Head Shape Evaluation in Infants and Young Children

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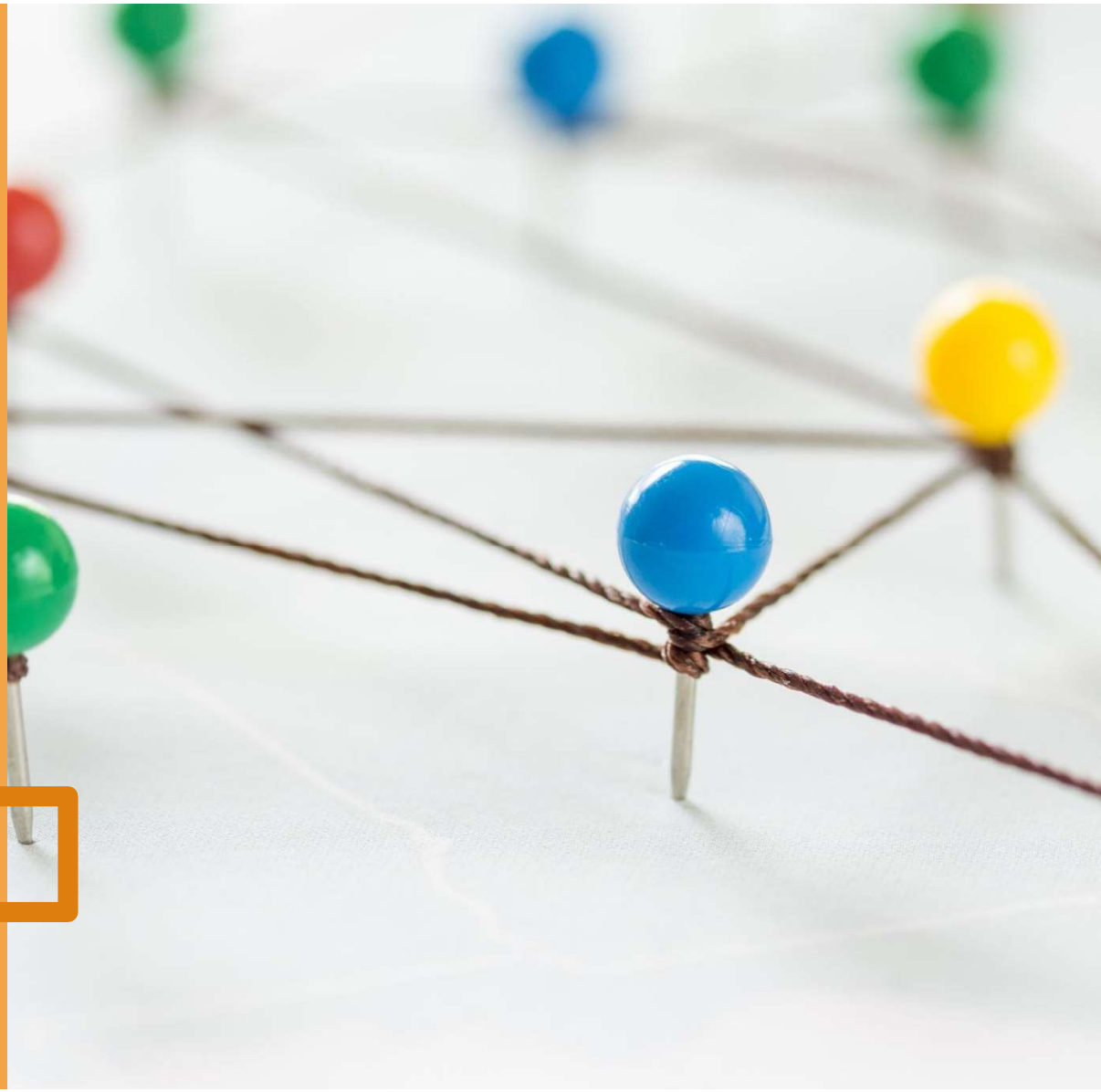

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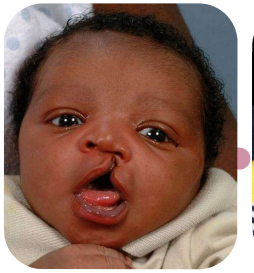
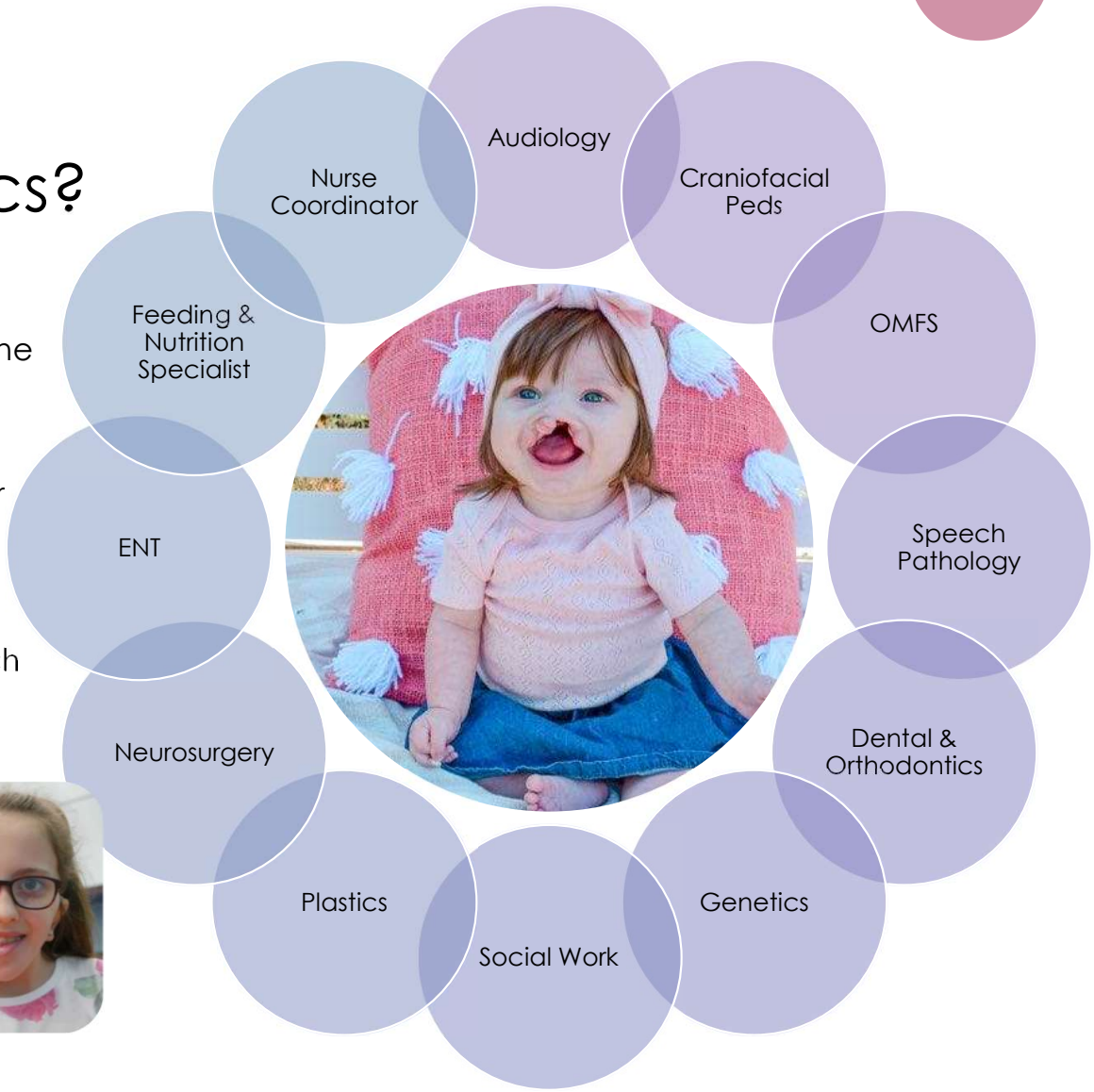
Craniofacial Pediatrics?

