Cranial Sutures & Craniosynostosis

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Objectives

• The objectives of this presentation are to:
  – Review the imaging features of normal cranial sutures
  – Identify the characteristics of abnormal skull shape on imaging
  – Review the characteristics of the most common non-syndromic and syndromic causes of craniosynostosis
Anatomical Review
Anatomical Review

- The bony plates of the skull communicate at the cranial sutures
- The anterior fontanelle occurs where the coronal & metopic sutures meet
- The posterior fontanelle occurs where the sagittal & lambdoid sutures meet
Anatomical Review

- The main cranial sutures & fontanelles include:

  - Coronal Sutures
  - Metopic Suture
  - Sagittal Suture
  - Lambdoid Sutures
  - Squamosal Sutures
  - Anterior Fontanelle
  - Posterior Fontanelle
Anatomical Review

- Growth of the skull occurs perpendicular to the cranial suture
- This is controlled by a complex signalling system including:
  - Ephrins (mark the suture boundary)
  - Fibroblast growth factor receptors (FGFR)
  - Transcription factor TWIST
Anatomical Review

- The cranial sutures are important for rapid skull growth in-utero & infancy
- The cranial sutures can usually be visualised on imaging into late adulthood
Normal Radiological Appearances
Normal Radiological Appearances

• The cranial sutures can be visualised on plain radiographs

• Standard views include:
  – PA
  – Lateral
  – Townes
Lateral Skull Radiograph

Right Lambdoid Suture

Left Lambdoid Suture

Sagittal Suture

Left Coronal Suture

Right Coronal Suture

Squamosal Sutures

Left Lambdoid Suture

Right Lambdoid Suture
Axial Computed Tomography

- On axial CT the cranial sutures can be visualised clearly
- On the following imaging a wide anterior fontanelle can be seen at the skull vertex
Axial CT

Cranial Sutures:
- Coronal
- Metopic
- Sagittal
- Lambdoid
3D CT

- With 3D reconstruction of the CT images, the cranial sutures can be visualised throughout their paths
Abnormal Head Shape
Craniosynostosis

• Premature fusion of one or more of the cranial sutures (craniosynostosis) occurs in approximately 1 in 2500 live births

• Lack of growth at the fused suture in combination with compensatory overgrowth at the normal sutures results in characteristic skull shape anomalies
Aetiology

- Isolated non-syndromic synostosis is thought to arise from mechanical pressure in-utero at a critical point during development.
- Genetic causes of craniosynostosis include mutations in FGFR1, 2 & 3 and TWIST1.
Sagittal Suture

• The sagittal suture is responsible for growth that results in widening of the skull

• The sagittal suture follows the path of the underlying sagittal sinus
Sagittal Synostosis

• Premature fusion of the sagittal suture results in a long shaped head (scaphocephaly = “boat-shaped”)

• This is the most common type of synostosis, accounting for 50-60% of all synostoses

• Radiological Features include:
  – Absence of sagittal suture
  – Decreased cephalic index (ratio of bicoronal distance to AP distance <75%)
Sagittal Synostosis on axial CT
Cranial Sutures:
- Coronal
- Metopic
- Sagittal
- Lambdoid
- Squamosal

Fused Sagittal Suture
Metopic Suture

- The metopic suture is responsible for horizontal growth of the forehead bones
- It is the only suture whose function is complete by birth
- Complete obliteration may therefore be seen at birth or within the first year of life, without pathological sequelae
Metopic Synostosis

• Premature fusion of the metopic suture results in a triangular shaped deformity (trigonocephaly)
• It accounts for 5-10% of synostoses
• Radiological features include:
  – Hypotelorism
  – Trigonocephaly
  – Upward elongation & medial rotation of orbits
  – Absence of the metopic suture (in presence of above features)
Metopic Synostosis on axial CT
Cranial Sutures:

- Lambdoid
- Squamosal
- Coronal
- Metopic
- Sagittal

Fused Metopic Suture
Coronal Suture

- The coronal suture is responsible for growth in the AP direction
- Premature fusion of the coronal sutures may be unilateral or bilateral
- Unicoronal synostosis is more likely to be an isolated non-syndromic event than its bilateral counterpart
Unicoronal Synostosis

