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Positional Molding vs. Craniosynostosis

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Objectives

- Understand and be able to discuss the basic pathology underlying positional molding.
- Be able to discuss the circumstances and timing under which it is necessary and appropriate to refer for neurosurgical consultation.
- Be able to discuss treatment options for positional molding and when they should be employed.
- Understand and be able to discuss the basic pathologies underlying nonsyndromic and syndromic craniosynostosis.
- Be able to discuss the circumstances and timing under which it is necessary and appropriate to refer for neurosurgical consultation.
- Be able to discuss treatment options for syndromic and nonsyndromic craniosynostosis and when they should be employed.

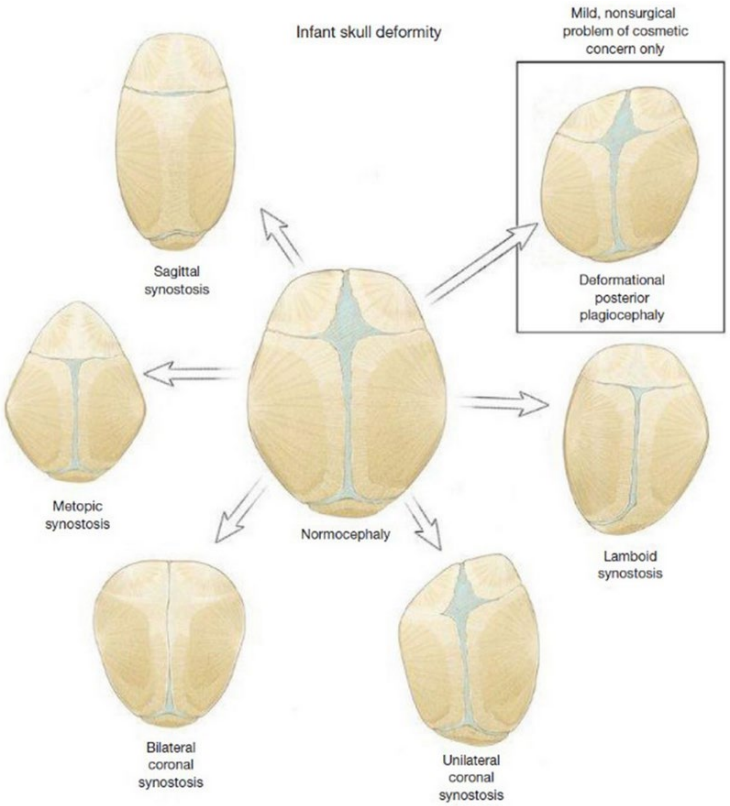
Abnormal Head Shape

- Abnormal head shape is due to the effects of either:

  Intrinsic factors, such as prematurely closed suture(s) restricting growth in a given direction

  or

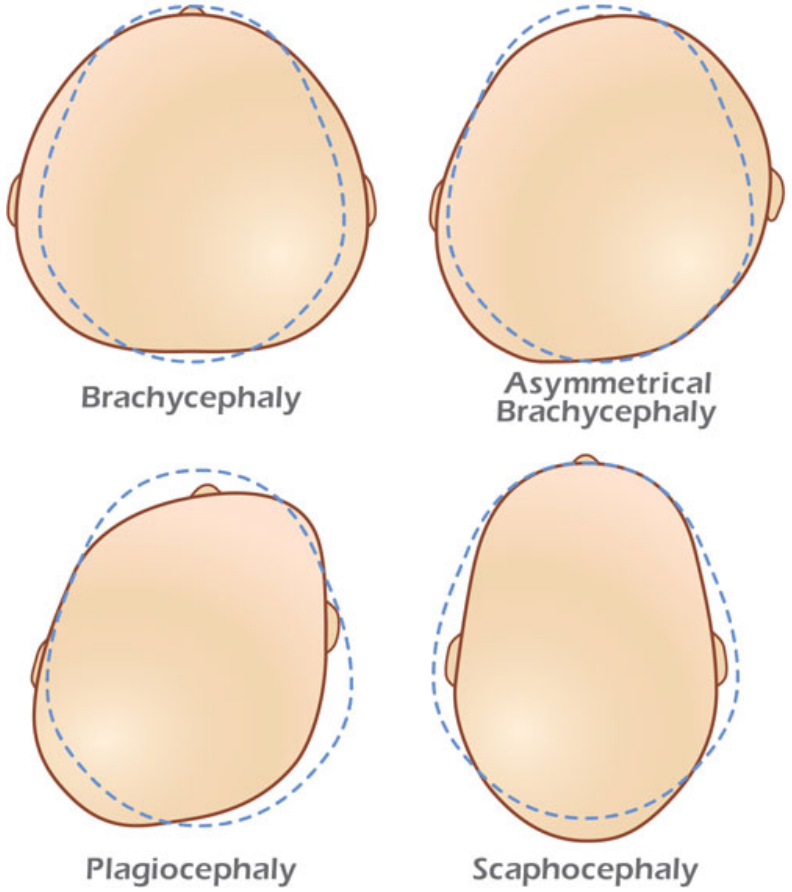
  Extrinsic factors, such as an infant lying on the same area of his or her head all the time



Modified from the cover of *Journal of Neurosurgery: Pediatrics* 2016;17(5). Original illustrator: Stacey Krumholtz. https://thejns.org/pediatrics/view/journals/j-neurosurg-pediatr/17/5/j-neurosurg-pediatr.17.issue-5.xml

