Craniosynostosis

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25 May 2006

Definition

- Heterogeneous group of disorders with premature fusion of cranial sutures
  - Non-syndromic most common
  - Also associated with >150 syndromes

Incidence

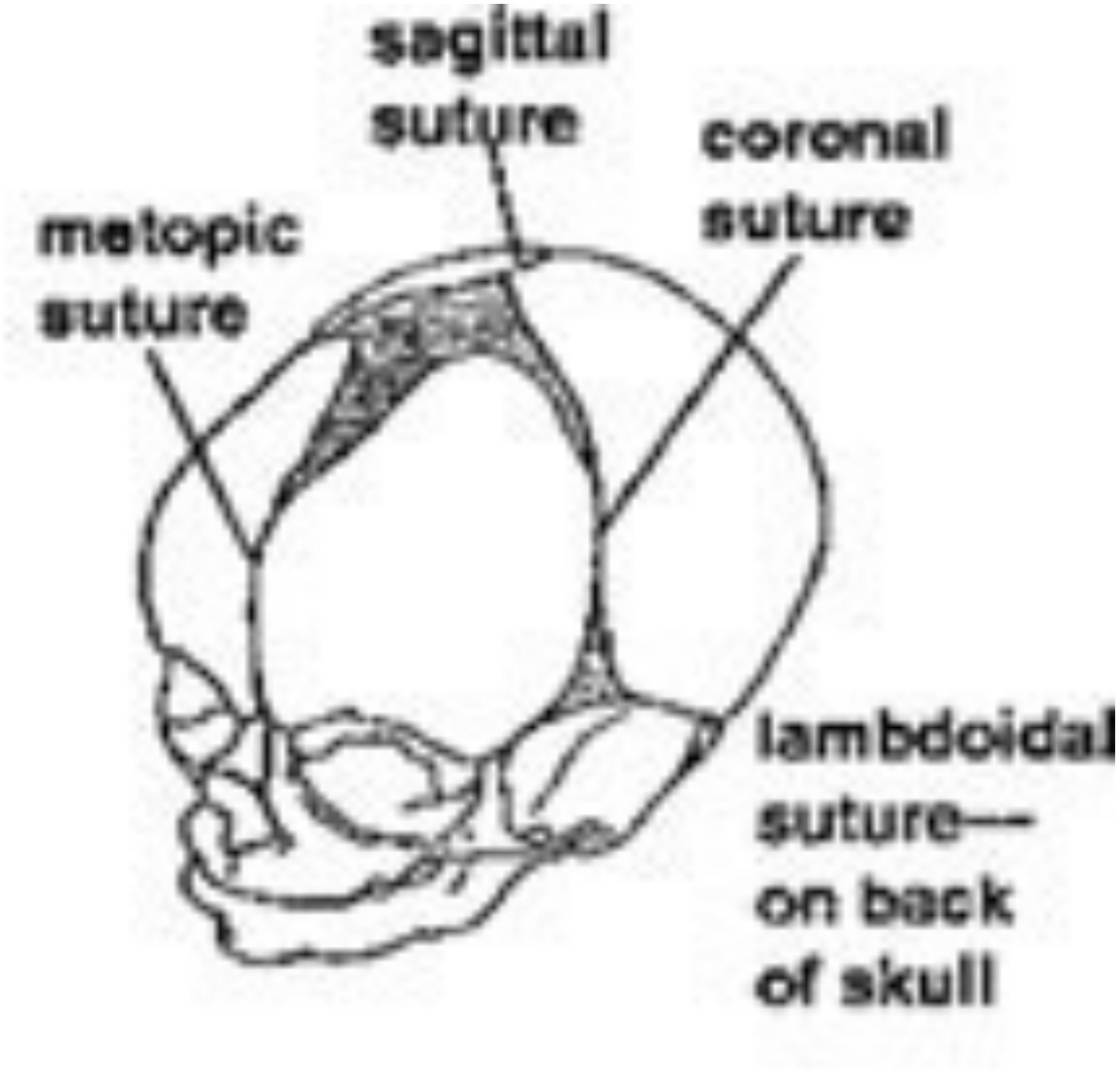
- 5-6 cases per 10,000 live births
  - Saggital synostosis most common
  - Followed by unilateral coronal, bilateral coronal, metopic, then lambdoid

Etiology

- Non-syndromic most common
  - Syndromic in 10-20% of cases
- Approx 14% of pedigrees with Craniosynostosis demonstrated familial aggregation
  - Predominantly autosomal dominant with variable penetrance
  - 60% penetrance and 61% sporadic incidence

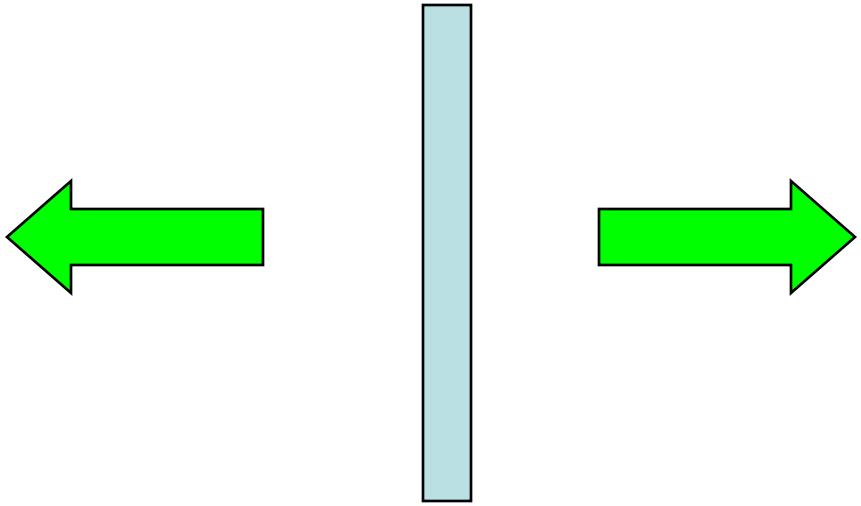
(Lajeunie *et al*, Am J Med Genet **55**:500-504, 1995)

