Evaluation and Treatment of Abnormal Head Shape in Pediatric Neurosurgery Patients

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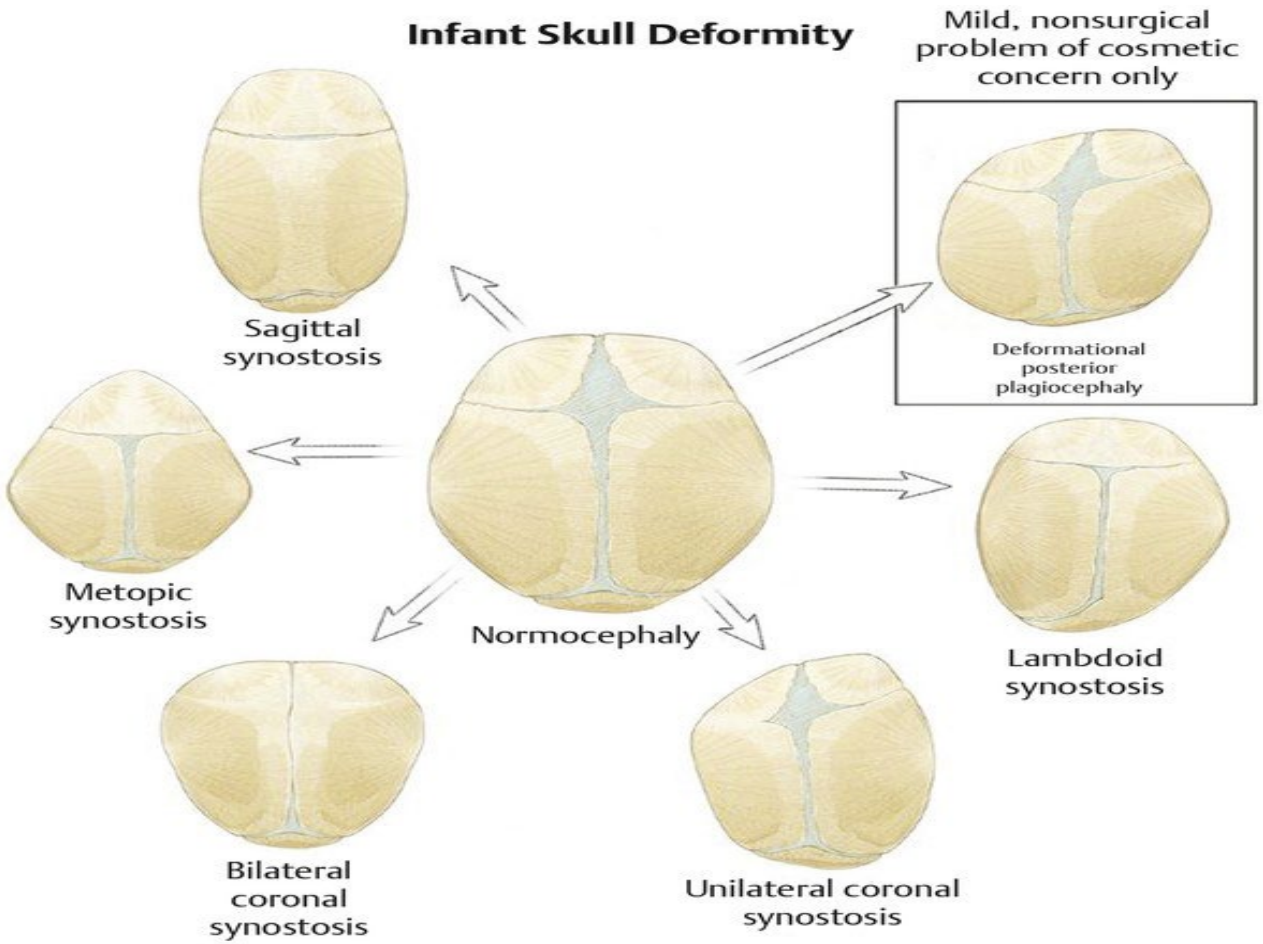
Ochsner Children's

Disclosure

- Nothing to disclose


Infant Skull Deformity
Mild, nonsurgical problem of cosmetic concern only
Sagittal synostosis
Deformational posterior plagiocephaly
Metopic synostosis
Normocephaly
Lambdoid synostosis
Bilateral coronal synostosis
Unilateral coronal synostosis

Epidemiology

- Craniosynostosis is seen in every 1 out of 2,000-2,500 births.
- Most commonly only single suture synostosis is seen.
- White non-Hispanic mothers have increased frequency.

Pathophysiology

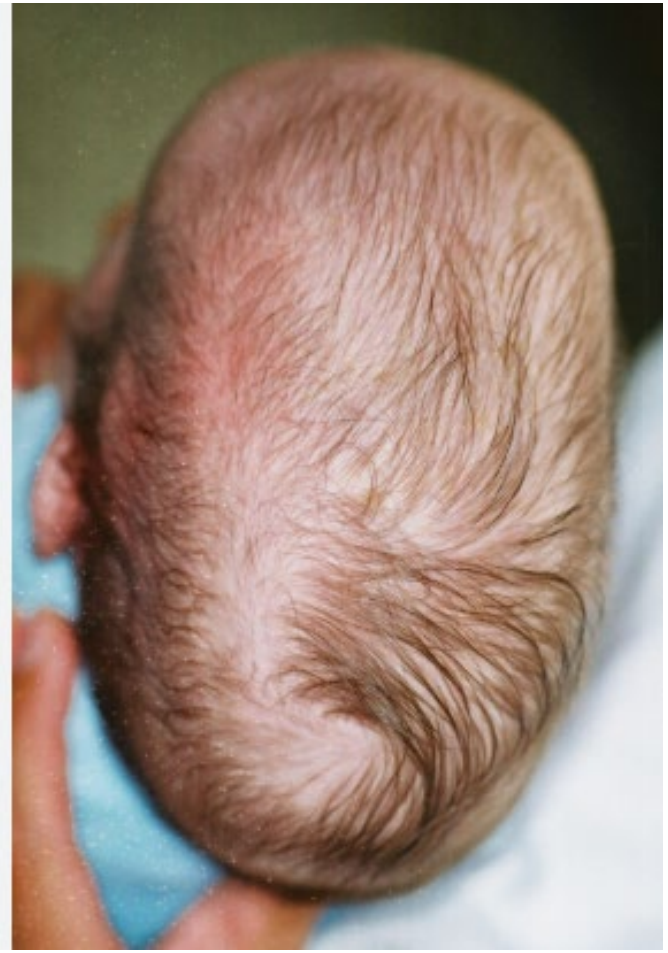
- The etiology is not completely understood and is most commonly sporadic.
- Advanced paternal age and maternal smoking have been implicated.
- Certain genetic mutations have been associated with craniosynostosis.
  - *FGFR1-3*, *NELL1*, *MSX2*

Surgical Indication

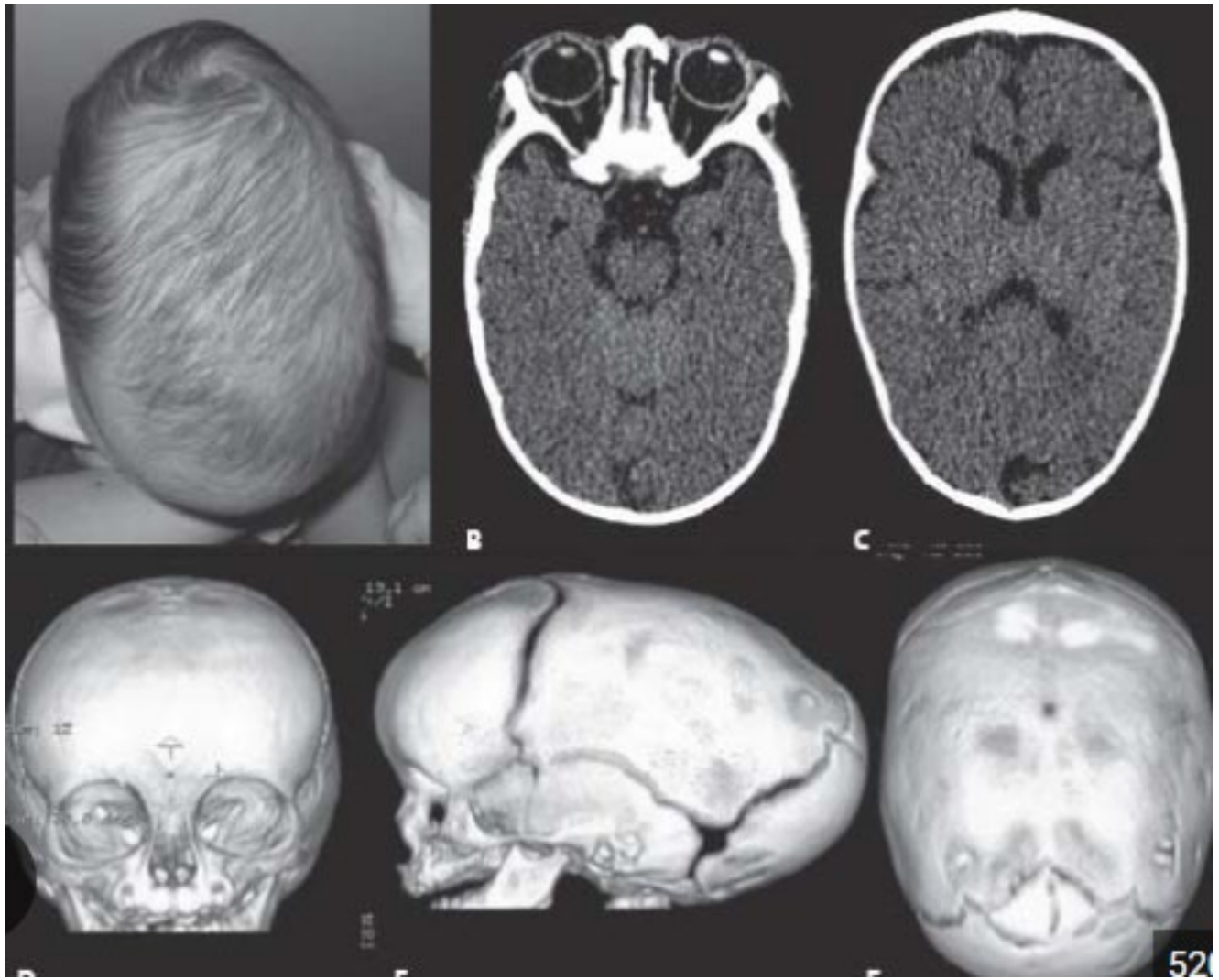
- The two main surgical indications are:
  - Correct skull shape for aesthetic and psychosocial considerations.
  - Make sure there is adequate space for normal brain growth to occur.
- There is a low percentage risk of increased intracranial pressure in single suture synostosis (4-14%).
  - That percentage is much higher in multi-suture synostosis (47-67%)