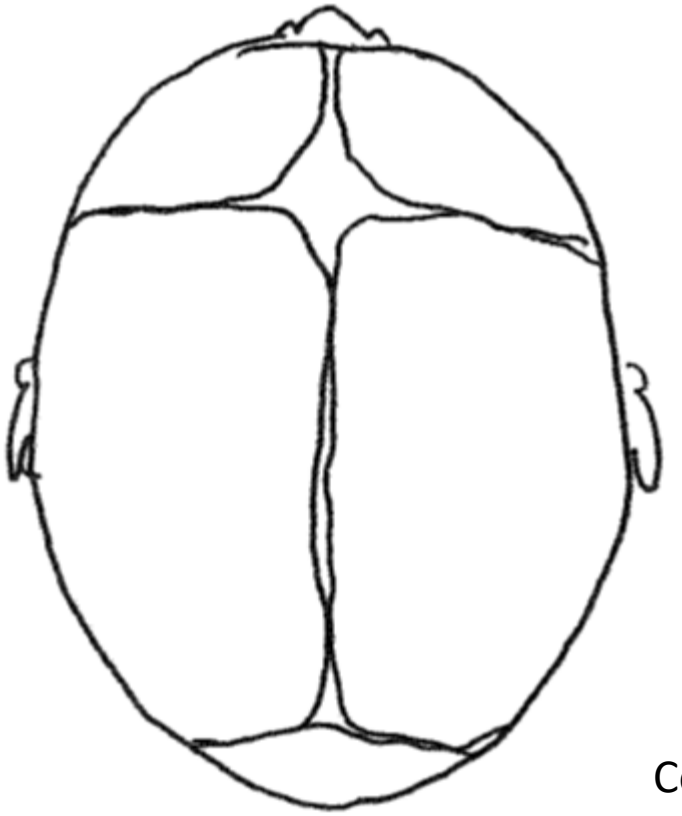
Positional Molding

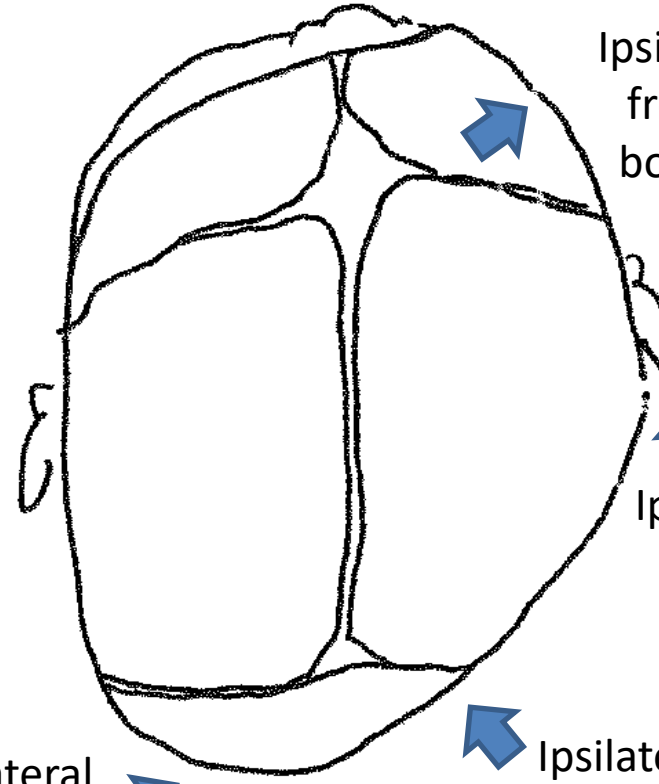
- *Plagiocephaly* – Asymmetric flattening of the occiput. With progressive deformity, ipsilateral anterior displacement of the ear and hemiface can be appreciated.
- *Brachycephaly* – Symmetric flattening of the occiput with shortening in the anterior-posterior diameter (APD) and widening of the biparietal diameter (BPD)
- *Dolichocephaly* – Elongation of the skull (increased APD) or biparietal narrowing that can be seen in NICU plagiocephaly and other benign conditions
- *Blending* of these head shapes can be seen.

