

CT Children's CLASP Guideline

Craniosynostosis

INTRODUCTION

Craniosynostosis is the result of the early fusion of one or more cranial sutures. The incidence of craniosynostosis is approximately 1 in 1700 births, with the sagittal suture being the most affected suture. An infant with prematurely fused sutures will display an abnormal skull morphology, with or without cranial growth deceleration. Early identification and referral to Neurosurgery is important for optimal intervention outcomes and can ensure least invasive surgical intervention.

INITIAL EVALUATION AND MANAGEMENT

TARGETED HISTORY & PHYSICAL EXAM:

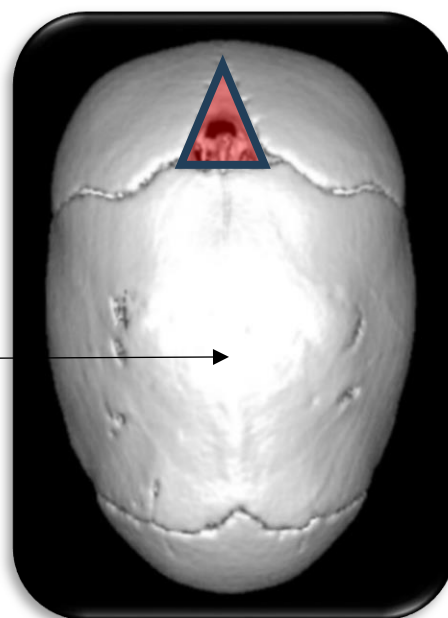
Physical exam findings can provide a diagnosis in the majority of cases

- **Assess:**
 - Gestational age adjusted head circumference growth chart for growth arrest
 - Anterior fontanelle size/shape for closure prior to 4 months of loss of diamond shape
 - Cranial and facial appearance

Types of Non-Syndromic Craniosynostosis:

- **Sagittal Craniosynostosis**
 - Elongated, narrow head
 - Frontal bossing and more tapered parietally and occipitally
 - Anteriorly displaced vertex
 - Triangular anterior fontanelle pointing anteriorly

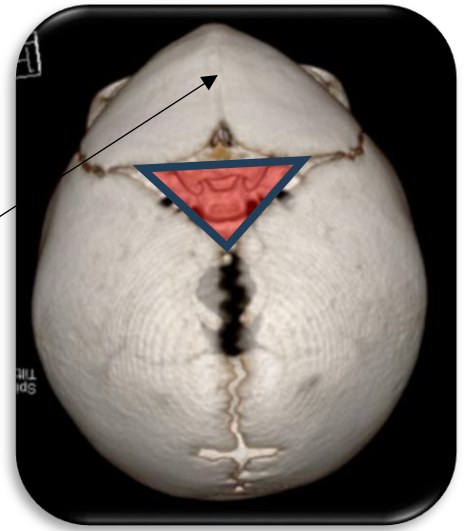
Sagittal Craniosynostosis



- **Metopic Craniosynostosis**

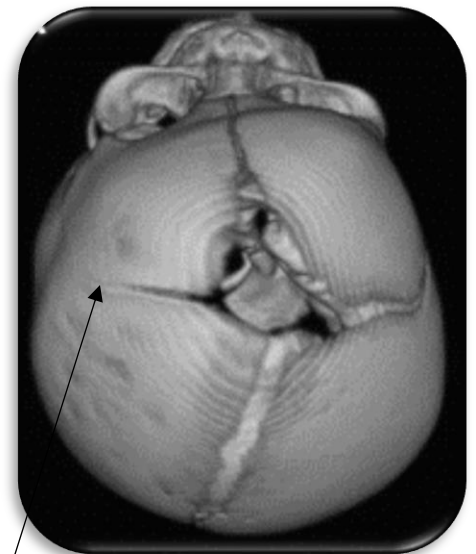
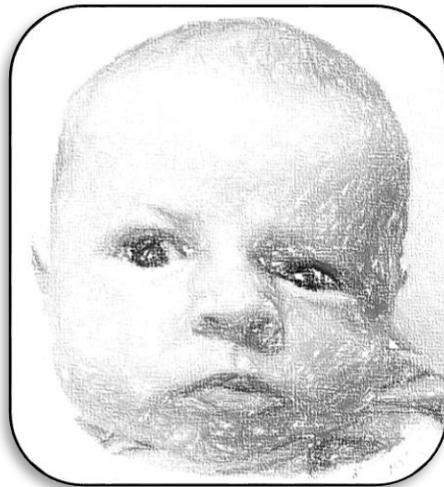
- Triangular forehead
- Hypotelorism
- Triangular fontanelle pointing posteriorly