Infant Skull Anatomy

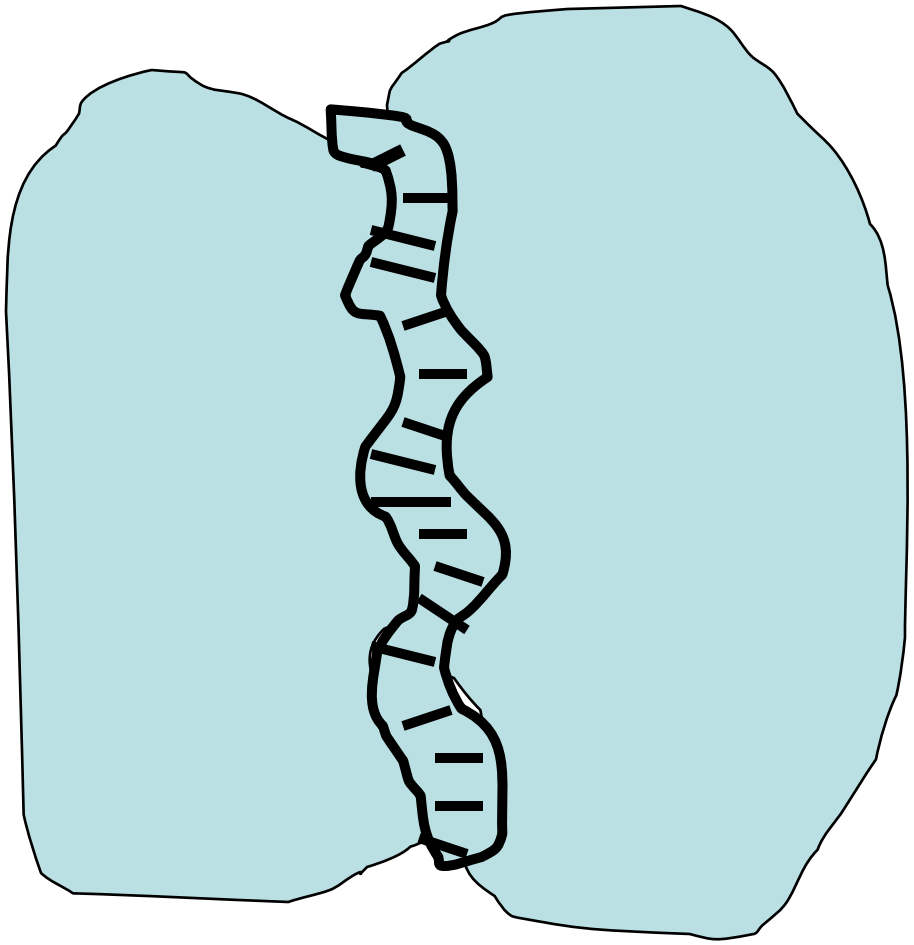


Suture Growth

- Sutures allow growth perpendicular to them
  - Growth at suture lines related to brain growth