Sagittal synostosis

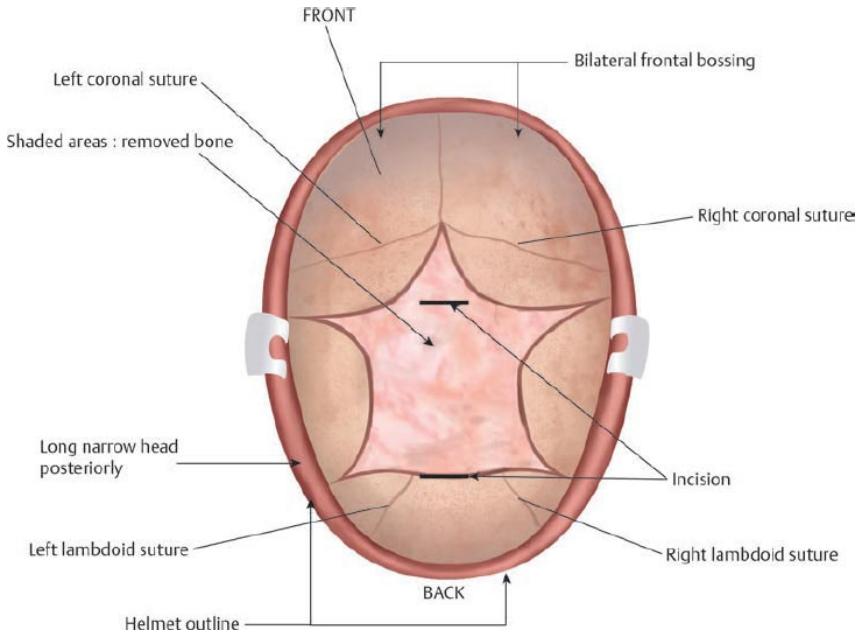
- Sagittal synostosis is the most common form of craniosynostosis.
  - Accounts for 53-60% of cases
- Patients have a narrow, elongated head shape (Scaphocephaly).
  - Biparietal narrowing
  - Occipital Bullet
  - Delayed anterior fontanelle closure
  - Peri-sutural ridging

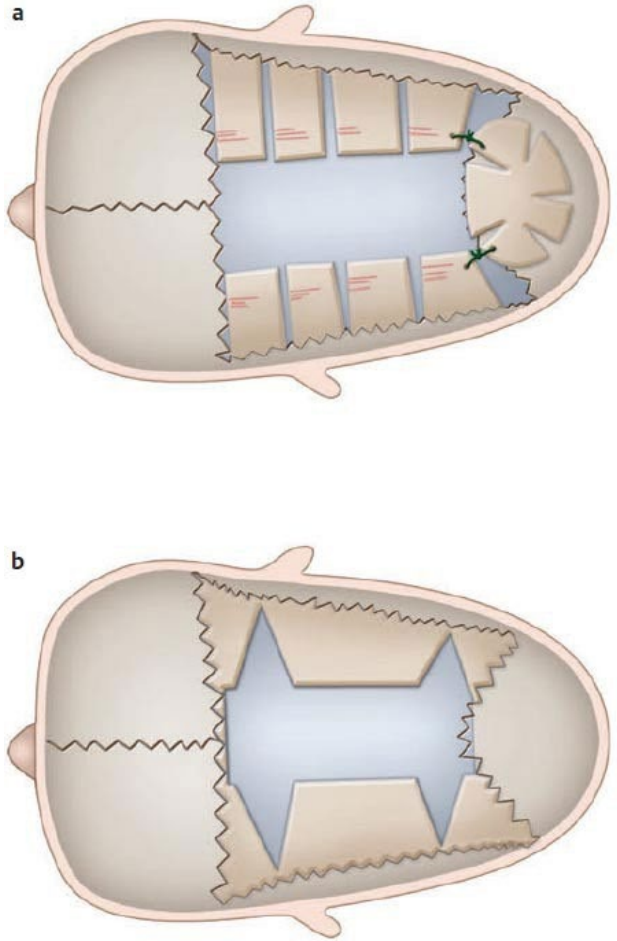


Sagittal synostosis



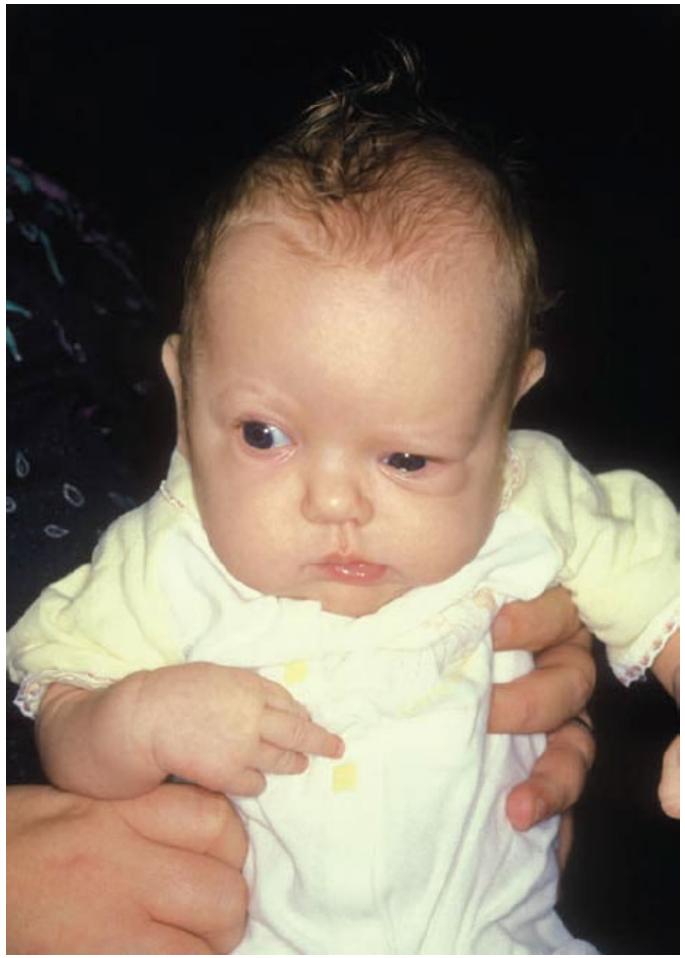
Sagittal Synostosis





Coronal Synostosis

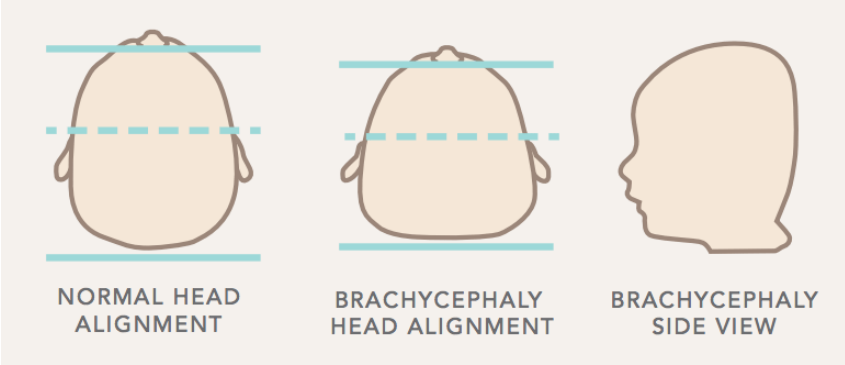
- 2nd most common type
  - Accounts for 17-29% of cases
  - Unilateral coronal synostosis is more common than bilateral