- Complex Care Pediatrics that specializes in the non-surgical & medical management of children born with craniofacial conditions or anomalies.
  - Provide a medical home at CHNOLA for these complex patients
  - Supplement to their primary care pediatrician in the community
- Patient and family centered holistic approach with the large multi-disciplinary team



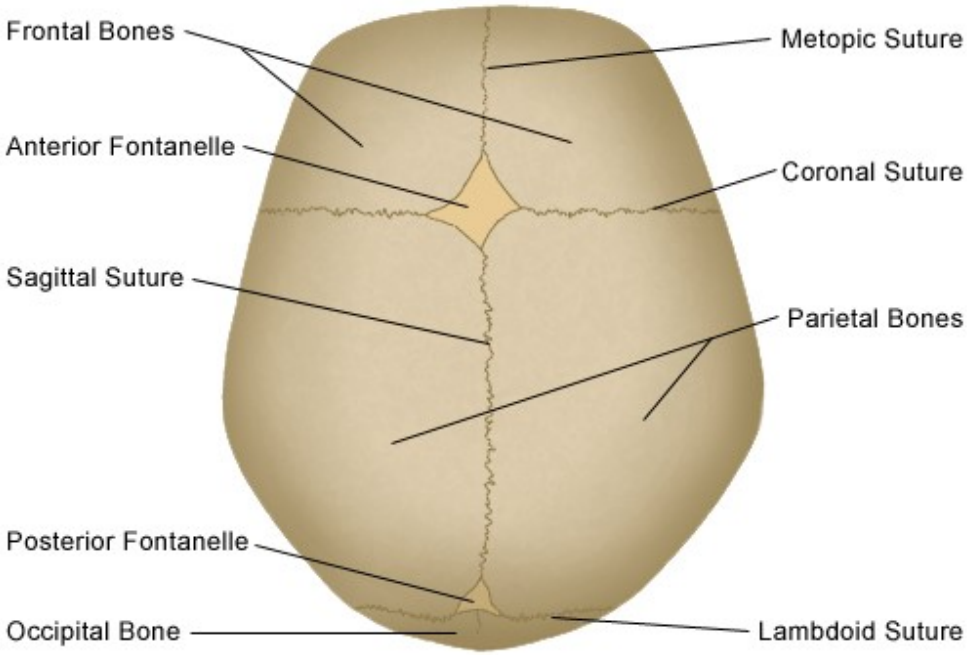


Head Shape Objectives

- Perform and document a systematic head shape exam
- Identify classic head shapes for plagiocephaly and single suture craniosynostosis
- Recommend appropriate and timely treatment of common head shape anomalies

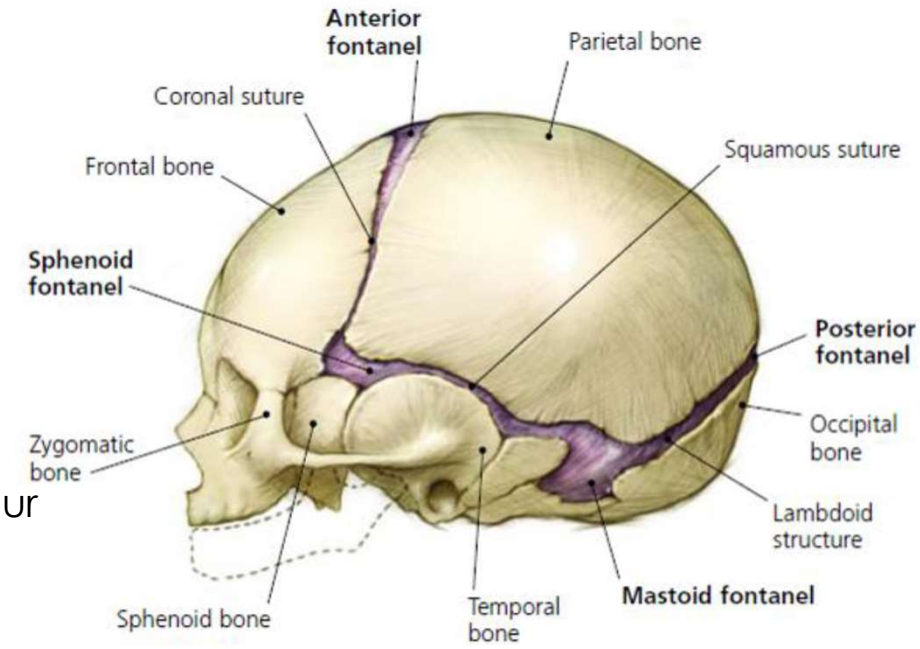
Normal Skull Anatomy and Growth

- Skull made of 5 major bones
  - Separated by 6 major sutures
  - Fontanelles are suture junctions
- Sutures allow for:
  - Deformation of the skull during birth
  - Rapid growth of the skull
- Skull growth happens in response to increasing brain and CSF volume
- Brain triples by one year and is 85% of adult size by 3yo



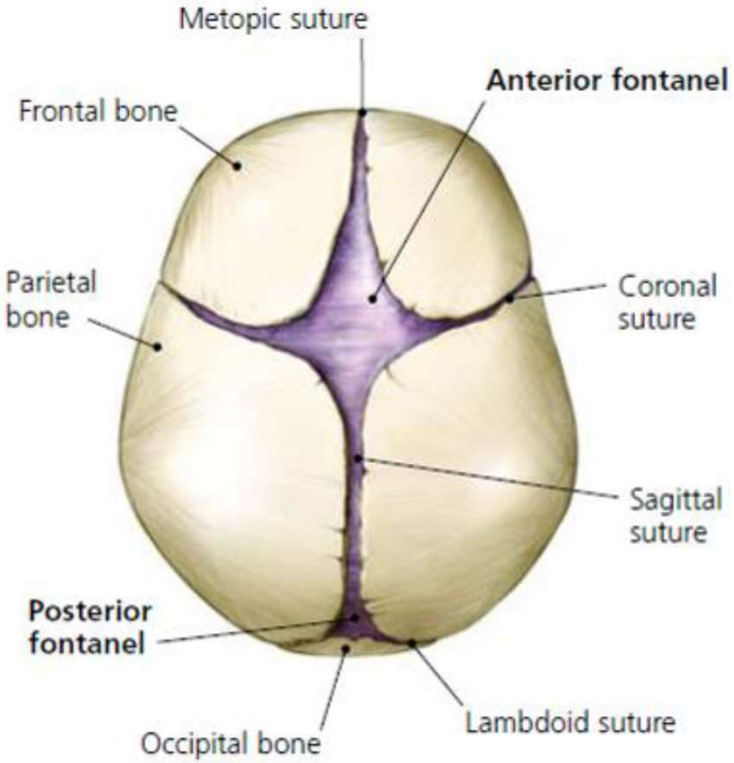
Normal Fontanelle Closure

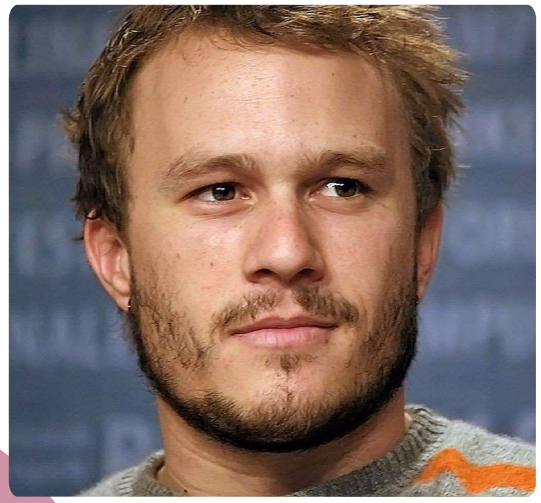
- Anterior fontanelle typical timing
  - Around 9-15 months of age
  - Range from 3-26 months
- Posterior fontanelle typical timing
  - By 2 months
  - Can be difficult to find in newborns at times
- Closed fontanelle means you can't feel the space any more
  - Doesn't mean the sutures are closed
  - Doesn't mean that no more growth can occur



Normal Fontanelle Closure

- Normal: head growth, head shape, development
  - I am not concerned for craniosynostosis
- Timing of fontanelle closure can be associated with medical conditions
  - Early Closure
    - Hyperthyroidism
    - Hyperparathyroidism
    - Hypophosphatasia
    - Rickets
  - Delayed closure or abnormally large fontanelle
    - Hypothyroidism
    - Malnutrition
    - Trisomy 21
    - Hydrocephalus