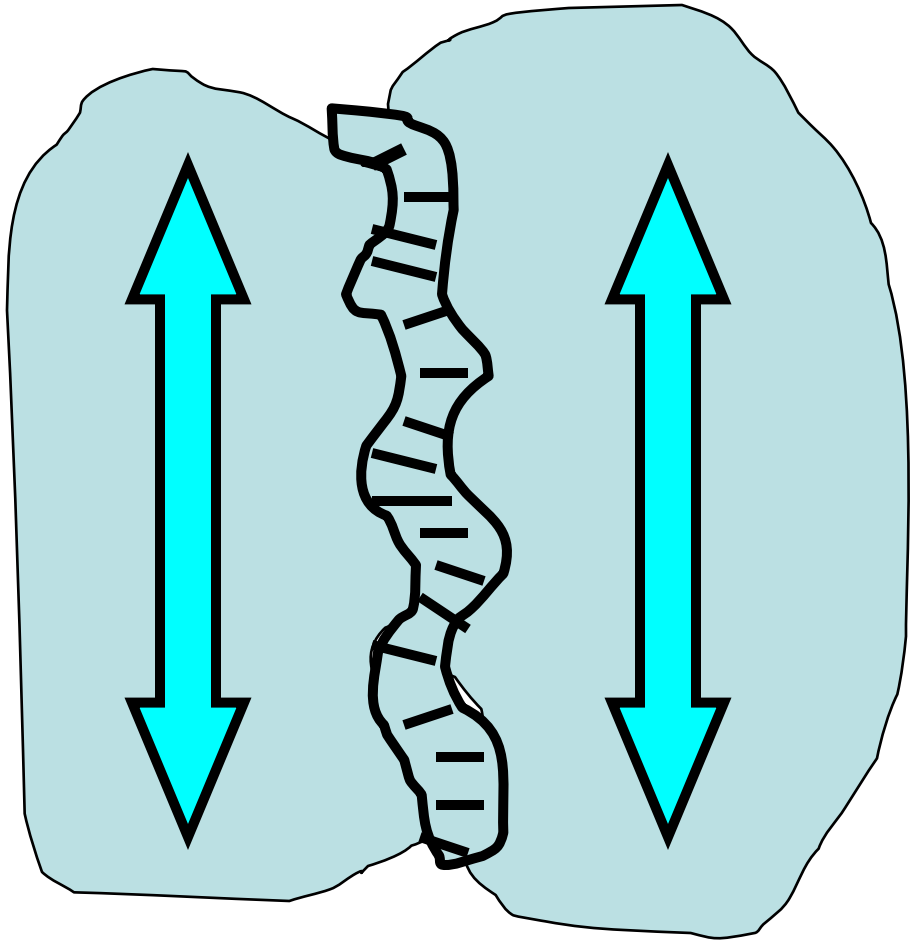
Suture Closure



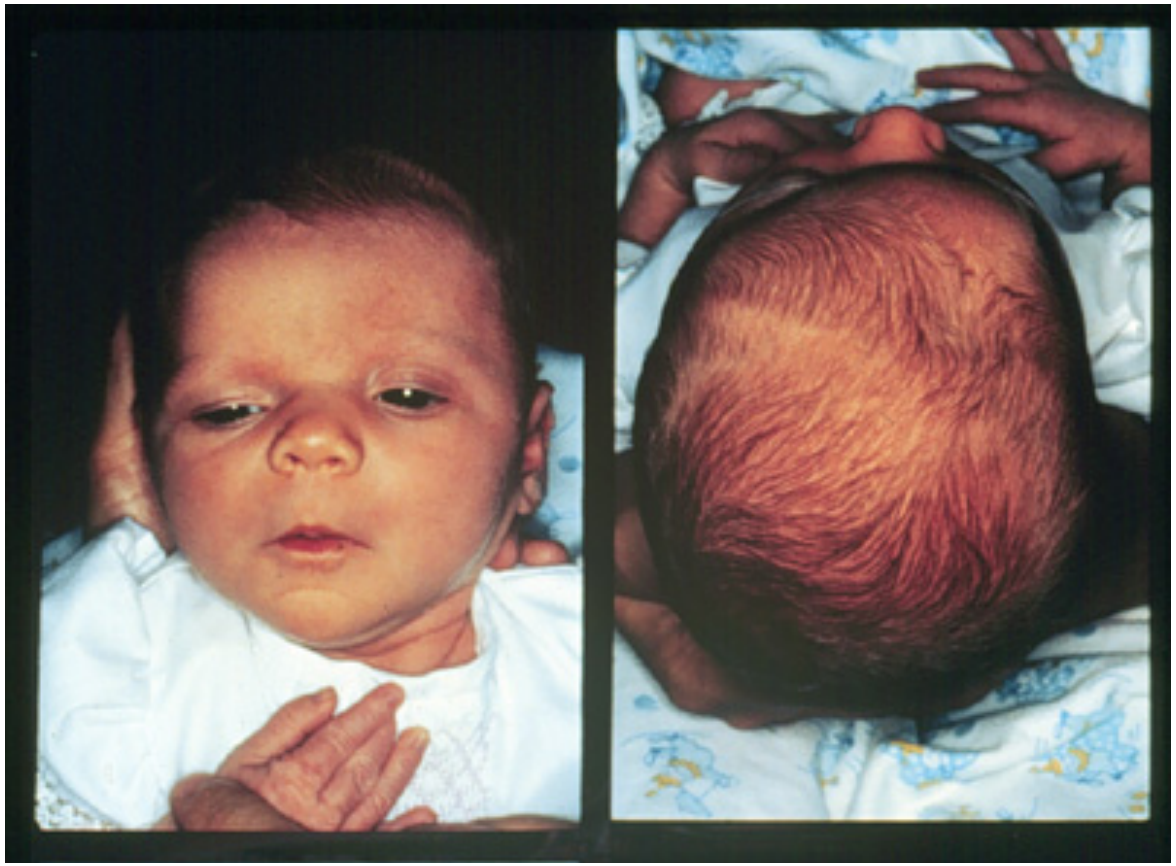
Ossification
By Age 8

Bone Union
By Age 20