Metopic craniosynostosis



- **Coronal craniosynostosis**

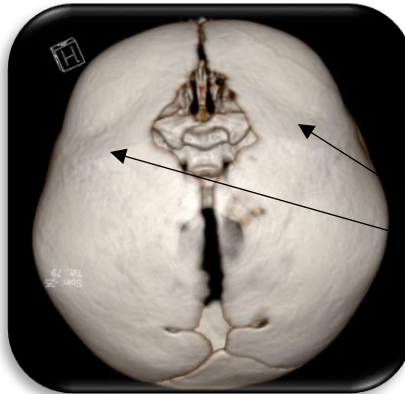
- Ipsilateral forehead recession
- Harlequin sign- elevation of ipsilateral orbit
- Frontal bossing on the unaffected side
- Nasal deviation towards the normal side
- Triangular fontanelle pointing away from the affected side



Unicoronal Craniosynostosis

- **Bicoronal craniosynostosis**

- Round, high head shape that is shallow front to back
- Proptosis
- Recessed brows

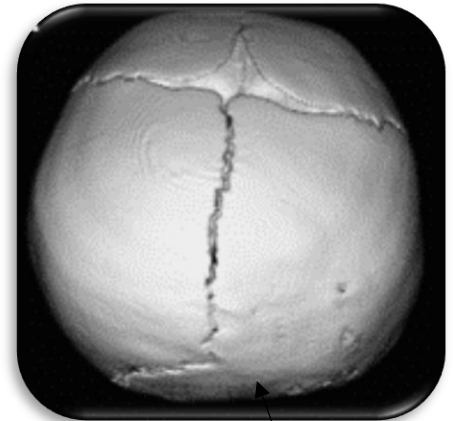


Bicoronal Craniosynostosis



- **Lambdoid Craniosynostosis**

- Contralateral frontal bossing
- Posterior and inferior displacement of the ipsilateral mastoid and rear
- Trapezoid appearance on vertex view



Lambdoid Craniosynostosis

RED FLAGS:

- Arrested cranial growth curve
- Small or closed fontanelle prior to 4 months of age
- Seizure, developmental regression
- Worsening head shape with age

	<p>INITIAL MANAGEMENT:</p> <ul style="list-style-type: none"> • Head circumference measurement • No management apart from referral to Neurosurgery is needed. • Imaging by the primary care provider is NOT indicated
WHEN TO REFER	<p>URGENT REFERRAL (seen within 2 weeks): Infants with suspected craniosynostosis should be referred as early as possible to ensure optimal management.</p> <ul style="list-style-type: none"> • Abnormally shaped or sized anterior fontanelle prior to 4 months of age • Abnormal head shape or facial morphology (as noted above) • Concern for craniosynostosis on imaging • Parental/provider concern for abnormal cranial growth
HOW TO REFER	<p>Referral to Neurosurgery Department via CT Children's One Call Access Center Make a Referral - Connecticut Children's (connecticutchildrens.org) Phone: 833.733.7669 Fax: 833.226.2329</p> <p><i>Information to be included with the referral:</i></p> <ul style="list-style-type: none"> ▪ Head circumference ▪ Growth chart
WHAT TO EXPECT	<p>What to expect from CT Children's Visit:</p> <ul style="list-style-type: none"> ▪ Growth measurements ▪ Photos for craniometrics ▪ If suspicious for craniosynostosis, may have an ultrasound ▪ If not craniosynostosis, then follow up for plagiocephaly ▪ If craniosynostosis, then will determine OR and pre op visit dates