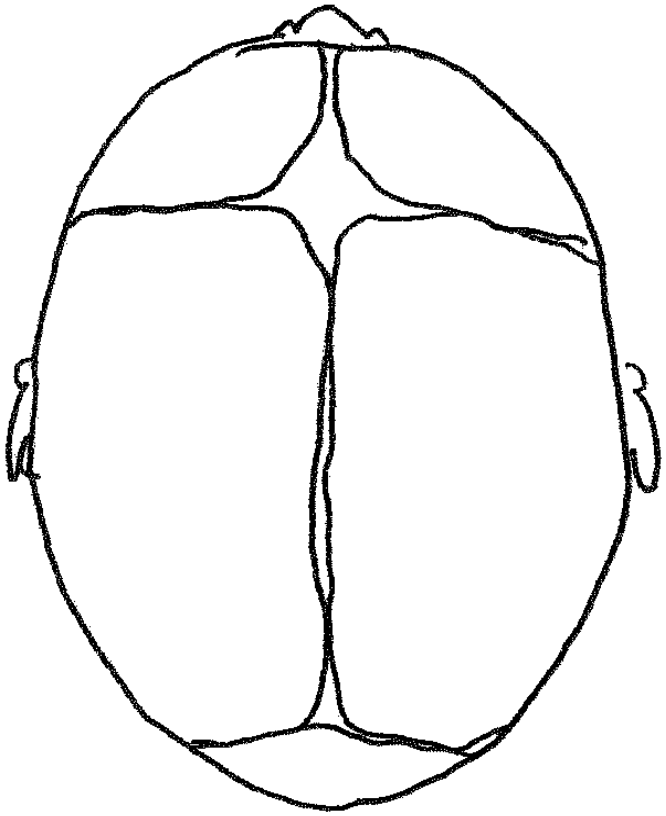


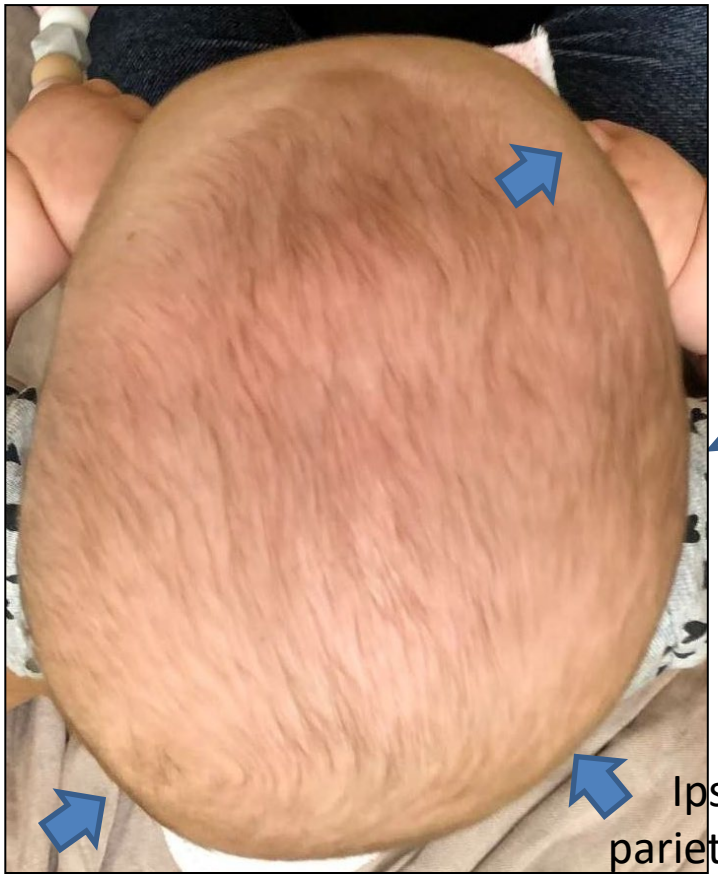
Positional Molding

- Epidemiology
  - Increased prevalence since “Safe Sleep Campaign” instituted in 1992
  - Subset with asymmetry at birth (13%–31%)
  - Most develop in first 2 months
  - Highest prevalence at 4 months
  - High association with torticollis
  - More often on the right side (54%–71%)
  - Risk factors
    - Primiparity, maternal age >35, breech position, prolonged labor and assisted vaginal delivery, oligohydramnios, cephalohematoma, male sex









Ipsilateral frontal bossing

Ipsilateral ear displaced anteriorly

Ipsilateral parieto-occipital flattening

Contralateral occipital bossing

Positional Molding

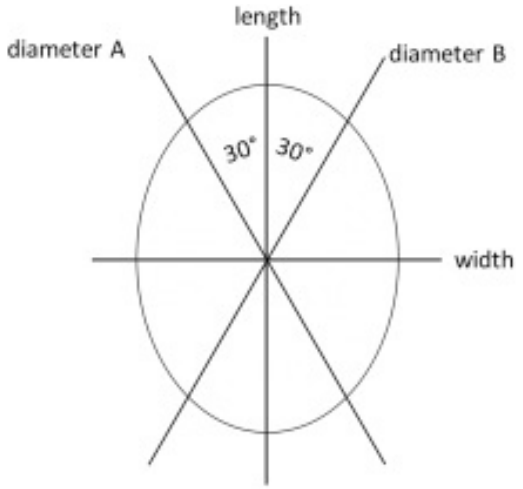
- Diagnosis made based on history and physical
  - Head shape usually normal at birth
  - Classic appearance
- If concern for synostosis, early referral to allow for time-dependent interventions
- Peaks at 4 months and often improves on own
  - Minimize with positioning program
  - Often do not helmet prior to 6 months given likelihood of natural resolution

Positional Molding

How is positional molding evaluated?

Standardized cranial measurements (anthropometry) are employed.

- Plagiocephaly:
  - Cranial Vault Asymmetry (CVA)
  - CVA Index (CVAI)
- Brachycephaly:
  - Cranial or Cephalic Index (CI)



$$\text{Cranial Vault Asymmetry (CVA)} = \text{longest diameter} - \text{shortest diameter}$$

$$\text{Cranial Vault Asymmetry Index (CVAI)} = \frac{\text{CVA}}{\text{shortest diameter}} \times 100$$

$$\text{Cranial (craniproportional) Index (CI)} = \frac{\text{width}}{\text{length}} \times 100$$

Positional Molding

- Helmet Therapy
  - Worn 23 hours per day for at least 3 months
  - Rarely see improvement if initiated after 1 year
    - 85% of head growth in first year
  - Expensive, potential lack of insurance reimbursement
  - Complications
    - Skin breakdown, pressure ulcers, contact dermatitis
  - In *mild* and *moderate* cases helmeting does NOT provide significant improvement in outcome compared to behavioral modification alone. (van Wijk RM, van Vlimmeren LA, Groothuis-Oudshoorn CG, Van der Ploeg CP, Ijzerman MJ, Boere-Boonekamp MM. Helmet therapy in infants with skull deformation: randomised controlled trial. *BMJ.* 2014;348:g2741)
  - Helmet therapy is recommended for infants with persistent *moderate* to *severe* plagiocephaly after a course of conservative treatment (repositioning and/or physical therapy). *Strength of Recommendation: Level II—uncertain clinical certainty.* (Tamber MS, Nikas D, Beier A, et al. Congress of Neurological Surgeons systematic review and evidence-based guideline on the role of cranial molding orthosis (helmet) therapy for patients with positional plagiocephaly. *Neurosurgery.* 2016;79[5]:E632–E633)

Lipira AB, Gordon S, Darvann TA, et al. Helmet versus active repositioning for plagiocephaly: a three-dimensional analysis. *Pediatrics.* 2010;126(4):e936–e945