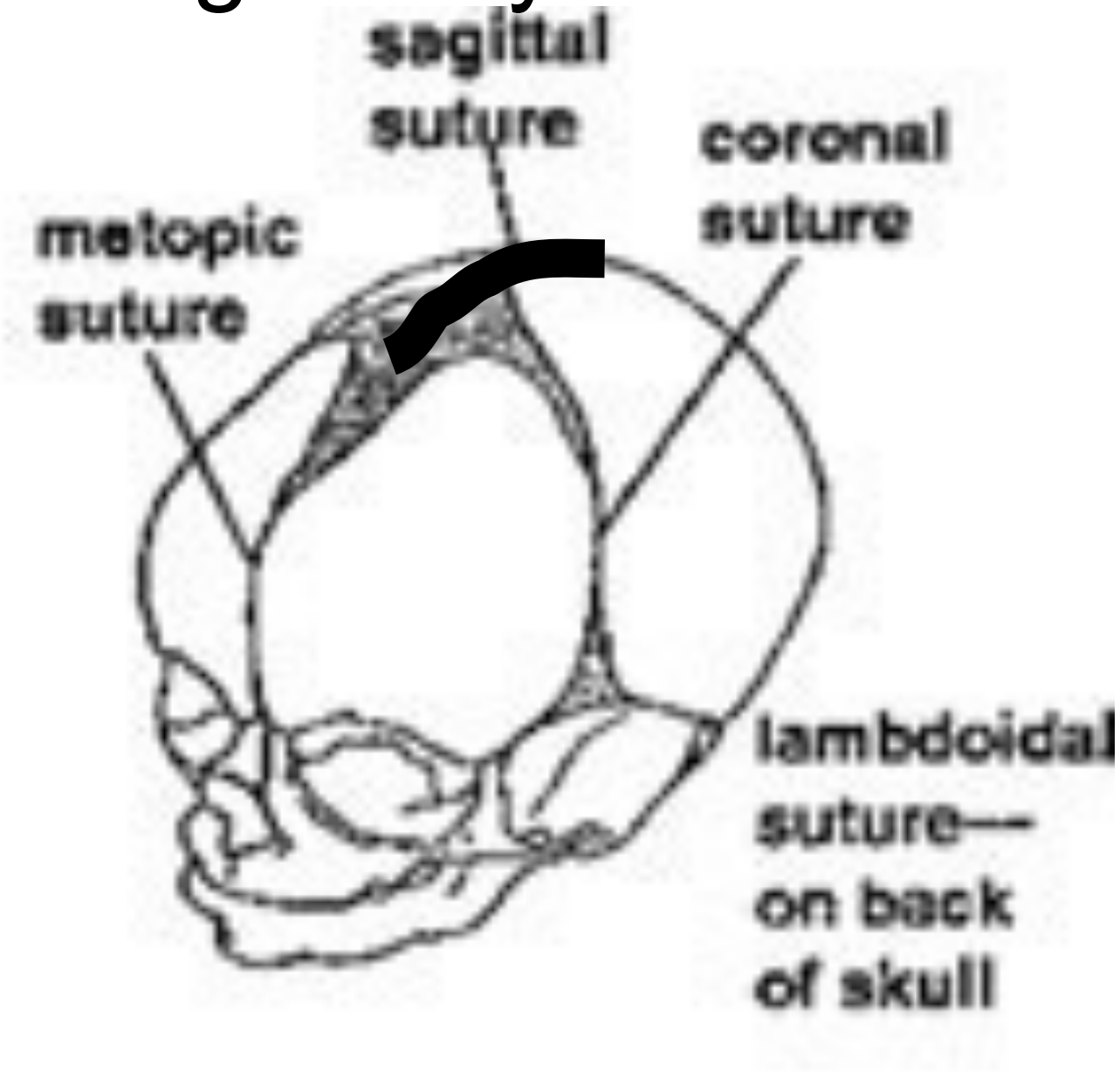
Early Closure Causes Growth Parallel to the Suture



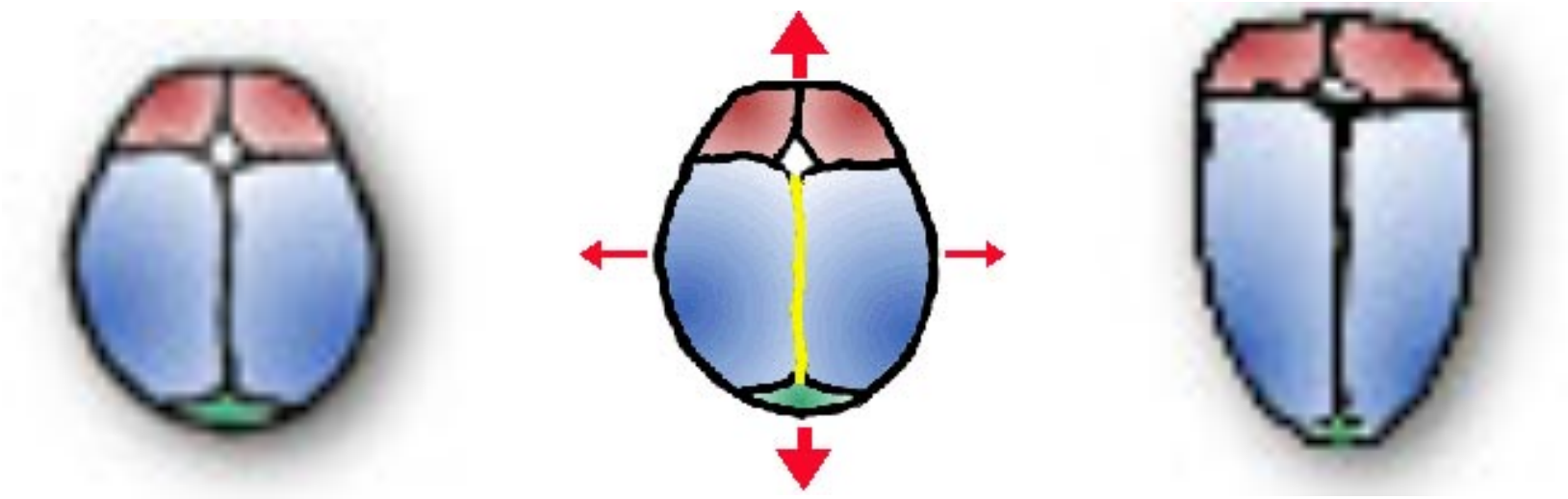
Craniosynostosis: Early Fusion of a Suture



