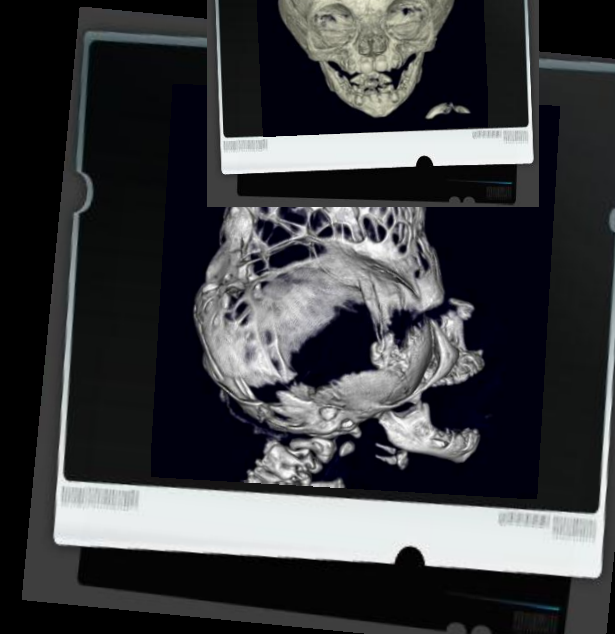
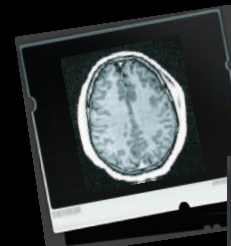
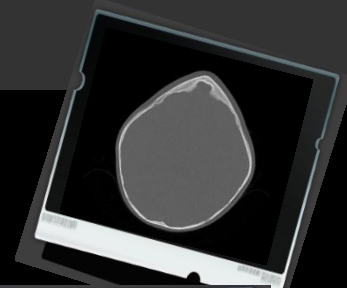


Cranial Sutures & Craniosynostosis

Dr Karen A Eley
Mr David Johnson
Dr Fintan Sheerin

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1	Dr Karen A Eley	
2	Mr David Johnson	
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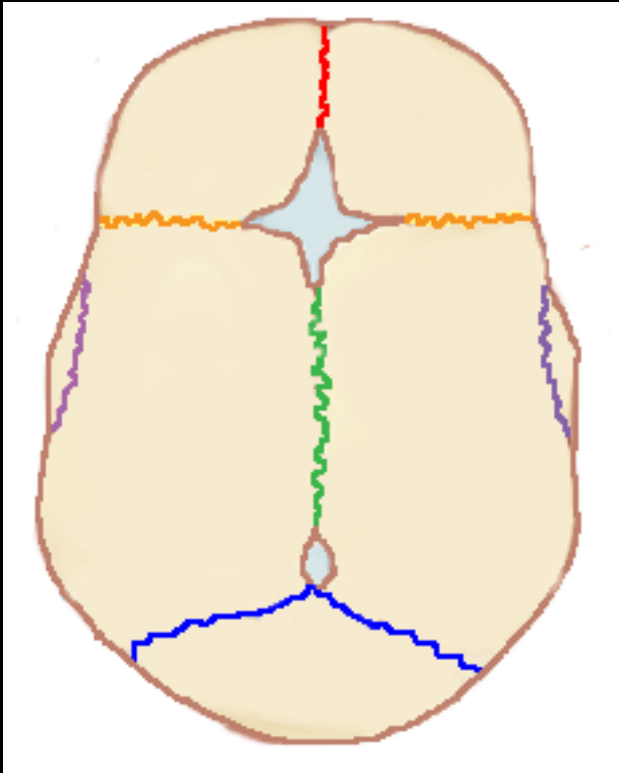


Objectives

- 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

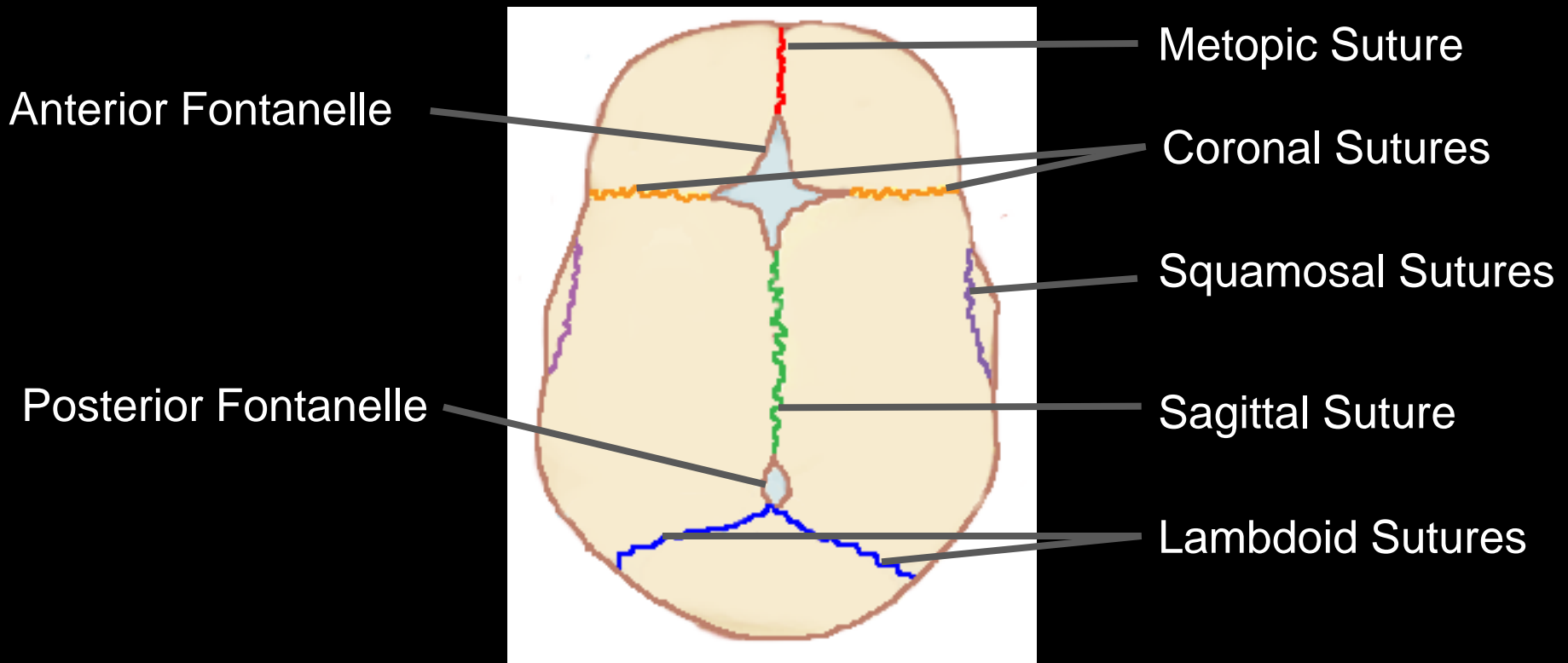
Anatomical Review



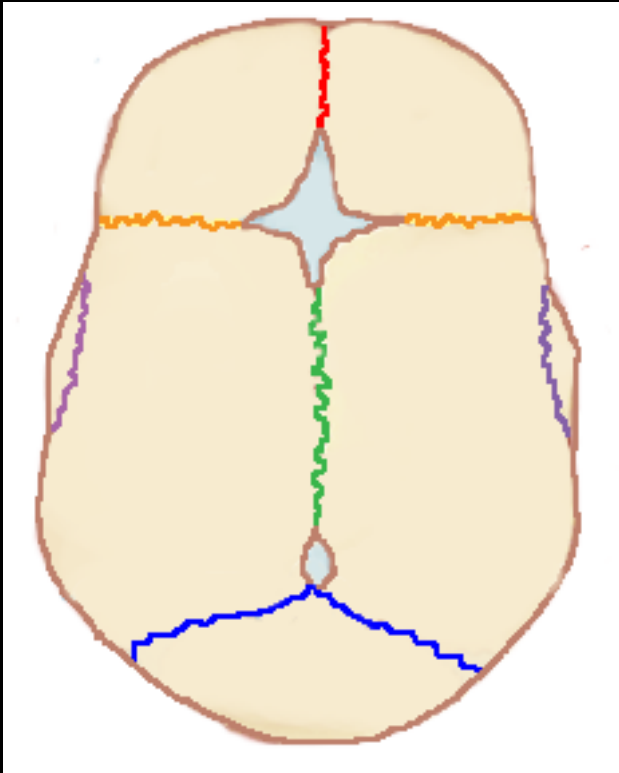
- The bony plates of the skull communicate at the cranial sutures
- The anterior fontanelle occurs where the coronal & metopic sutures meet
- The posterior fontanelle occurs where the sagittal & lambdoid sutures meet

