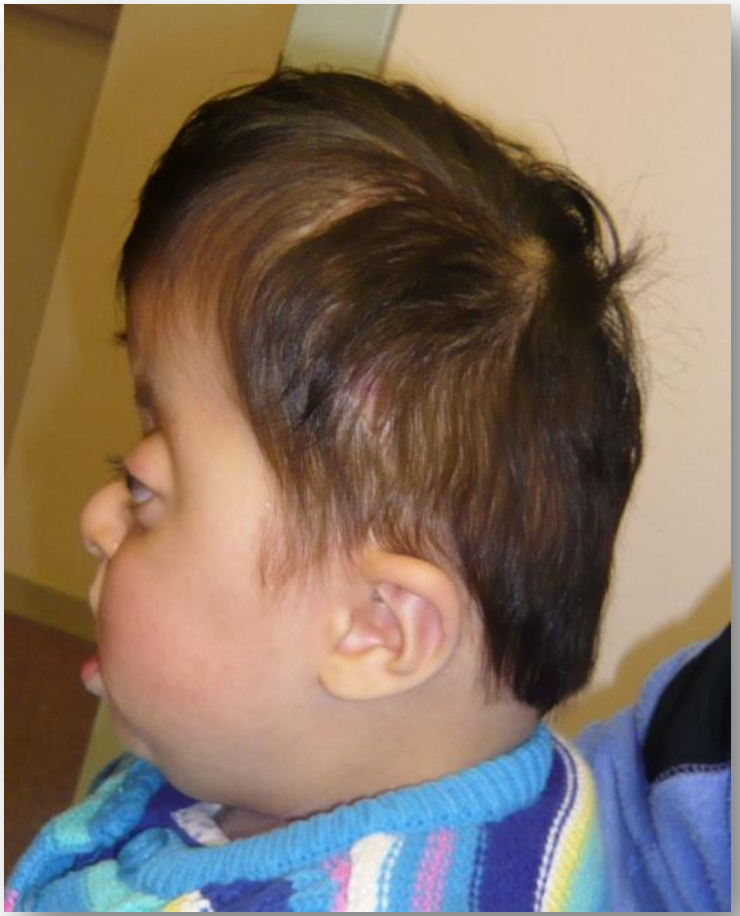
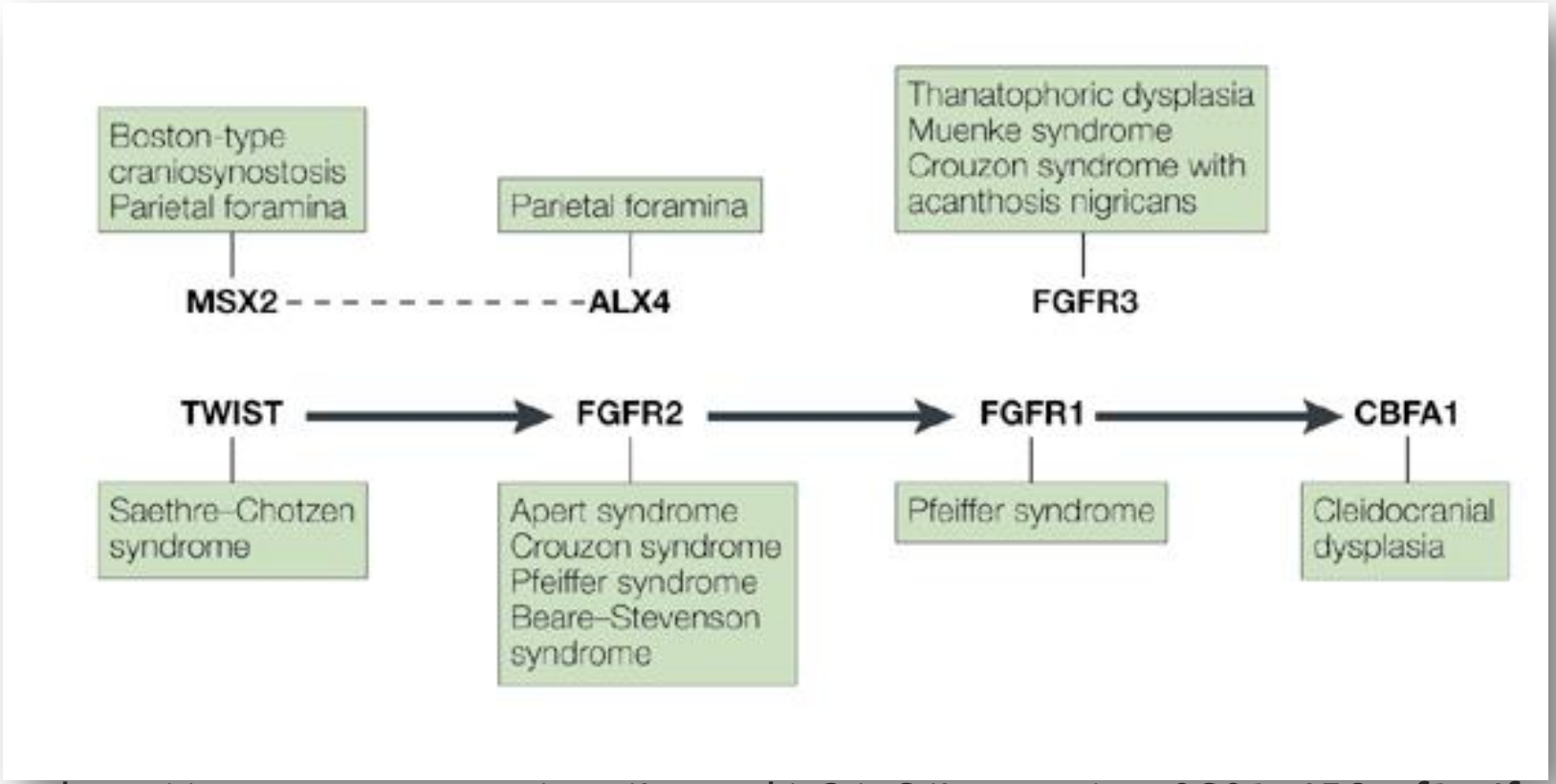
Syndromic craniosynostosis





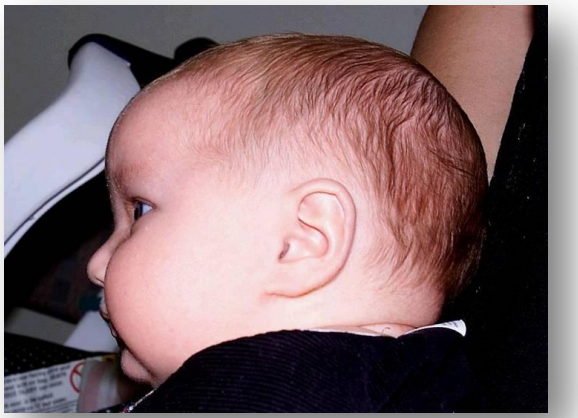
Genetic identification



http://www.nature.com/nrg/journal/v2/n6/images/nrg0601_458a_f1.gif

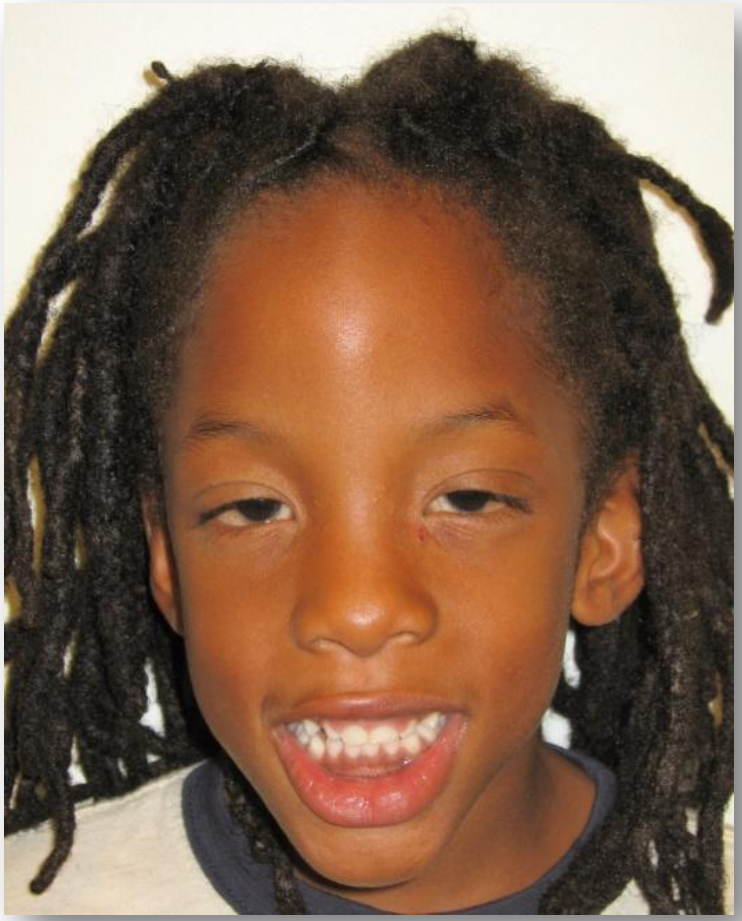
Craniosynostosis –Presentation

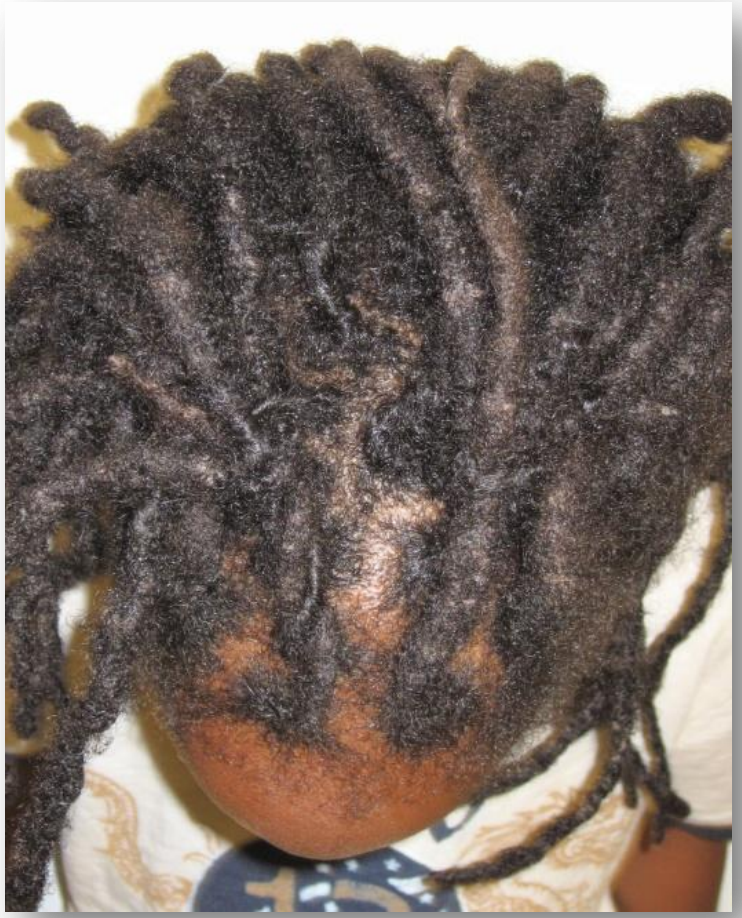
- Usually clinical identification
- Irregular head shape that worsens
- Ridging along sutures
- Falling off head circumference growth curve