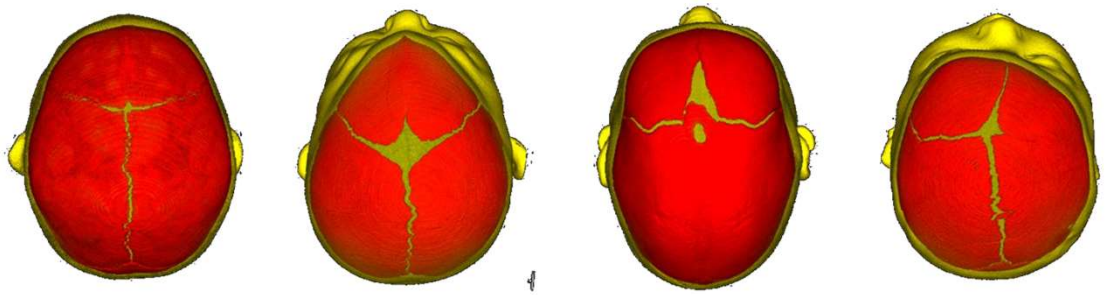
Normal Suture Closure

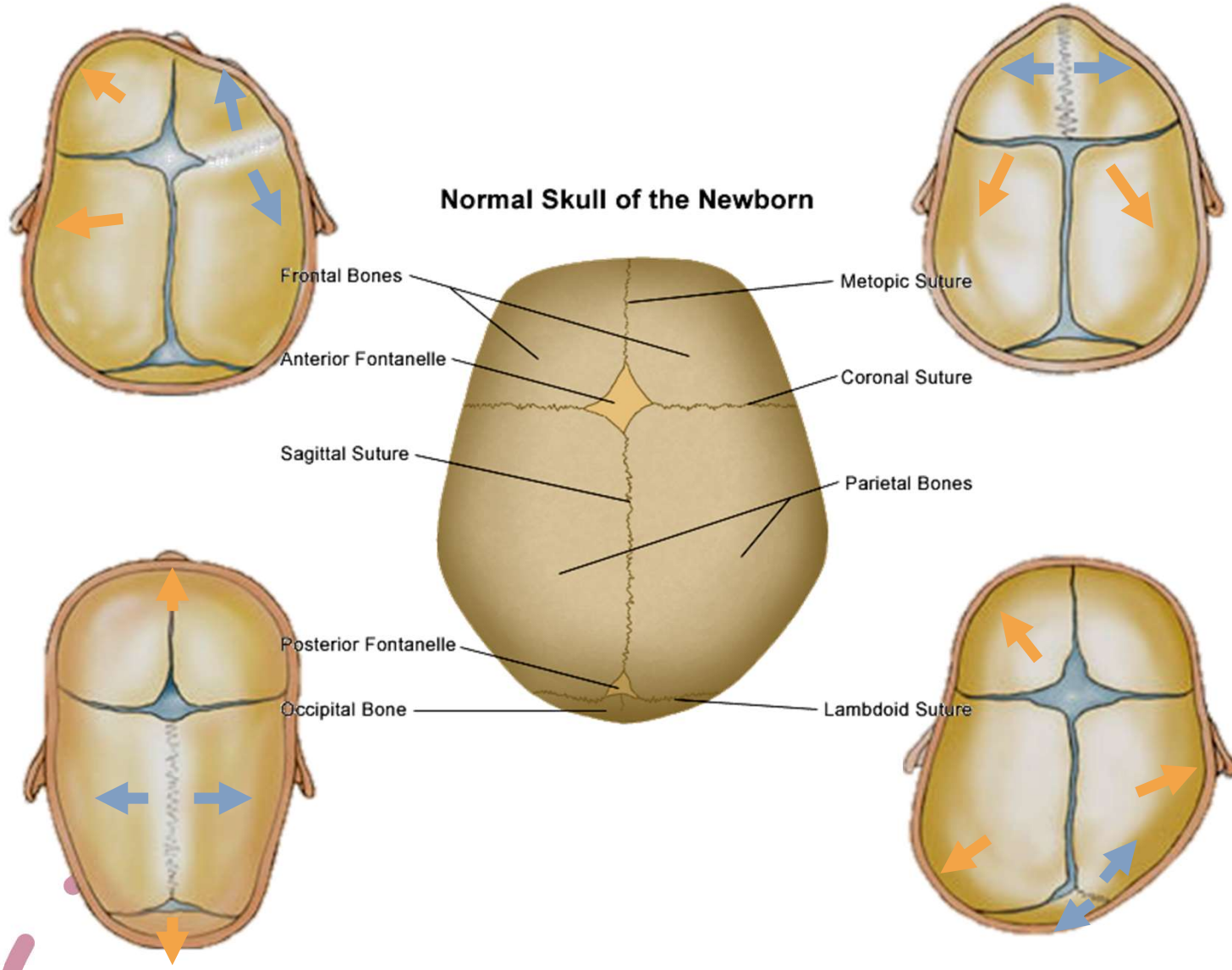
- Metopic
  - 3-9 months
    - At times even 2 months
  - Commonly forms a palpable or visible ridge
    - Benign normal variant if they have normal headshape
- Sagittal
  - 22 years
- Coronal
  - 24 years
- Lambdoid
  - 26 years

Craniosynostosis

- Early fusion of cranial sutures
  - Typically happens in utero
- Incidence: ~1:2500 births
- ~97% of craniosynostosis cases
  - Single suture
- 85% are non-syndromic
- 6-10% are familial

- Increased Risk of:
  - ↑ intracranial pressure
    - Single suture 4-42%
    - Multi-suture 50-68%
  - language delay
  - learning disability
    - Single suture 10-30%
  - other developmental delays





Mechanism of Skull Growth

Craniosynostosis results in the limitation of skull growth perpendicular to the area of fusion and compensatory overgrowth in other sutures

How to approach a misshapen head

Head Shape History

Birth History

- Gestational Age
- Maternal Gs & Ps
- Route of Delivery
  - Reason for c-section
- Prenatal Concerns:
  - Constraint
  - Breech
  - Oligohydramnios
  - Multiples
- Maternal Health & Medications
  - Diabetes: gestational vs. Type I/II
  - Thyroid Disease
  - Tobacco use

Why it matters

- Premature infants ↑ risk of deformational plagiocephaly or brachycephaly
  - Also with ↑ risk of vitamin D deficiency
- CS has ↑ incidence of unplanned c-section
- Risk factors for Craniosynostosis:
  - Intrauterine head constraint
  - Twin+ pregnancy
  - Maternal smoking
  - Maternal thyroid disease
- Breech skull molding can mimic sagittal craniosynostosis

Head Shape History

Specific Head Shape Questions

- Head shape at birth
- First notice a difference
- Changes over time
- Sleep position
- Head tilt or Torticollis
  - Changes over time
- Developmental history

Why it matters

- CS present at birth
  - Typical newborn molding improves with time
  - Craniosynostosis doesn't improve
- Plagio not present at birth
  - May worsen or improve over time
- Sleep position typically dictates plagiocephaly
- Head tilt and torticollis make any headshape worse
- Delayed motor milestones increase risk for plagiocephaly

Systematic Head Shape Exam

PHYSICAL EXAMINATION of the HEAD:

Patient: _____ Date: ___/___/___

Examiner: _____

Weight _____ kg, ____ to ____ percentile

Height _____ cm, ____ to _____ percentile

OFC _____ cm, ____ to _____ percentile (Nellhaus curve)

General: active alert engaging

Other: _____

Palpation:

Anterior fontanelle:

Size: Typical Small large

Shape: Diamond Triangular

Position: Typical Anteriorly placed Posteriorly placed

Quality: Flat Full

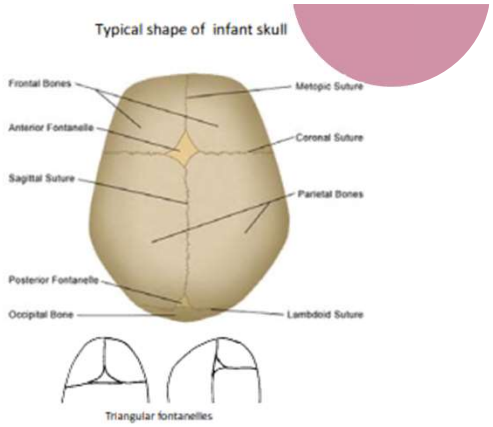
Soft Tense

Palpable ridge(s): Sagittal No Yes Mobile? Yes No

(if yes, side?) Coronal No Yes Right Left

Metopic No Yes

(if yes, side?) Lambdoid No Yes Right Left



Top down view:

Forehead shape: Typical Triangular Other: _____

Flattening/retrusion: No symmetric R L

Prominence: No symmetric R L

Occipital shape: Typical Other: _____

(if yes, side?) Flattening/retrusion No symmetric R L

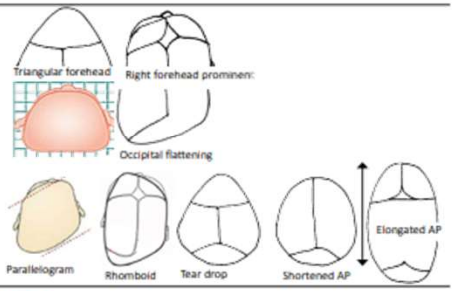
(if yes, side?) Prominence: No symmetric R L

Overall shape: Typical parallelogram rhomboid

Tear drop Long AP Short AP Brachy

Widest point: Bi-Parietal Bi-Temporal Other:____

Ear position (horizontal): Symmetric Right anterior Left



Side View:

AP diameter: Typical long short

Height: Typical Turricephalic Tall forehead Tall occiput

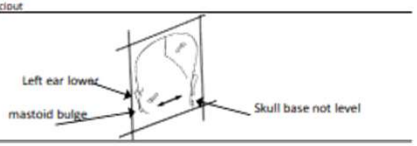
Other: Downsloping occiput

Posterior view:

Skull base: Level right lower left lower

Mastoid bulge: None Left mastoid bulge Right mastoid bulge

Ear position (vertical): symmetric right lower left lower



Front view:

Overall facial appearance: Symmetric Asymmetric

Eye opening Symmetric Right larger Left larger

Eye position: Symmetric Right lower Left lower

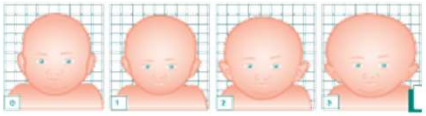
Eye spacing: Typical narrow wide

Facial width: Typical Wide Narrow

Nasal Twist: None to right to left

Head Tilt: Right Left

Torticollis: Right Left



Head Shape Exam

Findings

- Measurements
  - Wt %ile, Ht %ile, HC %ile
  - Proportional?
  - Who measured it?