Anatomical Review

- The main cranial sutures & fontanelles include:



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



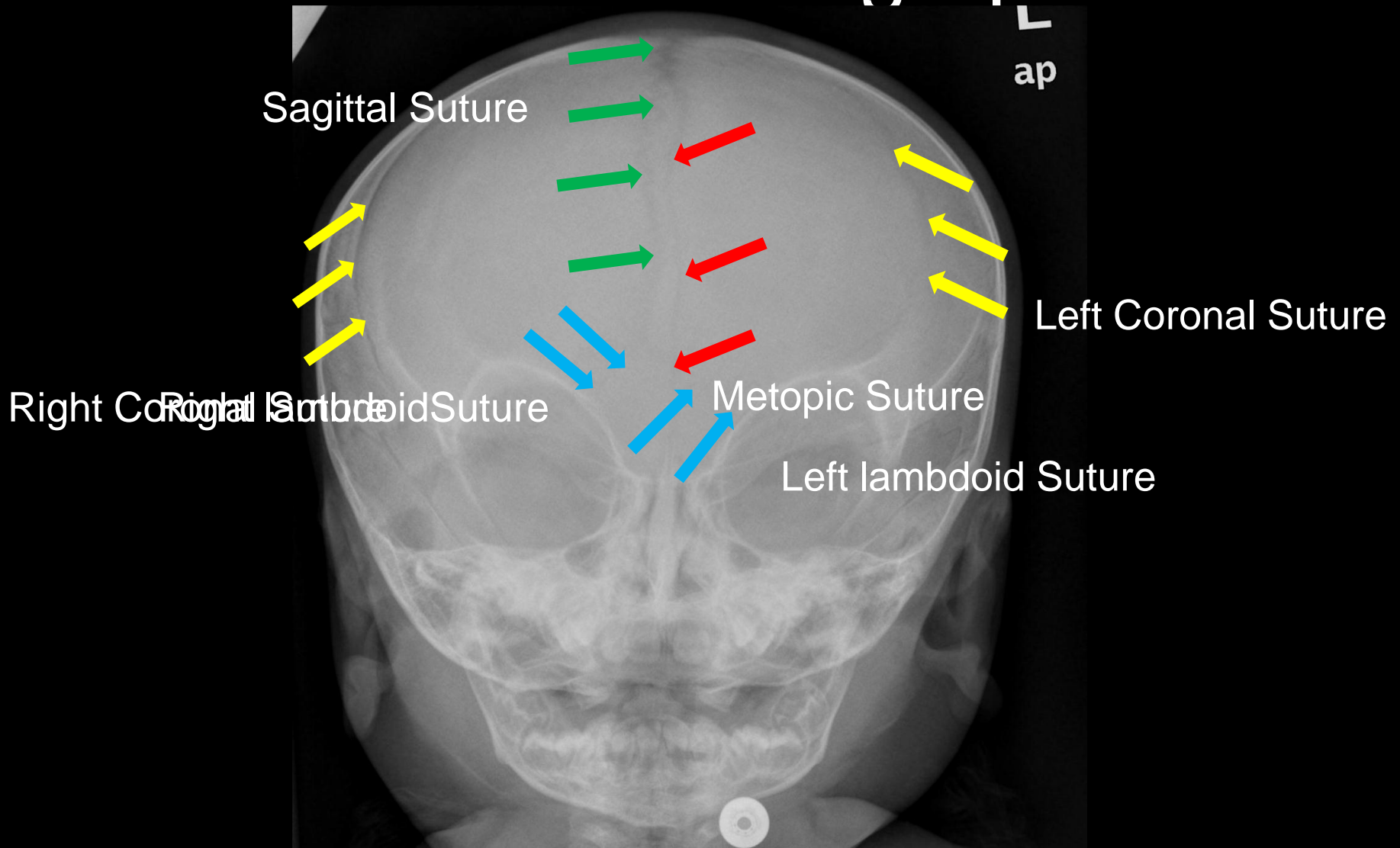
- The cranial sutures are important for rapid skull growth in-utero & infancy
- The cranial sutures can usually be visualised on imaging into late adulthood

Normal Radiological Appearances

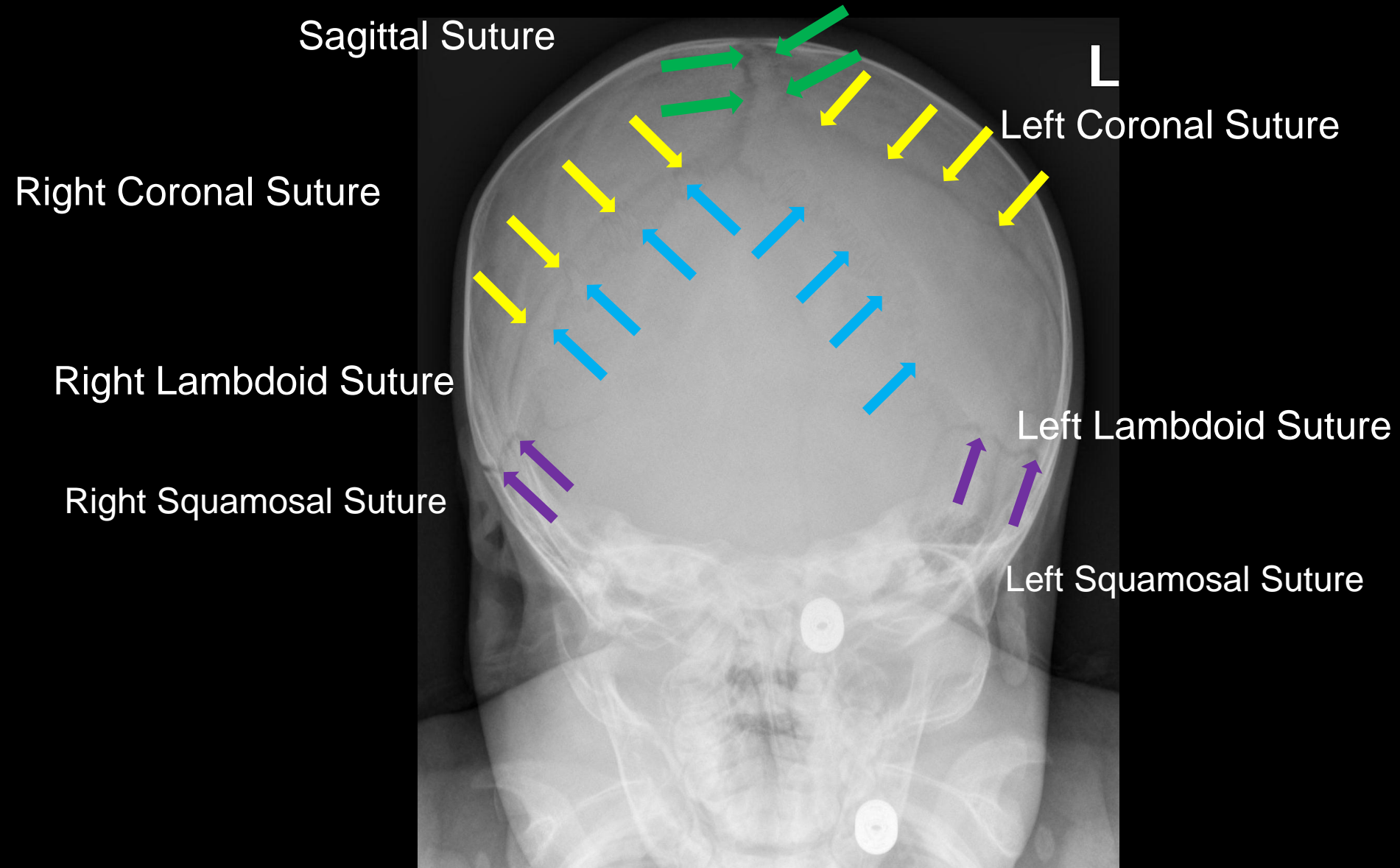
Normal Radiological Appearances

- The cranial sutures can be visualised on plain radiographs
- Standard views include:
 - PA
 - Lateral
 - Townes