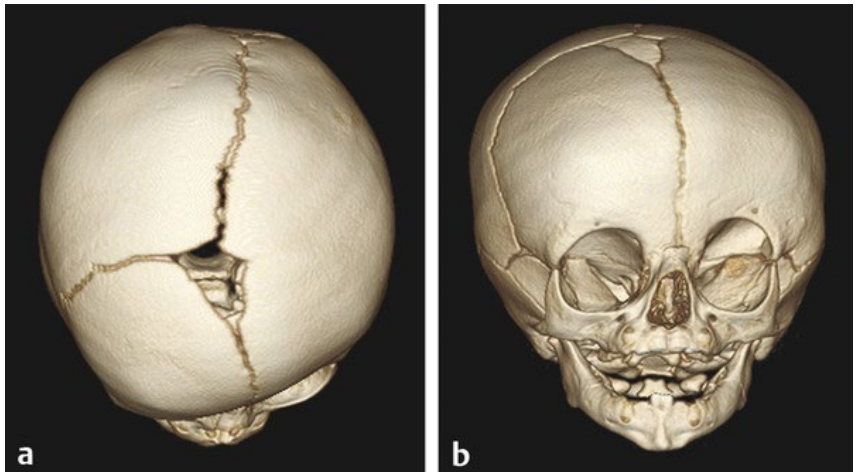
Coronal Synostosis

Coronal Synostosis

- Bilateral Coronal Synostosis (Brachycephaly)
  - Shortened anterior fossa
  - Widened biparietal diameter
  - Frontal towering (Turricephaly)
  - Recessed supraorbital rims
  - Flattened occiput

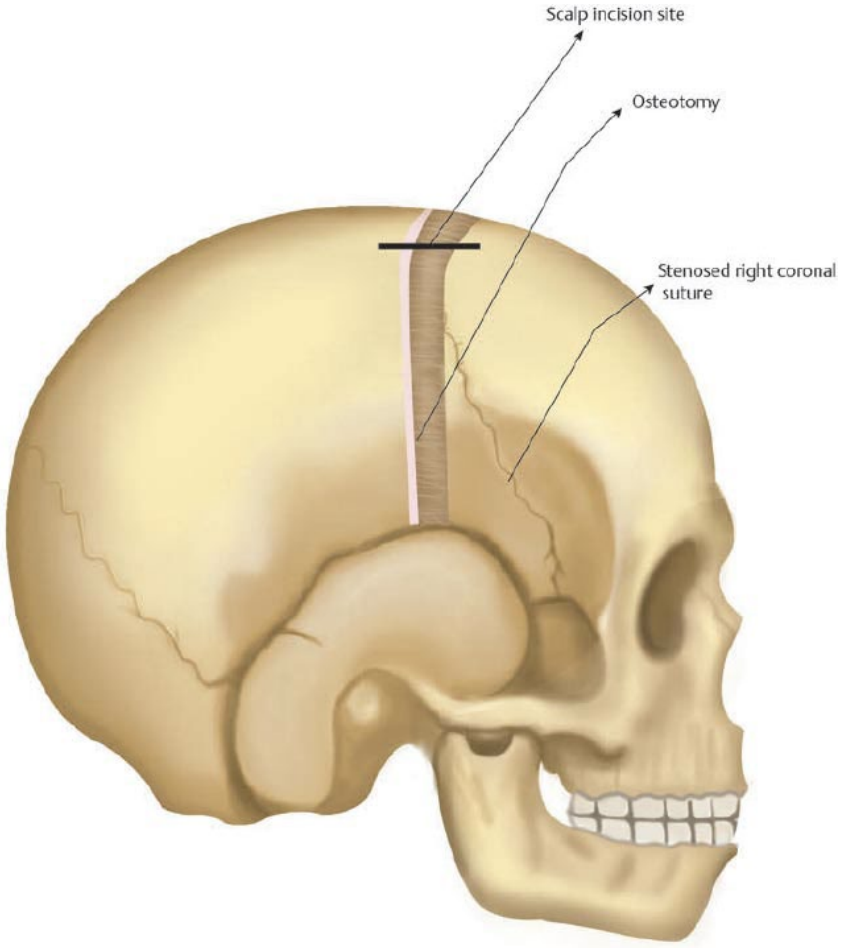


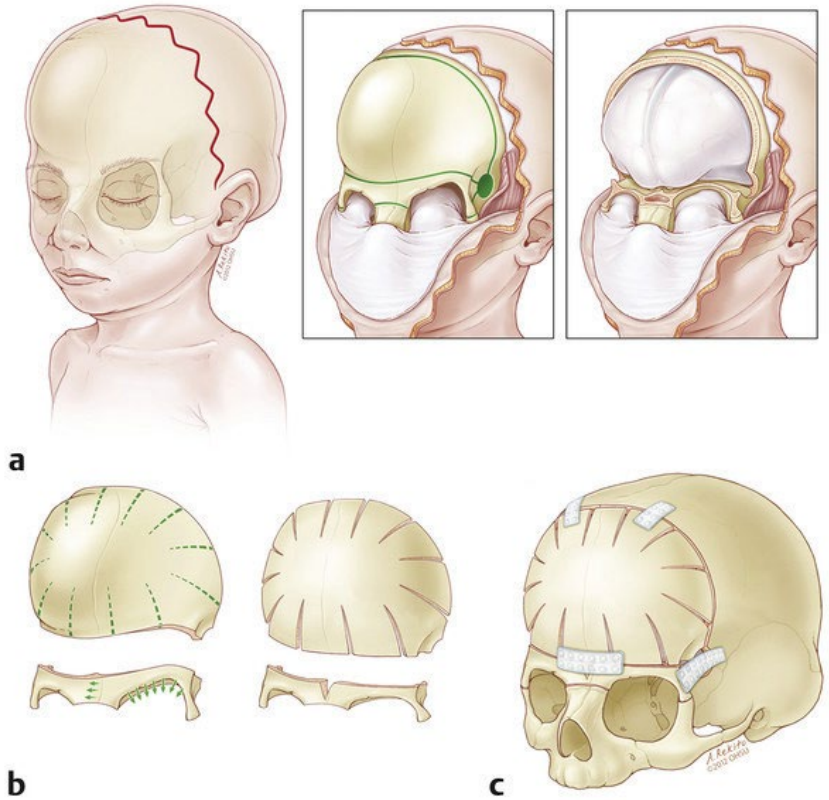
Coronal synostosis



- Unilateral Coronal synostosis (Anterior Plagiocephaly)
  - Flattening of the frontal bone
  - Raised supraorbital rim
  - Nasal radix deviation
  - Anterior ear displacement
  - Harlequin deformity (raised sphenoid wing)

Coronal Synostosis

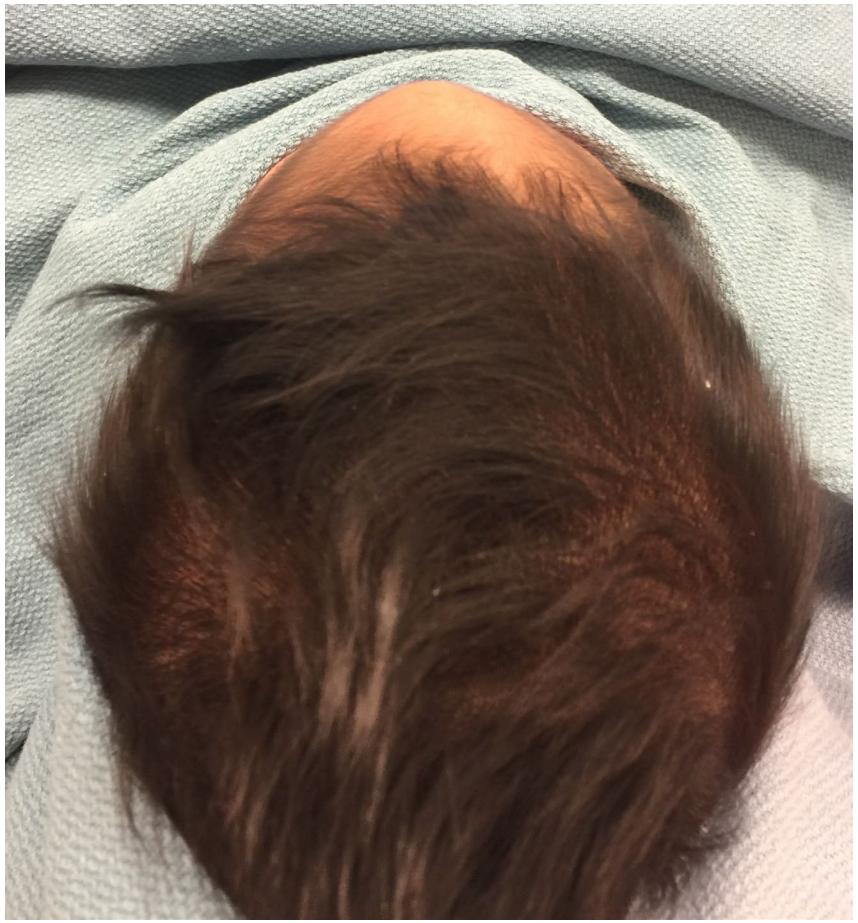




Metopic Synostosis

- Known as trigonocephaly.
- The metopic suture normally closes around 9 months of age, but it can close as early as 3 months.
- 3rd most common type:
  - Accounts for 4-10% of cases.