- Unicoronal synostosis results in anterior plagiocephaaly
- It accounts for 10-15% of synostoses
- Radiological features of unicoronal synostosis include:
  - Absence of one of the coronal sutures
  - Flattening of the forehead on the affected side
  - Deviation of the nose to the opposite side
  - Elevation of the orbit on the affected side
  - Upward rotation of lesser wing of sphenoid – on AP skull radiograph = Harlequin sign
Unicoronal Synostosis on axial CT
Bicoronal Synostosis

- Bicoronal synostosis results in brachycephaly
- It accounts for 5-10% of synostoses
- Radiological features of bicoronal synostosis include:
  - Absence of both coronal sutures
  - Shortening in the AP direction
  - Bilateral Harlequin eye signs (on skull radiograph)
Bicoronal Synostosis on axial CT
Cranial Sutures:
- Lambdoid
- Squamosal
- Coronal
- Metopic
- Sagittal

Fused Coronal Sutures
Lambdoid Synostosis

- Rarest of the isolated synostoses
- It accounts for 1% of all synostoses
- Radiological signs:
  - Absence of suture
  - Flattening over synostosed suture
  - Mastoid overgrowth resulting in bulge behind involved ear
  - Temporo-parietal bossing on opposite side
Right Lambdoid Synostosis on axial CT
Cranial Sutures:

- Coronal
- Metopic
- Sagittal
- Lambdoid
- Squamosal

Fused Right Lambdoid suture
Multiple Suture Synostosis

• Involvement of multiple sutures is often as a result of an identifiable genetic cause

• The most common syndromic causes of craniosynostosis include:
  – Muenke Syndrome
  – Saethre-Chotzen Syndrome
  – Crouzon Syndrome
  – Apert Syndrome
  – Pfeiffer Syndrome
Apert Syndrome

- Mutation in FGFR2 on Chromosome 10
- Multiple suture synostosis – usually coronal sutures with enlarged anterior fontanelle
- Mid-face hypoplasia
- Complex acrosyndactyly of the hands & feet

AP radiographs of hands (top) and feet (bottom) showing complex syndactyly in child with Apert Syndrome
Crouzon Syndrome

- Mutation in FGFR2 or FGFR3
- Bicoronal synostosis
- Mid-face hypoplasia
- Normal hands

Lateral cephalogram demonstrating mid-face hypoplasia in child with Crouzon syndrome
Muenke Syndrome

- P250R FGFR3 mutation on chromosome 3
- Coronal synostosis
- Bitemporal bossing
- Sensorineural hearing loss

Lateral 3D CT view of bicoronal synostosis in child with Muenke syndrome
Saethre-Chotzen Syndrome

• Mutation or deletion of TWIST1 gene on Chromosome 7
• Most frequently bicoronal or unicoronal synostosis
• Low frontal hairline
• Eyelid ptosis
• Soft tissue syndactyly (variable)
Pfeiffer Syndrome

- Mutation in FGFR1 or FGFR2
- Usually bicoronal synostosis
- Broad thumbs/ great toes
- Syndrome most commonly associated with pansynostosis (clover-leaf deformity)
Clover-leaf Deformity

3D CT in child with Pfeiffer syndrome

Note the clover-leaf deformity with multiple areas of bony defects
Management

• Surgical management is indicated in:
  – Emergency Situations:
    • Airway protection
    • Eye protection
    • Raised intracranial pressure
  – Elective:
    • To prevent progressive deformity
    • To prevent development of raised intracranial pressure
Management

• For metopic & coronal synostosis this is frequently with fronto-orbital advancement and remodelling

• For sagittal synostosis this is frequently with sub-total or total calvarial remodelling
Management

- Alternative surgical techniques include the use of distraction osteogenesis.
Plagiocephaly without Synostosis

• This is a clinical diagnosis

• Deformational/positional plagiocephaly occurs without premature fusion of the sutures

• There is a parallelogram deformity of the skull when viewed from vertex

• Radiological features include:
  – All cranial sutures visible
  – On AP skull radiograph there may be rotation due to positioning of the infant on the flattened side on radiography plate
Summary
Radiological Features of Synostosis

• Primary Changes:
  – Loss of sutural interdigitations
  – Loss of suture lucency
  – Sclerosis of suture
  – Raising (lipping) of suture

• Secondary Changes:
  – Abnormal skull shape
  – Copper beating (suggesting raised ICP)
  – Harlequin sign (on plain radiograph)
Further Reading


• Branson HM, Shroff MM. Craniosynostosis and 3-Dimensional Computed Tomography. 2011; 32(6): 569-577