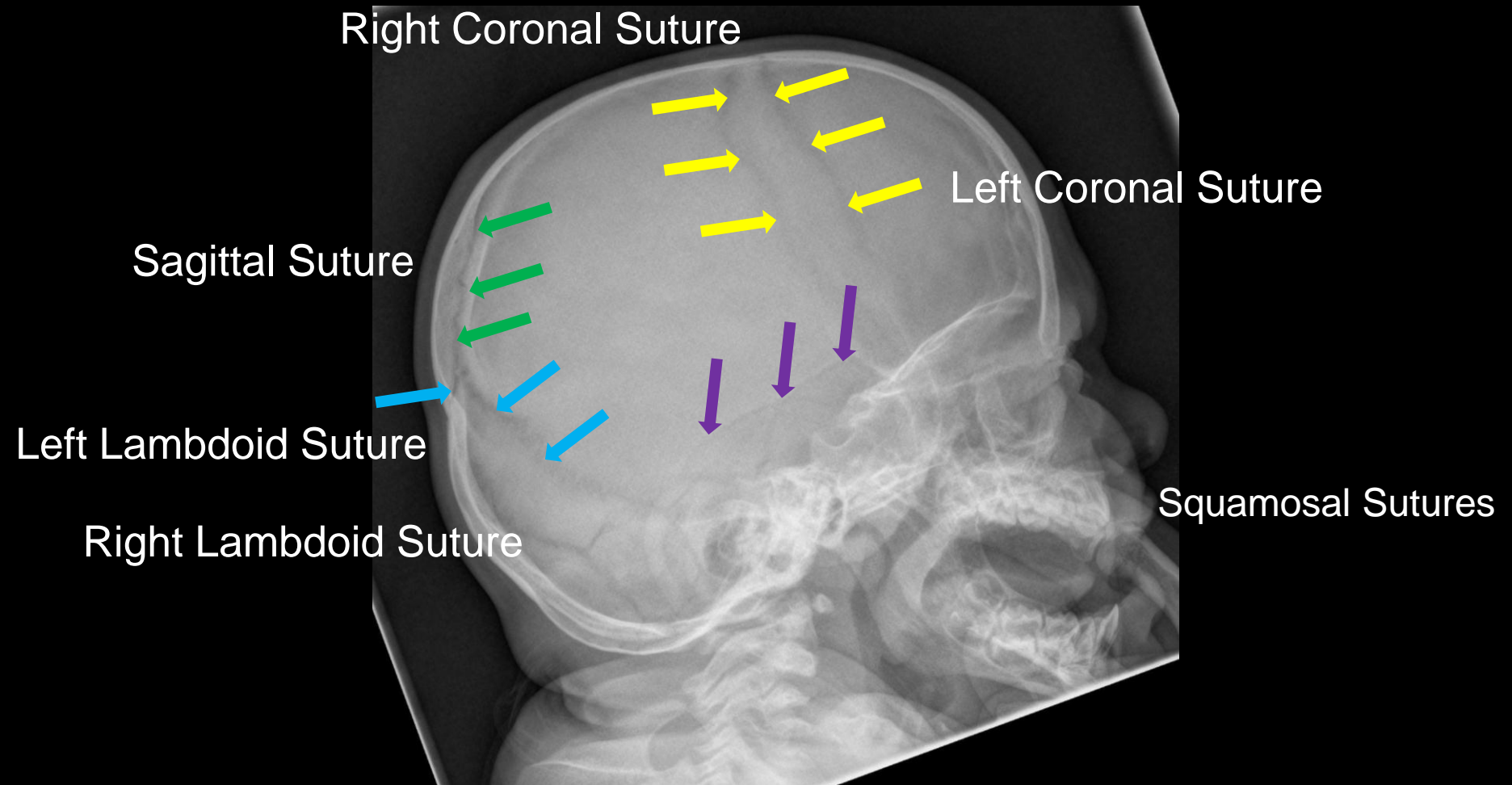
PA Skull radiograph



Townes View



Lateral Skull Radiograph



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

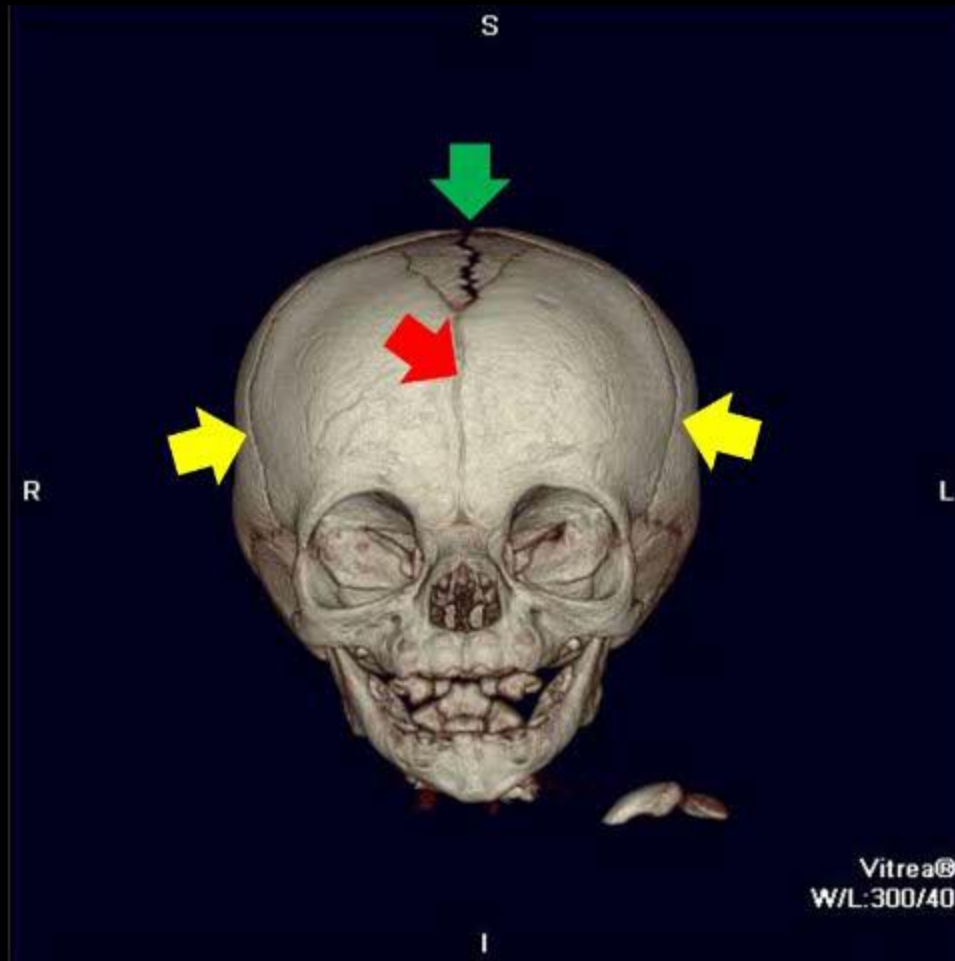


Lambdoid

3D CT

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

3D CT



Cranial Sutures:

	Coronal		Metopic		Sagittal		Lambdoid		Squamosal
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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