Positional Molding

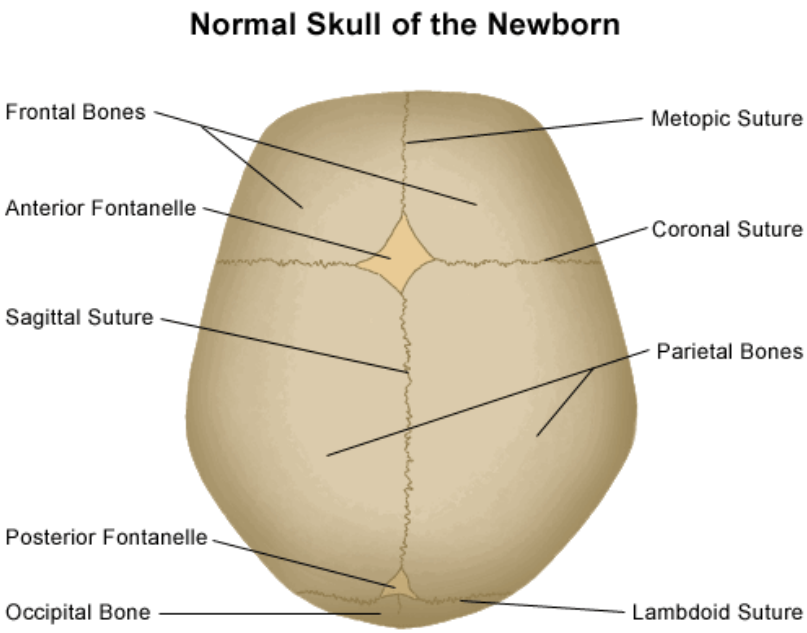
- Take home
  - Parental education and repositioning program
  - Supine to sleep, but tummy time while awake
  - If asymmetry +/- torticollis, recommend repositioning, physical therapy, and stretching)
  - If *severe* deformity (defined as a diagonal difference of >12 mm) at 6 months, will likely achieve greater improvement with helmet
  - Head shape is *unlikely* to affect cognitive development
  - Early referral if synostosis is suspected (the earlier the better)

If unsure, please refer!

Craniosynostosis

- The calvarium (skull) is comprised of 8 major bones (paired frontal, parietal, occipital, and temporosphenoidal) which are separated by fibrous joints called sutures.

- Craniosynostosis is a condition in which 1 or more sutures close prematurely resulting in problems with normal skull growth.



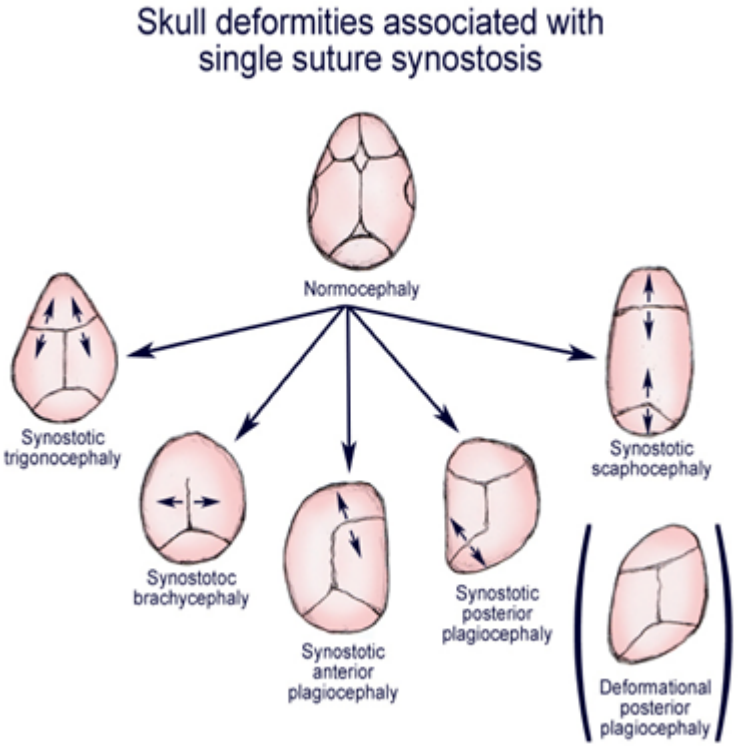
Craniosynostosis

- Sutures close in a sequential time-dependent manner.
  - Metopic 3–8 months
  - Sagittal 22 years
  - Coronal 24 years
  - Lambdoid 26 years
  - Squamosal 60 years

- Growth velocity of the skull changes with age.
  - The head typically doubles in size in the first 6–9 months, with another doubling by 2 years.

Craniosynostosis

- Normal skull growth is perpendicular to the suture.
  - Understanding this principle can predict the site and shape of the skull deformity associated with premature sutural closure (Virchow's hypothesis).
- Craniosynostosis can adversely impact the intracranial pressure, developmental delay, and visual disturbances in some instances.



Craniosynostosis

- Epidemiology
  - Incidence is 1 in 2,000–2,500 live births.
  - Single suture, nonsyndromic synostosis accounts for up to 95% of all cases.
  - Most cases of nonsyndromic craniosynostosis are thought to be sporadic but the following have been implicated to some degree:
    - Metabolic (hyperthyroidism) or hematologic syndromes
    - Clomiphene citrate, valproic acid, phenytoin and retinoic acid
    - Maternal smoking
    - Advanced maternal age
    - >100 mutations, including those affecting *FGFR1-3, NELL1, MSX2, TWIST,* and *GLI3*

Craniosynostosis

- Head shape abnormality is usually present at birth (most synostosis cases occur in utero).
- The diagnosis of craniofacial deformity is primarily based upon physical examination.
- Radiographic studies are best performed at a center with a craniofacial team.
- 3D computed tomography (CT) scan imaging
  - Confirm diagnosis and rule out mimics
  - Surgical planning
  - Very low dose CT
    - 85% reduction in radiation dose compared to CT