- Later increased ICP signs
  - Irritability/vomiting
  - Papilledema

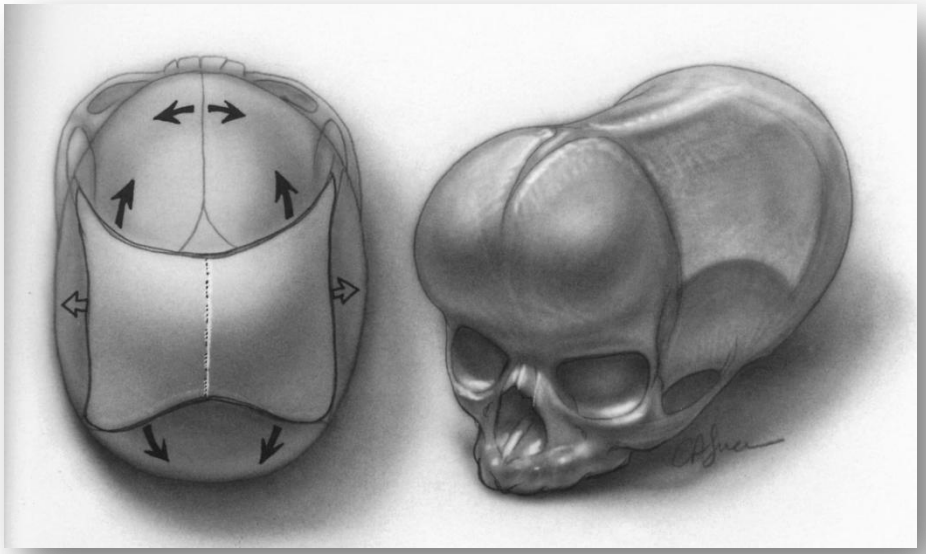
Untreated craniosynostosis



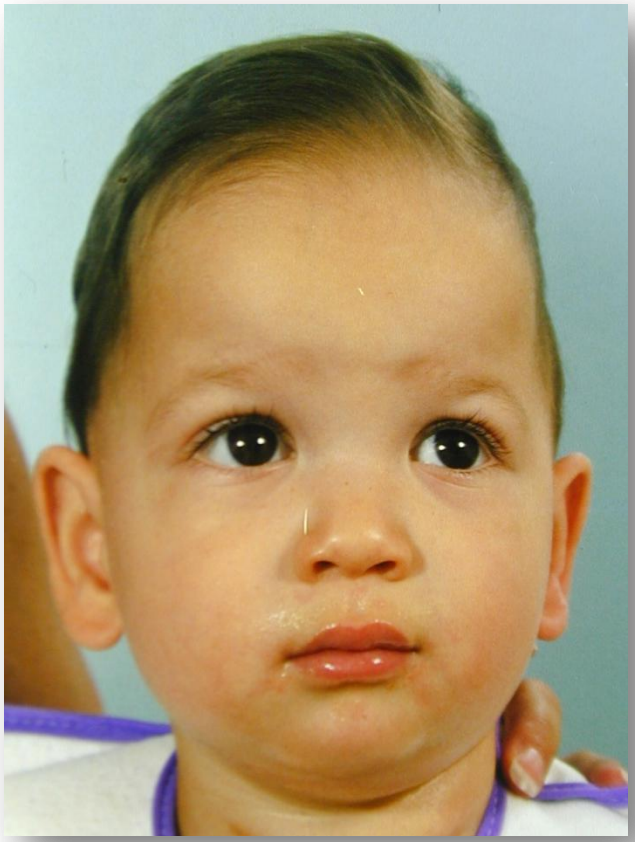


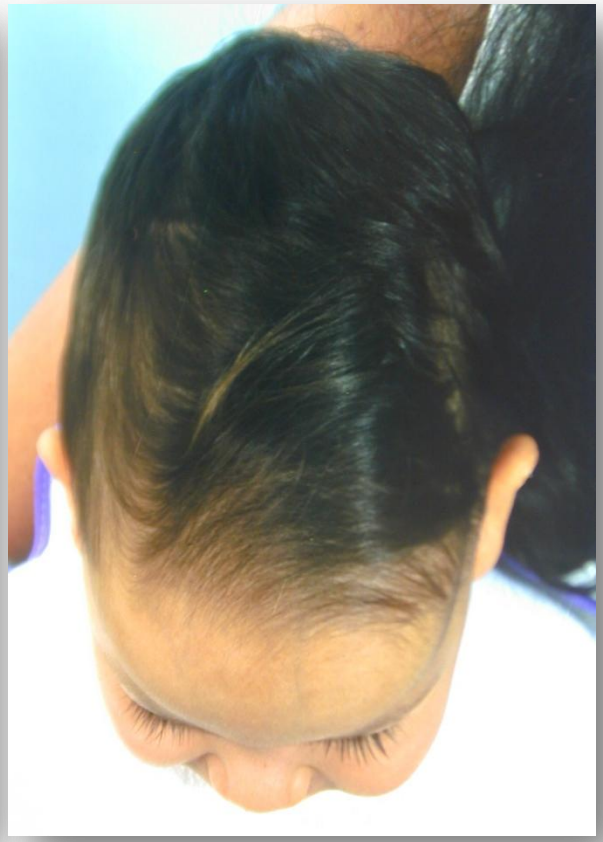
Sagittal Synostosis

- Most common type
- Male to Female 4 : 1
- ↑ AP length
- ↓ Width

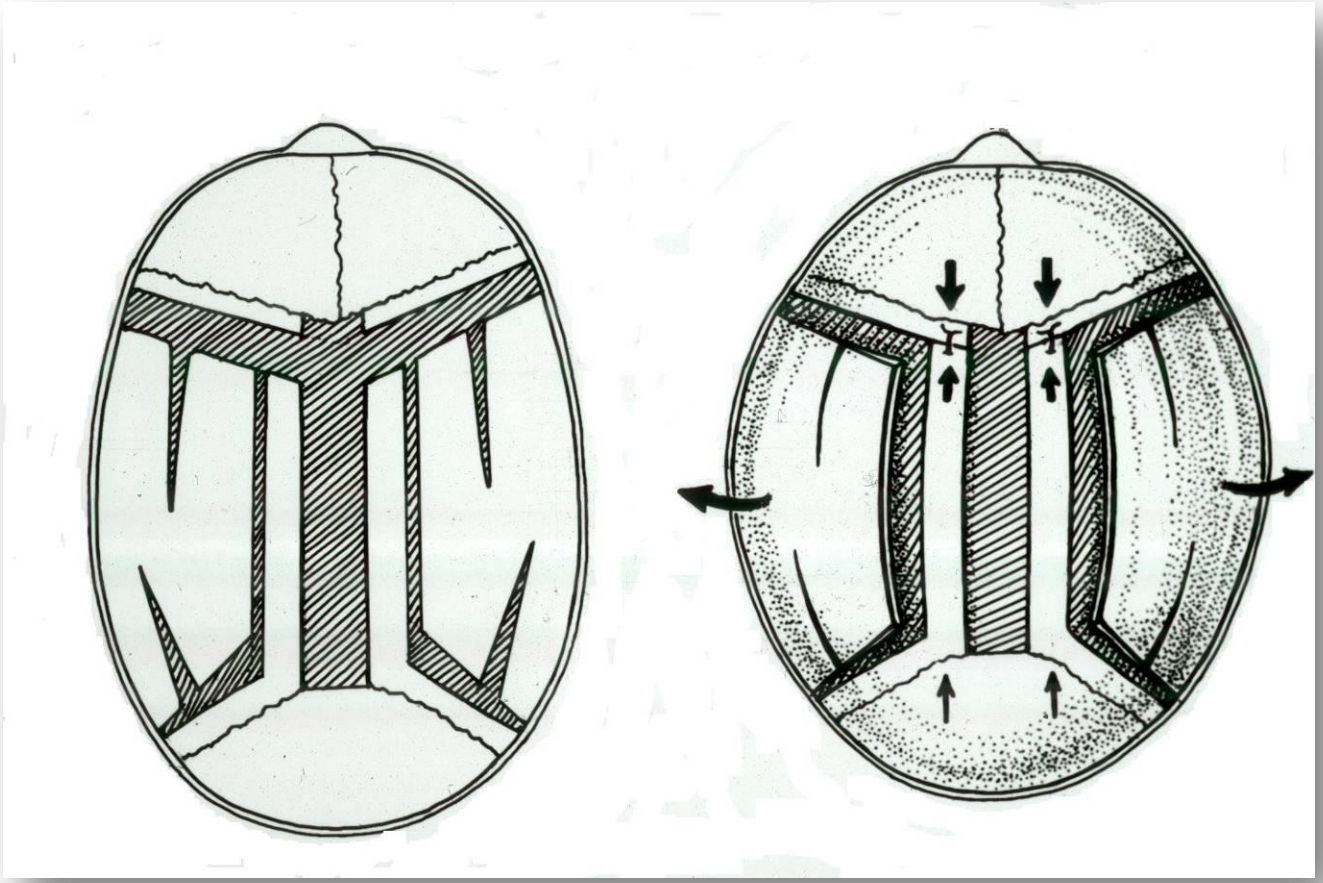


Craniosynostosis – surgery

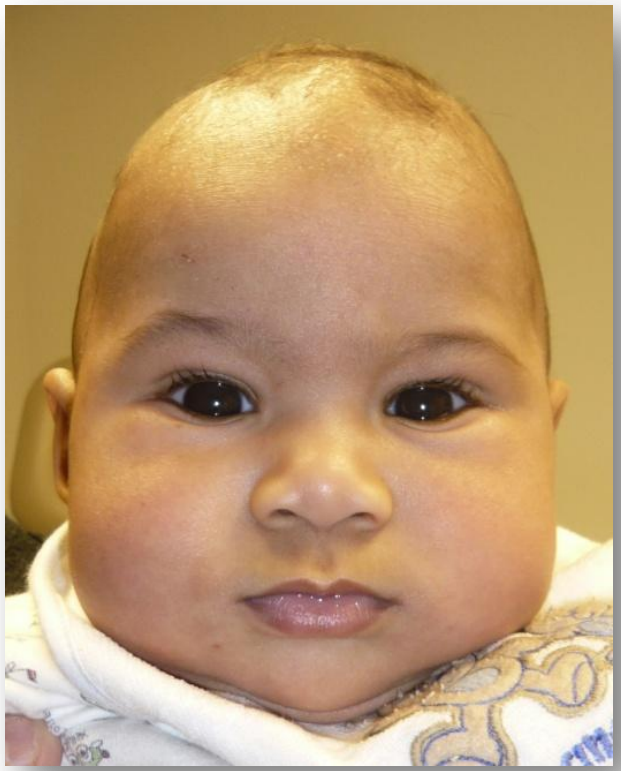


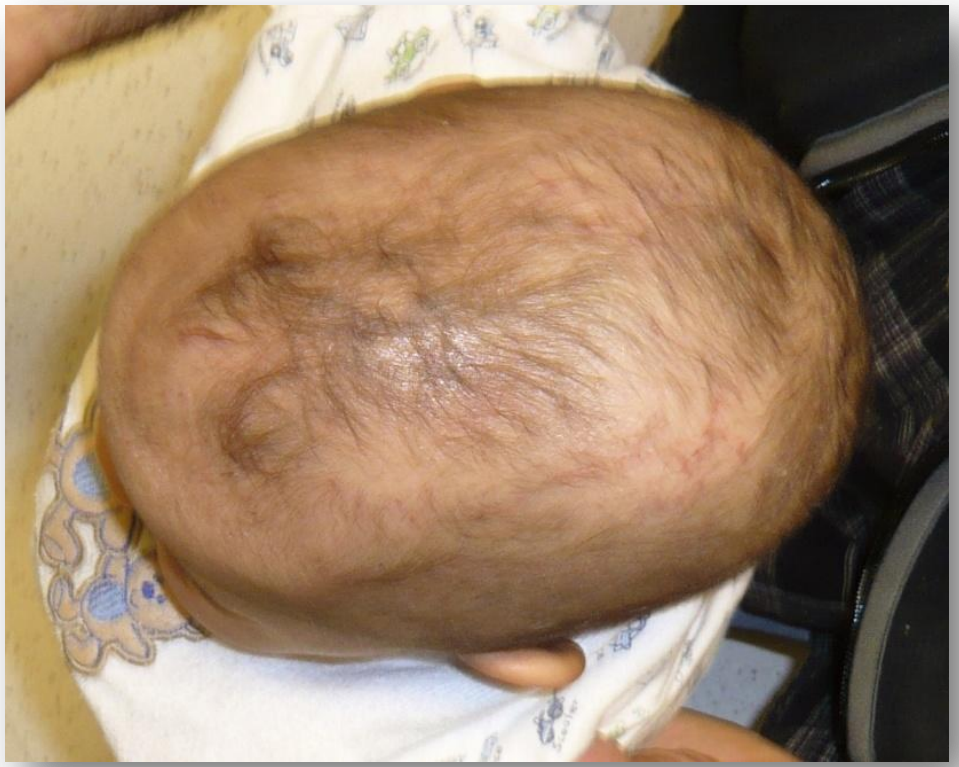


Craniosynostosis – surgery