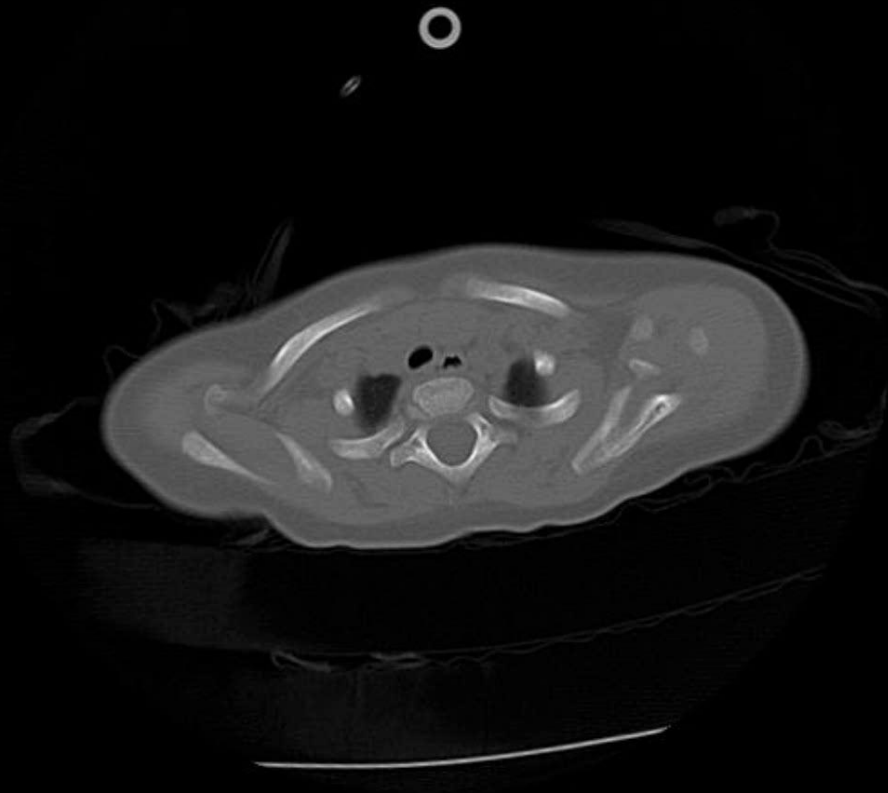
Sagittal Suture

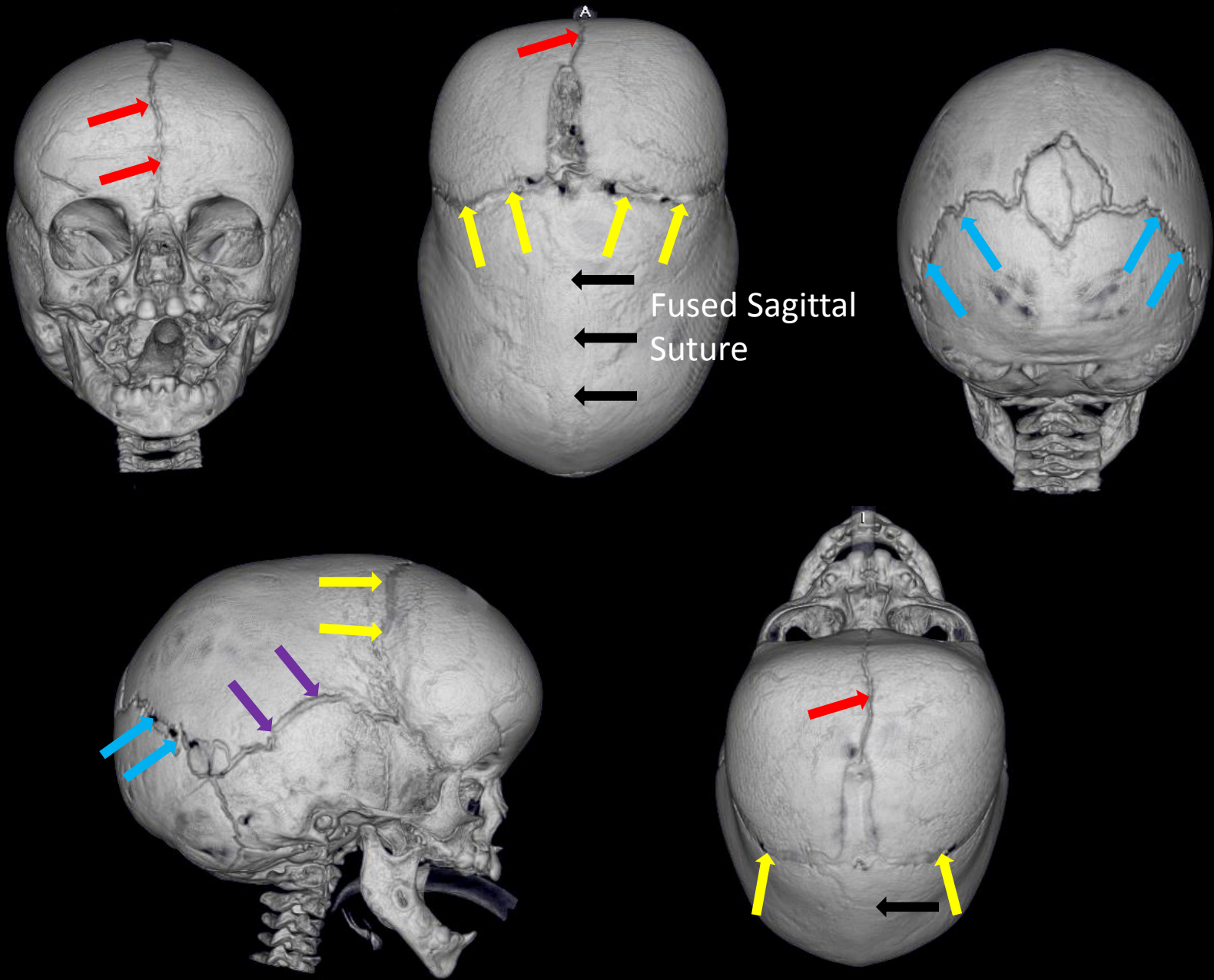
- The sagittal suture is responsible for growth that results in widening of the skull
- The sagittal suture follows the path of the underlying sagittal sinus

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
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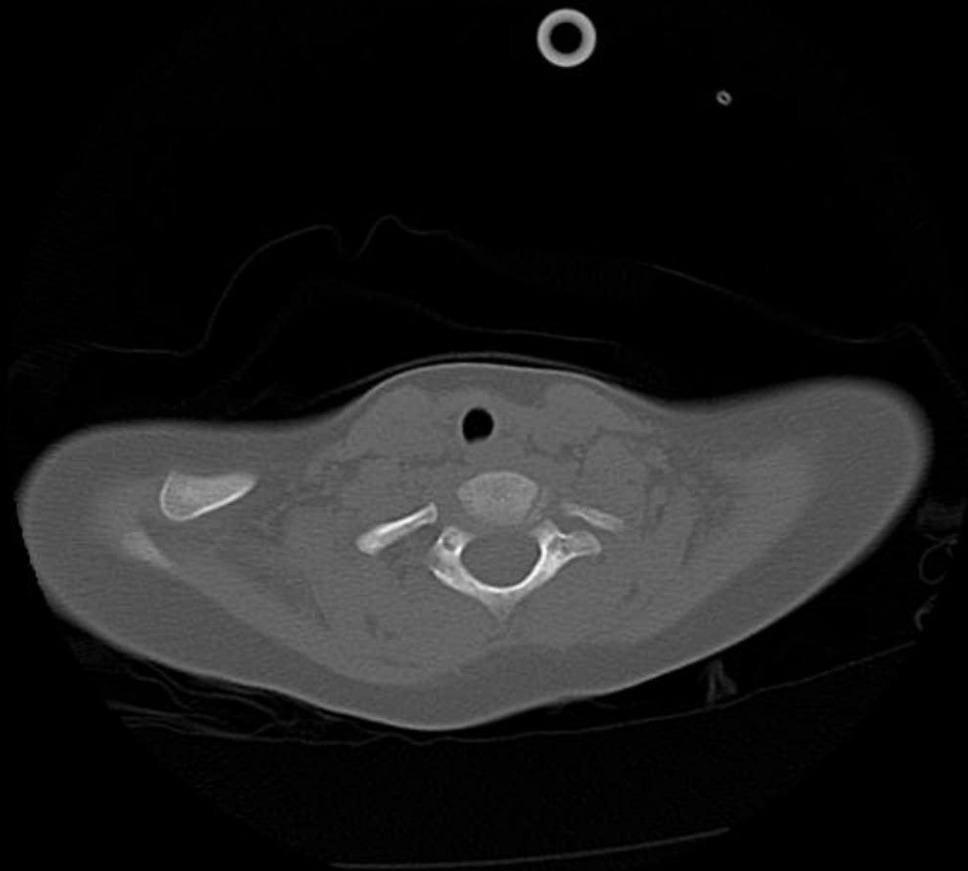
Metopic Suture