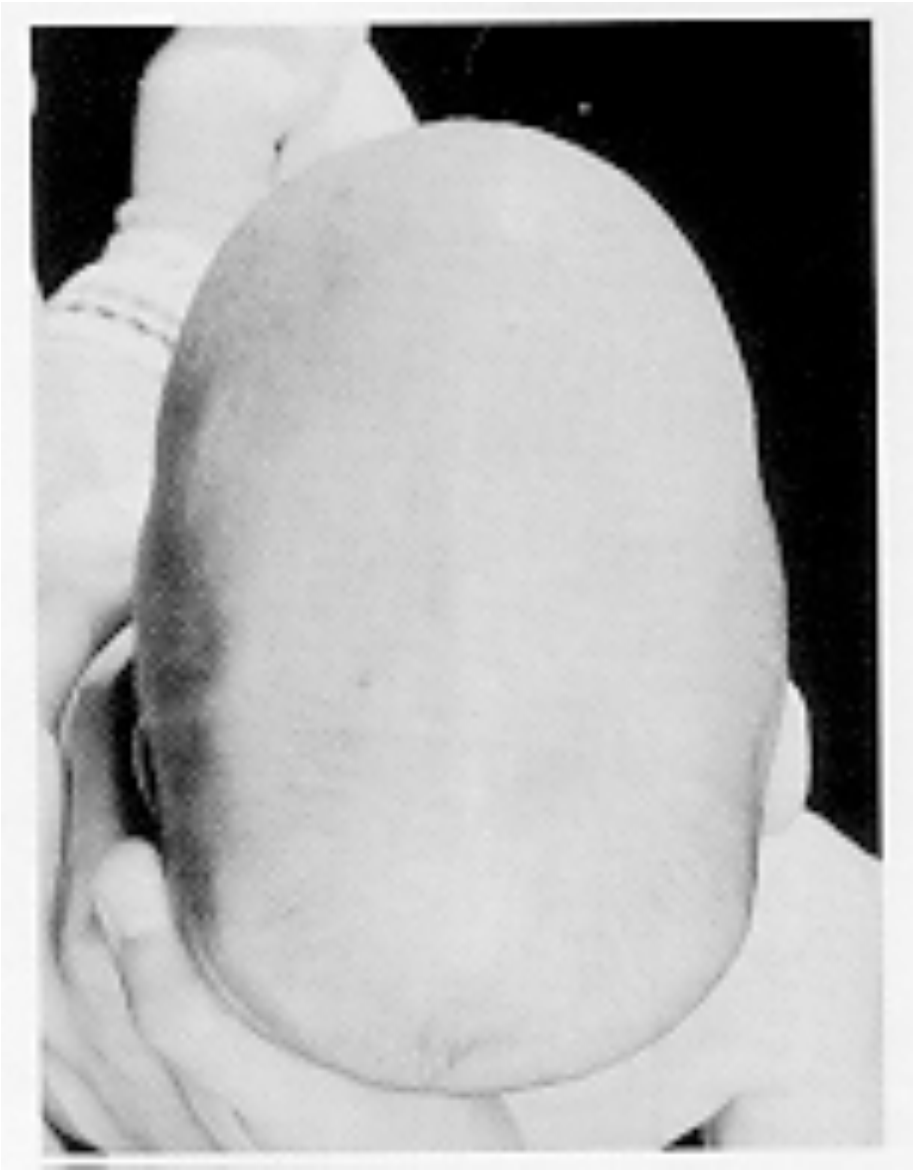
Sagittal Synostosis



Sagittal Synostosis

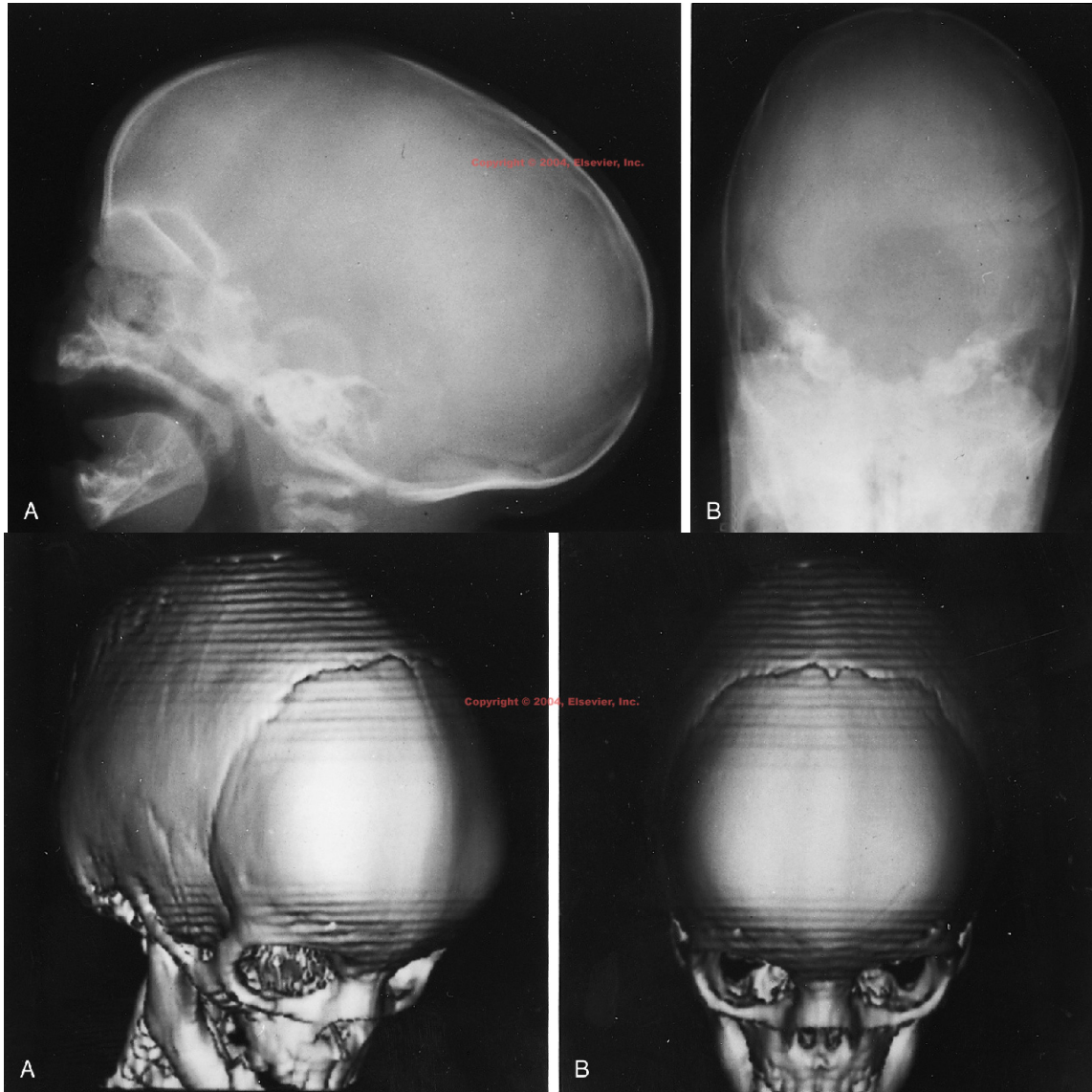


"Boat-Head"
(Scaphocephaly)