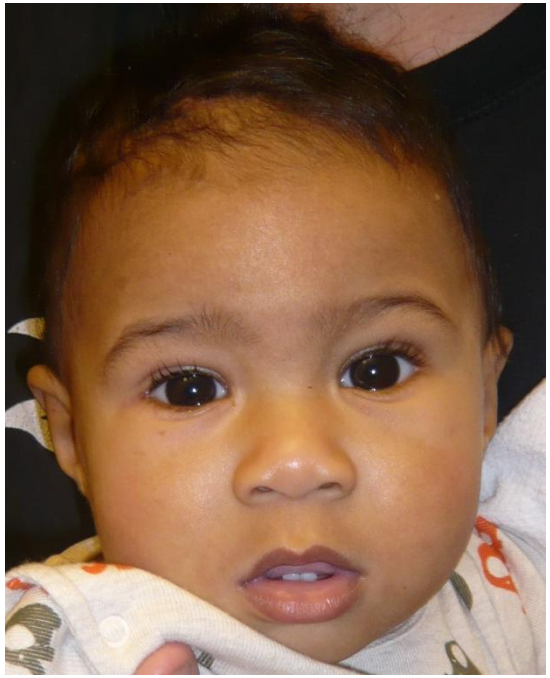


Sagittal synostosis

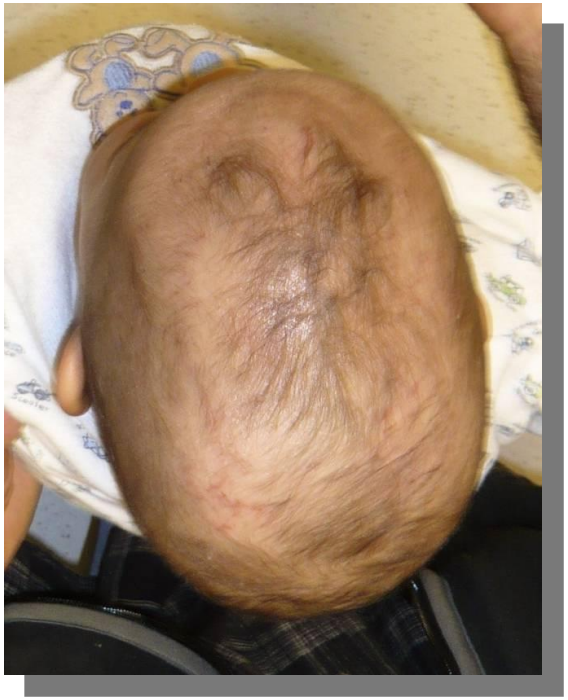


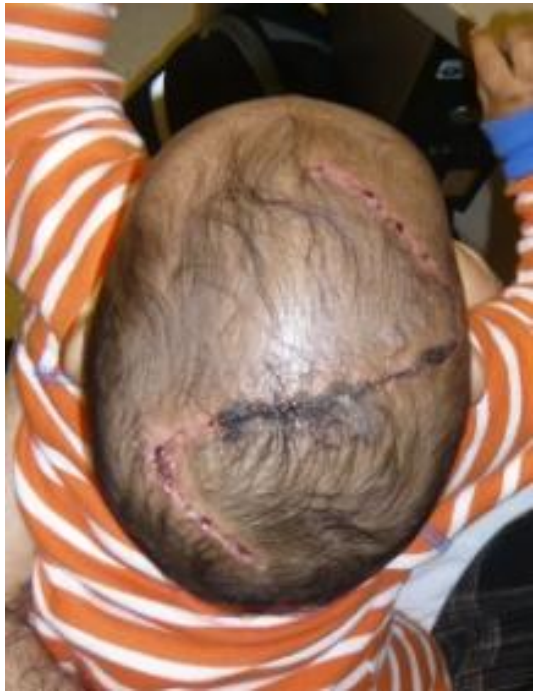


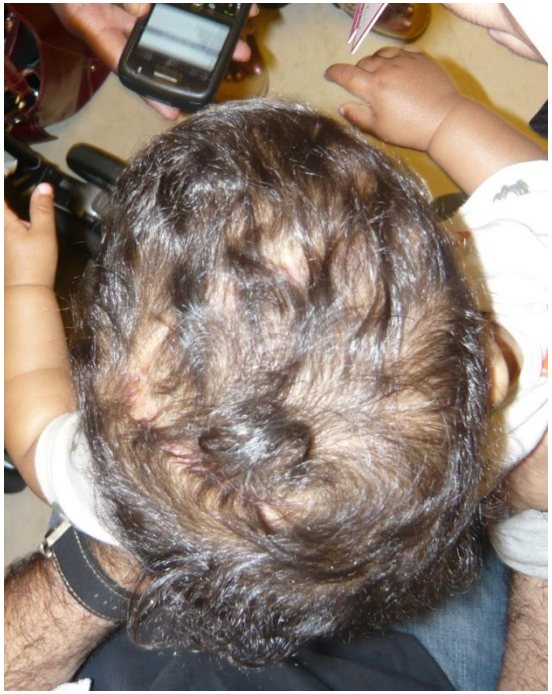
Sagittal synostosis



Sagittal synostosis

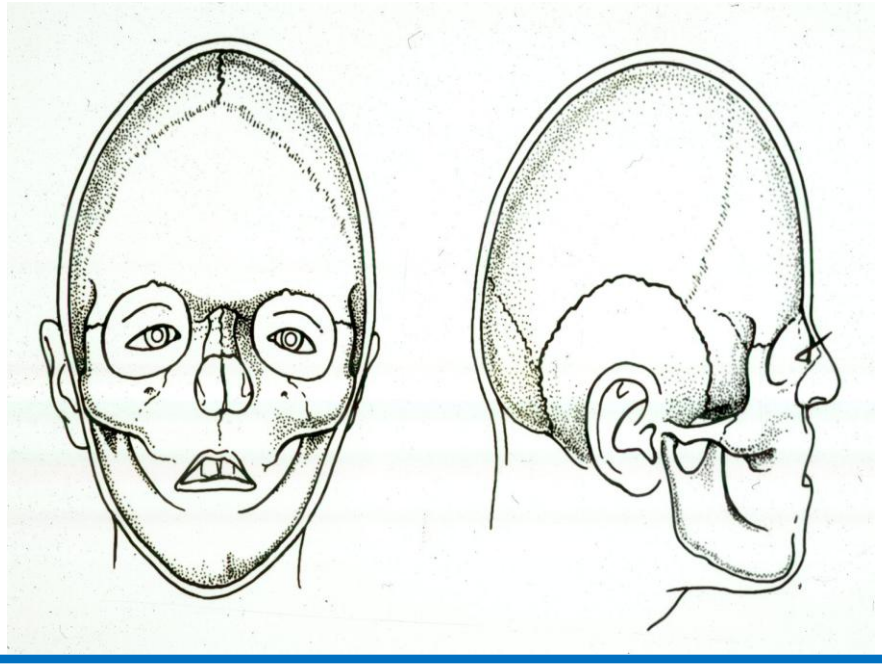






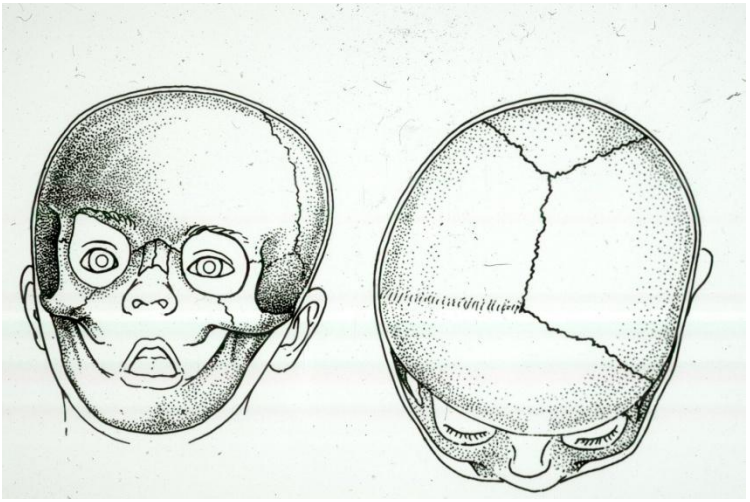
Bilateral Coronal Synostosis

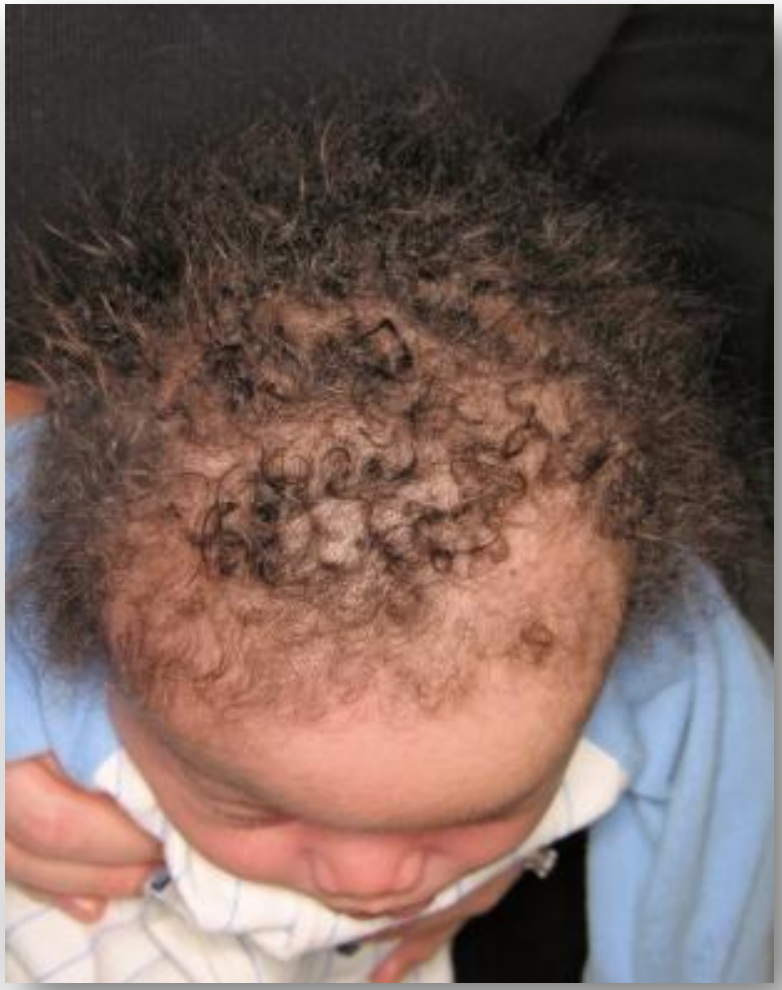
- ✓ Decrease A-P dimension
- ✓ Increase height (turricephaly)
- ✓ Common syndromic example
  - Crouzon
  - Apert