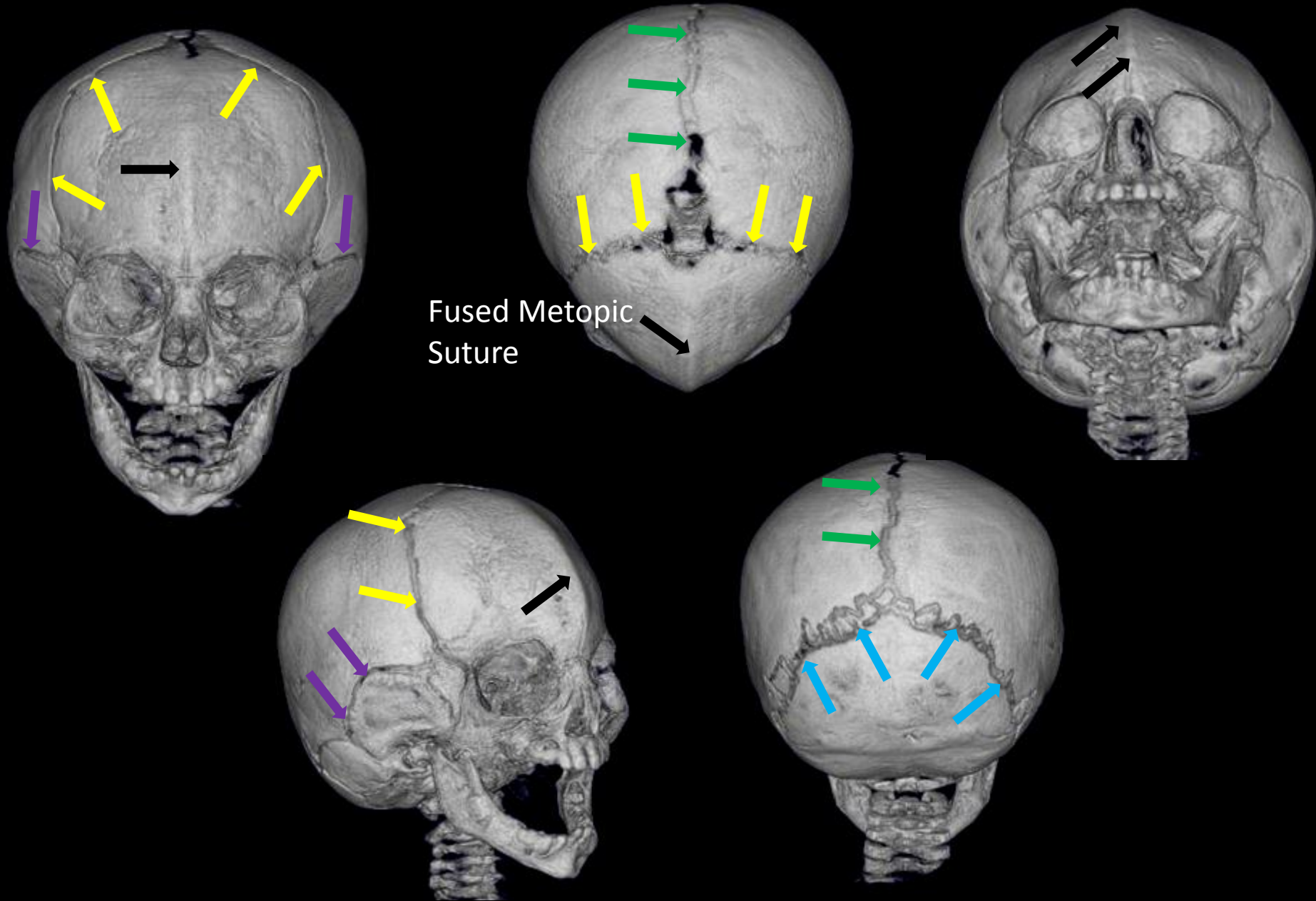
- The metopic suture is responsible for horizontal growth of the forehead bones
- It is the only suture whose function is complete by birth
- Complete obliteration may therefore be seen at birth or within the first year of life, without pathological sequelae

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





Fused Metopic
Suture

Cranial Sutures:									
	Coronal		Metopic		Sagittal		Lambdoid		Squamosal

Coronal Suture

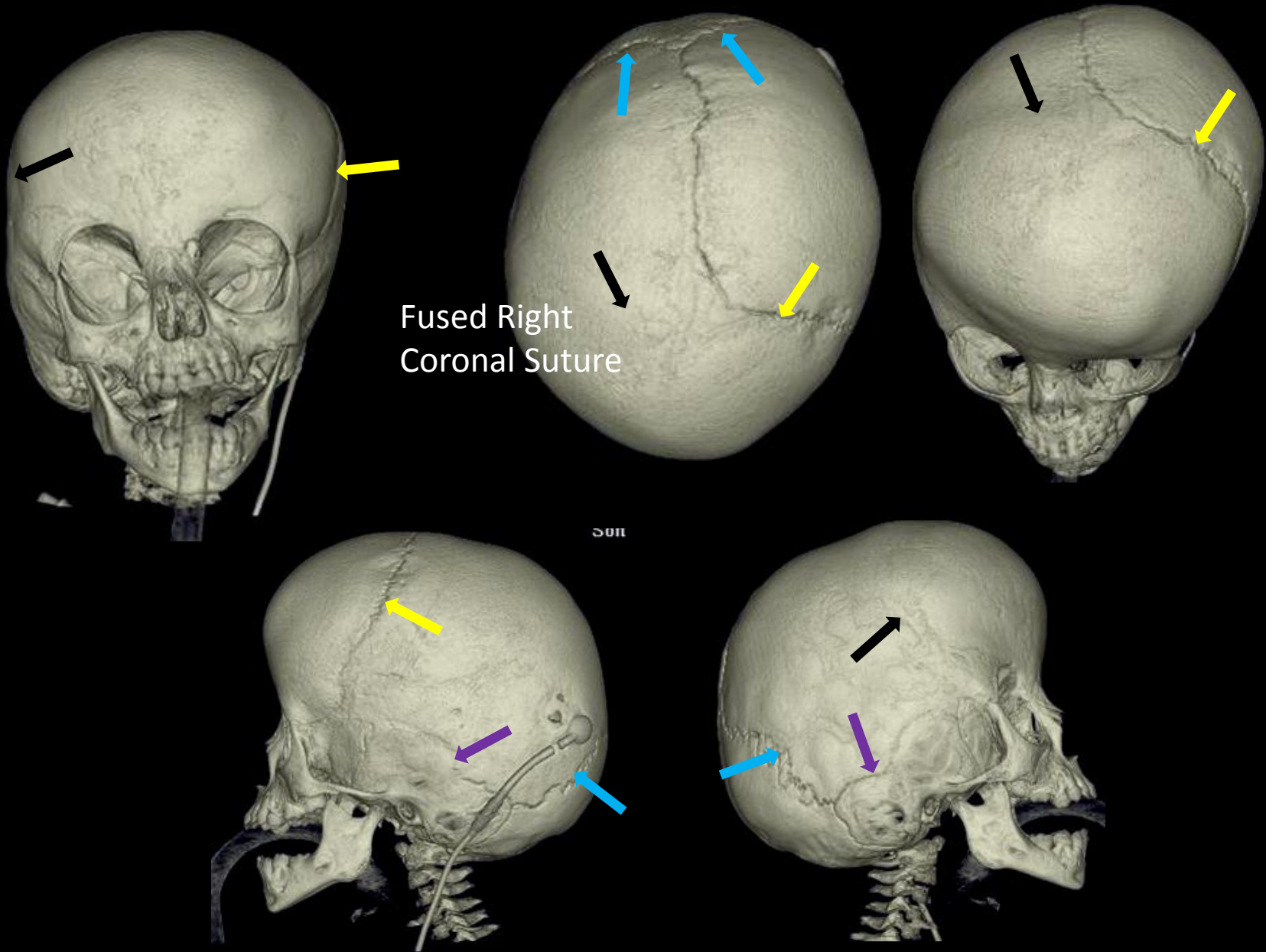
- The coronal suture is responsible for growth in the AP direction
- Premature fusion of the coronal sutures may be unilateral or bilateral
- Unicoronal synostosis is more likely to be an isolated non-syndromic event than its bilateral counterpart

Unicoronal Synostosis

- Unicoronal synostosis results in anterior plagiocephaly
- 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