Why it matters

- Microcephaly not generally associated with single suture craniosynostosis
- Macrocephaly commonly seen with abnormal head shapes

Normal | Plagiocephaly | Brachycephaly | Scaphocephaly

Systematic Head Shape Exam

- Palpation
- Top Down View
- Side View
- Posterior View
  - *Correct for head tilt*
- Frontal/Facial view

~~**Frontal Bossing**~~

PHYSICAL EXAMINATION of the HEAD:
Patient: _____ Date: ___/___/___
Examiner: _____
Weight _____ kg, ____ to ____ percentile
Height _____ cm, ____ to ____ percentile
OFC _____ cm, ____ to ____ percentile (Nellhaus curve)
General: active alert engaging
Other: _____

Palpation:
Anterior fontanelle:
Size: Typical Small large
Shape: Diamond Triangular
Position: Typical Anteriorly placed Posteriorly placed
Quality: Flat Full
Soft Tense

Palpable ridge(s): Sagittal No Yes Mobile? Yes No
(if yes, side?) Coronal No Yes Right Left
Metopic No Yes
(if yes, side?) Lambdoid No Yes Right Left

Top down view:
Forehead shape: Typical Triangular Other: _____
Flattening/retrusion: No symmetric R L
Prominence: No symmetric R L

Occipital shape: Typical Other: _____
(if yes, side?) Flattening/retrusion No symmetric R L
(if yes, side?) Prominence: No symmetric R L

Overall shape: Typical parallelogram rhomboid
Tear drop Long AP Short AP Brachy

Widest point: Bi-Parietal Bi-Temporal Other:____
Ear position (horizontal): Symmetric Right anterior Left

Side View:
AP diameter: Typical long short
Height: Typical Turricephalic Tall forehead Tall occiput
Other: Downsloping occiput

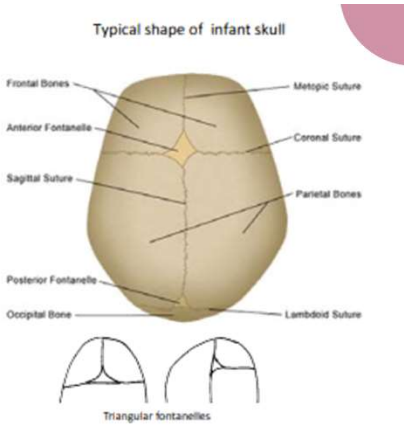
Posterior view:
Skull base: Level right lower left lower

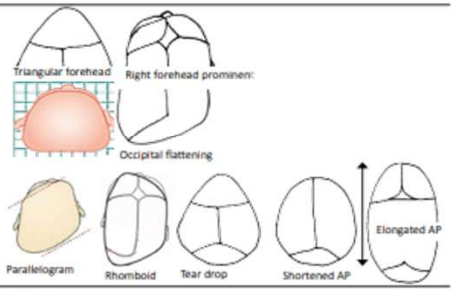
Mastoid bulge: None Left mastoid bulge Right mastoid bulge

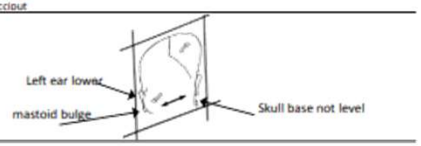
Ear position (vertical): symmetric right lower left lower

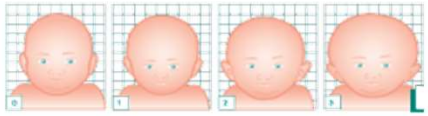
Front view:
Overall facial appearance: Symmetric Asymmetric
Eye opening Symmetric Right larger Left larger
Eye position: Symmetric Right lower Left lower
Eye spacing: Typical narrow wide

Facial width: Typical Wide Narrow
Nasal Twist: None to right to left
Head Tilt: Right Left
Torticollis: Right Left





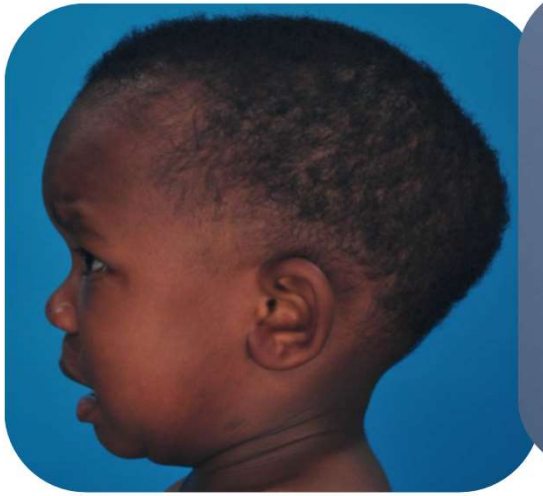




Head Shape Exam

Side View

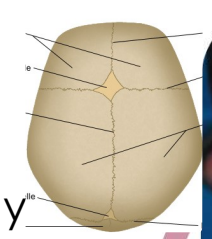
- AP Diameter
  - Typical Short Long
- Height
  - Typical Turricephalic
    Tall Forehead Tall Occiput
- Prominence
  - Prominent Forehead
  - Prominent Occiput

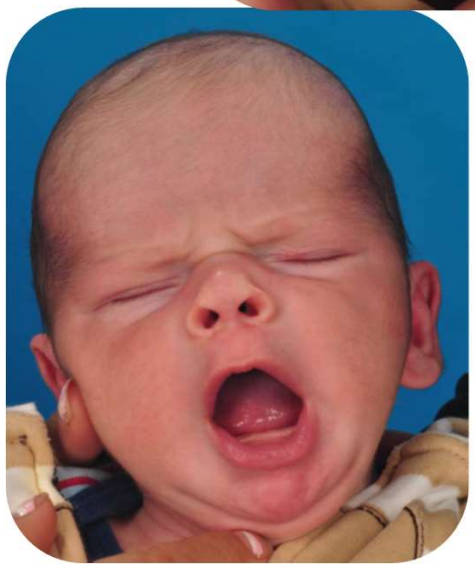


Single Suture Craniosynostosis vs. Plagiocephaly

Sagittal Craniosynostosis

- HC Measurement: relative macrocephaly
- Palpation:
  - Anterior Fontanelle: Triangular in normal position
    - Often larger than expected
  - Palpable ridge over the sagittal suture
    - non-mobile
- Top-Down View:
  - Overall Shape: narrow & elongated AP
    - **Widest point: not Bi-Parietal**
      - At or anterior to the ears
      - May not have a widest point
      - Bi-temporal
- Side View:
  - AP Diameter: long
  - **Prominent Forehead & Occiput**
- Facial/Frontal View:
  - Overall face: symmetric, large forehead

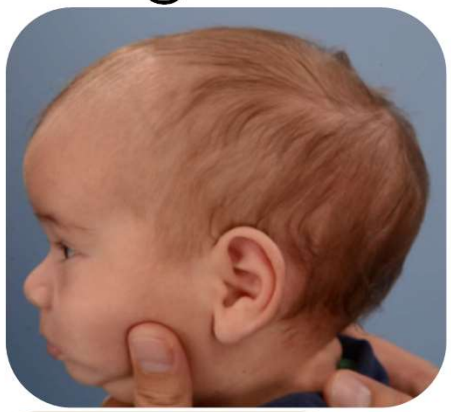


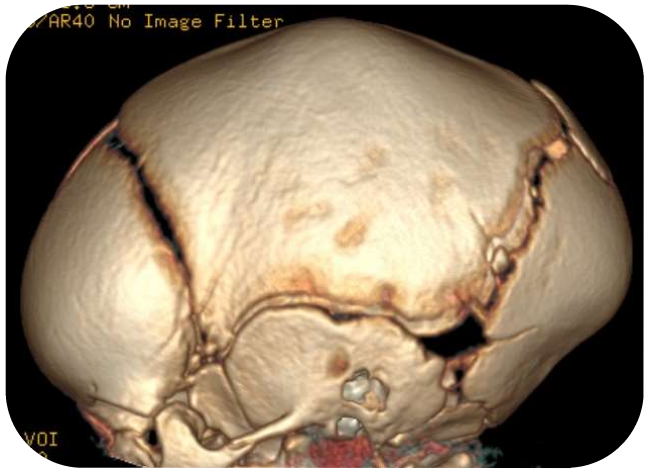


Sagittal Craniosynostosis

- Incidence 2-3.2:10,000
  - 1:4,200
  - 40-50% of Craniosynostosis
- Male:Female – ~3.5:1 ratio
- Age for treatment:
  - Endoscopic Repair: 1.5-4mo
  - Open Repair: 4-8mo