Craniosynostosis

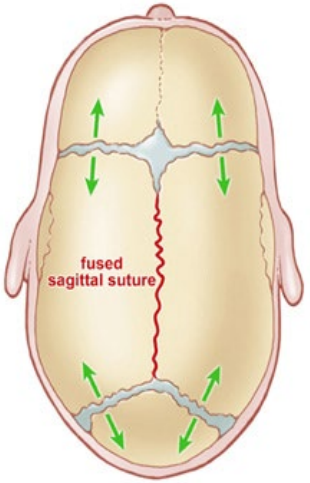
- Treatment
  - Goal of treatment is complete release of fused suture and correction of compensatory skull changes.
  - Timing of surgery is important to the outcome.
    - Early surgery utilizes the rapid brain growth potential for skull remodeling.
      - Birth = 400g ➔ 6 months = 800g ➔ 2.5 years = 1600g

Craniosynostosis

- Surgical techniques
  - Endoscopic/minimally invasive
    - Done early—hence, timely referral is key (average age at surgery is 3 months; may be considered up to 6 months of age)
    - Takes advantage of the rapid brain/skull growth in this age group
    - Less blood loss, shorter operating room and hospital stay, lower cost, and smaller incisions
    - Requires springs or postoperative helmeting (for up to 1 year)
    - Limited data for nonsagittal suture synostosis (Yan H, Abel TJ, Alotaibi NM, et al. A systematic review of endoscopic versus open treatment of craniosynostosis. Part 2: the nonsagittal single sutures. *J Neurosurg Pediatr.* 2018;22[4]:361–368)
  - Open
    - Performed at 6–12 months of age (or later if child is older at diagnosis)
    - More blood loss, longer operating room and hospital stay, bigger incision but immediate correction, and no postoperative molding orthosis
  - Long-term prospective registries needed to establish timing and intervention type with respect to complications, cost, and equivalent cosmetic outcomes (Yan H, Abel TJ, Alotaibi NM, Yan, et al. A systematic review and meta-analysis of endoscopic versus open treatment of craniosynostosis. Part 1: the sagittal suture. *J Neurosurg Pediatr.* 2018;22[4]:352–360)

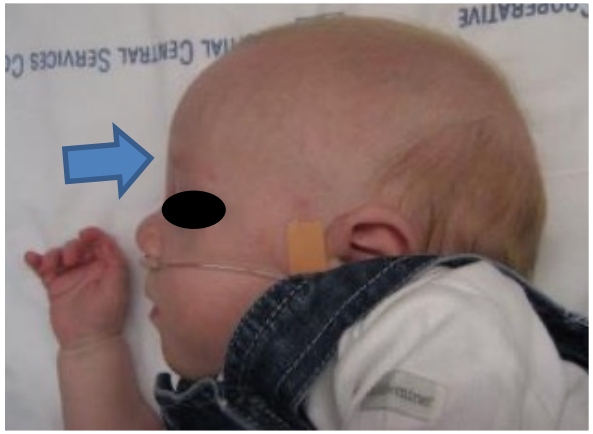
Sagittal Craniosynostosis

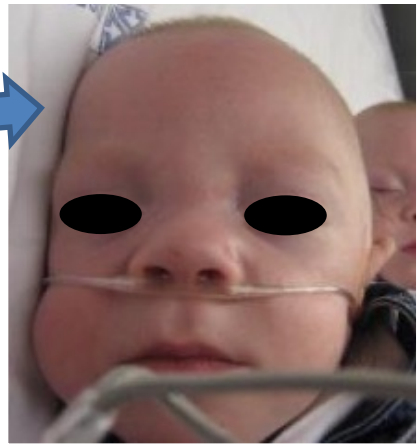
- Scaphocephaly
  - Most common form of single suture synostosis
    - 50%–60% of craniosynostosis cases
  - Characterized by a narrow, elongated skull +/- frontal and/or occipital bossing
  - Can be associated with intracranial hypertension if left untreated



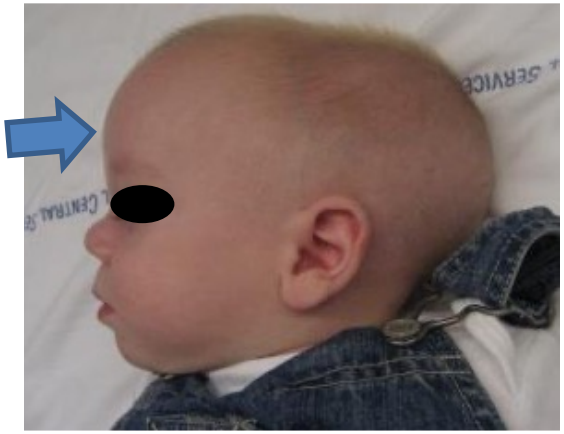
Dolichocephaly

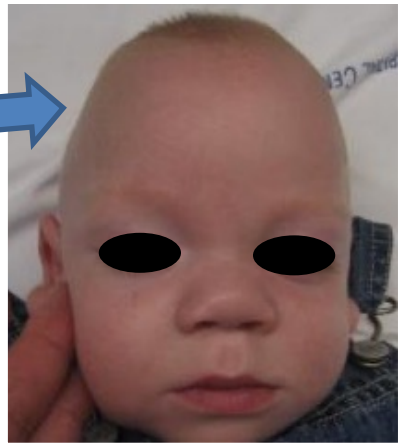
Modest elongated AP dimension with biparietal narrowing *without* frank frontal bossing or occipital bulging