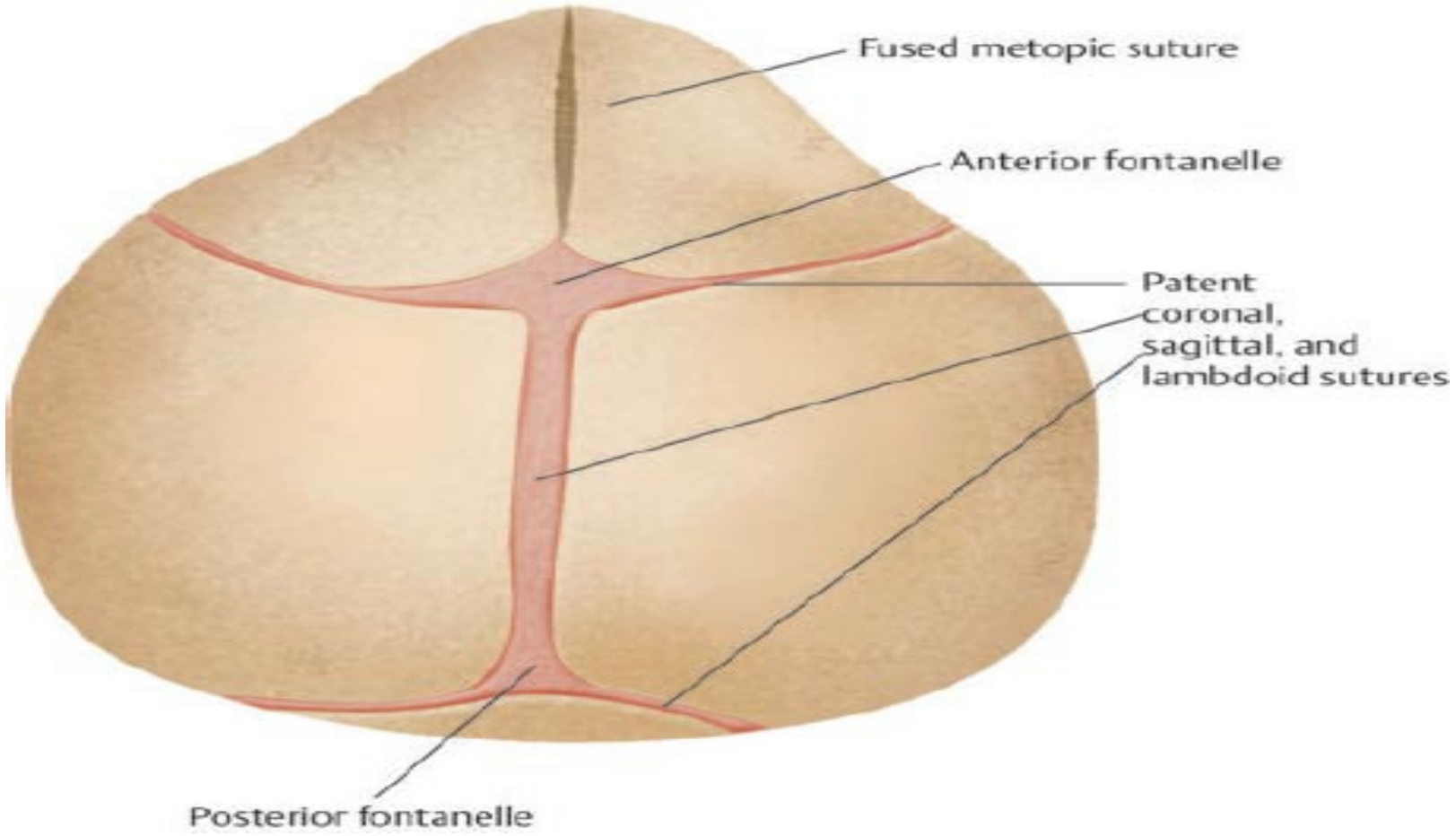
Metopic synostosis



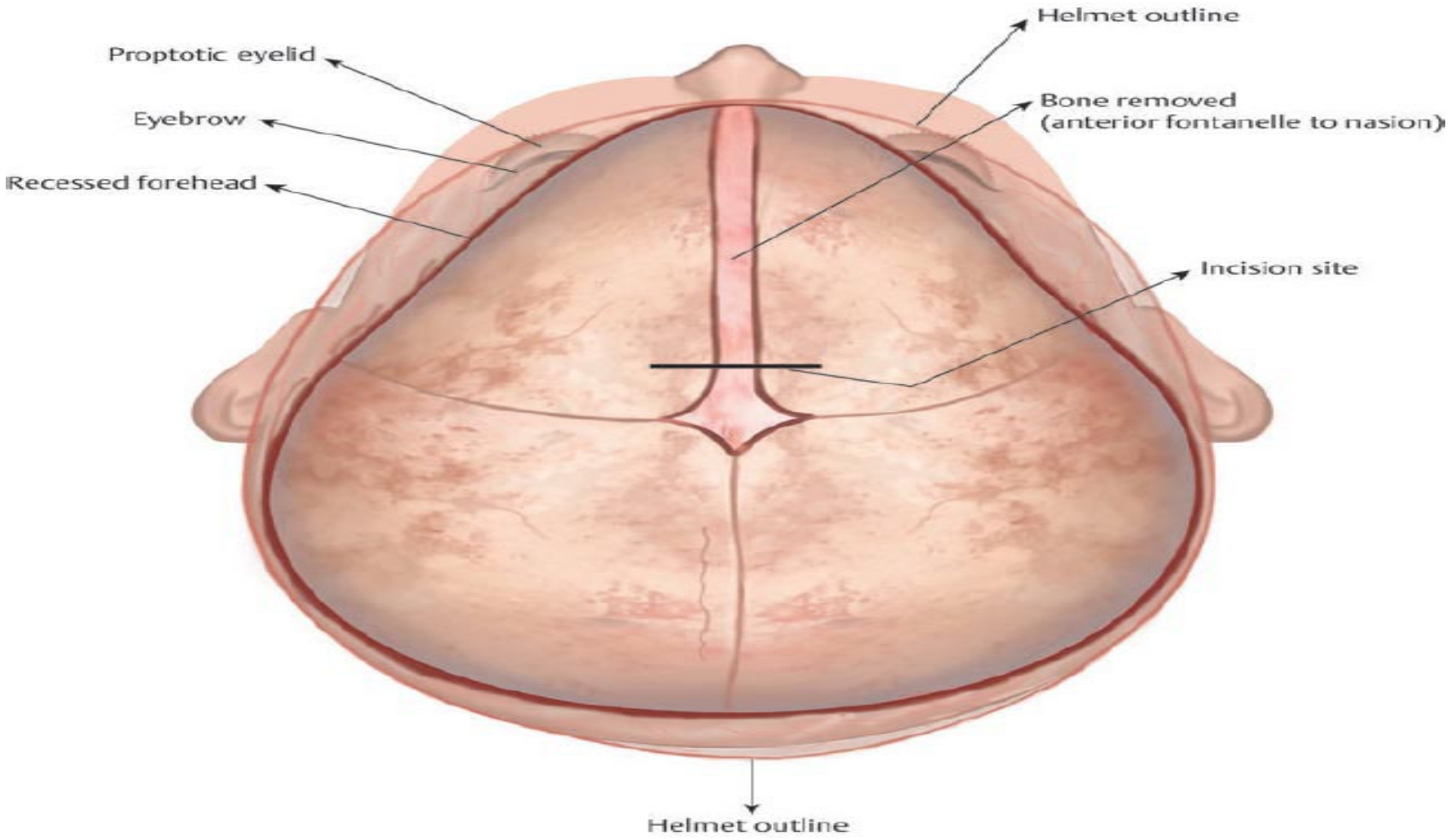
Metopic Synostosis

- Involves:
  - ridging along the metopic suture.
  - posterior displacement of the supraorbital rims
  - hypotelorism
  - flattening of the frontal bones
  - anterior displacement of the coronal suture
  - temporal narrowing