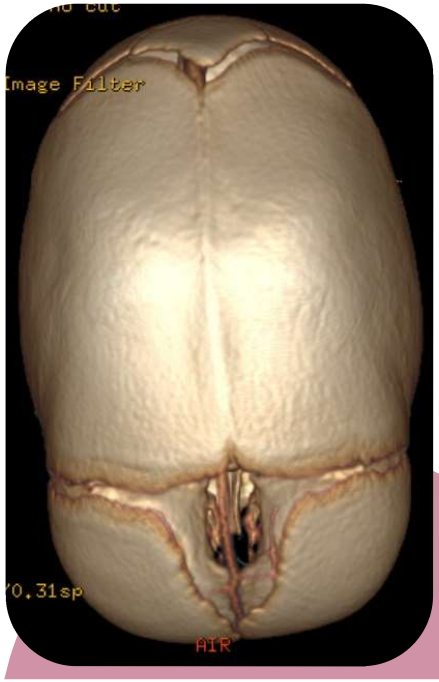
Sagittal Craniosynostosis



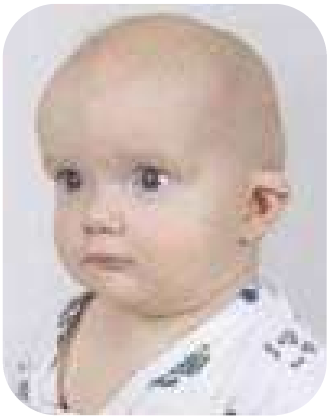






Sagittal Craniosynostosis












Versus Breech Molding

Metopic Craniosynostosis

- HC Measurement: normal to relative macrocephaly
- Palpation:
  - Anterior Fontanelle: small/closed
  - Palpable ridge over the metopic suture
- Top-Down View:
  - **Forehead shape: bilateral/symmetric retrusion**
    - **Lateral globes visible**
  - Overall Shape: teardrop shaped
- Facial/Frontal View:
  - Calvarium: symmetric, but more prominent posteriorly
  - Overall face: symmetric with bi-temporal constriction
  - **Eye Opening & Position: symmetric with bilateral arched eyebrows**
  - Eye Spacing: narrow
  - Facial Width: narrow compared to calvarium
    - Small face on a big head

Metopic Craniosynostosis

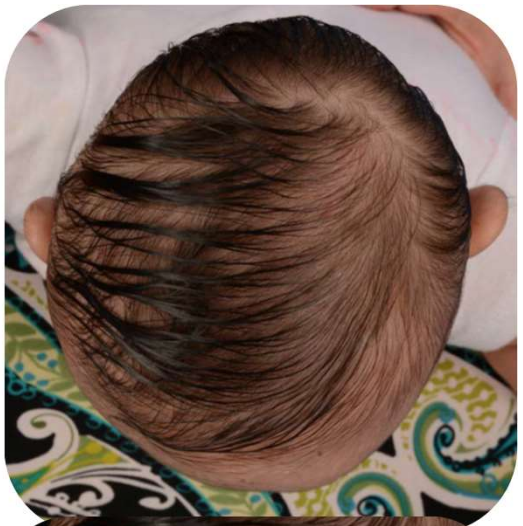
- Incidence 1:10,000
  - 19-28% of all craniosynostosis
- Male:Female – 3.2:1 ratio
- 25-30% are associated with a syndrome

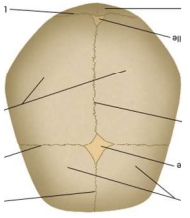
Metopic Craniosynostosis

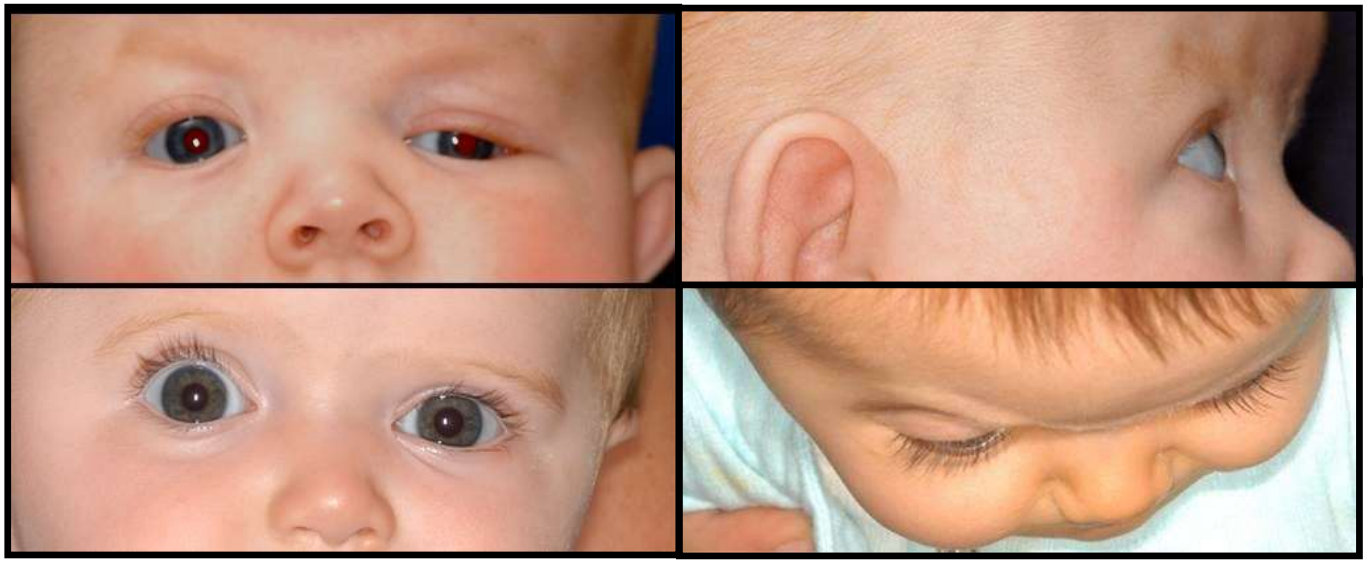
- Age for Treatment
  - Endoscopic: 1.5-4 mo
  - Open Repair: 9-14mo
- Isolated Metopic Ridge
  - Normal head shape
  - No Bi-temporal constriction
  - Normal forehead contouring

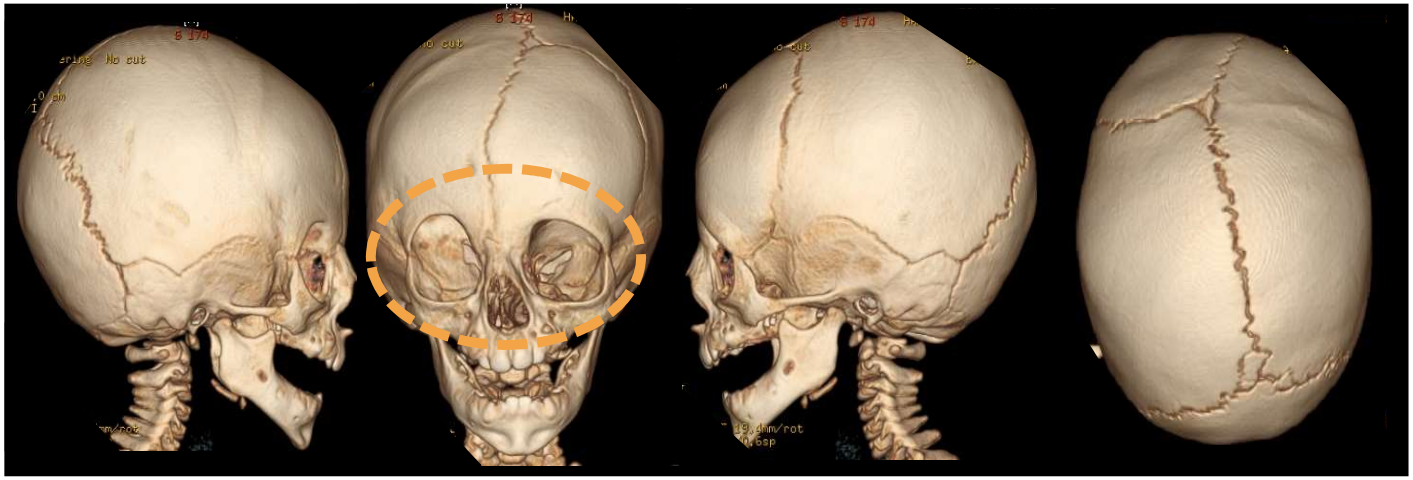
Unicoronal Craniosynostosis

- HC Measurement: more normal
- Palpation:
  - Anterior Fontanelle: triangular (blunted to side of CS)
  - +/- Palpable ridge over the effected coronal suture
- Top-Down View:
  - Forehead shape: effected sided retrusion
    - Effected side lateral globe and cheek visible
  - Overall Shape: rhomboid
    - Widest point: askew temporal to parietal
- Facial/Frontal View:
  - Calvarium: symmetric
  - **Overall face: effected side forehead swept back**
    - **Facial scoliosis/twist C-shaped away from defect**
    - **Unilateral raised eyebrow**
  - **Eye Opening: effected side larger**
  - Eye Position: symmetric vs. opposite side slightly lower
  - Eye Spacing: typical
  - Facial Width: normal
  - **Nasal Twist: tip away from defect**