Scaphocephaly





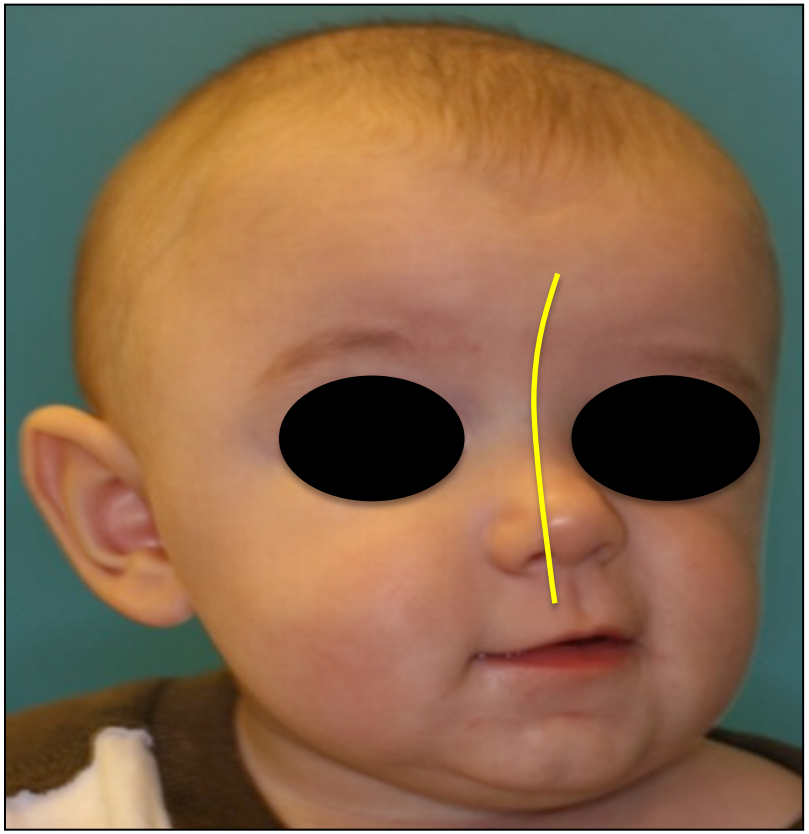
Elongated AP dimension with frontal bossing, occipital bulleting, and saddle deformity at vertex

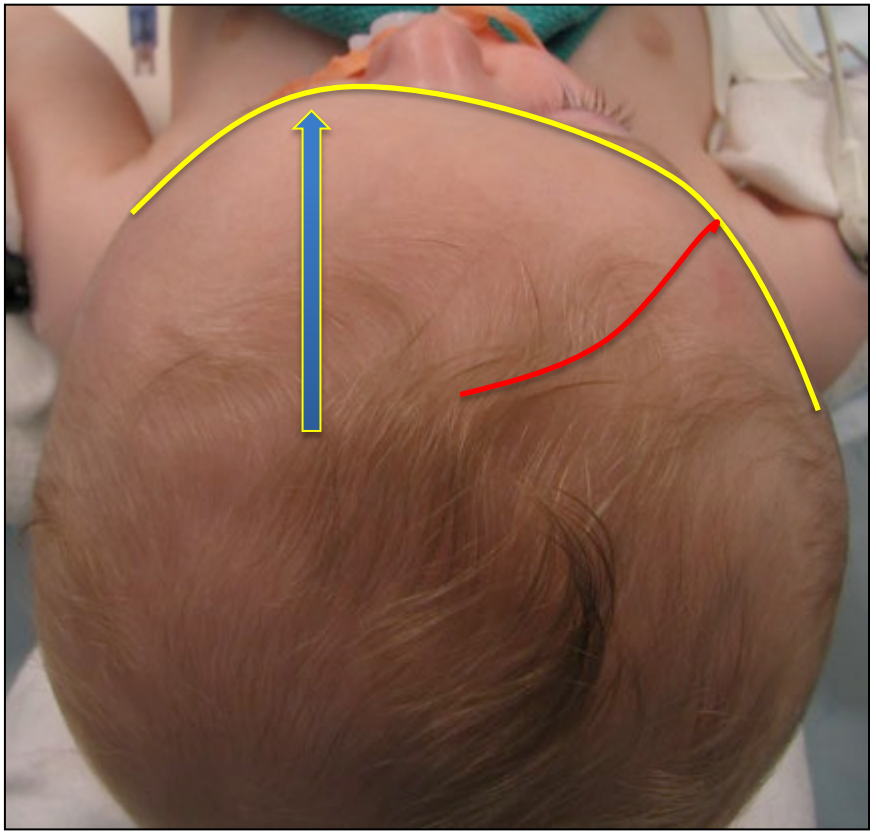
Dias MS, Samson T, Rizk EB, Governale LS, Richtsmeier JT, AAP Section on Neurologic Surgery, and Section on Plastic and Reconstructive Surgery. Identifying the misshapen head: craniosynostosis and related disorders. *Pediatrics.* 2020;146(3):e2020015511

Coronal Craniosynostosis

- Anterior Plagiocephaly
  - 15%–30% of synostosis cases
  - Unilateral coronal synostosis >> Bilateral coronal synostosis
  - Unilateral coronal synostosis
    - Flattening of the forehead on the affected side
    - Vertically elongated eye on the affected side—called a “Harlequin Eye”
    - Nose deviated to contralateral side
    - Ear displaced anteriorly on the affected side
    - Strabismus is seen in ~30% of cases
    - Can be associated with intracranial hypertension if left untreated