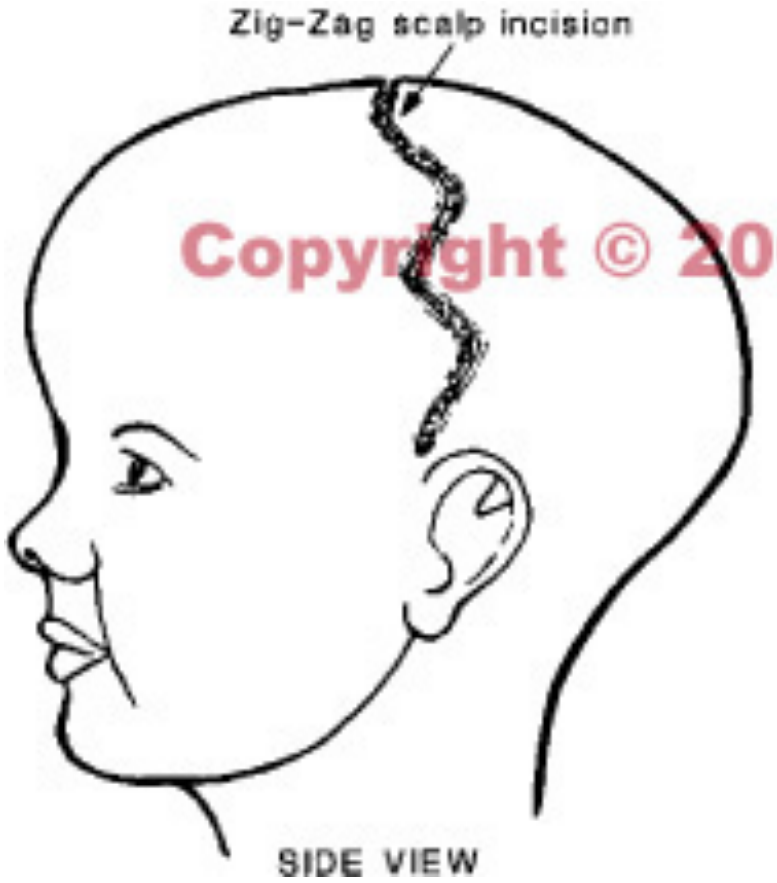
Sagittal synostosis

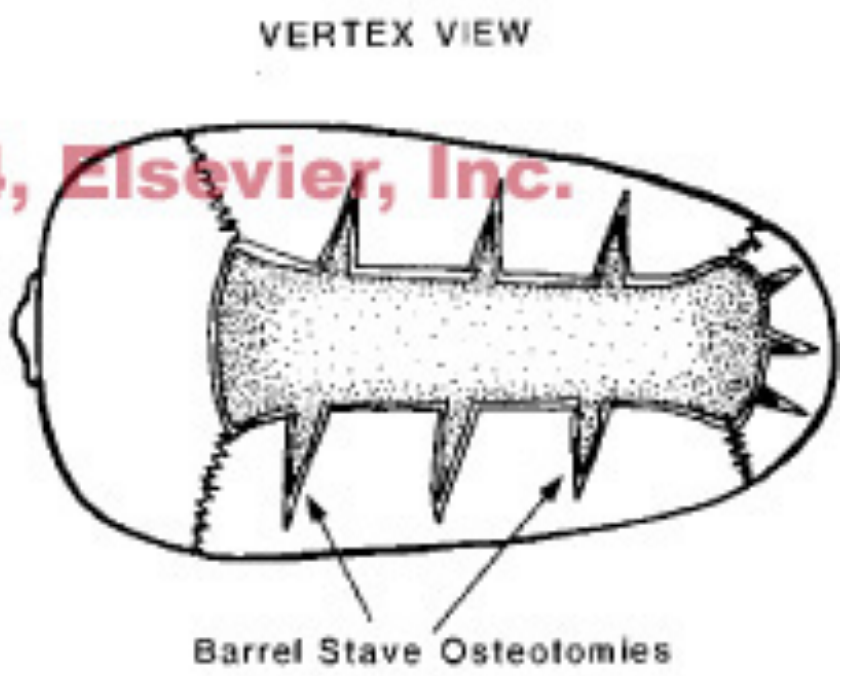
- May be diagnosed by clinical exam
- Characteristic appearance on plain films and CT