Uni-Coronal Craniosynostosis

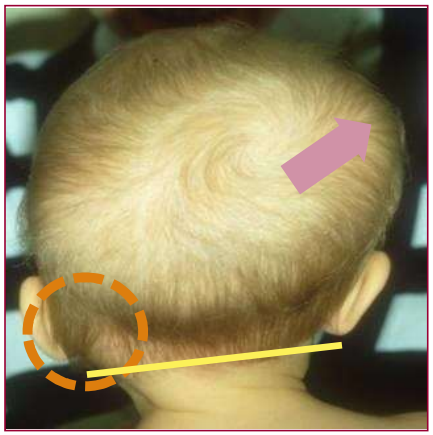
- Incidence 0.7-1:10,000
  - 12-24% of all craniosynostosis
- Male:Female
  - 1:3.6 ratio
- 30% have mutation
  - FGFR3 or TWST1
  - +/- c-spine changes
- Treatment Age:
  - Endoscopic: 1.5-4mo
    - Not as common
  - Open repair
    - 9-14mo

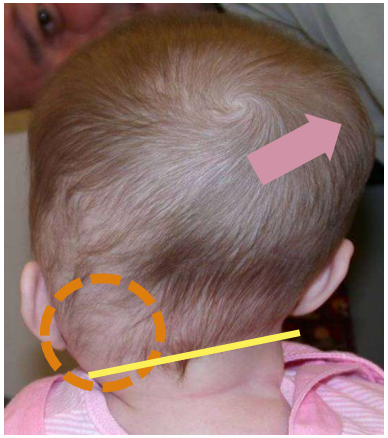
Uni-Coronal Craniosynostosis

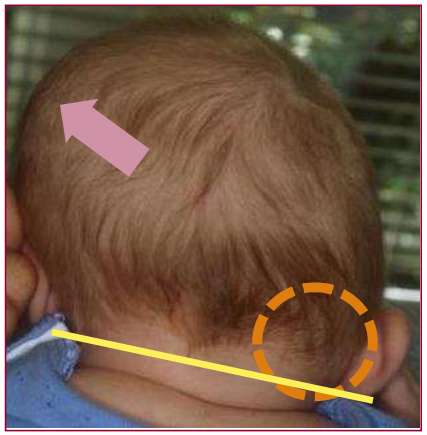
Lambdoid Craniosynostosis

- HC Measurement: normal to relative macrocephaly
- Palpation:
  - Palpable ridge over the effected lambdoid suture
- Top-Down View:
  - Forehead shape: effected sided retrusion
    - Vs opposite side prominence
  - Occipital Shape: same flattening vs. opposite prominence
  - Overall Shape: rhomboid
    - Widest point: askew temporal to parietal
  - Horizontal Ear Position: effected side posterior
- Side View:
  - AP Diameter: Typical
- Posterior View: (correct head tilt)
  - **Skull base: lower on effected side**
  - **Effected side Mastoid Bulge**
  - **Vertical Ear Position: effected side lower**
- Facial/Frontal View:
  - **Calvarium: opposite side parietal prominence**
  - Overall face: Effected ear more prominent
    - facial scoliosis C-shape away from the effected side
  - Eye Opening: Effected side larger
  - Eye Position: symmetric vs. opposite side slightly lower
  - Eye Spacing: typical
  - Facial Width: normal with effected side wider
  - Nasal Twist: tip away from defect