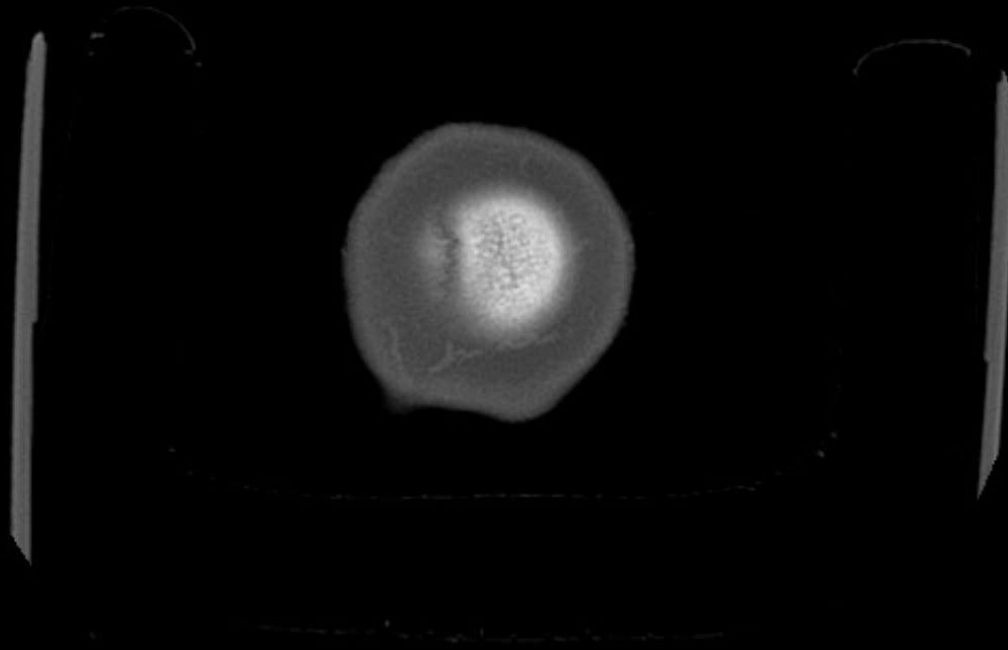
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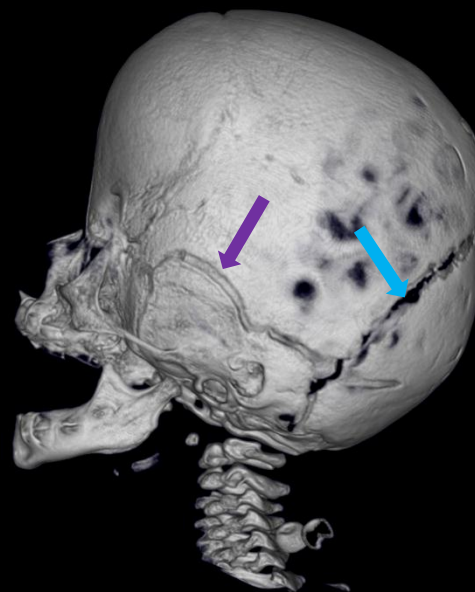
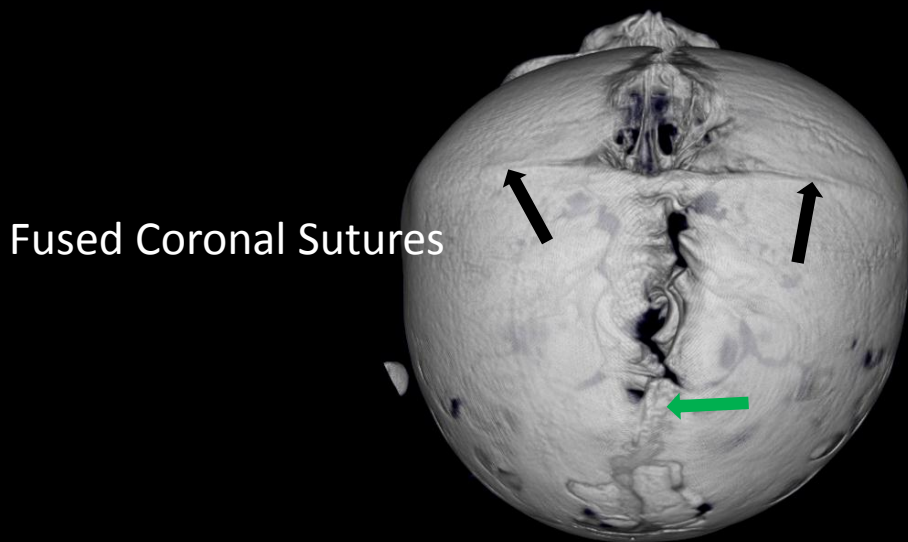
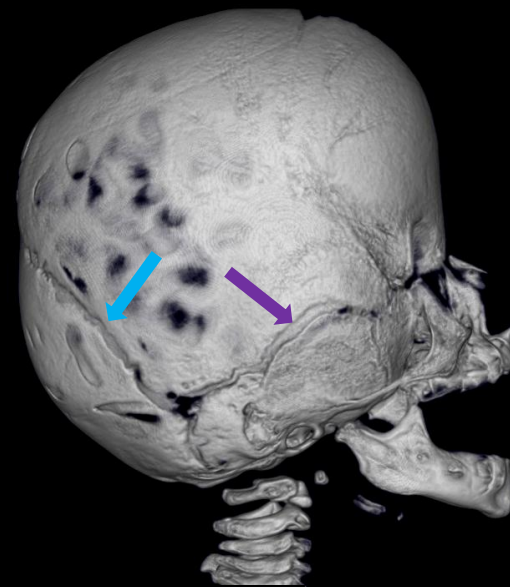
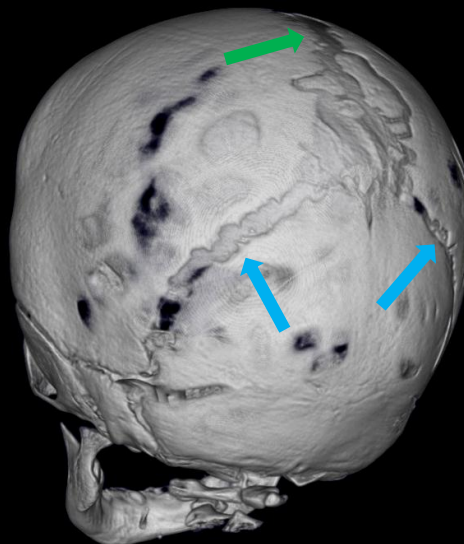
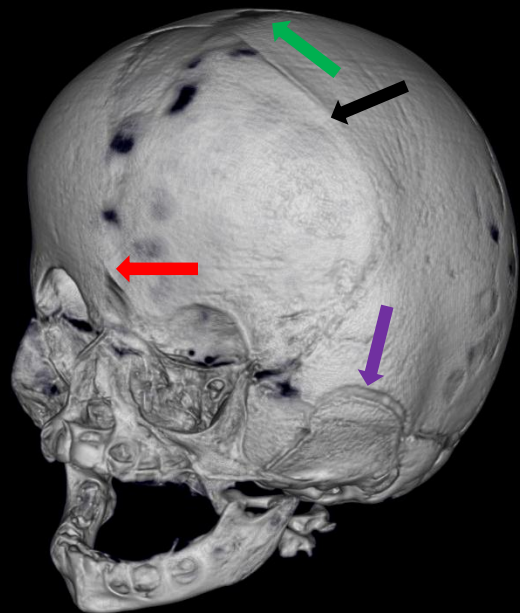
	Coronal		Metopic		Sagittal		Lambdoid		Squamosal
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Bicoronal Synostosis


- 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





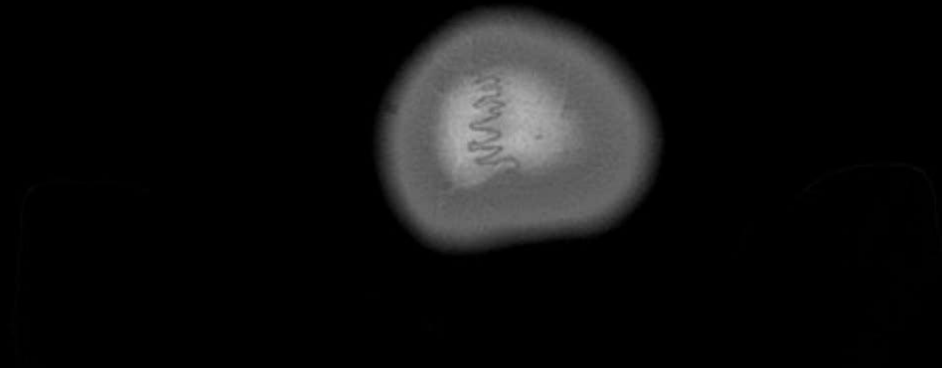
Cranial Sutures:

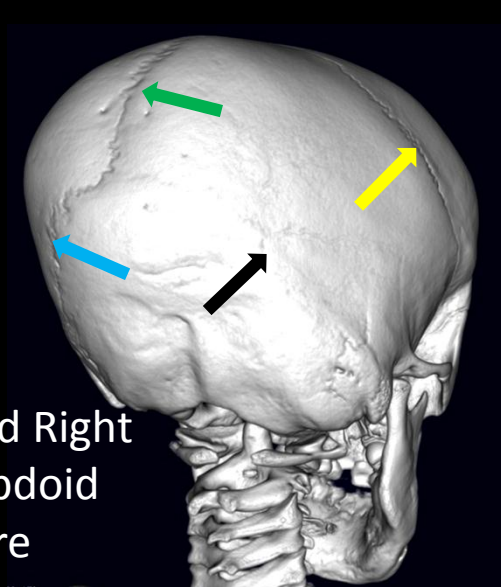
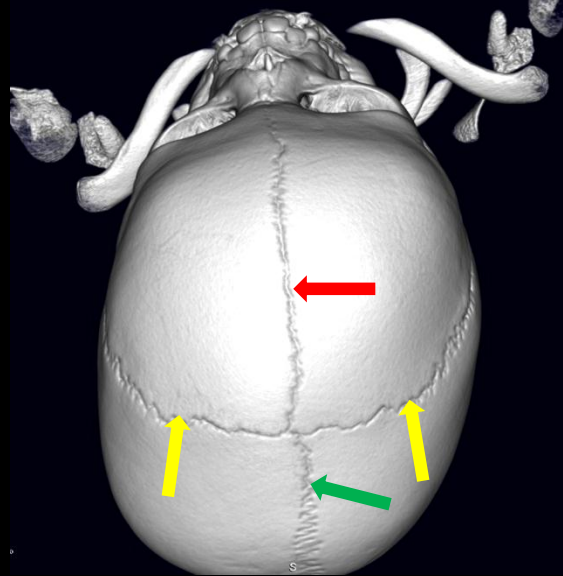
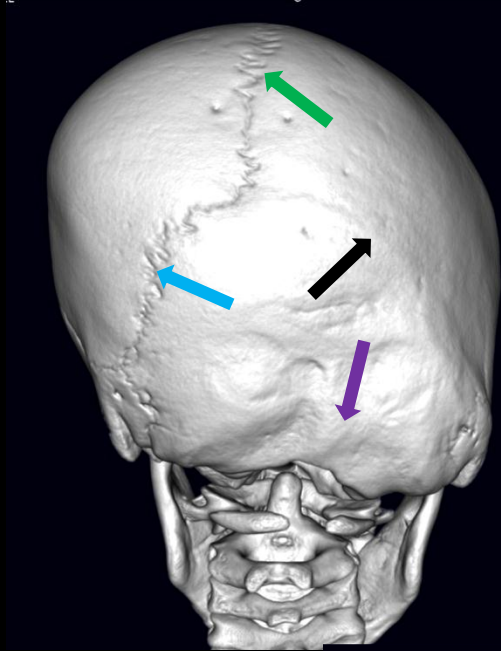
	Coronal		Metopic		Sagittal		Lambdoid		Squamosal
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Lambdoid Synostosis

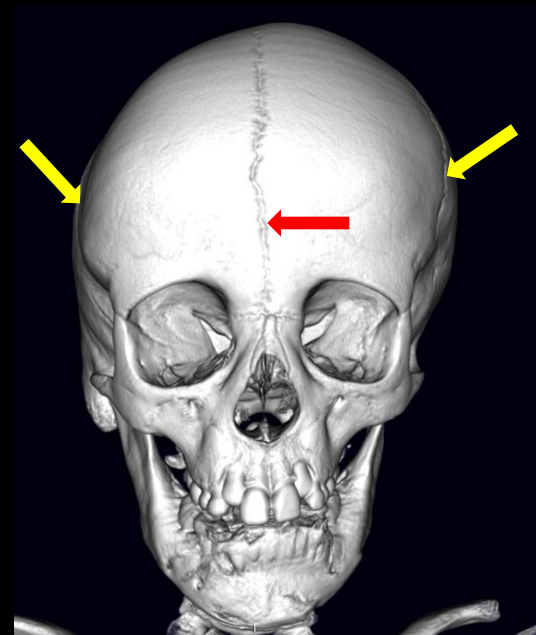
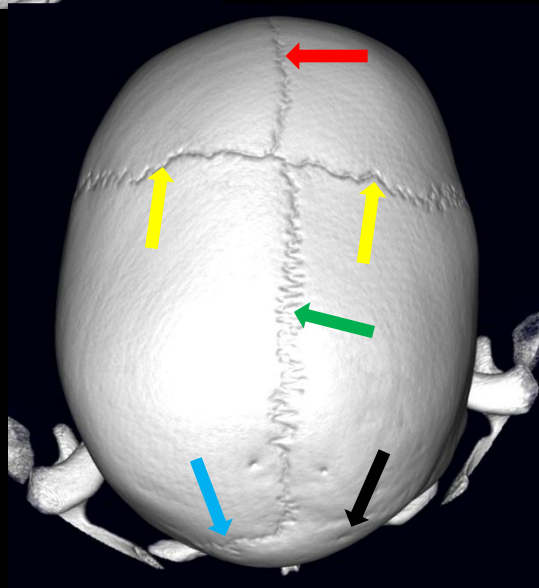
- Rarest of the isolated synostoses
- It accounts for 1% of all synostoses
- Radiological signs:
 - Absence of suture
 - Flattening over synostosed suture
 - Mastoid overgrowth resulting in bulge behind involved ear
 - Temporo-parietal bossing on opposite side

Right Lambdoid Synostosis on axial CT





Fused Right
Lambdoid
suture



Cranial Sutures:

	Coronal		Metopic		Sagittal		Lambdoid		Squamosal
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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

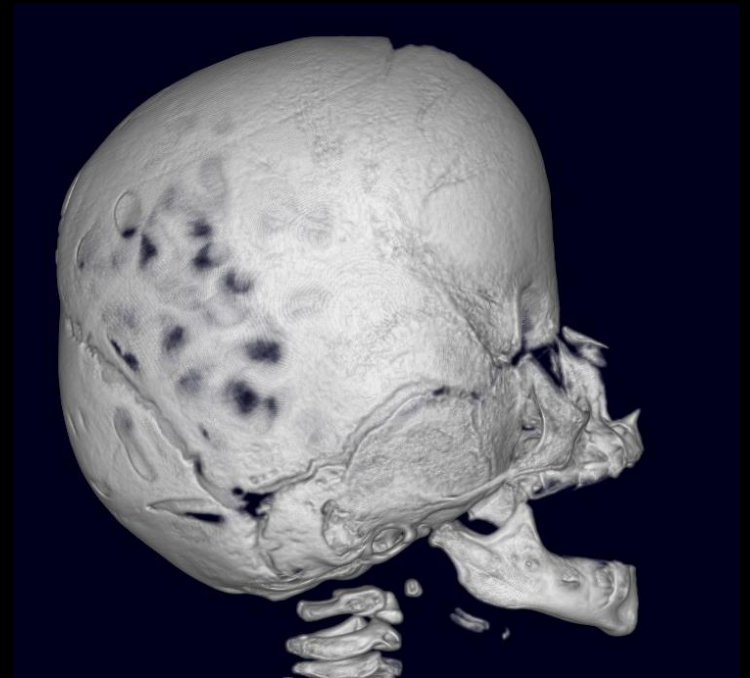
- Mutation in FGFR2 or FGFR3
- Bicoronal synostosis
- Mid-face hypoplasia
- Normal hands



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

Muenke Syndrome

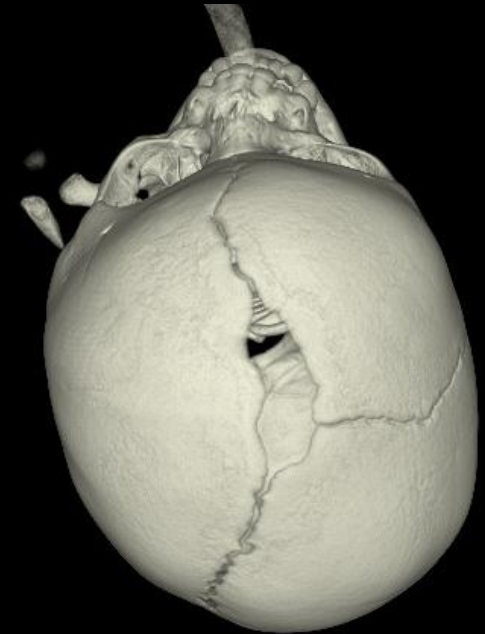
- P250R FGFR3 mutation on chromosome 3
- Coronal synostosis
- Bitemporal bossing
- Sensorineural hearing loss