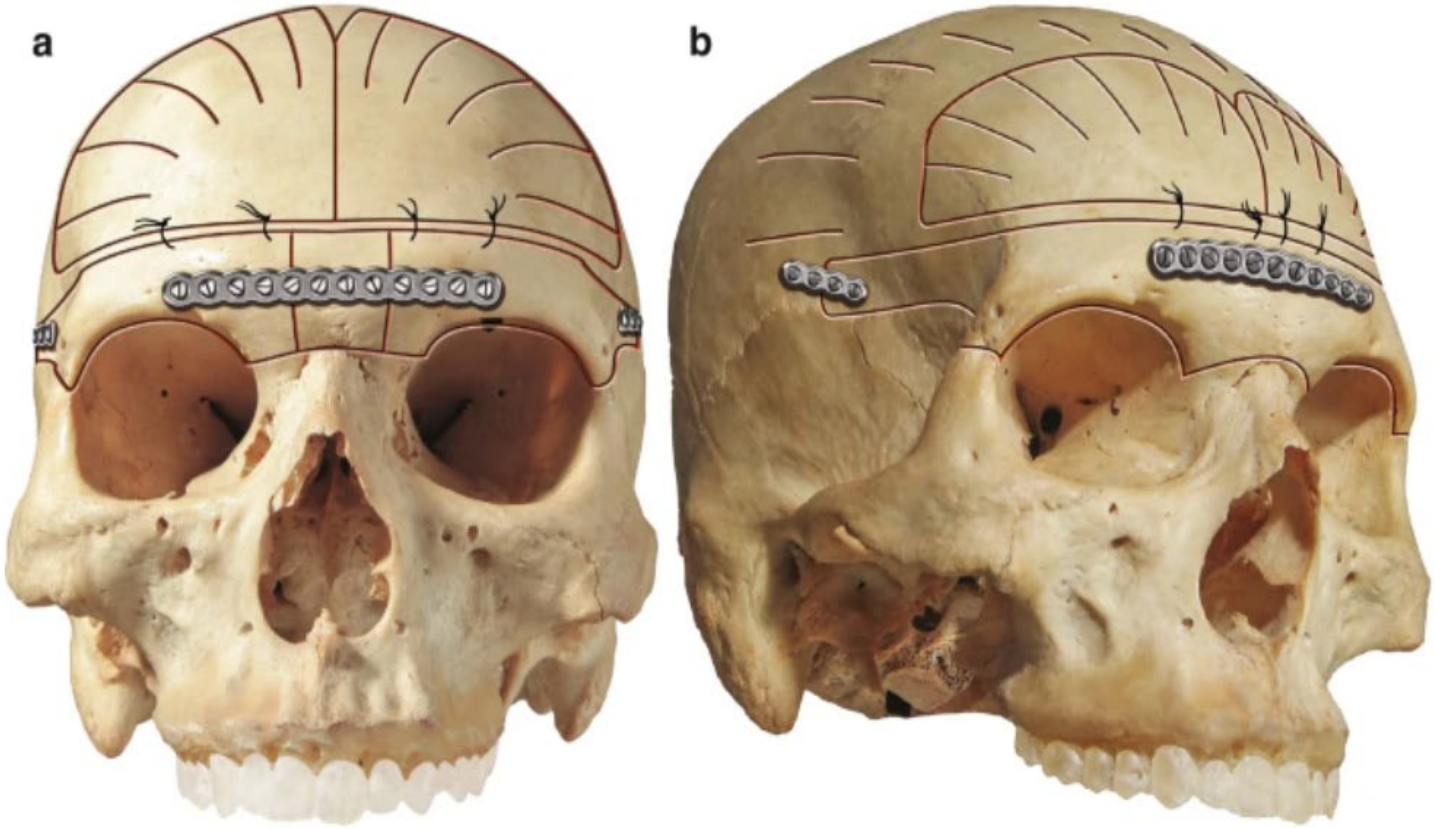
Metopic Synostosis



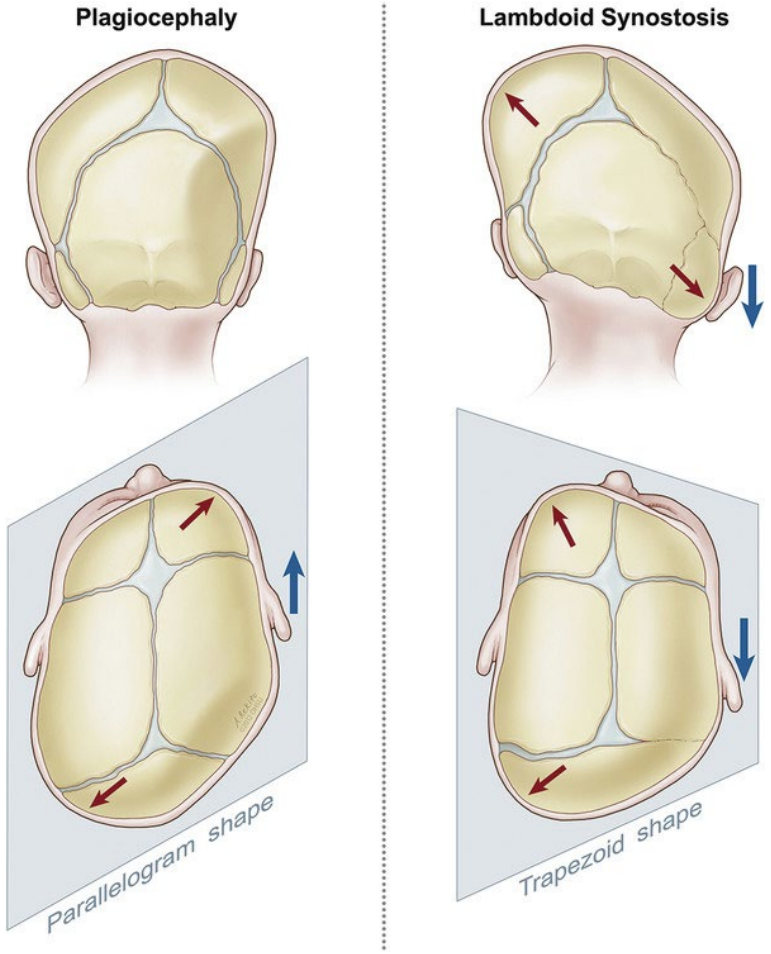
Metopic Synostosis



Metopic Synostosis

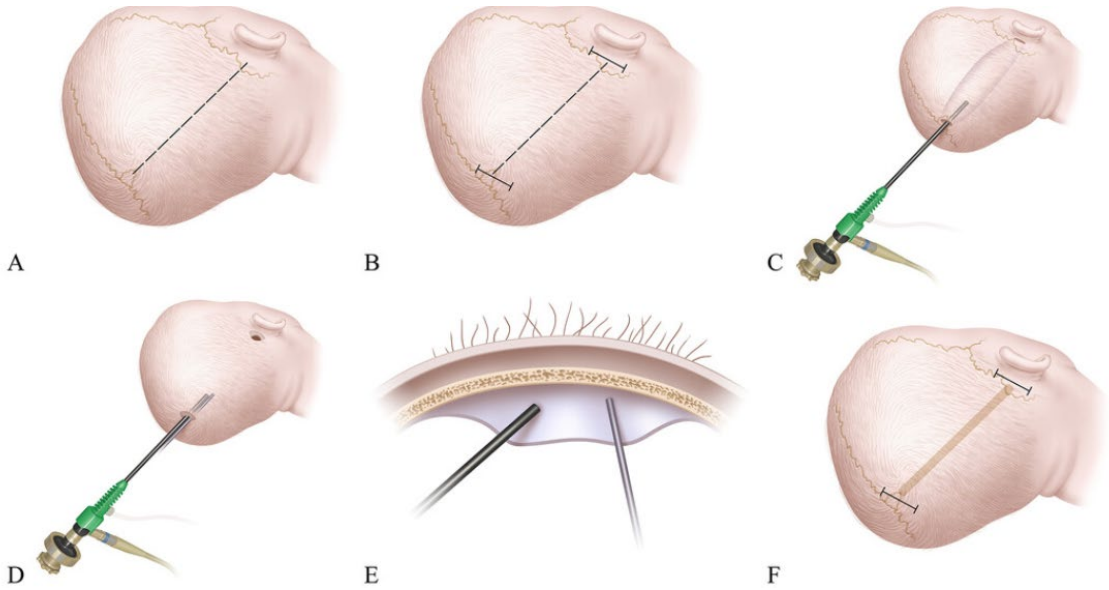


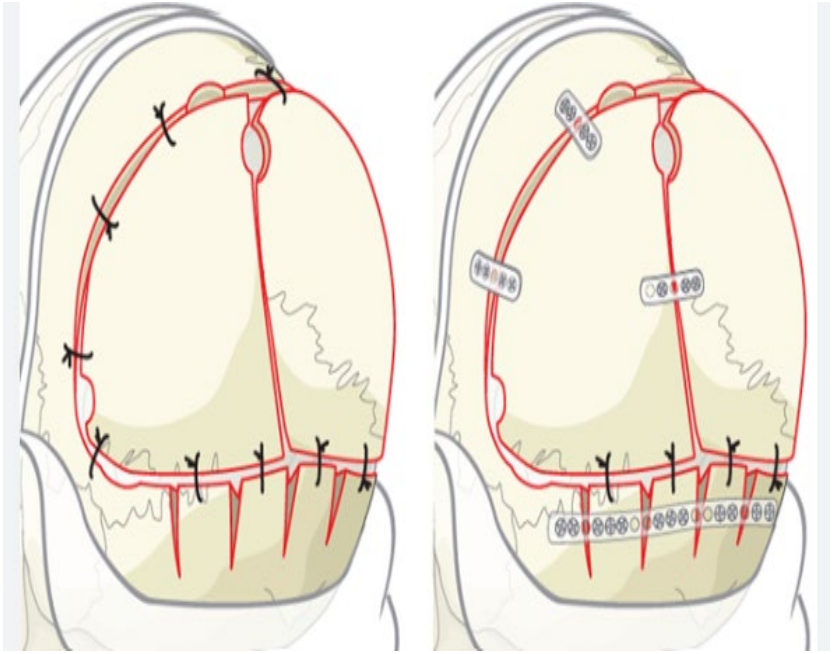
Lamboid synostosis



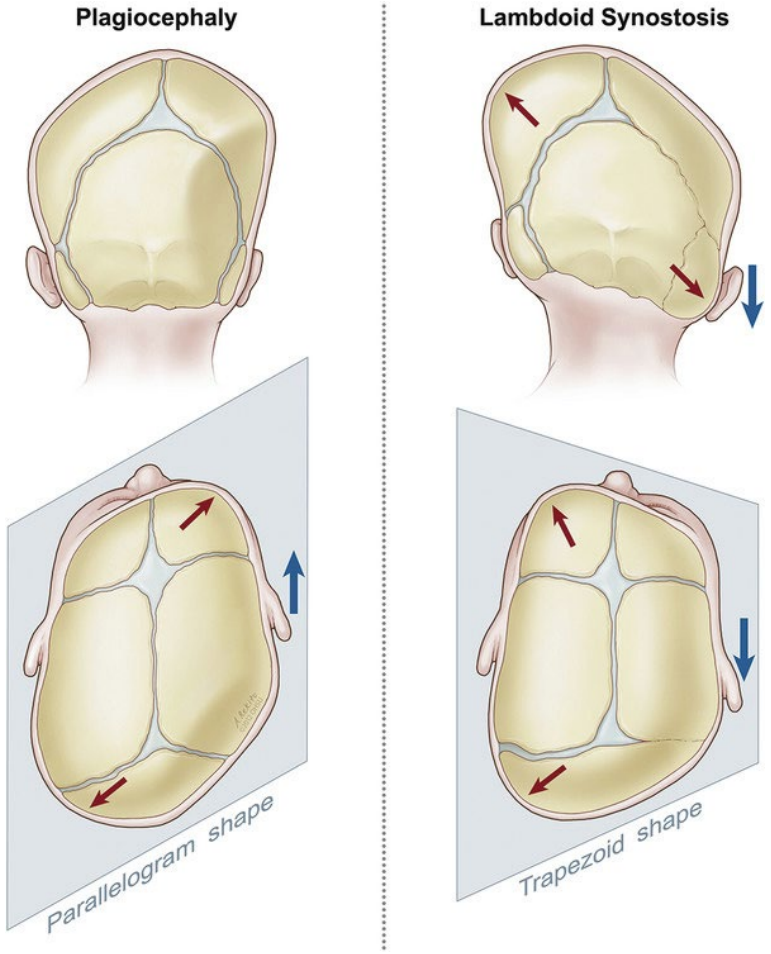
- Least common type
  - Accounts for less than 5% of cases
- Lambdoid Synostosis
  - Trapezoid shape
  - Posterior/inferior displacement of ipsilateral ear

Lambdoid synostosis





Positional Plagiocephaly



- Positional Plagiocephaly:
  - Parallelogram shape
  - Anterior ipsilateral ear displacement
  - Anterior ipsilateral frontal bossing

Positional Plagiocephaly

- Joint Guidelines Committee of the American Association of Neurological Surgeons (AANS) and the Congress of Neurological Surgeons (CNS) and American Academy of Pediatrics (AAP)
  - 2016
- Imaging:
  - Clinical examination is recommended for the diagnosis of plagiocephaly, and imaging is rarely necessary, except in cases in which clinical diagnosis is equivocal.

Positional Plagiocephaly

- Joint Guidelines Committee of the American Association of Neurological Surgeons (AANS) and the Congress of Neurological Surgeons (CNS) and American Academy of Pediatrics (AAP)

- Repositioning:
  - Repositioning is an effective treatment for deformational plagiocephaly.