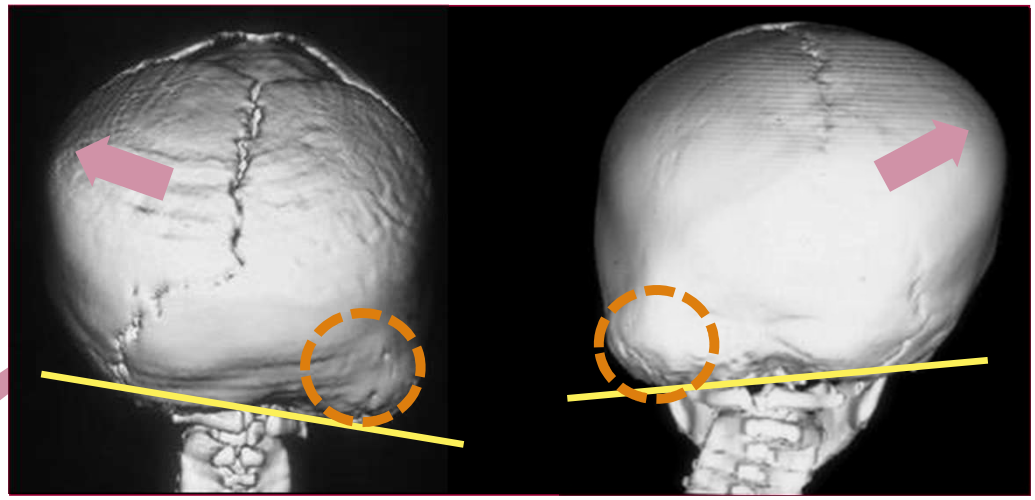
Lambdoid Craniosynostosis Hallmarks

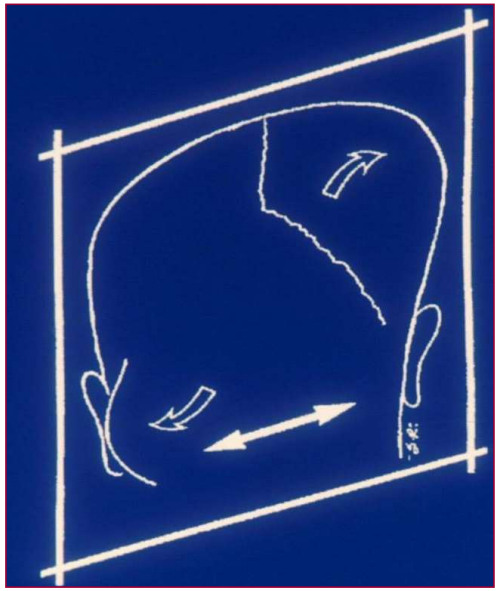


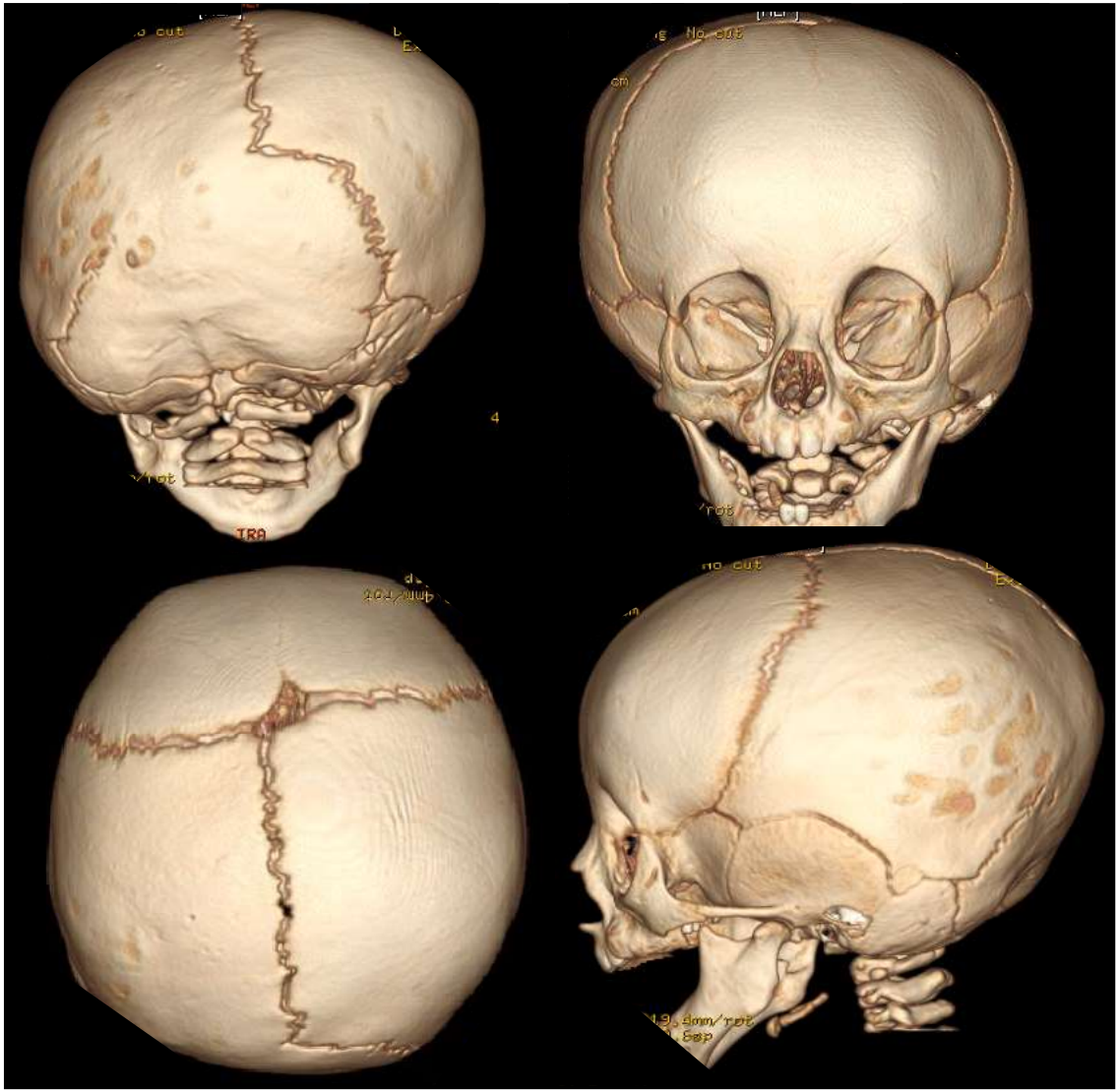




- Skull base tilt
- Ipsilateral mastoid bulge
- Vertical ear dystopia
- Contralateral parietal bulge

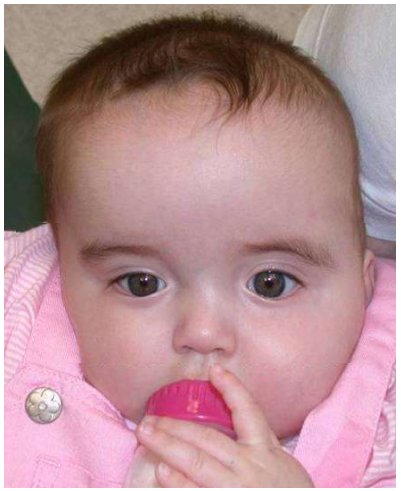


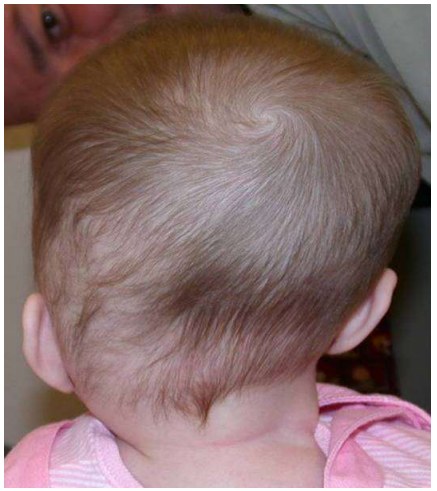




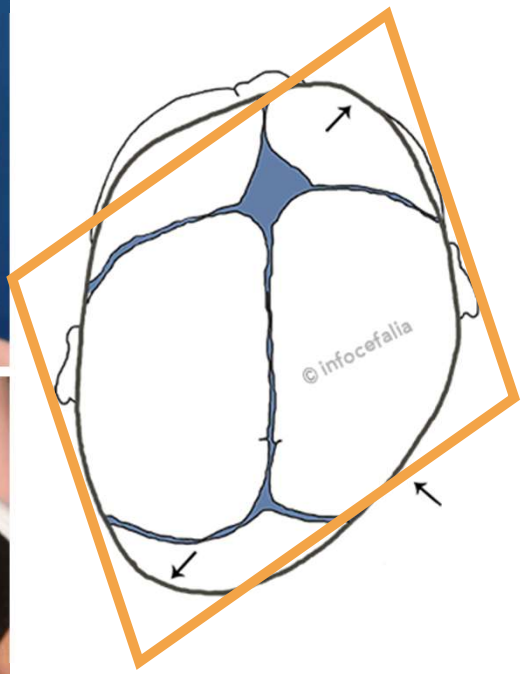
Lambdoid Craniosynostosis

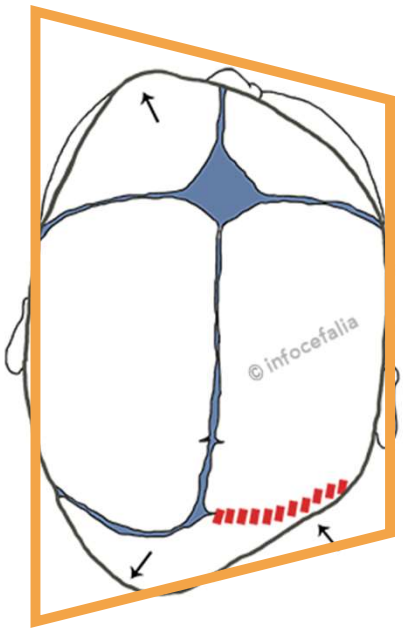
- Incidence: 0.1:10,000
  - Only 2% of all craniosynostosis
- Male:Female – 3.3:1 ratio
  - Some studies show 1:1
- Treatment Age:
  - Endoscopic: 1.5-4mo
    - Not as common
  - Open repair
    - 9-14mo





Lambdoid vs. Positional Deformational Plagiocephaly



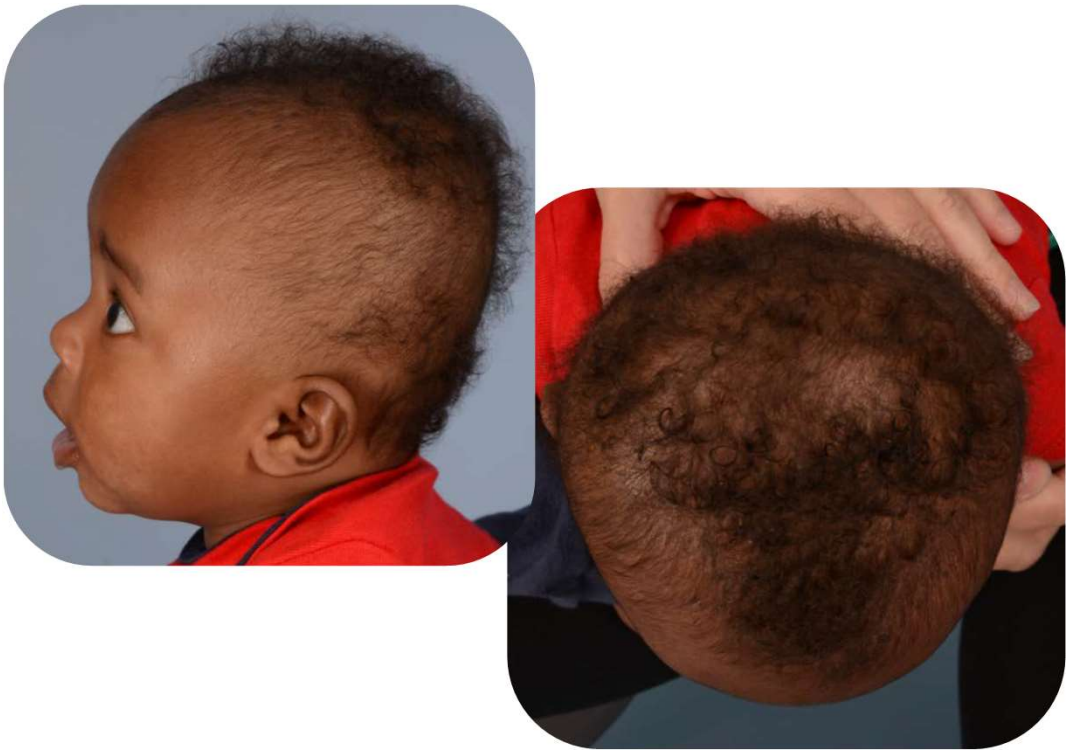


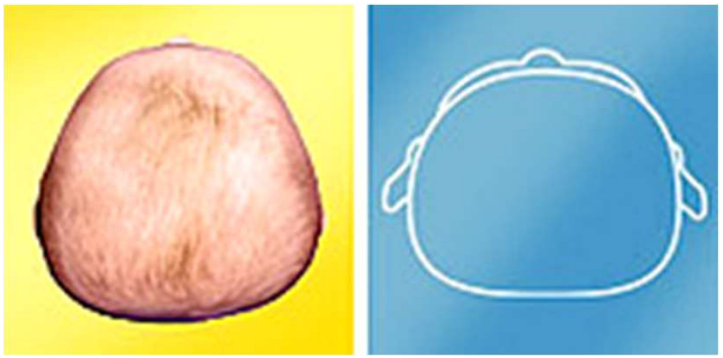
Plagiocephaly

- HC Measurement: normal
- Top-Down View:
  - **Forehead shape: slight same sided prominence**
  - **Occipital Shape: same side flattening**
  - **Overall Shape: parallelogram**
    - Widest point: askew but bi-parietal
  - **Horizontal Ear Position: effected side anterior**
- Side View:
  - AP Diameter: normal to short
  - Height: tall/flat occiput
  - No significant prominence
- Posterior View: (correct for head tilt)
  - Skull base: level
  - No Mastoid Bulge
  - Vertical Ear Position: symmetric
- Facial/Frontal View:
  - Calvarium: symmetric
  - Overall face: symmetric
    - Can have some asymmetry with significant plagiocephaly
  - Eye Opening, position, spacing: symmetric, typical
  - Facial Width: normal to slightly wider
  - Nasal Twist: none