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)



*Left unicoronal synostosis
in child with Saethre-
Chotzen syndrome*

Pfeiffer Syndrome

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

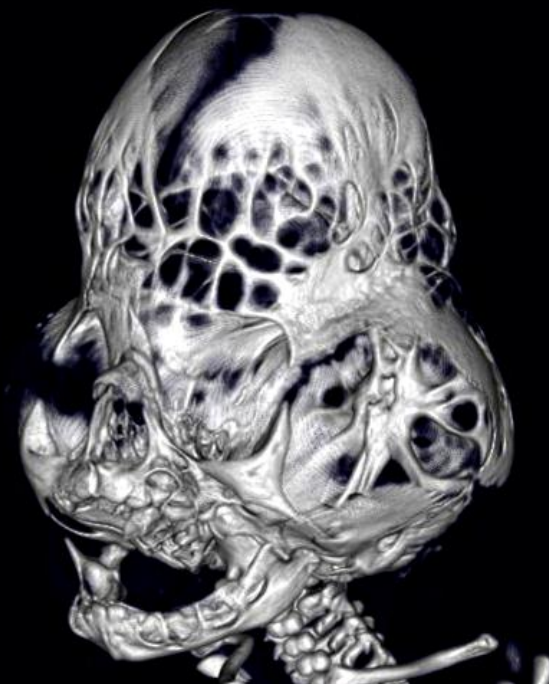
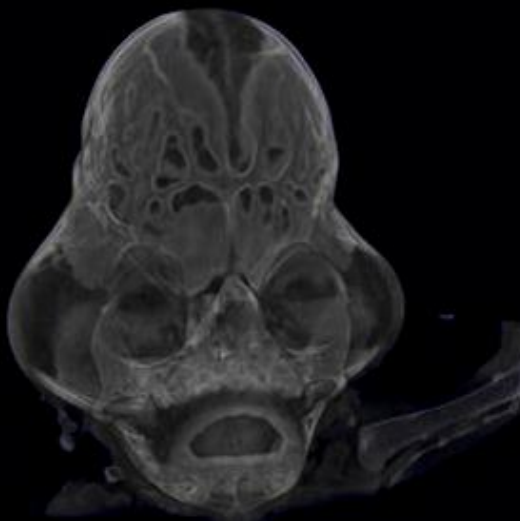
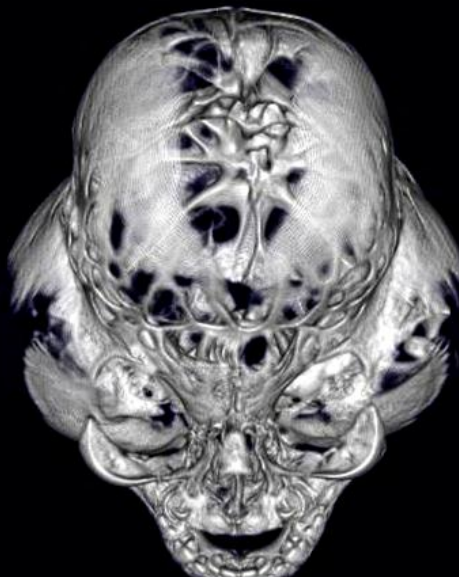


AP radiograph of right foot showing broad great toe in child with Pfeiffer syndrome

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

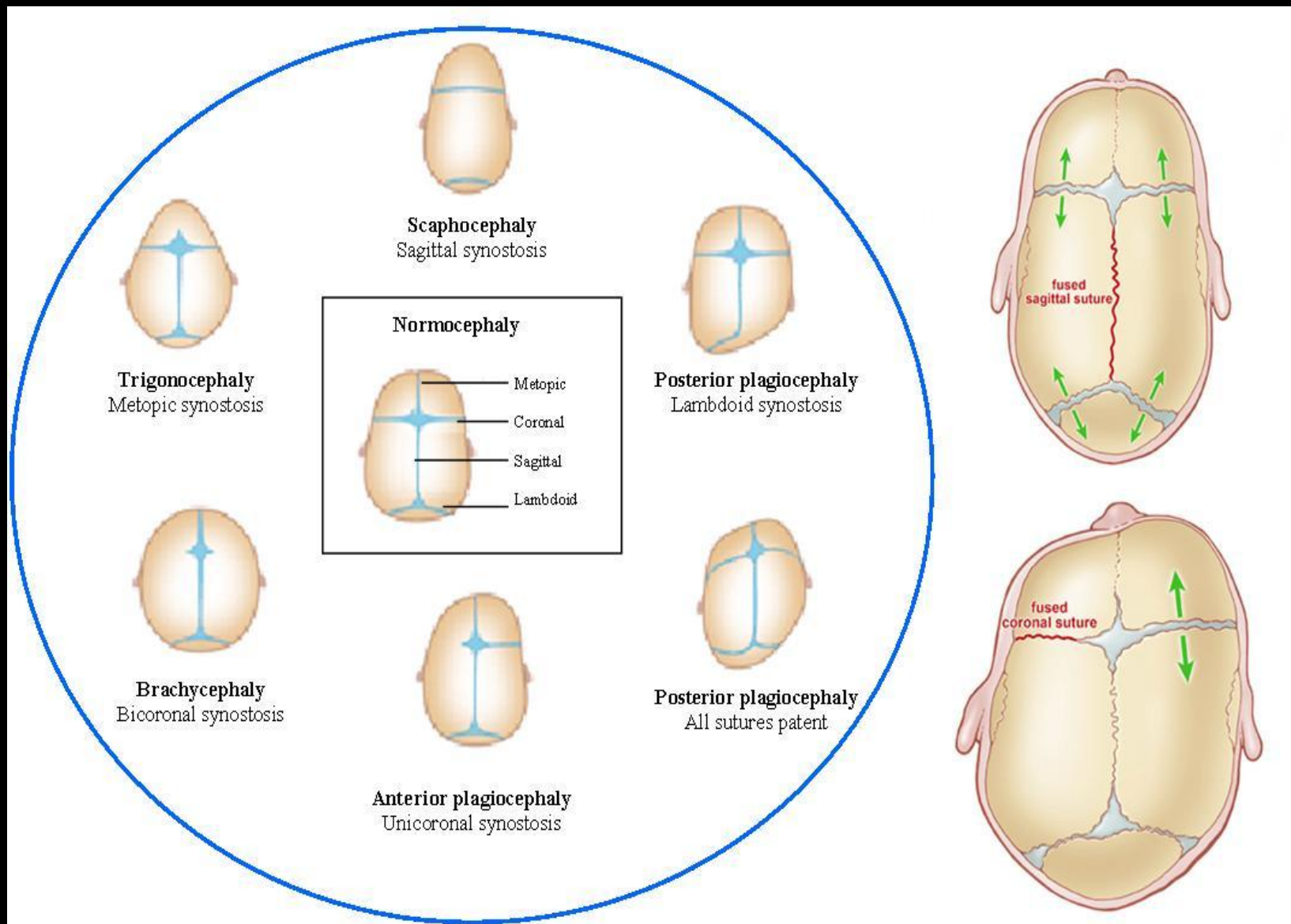


Lateral skull radiograph showing bilateral distractors and distraction gap. This is posterior distraction.

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



References:

<http://www.cincinnatichildrens.org/health/c/craniosynostosis/>

<http://www.ohsu.edu/xd/health/services/doernbecher/programs-services>

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

- Johnson D, Wilkie AO. Craniosynostosis. Eur J Hum Genet. 2011; 19(4): 369-76
- Branson HM, Shroff MM. Craniosynostosis and 3-Dimensional Computed Tomography. 2011; 32(6): 569-577
- Kirmi O, Lo SJ, Johnson D, Anslow P. Craniosynostosis: A radiological and surgical perspective. Semin ultrasound CT MR. 2009; 30(6):492-512