    - Although there is Level 1 evidence that PT is more effective.

Positional Plagiocephaly

- Joint Guidelines Committee of the American Association of Neurological Surgeons (AANS) and the Congress of Neurological Surgeons (CNS) and American Academy of Pediatrics (AAP)
- Physical Therapy:
  - Physical therapy is recommended over repositioning education alone for reducing prevalence of infantile positional plagiocephaly in infants 7 weeks of age (Level I).
  - Physical therapy is as effective for the treatment of positional plagiocephaly and recommended over the use of a positioning pillow in order to ensure a safe sleeping environment and comply with American Academy of Pediatrics recommendations (Level I).

Positional Plagiocephaly

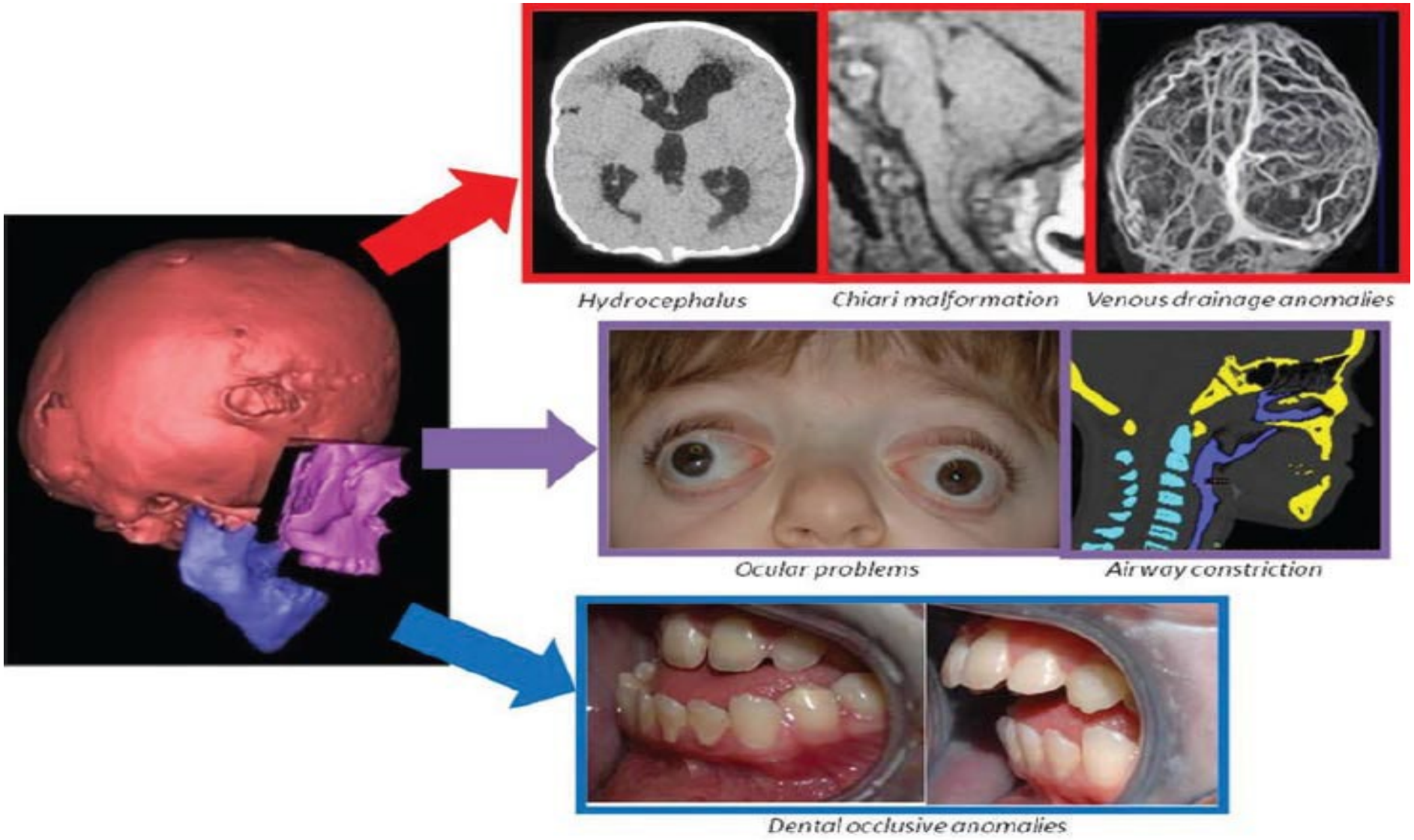
- Joint Guidelines Committee of the American Association of Neurological Surgeons (AANS) and the Congress of Neurological Surgeons (CNS) and American Academy of Pediatrics (AAP)

- Helmet Therapy:
  - Helmet therapy is recommended for infants with persistent moderate to severe plagiocephaly after a course of conservative treatment (Level II).
  - Helmet therapy is recommended for infants with moderate to severe plagiocephaly presenting at an advanced age (Level II).