Early surgical correction

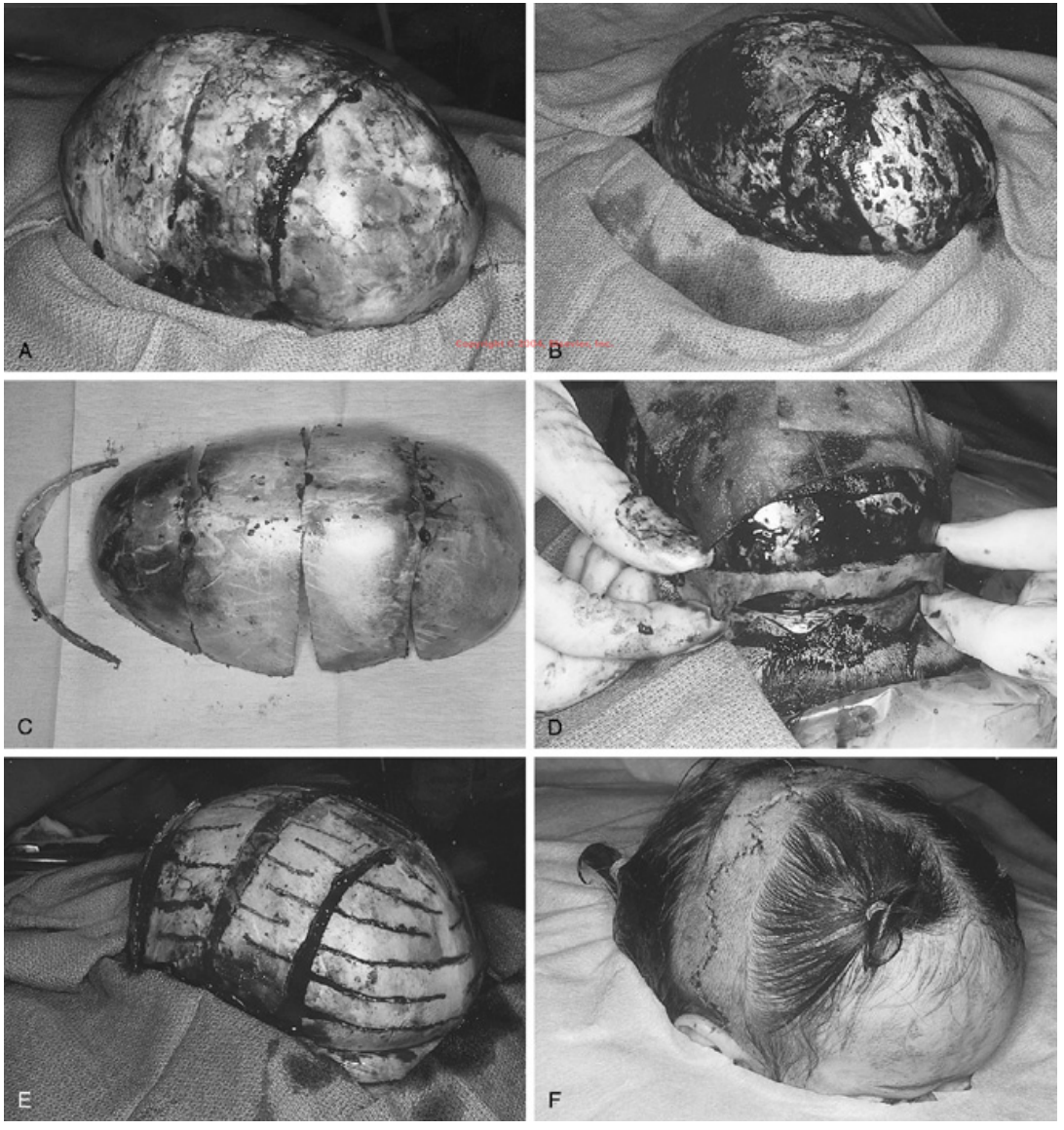
- Midsagittal craniectomy and barrel stave osteotomies
- Good success when less than 6 months of age