Plagiocephaly

- ~22% of 2-6 month olds
  - Some studies put it at almost 50%
- Male:Female – 1.5:1 ratio
- Acquired in the first 4-12 weeks
  - Intrauterine deformation can happen
- Flattening corresponds to side baby sleeps on
- ~20% with some degree of torticollis





Plagiocephaly

- Increased risk in infants with hypotonia or developmental delay
- Correlation of plagiocephaly and poorer cognitive outcomes

Brachycephaly

- Positional
- Symmetric posterior flattening
  - Occipital lift
- Common in NICU babies
  - Prolonged sedation or intubation

Plagiocephaly: Conservative Treatment

- Increased supervised tummy time
- Core strengthening
- Less time on their back
- Repositioning in crib/bed
- Physical therapy for torticollis
  - Early referral to PT
  - PM&R referral if not improved

Plagiocephaly: Helmet Treatment

- Hard shell filled with foam lining
  - Redirect skull growth
  - Fit closely to the infant's head in some spots
  - Allow excess space/growth in flattened areas
- **Only "work" with asymmetric plagiocephaly**
  - **Not brachycephaly**
- Started at 4-6mo
  - Won't work if started after 8mo
- Worn for 23 hours a day
  - For 4-6 months (until about a year old)
- Cost $2000 to $2500
  - Most insurances don't cover them unless significant asymmetry

Plagiocephaly: Helmet Treatment Randomized control trial

- 84 infants
  - 40 for conservative management
  - 30 for helmet therapy
  - 14 dropped out or disappeared
- Helmet group:
  - 10/30 worn for the entire treatment (until 12mo)
    - Parents stopped early due to: happy with results, side effects, dissatisfaction with results, and one would "spontaneously pop off"
  - 73% had issues with the helmet fitting and not shifting throughout the day

Van Wijk, RM, et al. Helmet Therapy in infants with positional skull deformation: randomized control trial. BMJ 2014.

Plagiocephaly: Helmet Treatment Randomized control trial

- Primary outcome: plagiocephaly score at 24mo
  - No significant difference between the groups
- Secondary outcomes:
  - Side effects: all parents reported at least one side effect
    - Problems with helmet acceptance (24%)
    - Skin irritation (96%)
    - Augmented sweating (71%)
    - Unpleasant odor (76%)
    - Pain (33%)
    - Feeling hindered when cuddling the child (77%)
- Conclusion:
  - Helmets had no increased benefit, but substantially higher costs than conservative management

Van Wijk, RM, et al. Helmet Therapy in infants with positional skull deformation: randomized control trial. BMJ 2014.

What to do with a misshapen head:

Don't get imaging

Plagiocephaly

- Conservative Management
- Referral Plagiocephaly Clinic
    - PM&R
    - Plastic Surgery
    - Craniofacial Peds
  - ASAP if significant asymmetry or torticollis
  - Ideally no later than 6mo

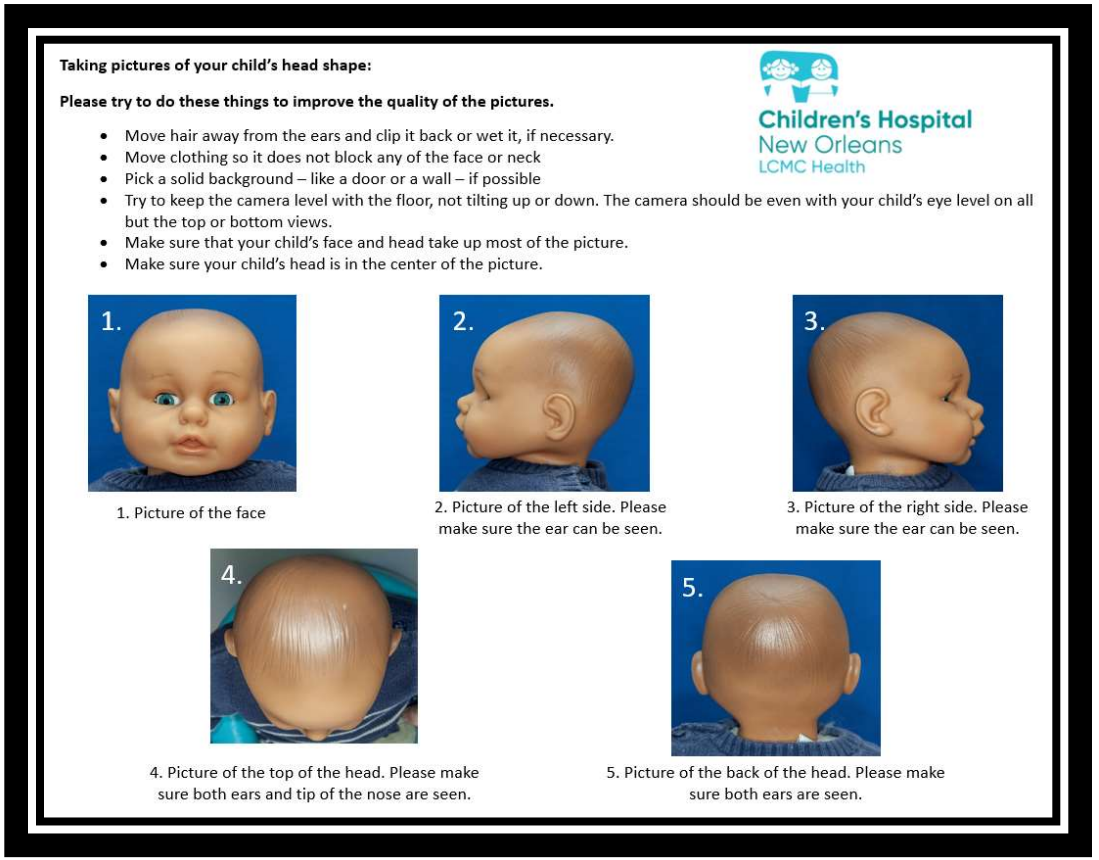
Craniosynostosis

- Referral Craniosynostosis Clinic
    - Neurosurgery
    - Plastic Surgery
    - Craniofacial Peds
- Call the Craniofacial Center
  - 504.896.9857

What to do with a misshapen head:

Unknown head shape or pretty sure its plagiocephaly

- Don't get imaging
- Refer to Plagiocephaly Clinic
- Take photos
  - Place them in your note
  - Send me an Epic message
  - Email me a couple photos
    - gfulto@lsuhsc.edu



Taking pictures of your child's head shape:

Please try to do these things to improve the quality of the pictures.

Children's Hospital New Orleans
LCMC Health

- Move hair away from the ears and clip it back or wet it, if necessary.
- Move clothing so it does not block any of the face or neck
- Pick a solid background – like a door or a wall – if possible
- Try to keep the camera level with the floor, not tilting up or down. The camera should be even with your child's eye level on all but the top or bottom views.
- Make sure that your child's face and head take up most of the picture.
- Make sure your child's head is in the center of the picture.

1. Picture of the face

2. Picture of the left side. Please make sure the ear can be seen.

3. Picture of the right side. Please make sure the ear can be seen.

4. Picture of the top of the head. Please make sure both ears and tip of the nose are seen.

5. Picture of the back of the head. Please make sure both ears are seen.

Questions or Comments?

- Call or email me:
  - Office: 504.896.9857
  - Desk: 504.894.5271
  - gfulto@lsuhsc.edu

- References and Further reading:

  - Dias MS, Samson T, Rizk EB, Governale LS, Richtsmeier JT; SECTION ON NEUROLOGIC SURGERY, SECTION ON PLASTIC AND RECONSTRUCTIVE SURGERY. Identifying the Misshapen Head: Craniosynostosis and Related Disorders. *Pediatrics*. 2020;146(3):e2020015511. doi:10.1542/peds.2020-015511
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  - Kiesler J, Ricer R. The abnormal fontanel. Am Fam Physician. 2003 Jun 15;67(12):2547-52. PMID: 12825844.
  - Birgfeld CB, Saltzman BS, Hing AV, Heike CL, Khanna PC, Gruss JS, Hopper RA. Making the diagnosis: metopic ridge versus metopic craniosynostosis. J Craniofac Surg. 2013 Jan;24(1):178-85. doi: 10.1097/SCS.0b013e31826683d1. PMID: 23348281.

- Thanks
  - Craniofacial Peds Team at Seattle Children's
    - ~95% of my knowledge
    - Many of the photos and diagrams

Seattle Children's
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