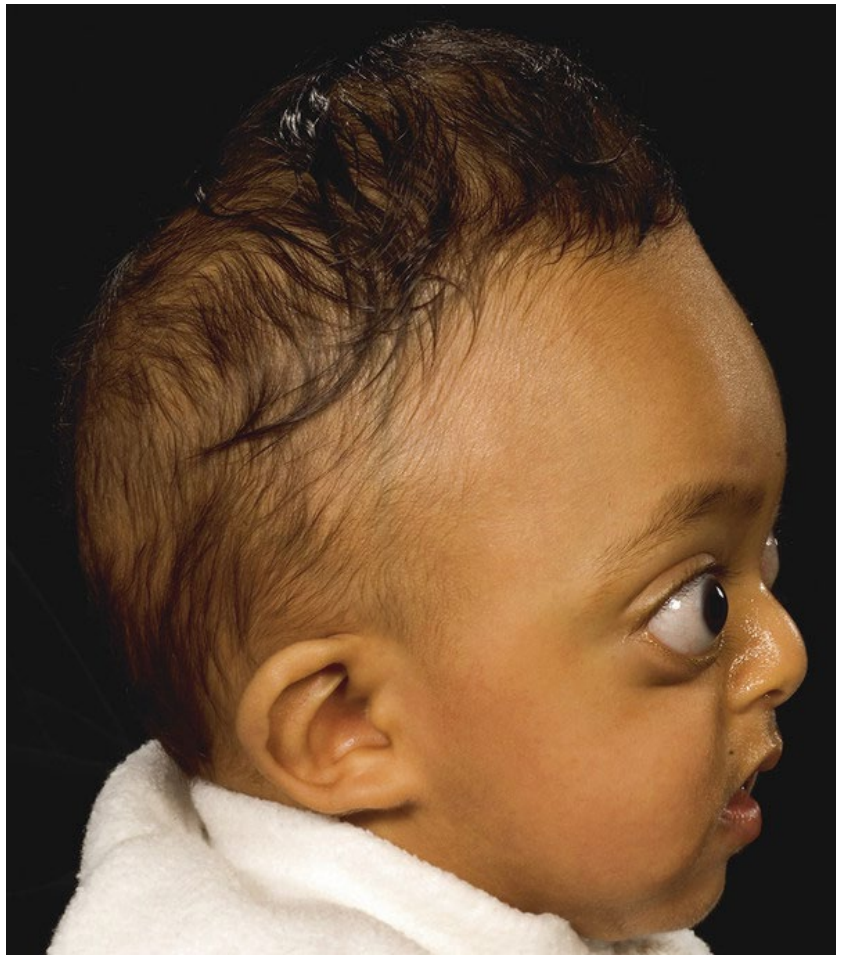
Syndromic Craniosyntosis

- Represents fewer than 5% of cases
- Raised ICP is common
- Can require more than one surgery
- Must have a team approach to treatment
  - Airway abnormalities
  - Cognitive delay
  - Orthopaedic abnormalities
  - Genetic abnormalities

Syndromic Craniosynostosis



Crouzan's Syndrome



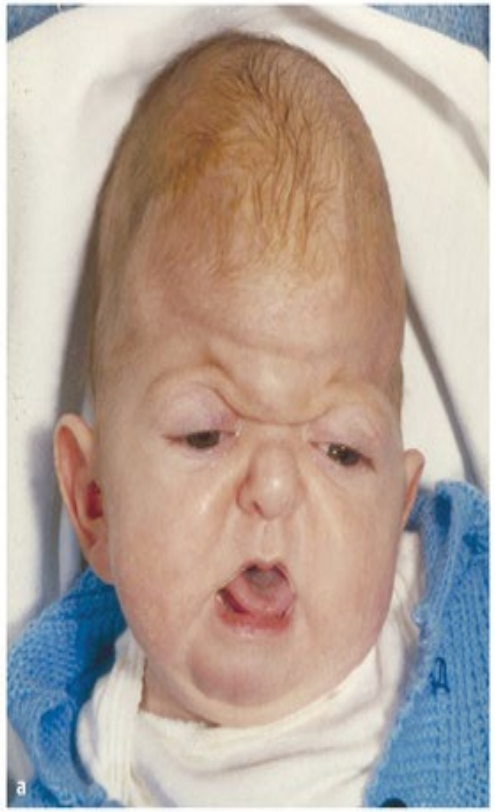
- First described in 1912
- Associated with autosomal dominant inheritance pattern
  - *FGFR2* gene
- Increased paternal age is a risk factor
- Normal intelligence
- 1.5 per 100,000 per year

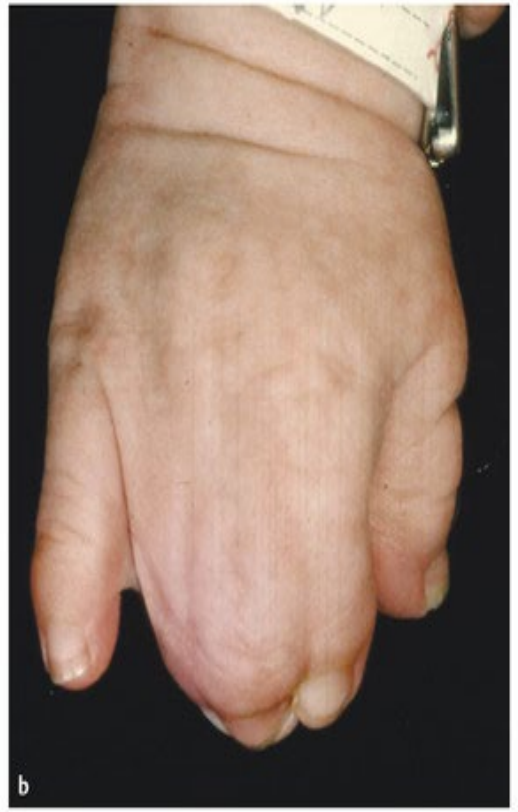
Crouzan's Syndrome

- a "beaky" nose
- a recessed frontal region due to bicoronal synostosis
- prominent eyes (exorbitism) due to the combined recession of the infra- and supraorbital regions
- retruded maxilla
- Extra-cranial findings:
  - Cervical vertebral fusion
  - Ankylosis affecting particularly the elbows
- Monitor for increased ICP.

Apert's Syndrome

- First described in 1906
- 1.5 per 100,000 per year
- Autosomal dominant inheritance
  - *FGFR2* gene
- Developmental and learning difficulties