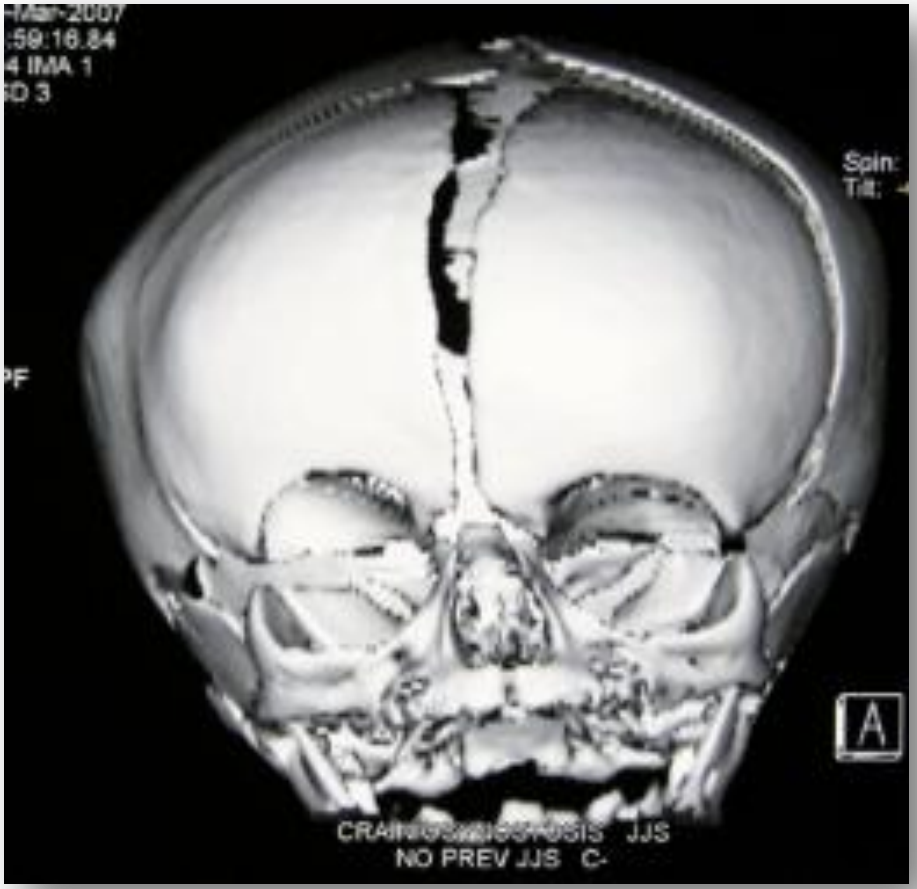


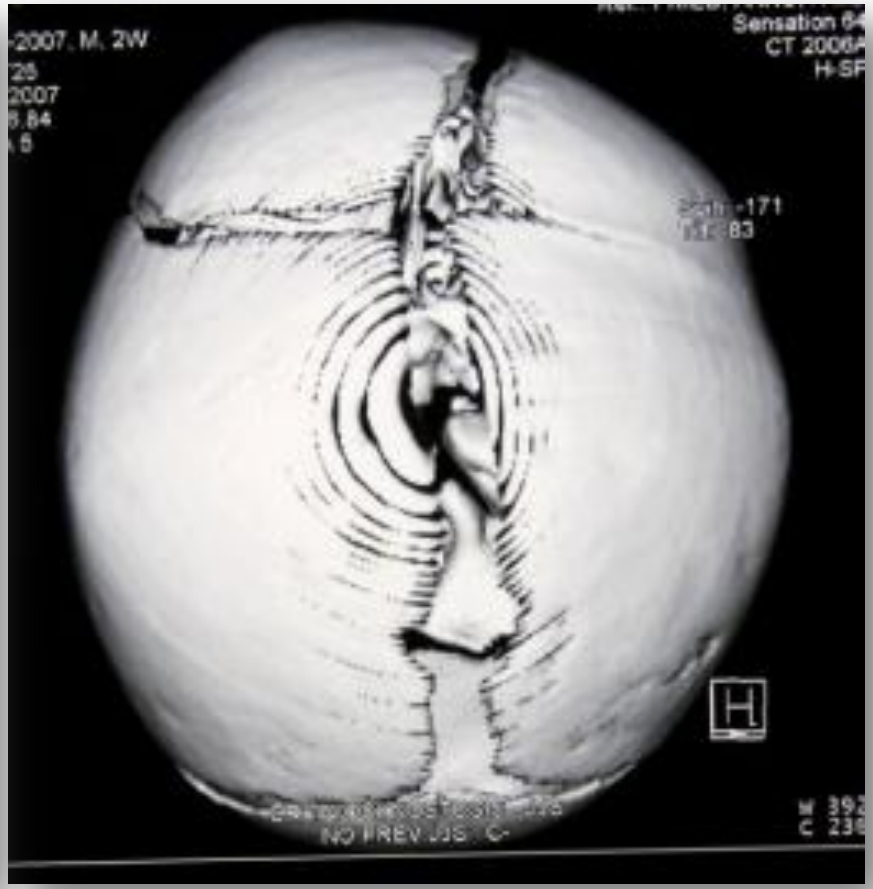
Unilateral Coronal Synostosis

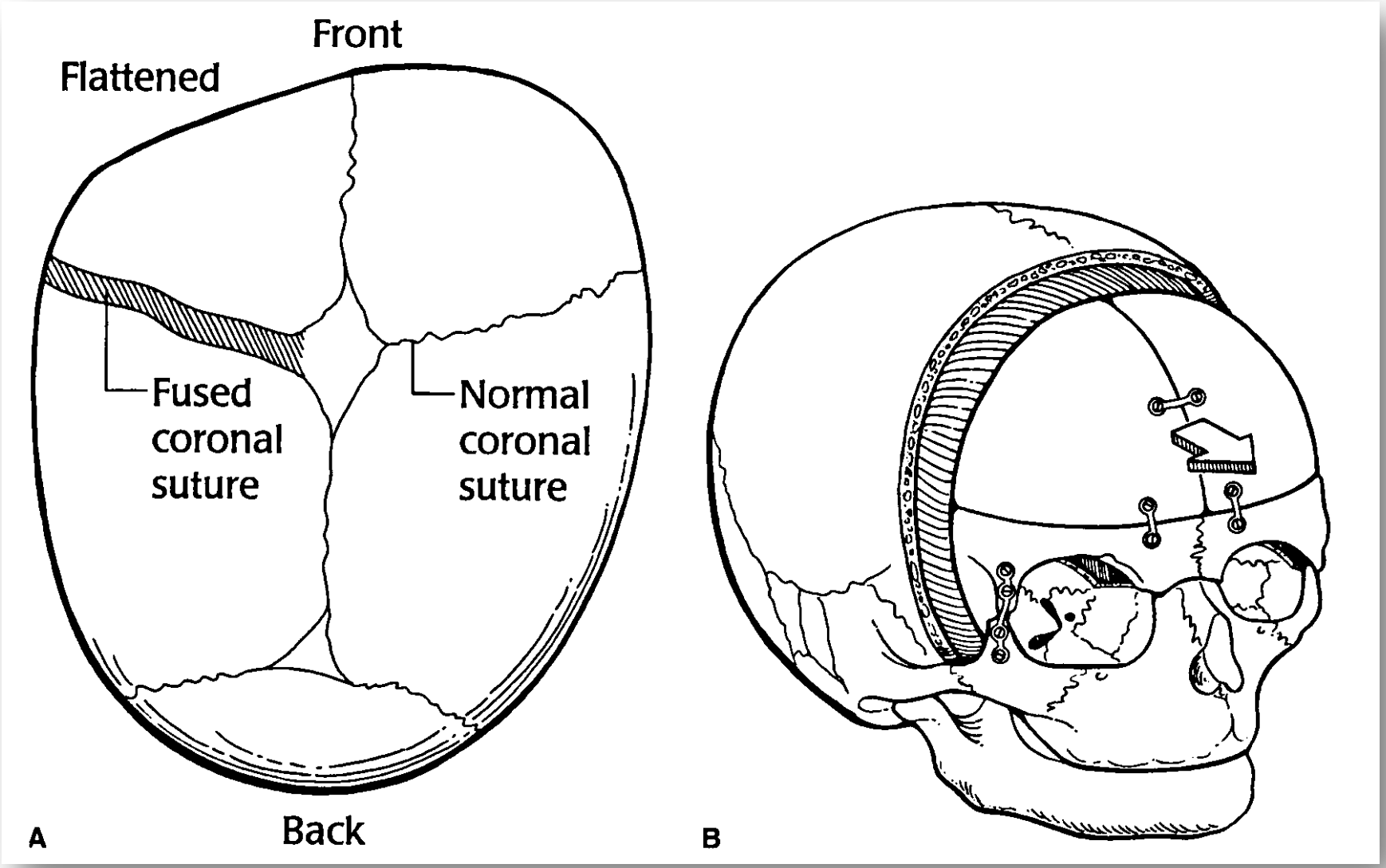
- ✓ Flatten, elongated ipsilateral forehead
- ✓ Contralateral bossing
- ✓ Harlequin orbit