Coronal Craniosynostosis





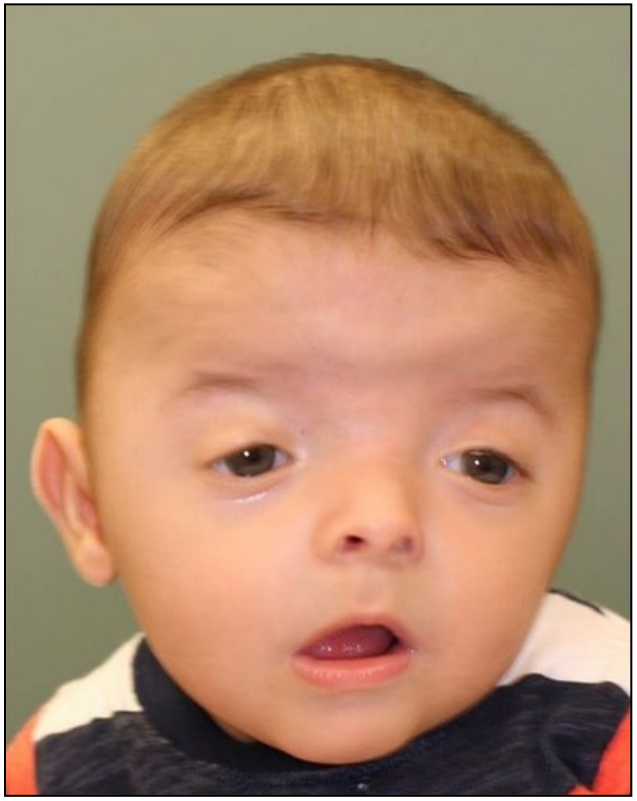
Fronto-orbital retrusion and elevated ipsilateral orbit (Harlequin Eye), ipsilateral nasal root and contralateral nasal tip deviation, and anterior displacement of ipsilateral ear

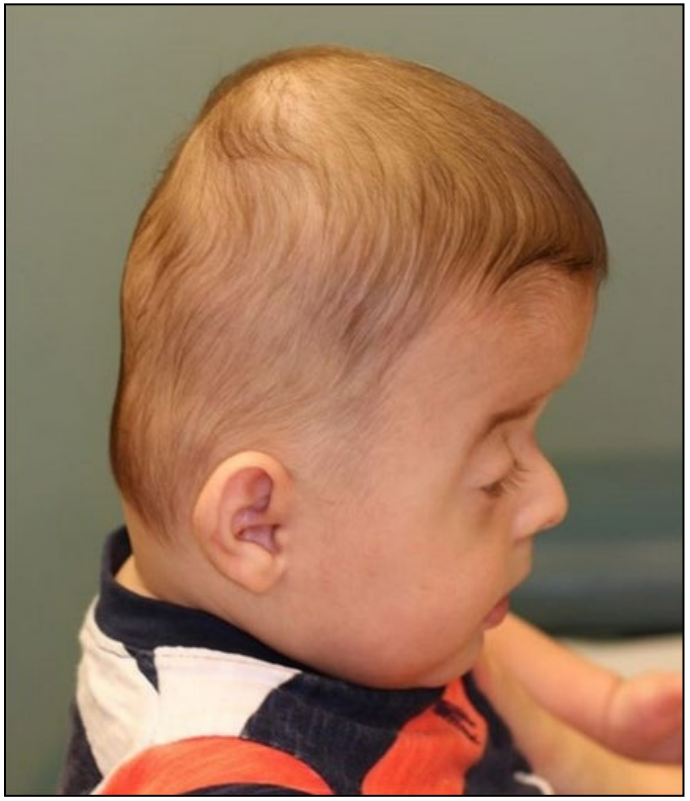
Dias MS, Samson T, Rizk EB, Governale LS, Richtsmeier JT, AAP Section on Neurologic Surgery, and Section on Plastic and Reconstructive Surgery. Identifying the misshapen head: craniosynostosis and related disorders. *Pediatrics.* 2020;146(3):e2020015511

Coronal Craniosynostosis

- Brachycephaly
  - 5%–10% of coronal synostosis cases
  - Bilateral coronal synostosis (turribrachycephaly)
    - Biparietal widening with AP shortening (brachycephaly)
    - Frontal towering (turricephaly)
    - Flattening of the occiput and the caudal frontal bone with bulging of the cephalic frontal bone can be seen
    - Bilateral Harlequin Eyes may be observed radiographically
  - Ridging of the coronal (or any) sutures without the skull deformity associated with synostosis of that suture is likely benign overriding of cranial bones commonly seen in infancy.