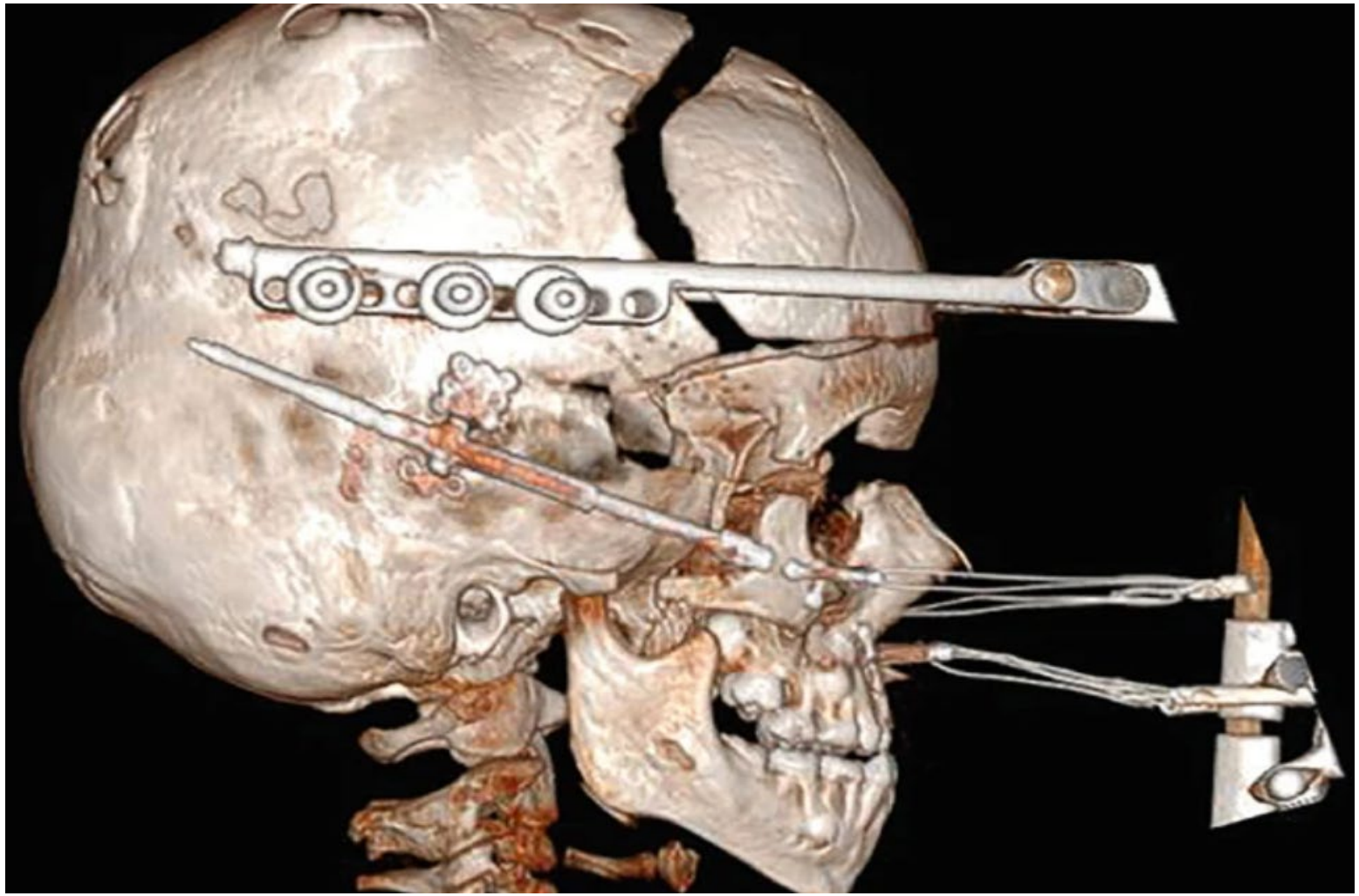




Apert's Syndrome

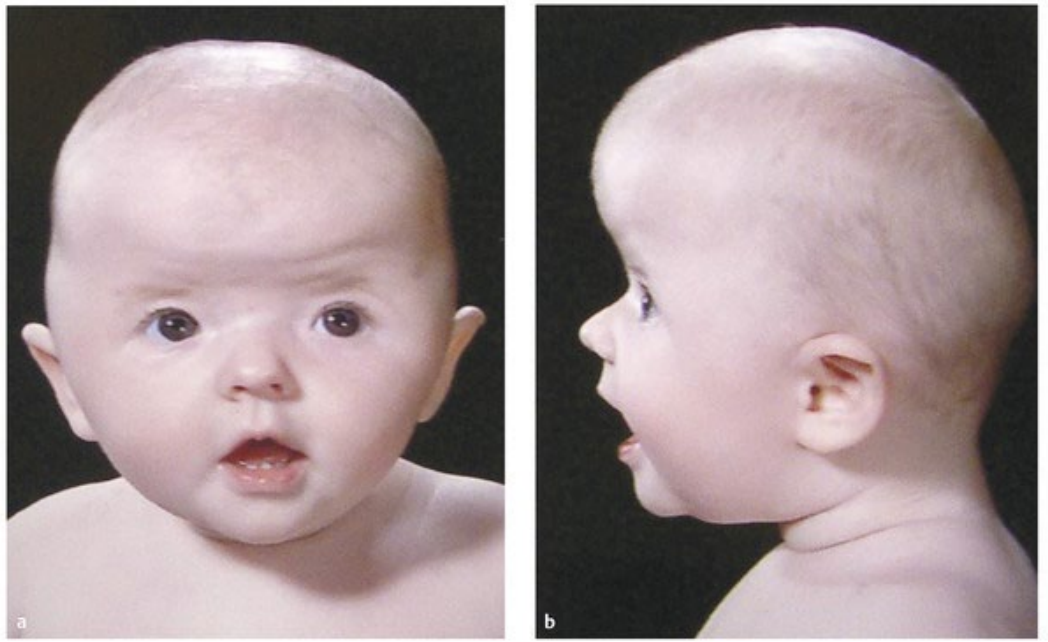
- Tall and shortened from front to back (turribrachycephaly)
- Midfacial (maxillary) retrusion
- Proptosis
- Downward cant to the palpebral fissures
- Hypertelorism
- Extra-cranial findings:
  - Syndactyly (fingers and toes)
  - Visceral and cutaneous abnormalities
  - Cervical spine fusion
- Initially just the coronal sutures are fused, but by 2 years of age all the sutures are fused.

Apert's Syndrome



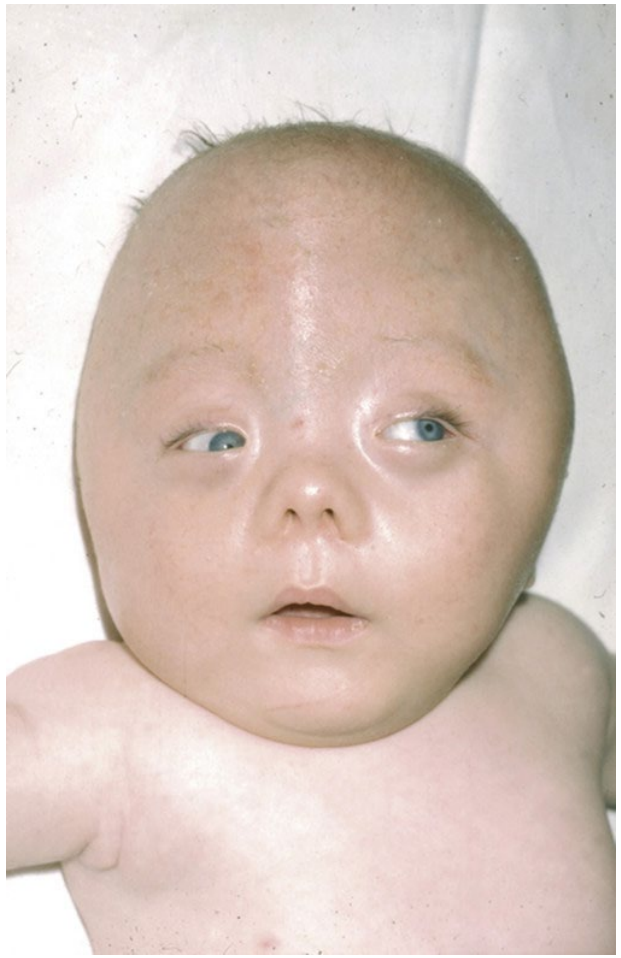
Muenke Syndrome

- 1 per 30,000 per year
- Autosomal dominant inheritance
  - *FGFR3 gene*
- Synostosis typically affects either one or both coronal sutures
- Raised ICP is uncommon
- Learning difficulty is not uncommon



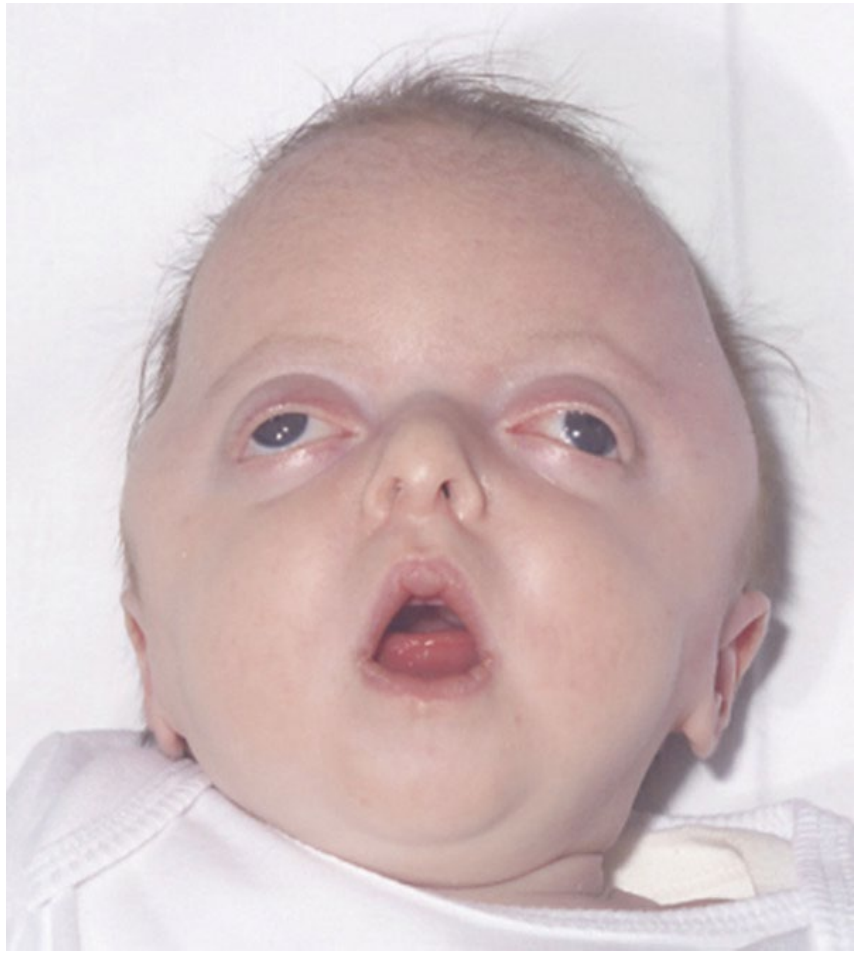
Pfeiffer Syndrome

- 1 per 100,000 per year
- Autosomal dominant inheritance
  - *FGFR1* & *FGFR2* gene
- Suture fusions that range from bicoronal synostosis alone to pan-synostosis
- Digital abnormalities
  - Usually shortened curved thumbs and great toes



- Pfeiffer Type I
  - Bicoronal synostosis
  - Midface retrusion
  - Digital abnormalities
  - Can have unaffected neurocognitive development

- Pfeiffer Type II and III
  - Severe midface and frontal retrusion
    - Airway obstruction
    - Ocular protrusion (corneal damage)
  - shortening of the skull base and crowding of the posterior fossa (lamboid synostosis)
    - Increased risk of hydrocephalus



Thank you

Ochsner Children's