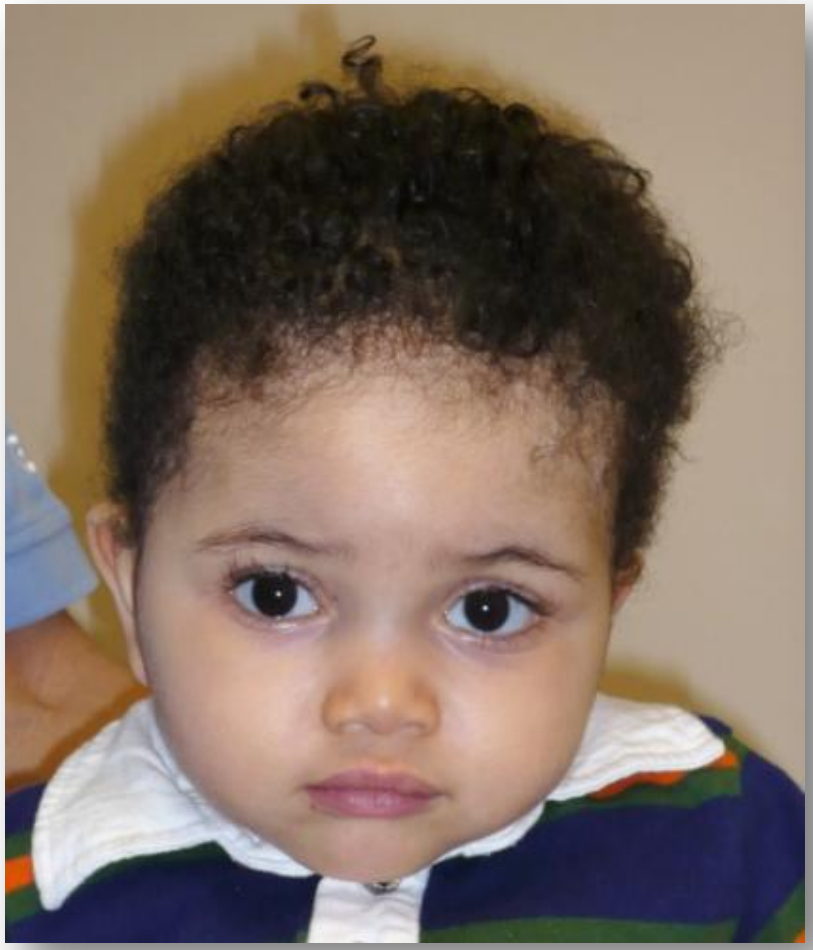





Spin:
Tilt:
A
JJS
NO PREV JJS C-

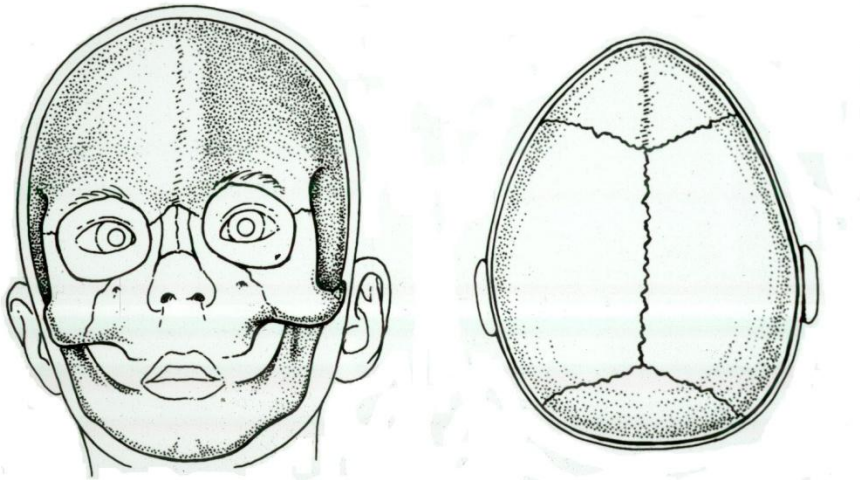

Sensation
CT
H-SF
H
NO PREV JJS C-

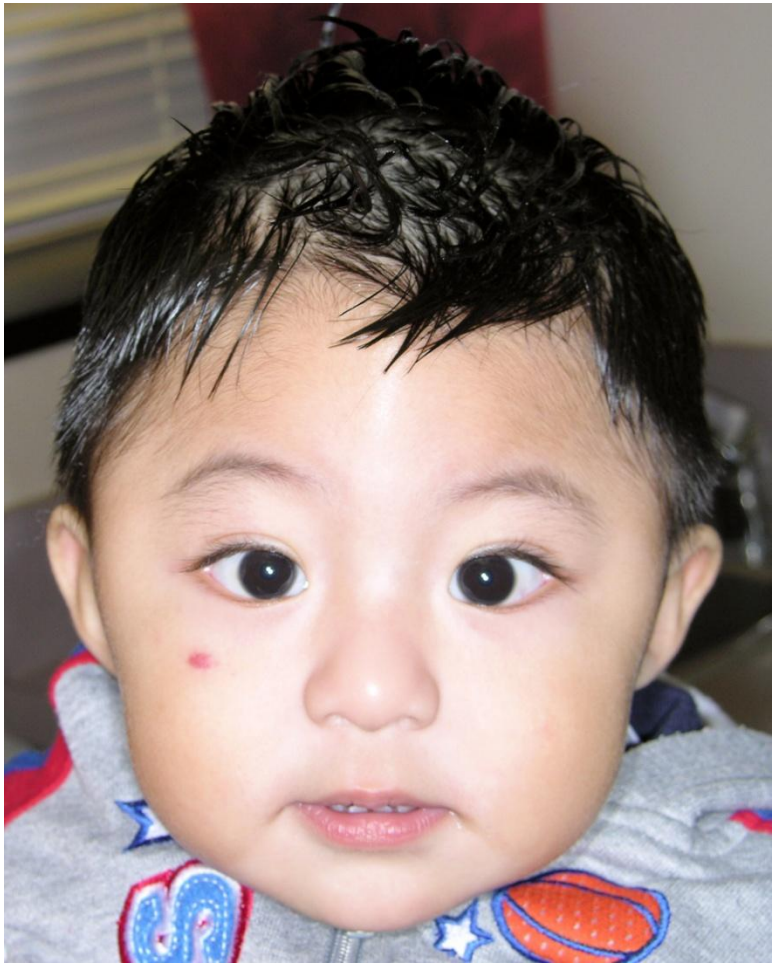

Front
Flattened
Fused coronal suture
Normal coronal suture
Back
A
B