Coronal Craniosynostosis





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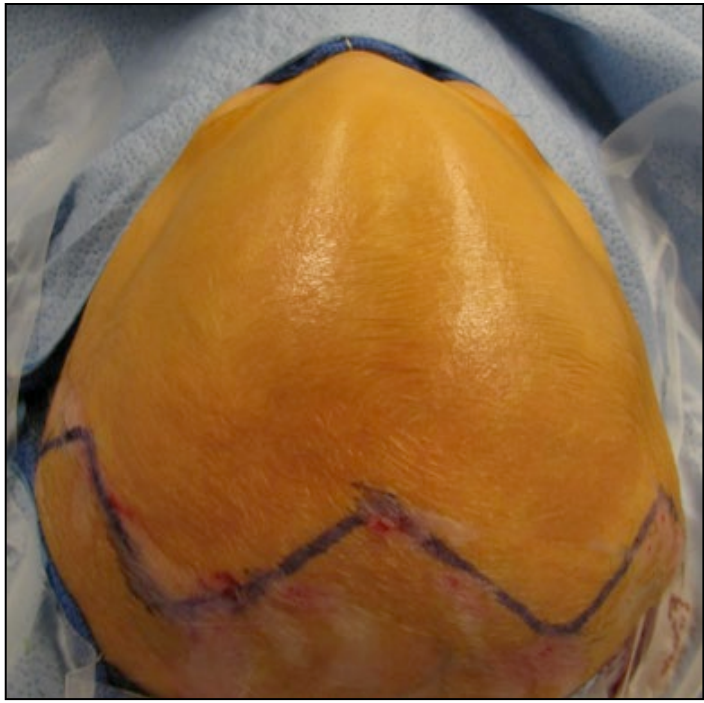
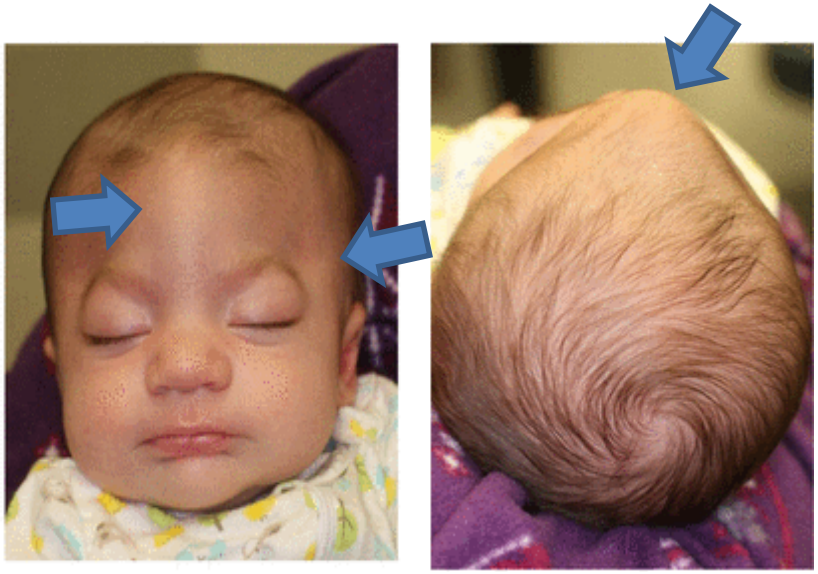
Metopic Craniosynostosis

- Trigonocephaly
  - 4%–10% of synostosis cases
  - Metopic synostosis
    - Triangular wedging of the frontal bones
    - Hypotelorism with inter-orbital and bitemporal narrowing with compensatory prominence of the parietal bosses
    - Posterior displacement of the superolateral orbital rims
    - Can be associated with intracranial hypertension if left untreated
    - Must be differentiated from benign metopic ridging, characterized by a prominence along the closed metopic suture (typically superior/adjacent to the anterior fontanelle) without trigonocephaly



Metopic synostosis



Pointed forehead, hypotelorism, and temporal retrusion

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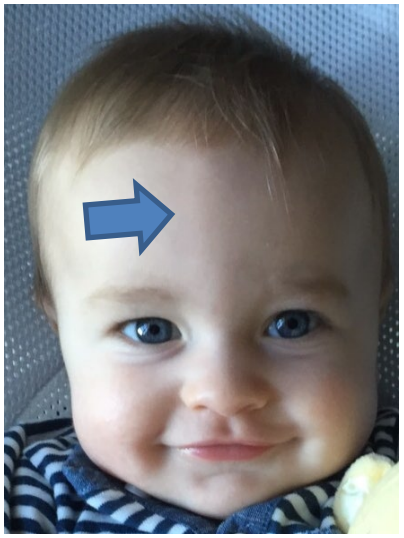
Metopic ridge

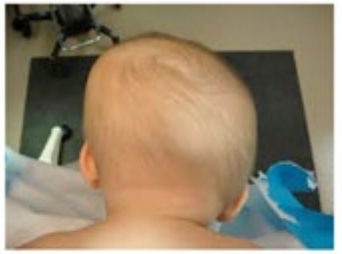


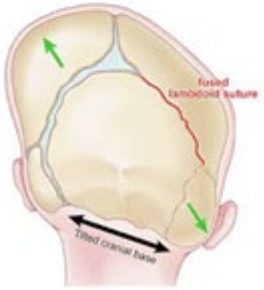
Rounded forehead, normally spaced eyes, and a prominence of the metopic ridge

Courtesy of Elias Rizk, MD, MSc, FAANS

Lambdoid Craniosynostosis

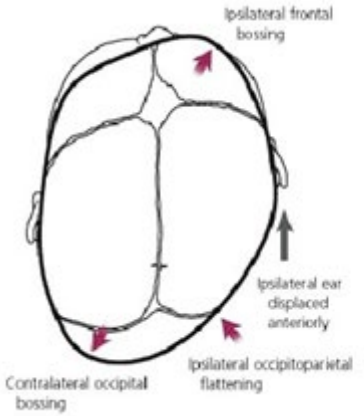
- Posterior Plagiocephaly
  - <2% of all cases
  - Unilateral lambdoid synostosis
    - Trapezoid shape with flattening of the frontal and occipital prominences on the affected side
    - Contralateral frontal and parietal bossing
    - Posterior and inferior displacement of the ear on the affected side
    - Must be differentiated from the far more common non-synostotic acquired positional molding previously discussed
    - Can be associated with intracranial hypertension if left untreated

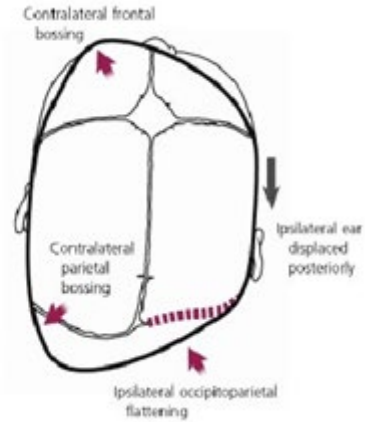




Parallelogram
Deformational plagiocephaly

Trapezoid
Lambdoid synostosis





Craniosynostosis

- Syndromic Craniosynostoses
  - <5% of craniosynostosis cases
  - Broad spectrum of genetic variability within a syndrome
  - Typically have much more pronounced facial dysmorphism
  - Crouzon, Apert, Pfeiffer, Saethre-Chotzen syndrome, Muenke syndrome, Carpenter syndrome, etc.

Questions?

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