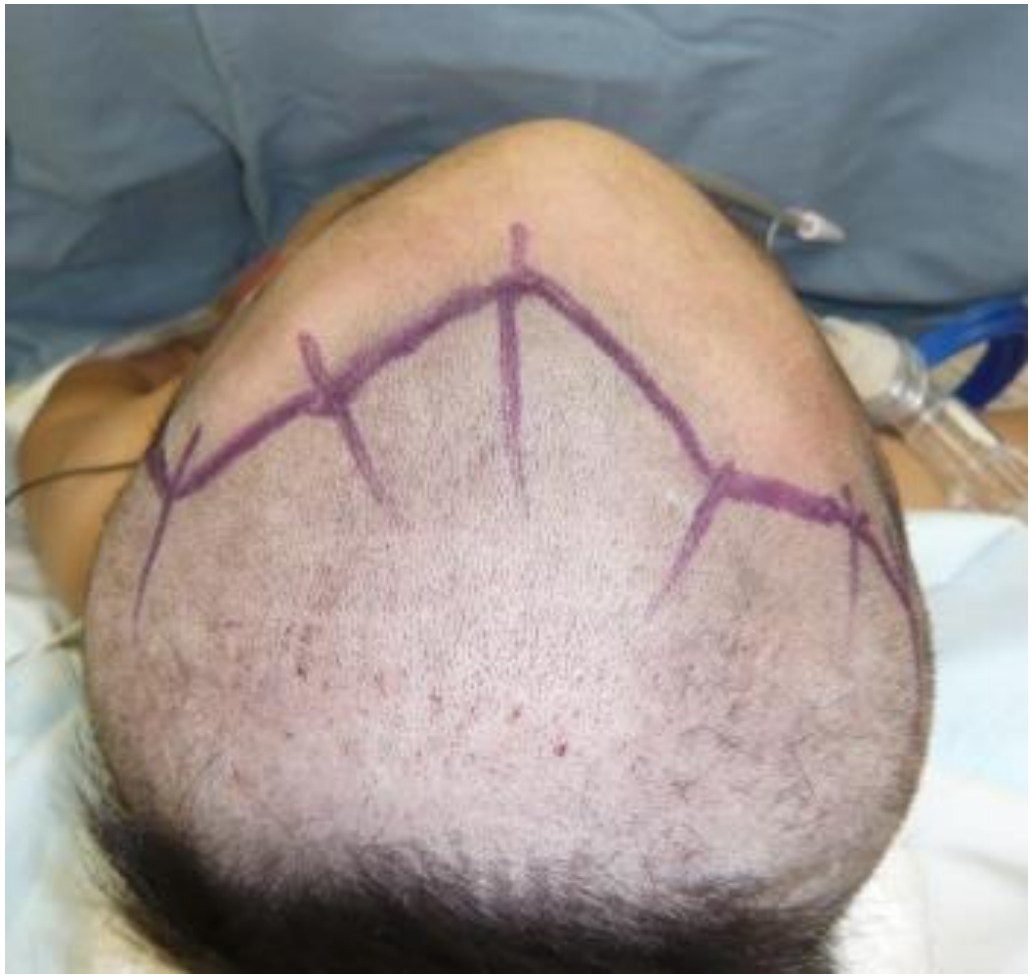


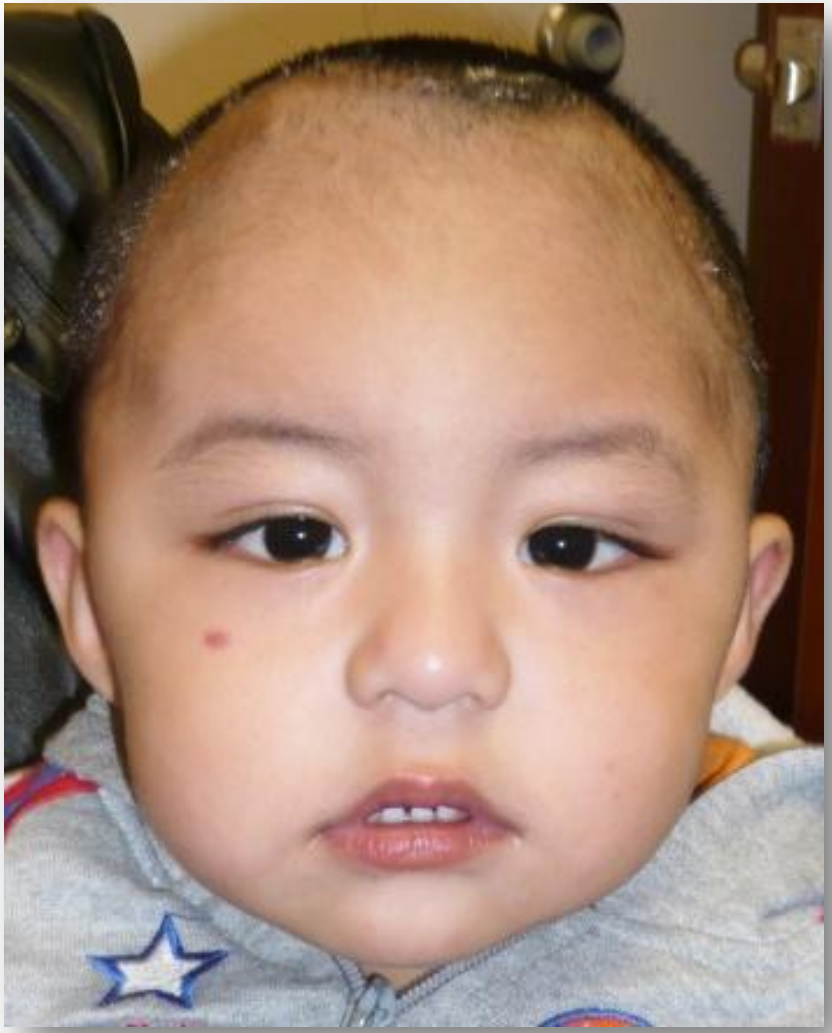
Metopic Synostosis

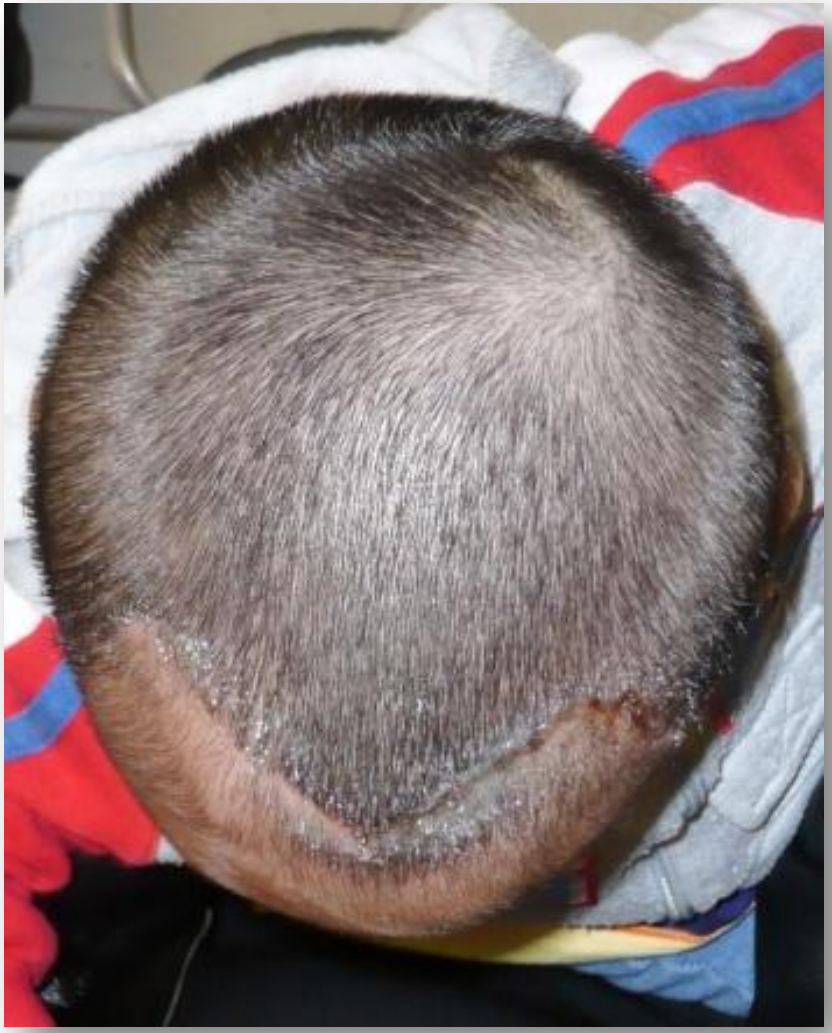
- Characteristic keel shaped deformity
- Receded superior orbital rims
- Bitemporal narrowing
- Hypotelorbitism
- Uncommon (<10%)

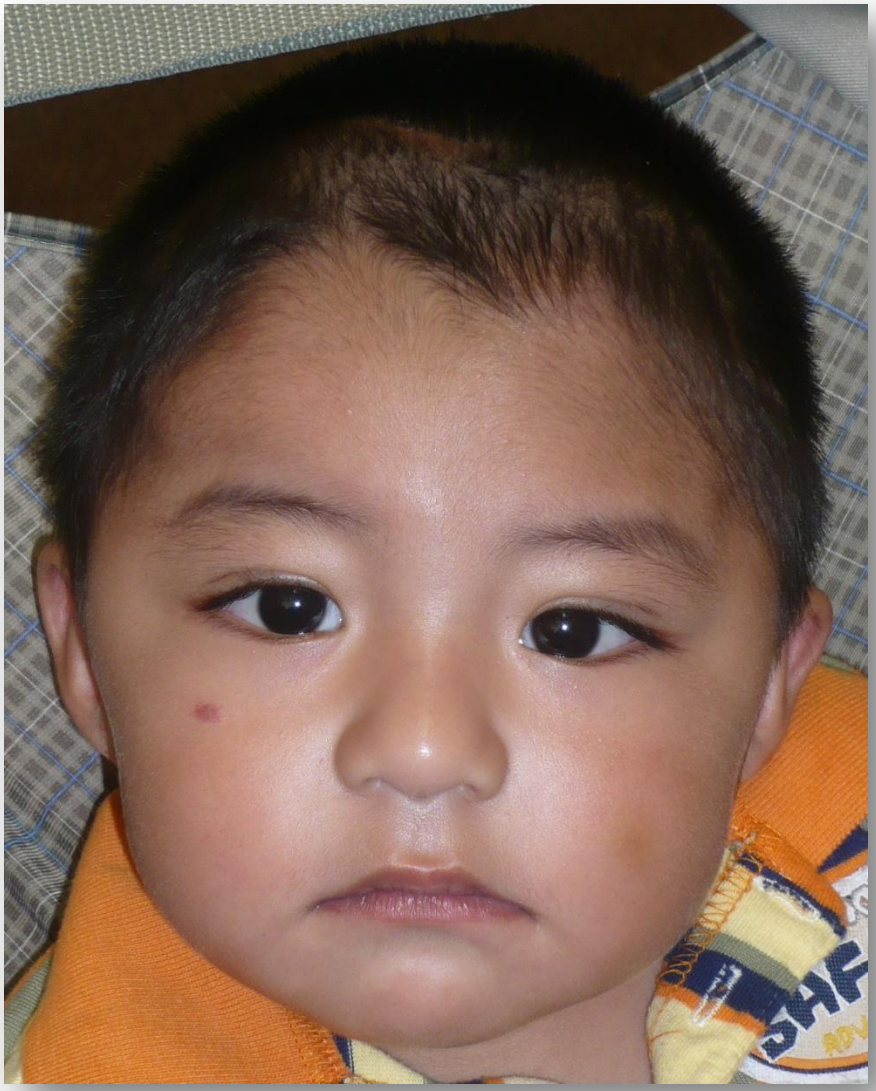


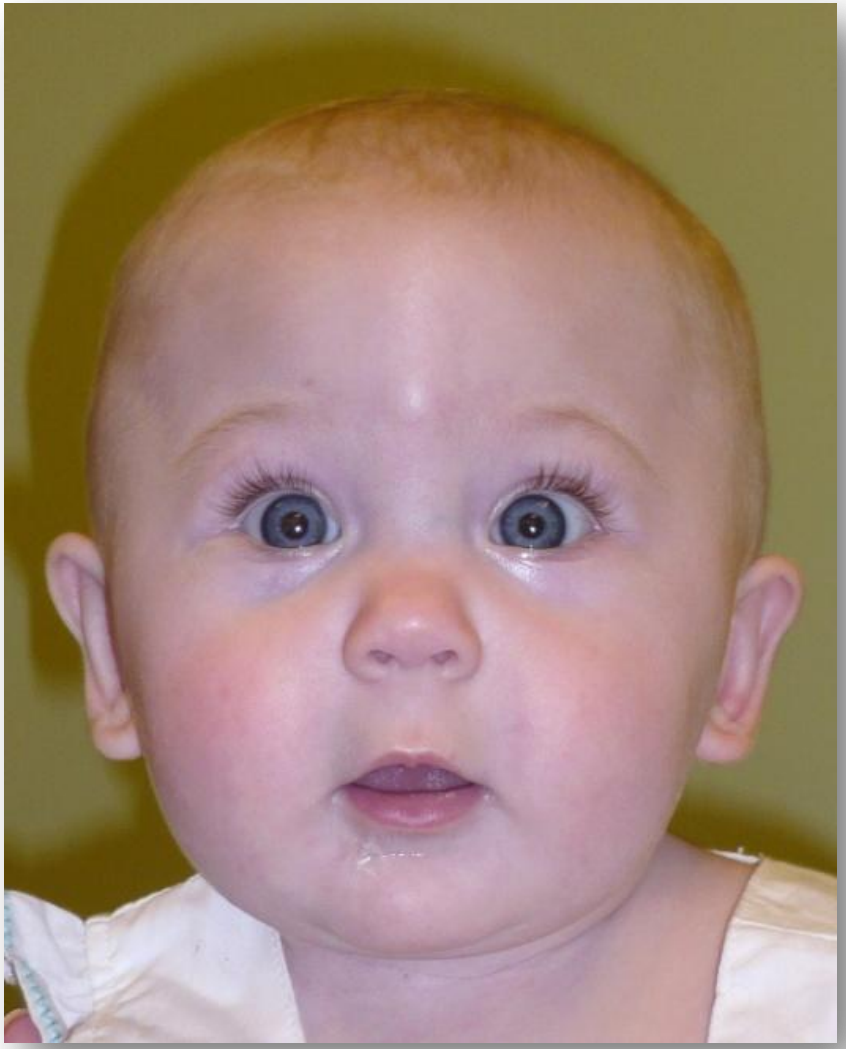


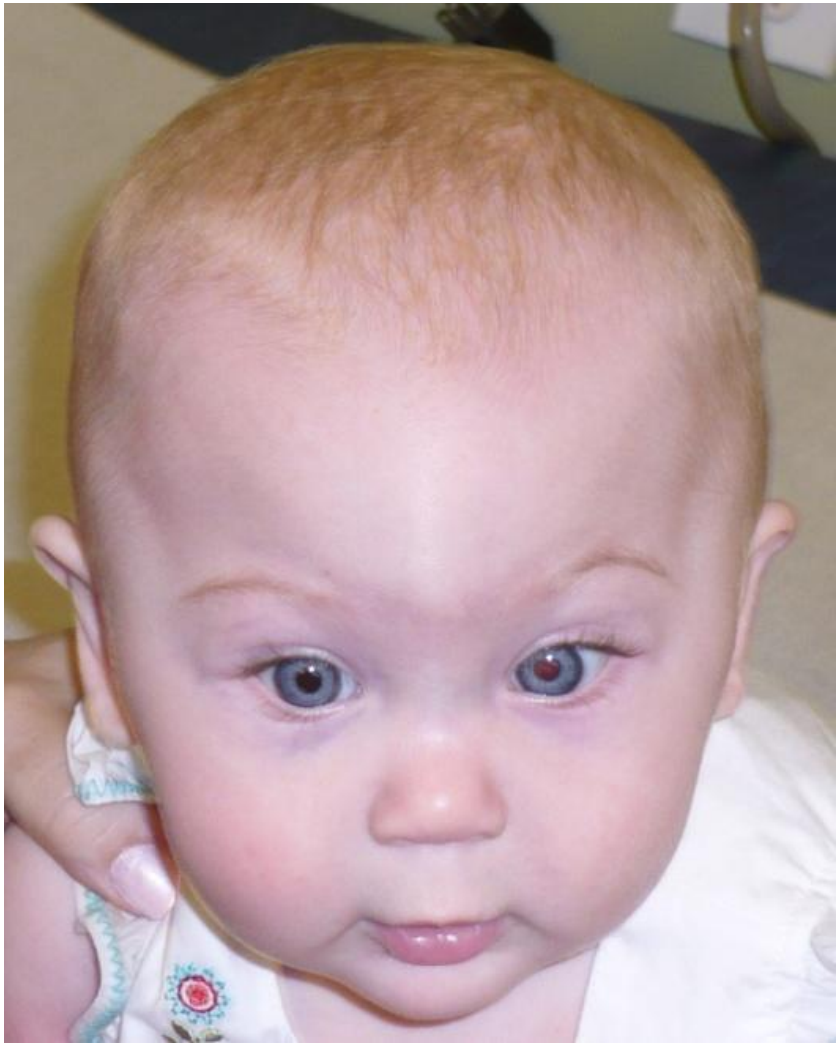




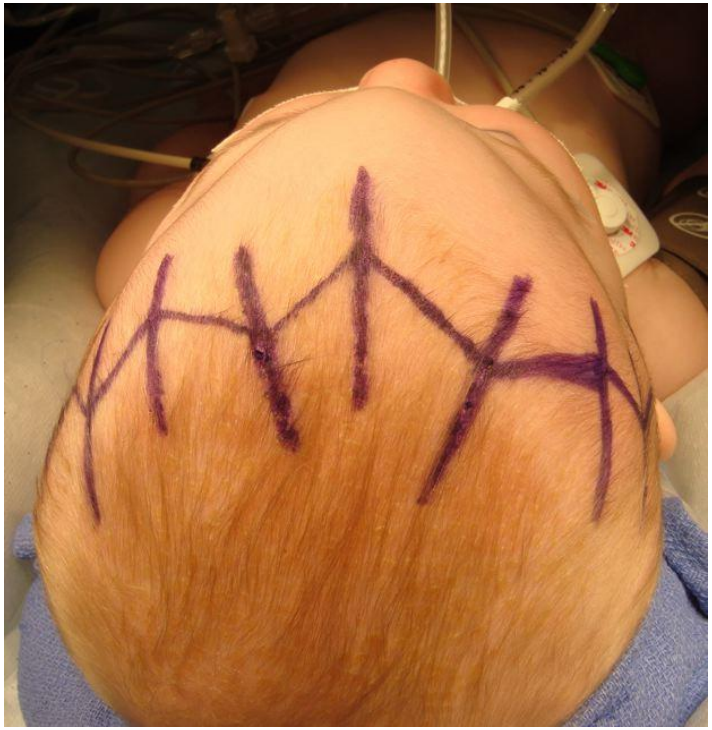


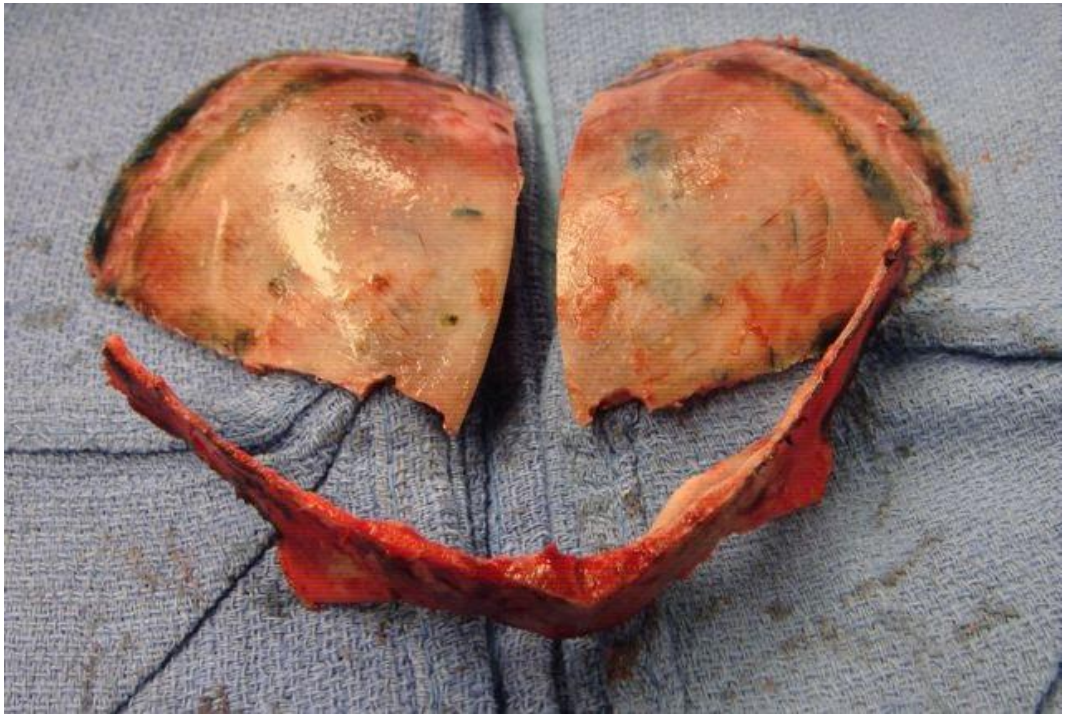


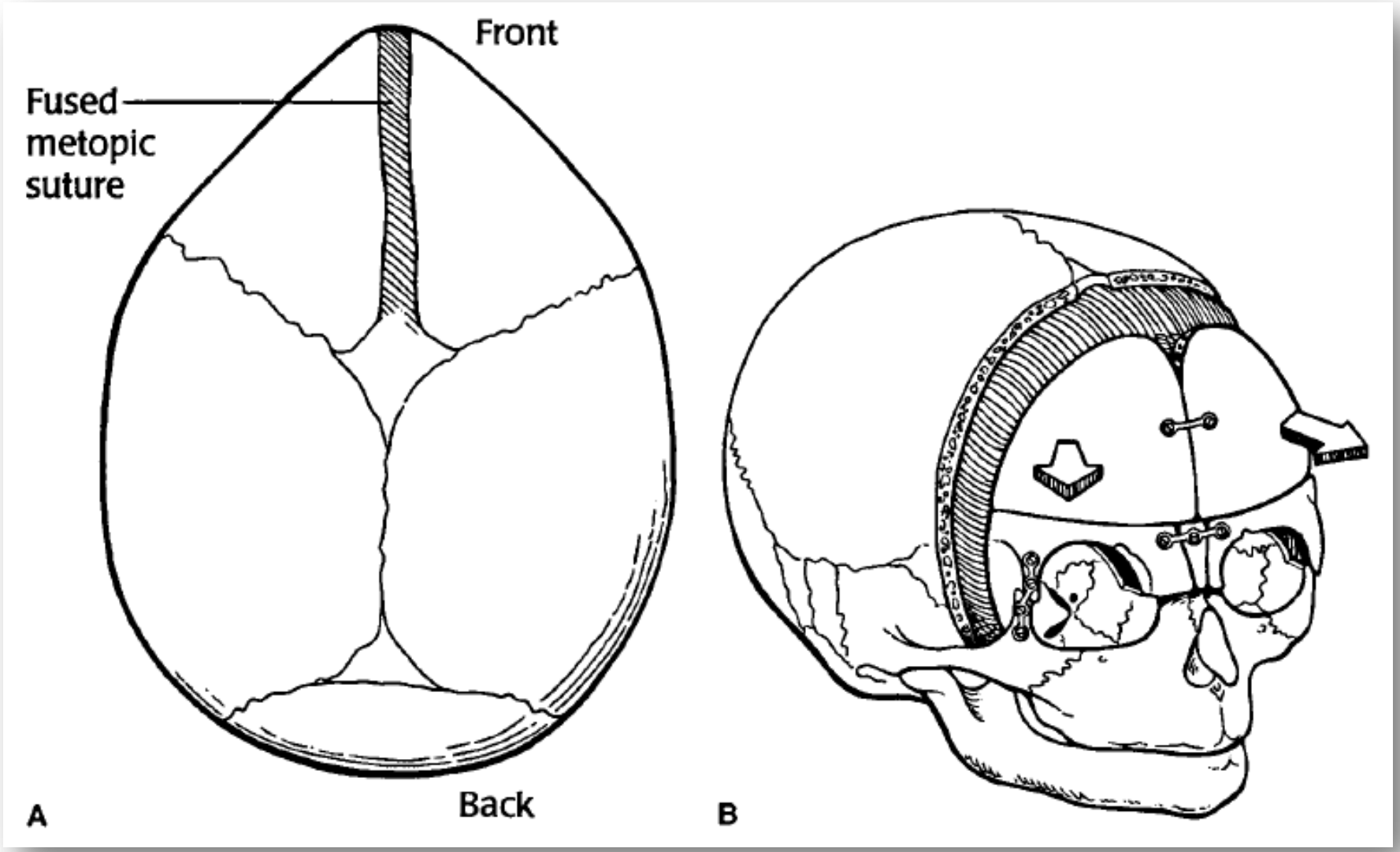




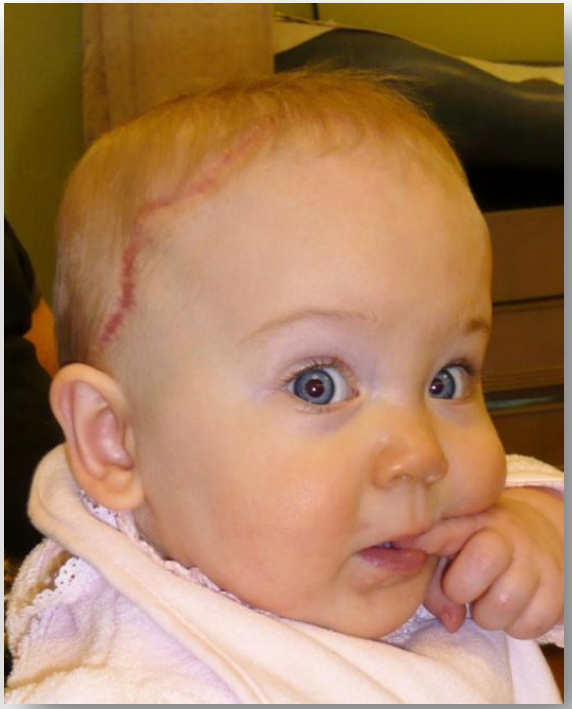
Intraoperative



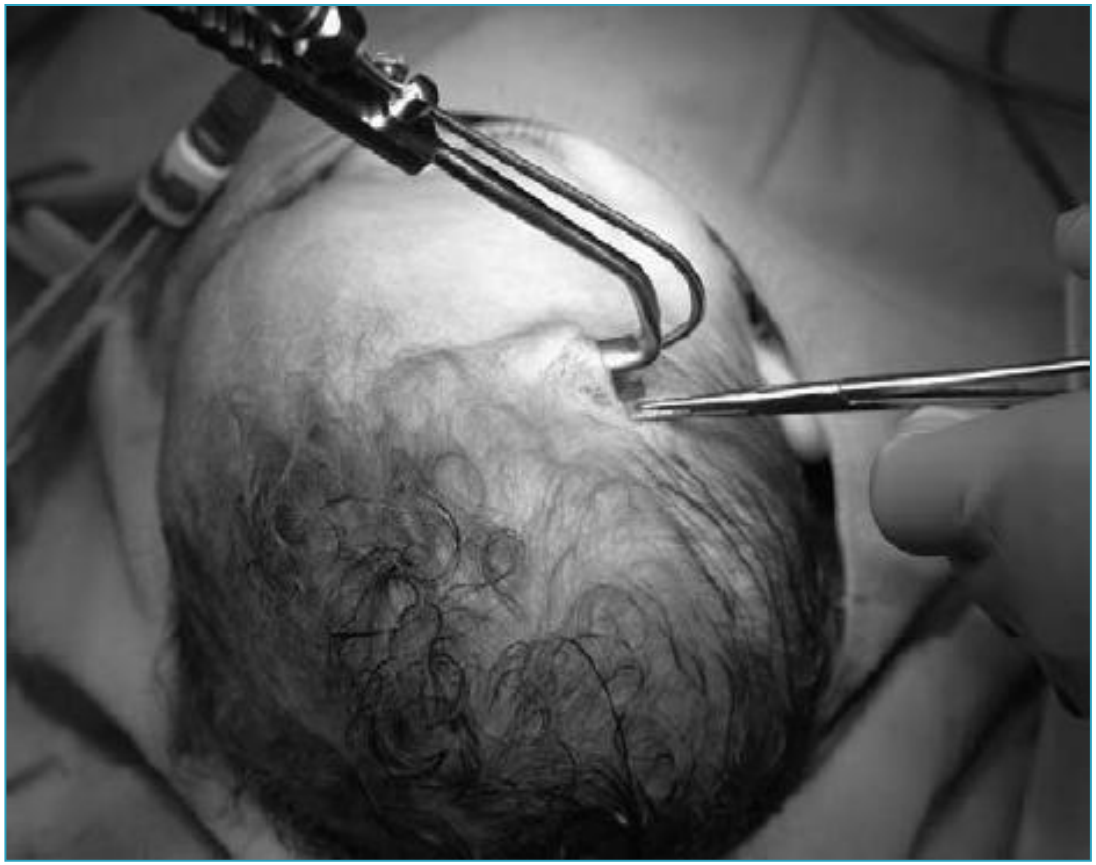



Front
Fused metopic suture
Back
A
B

Postoperative



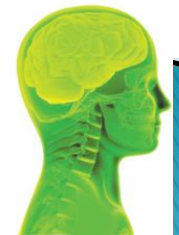
Minimally invasive surgery



Endoscopic approach to coronal craniosynostosis. Barone CM, David F. Jimenez DF, Clin Plastic Surg 31 (2004) 415– 422.

Summary

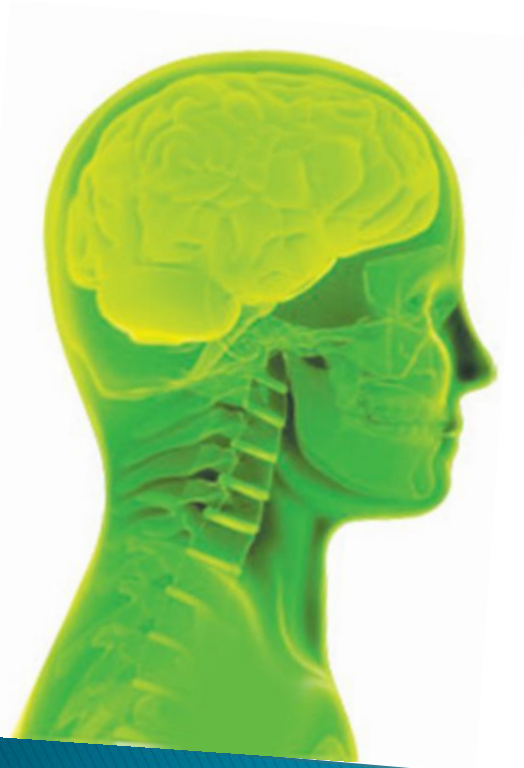
- Positional head deformity
  - May be difficult to distinguish from synostosis
  - Conservative management & PT, helmet optional
- Craniosynostosis
  - Evaluate for other factors such as microcephaly
  - CT scan for neurologic issues, not head shape
  - Surgery if indicated
- Minimally invasive surgery
  - Literature – good results in appropriate patients
  - Early referral (under 3 months of age if possible)



Weill Cornell Medical College

NewYork-Presbyterian
Phyllis and David Komansky
Center for Children's Health

Pediatric
Brain and Spine Center



Thank you

Samuel Rhee, MD

Recent Director, Craniofacial Surgery Program

Department of Surgery

Weill Cornell Medical College

New York-Presbyterian Medical Center