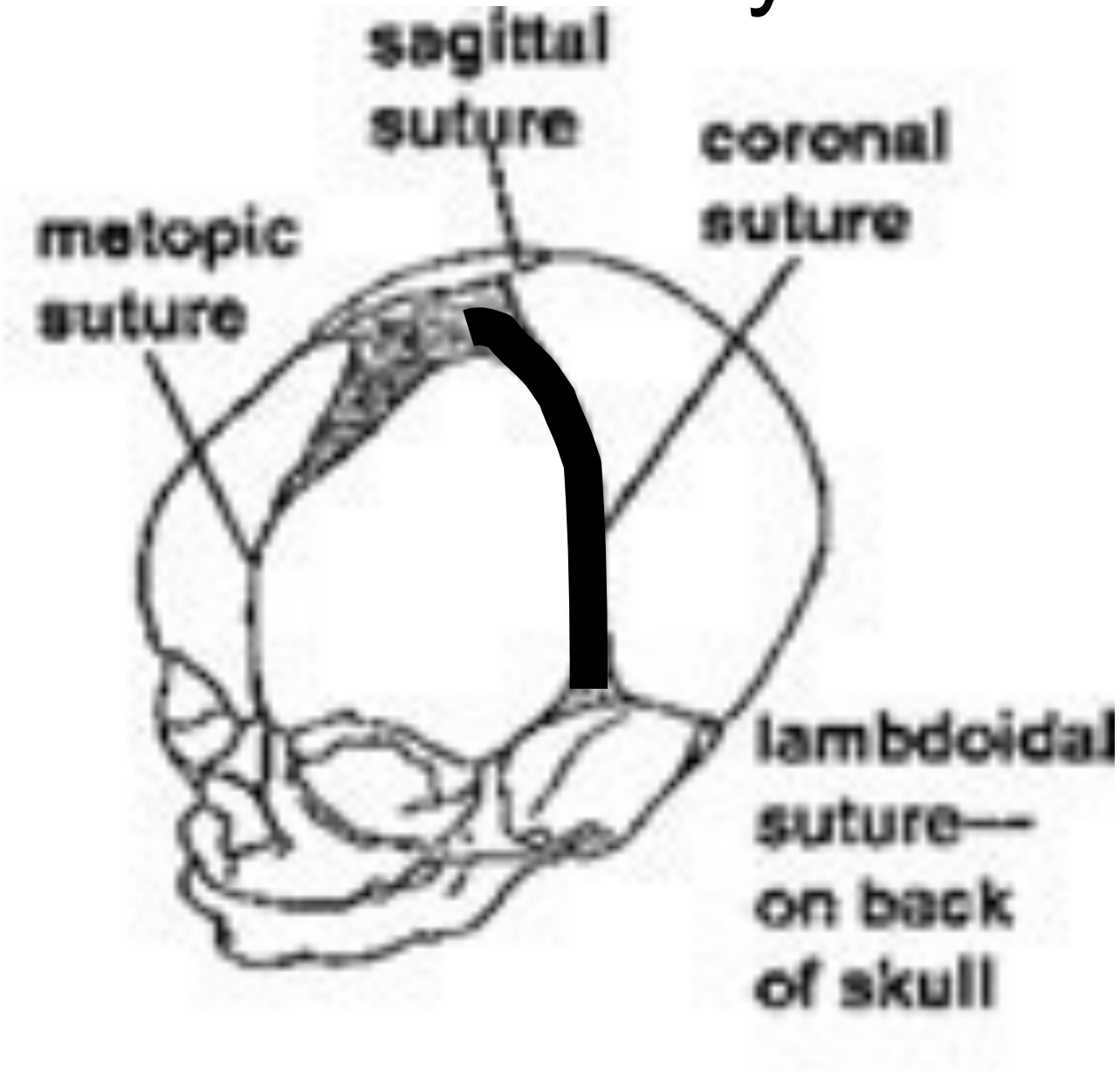


Late surgical correction

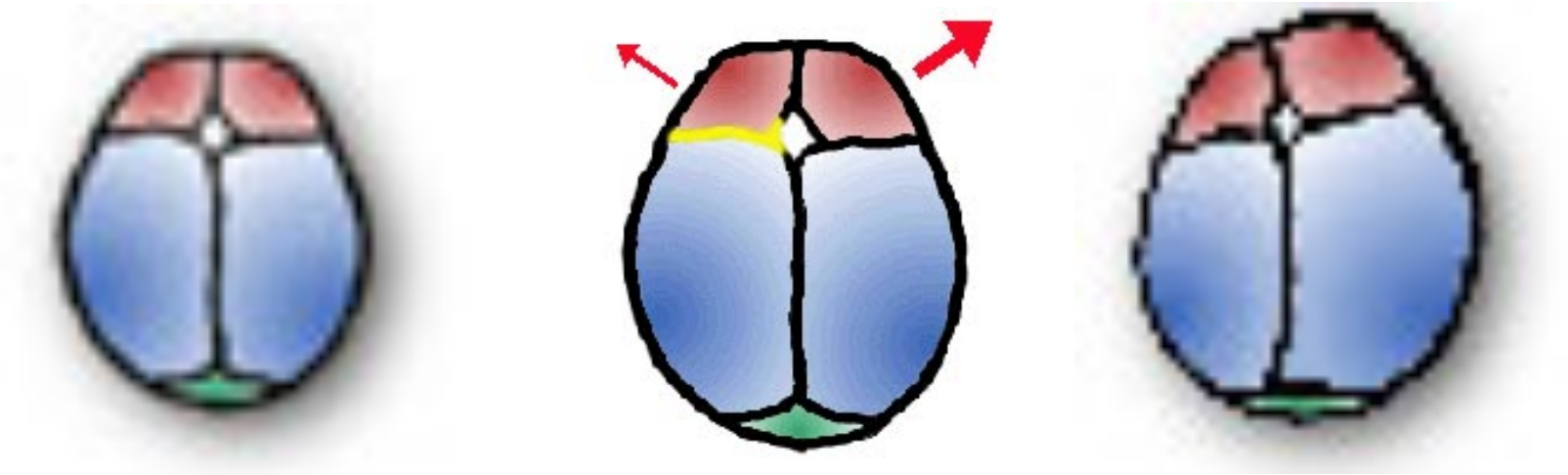
- More extensive cranial vault remodelling
- Usually in collaboration with plastic surgery



Unilateral Coronal Synostosis

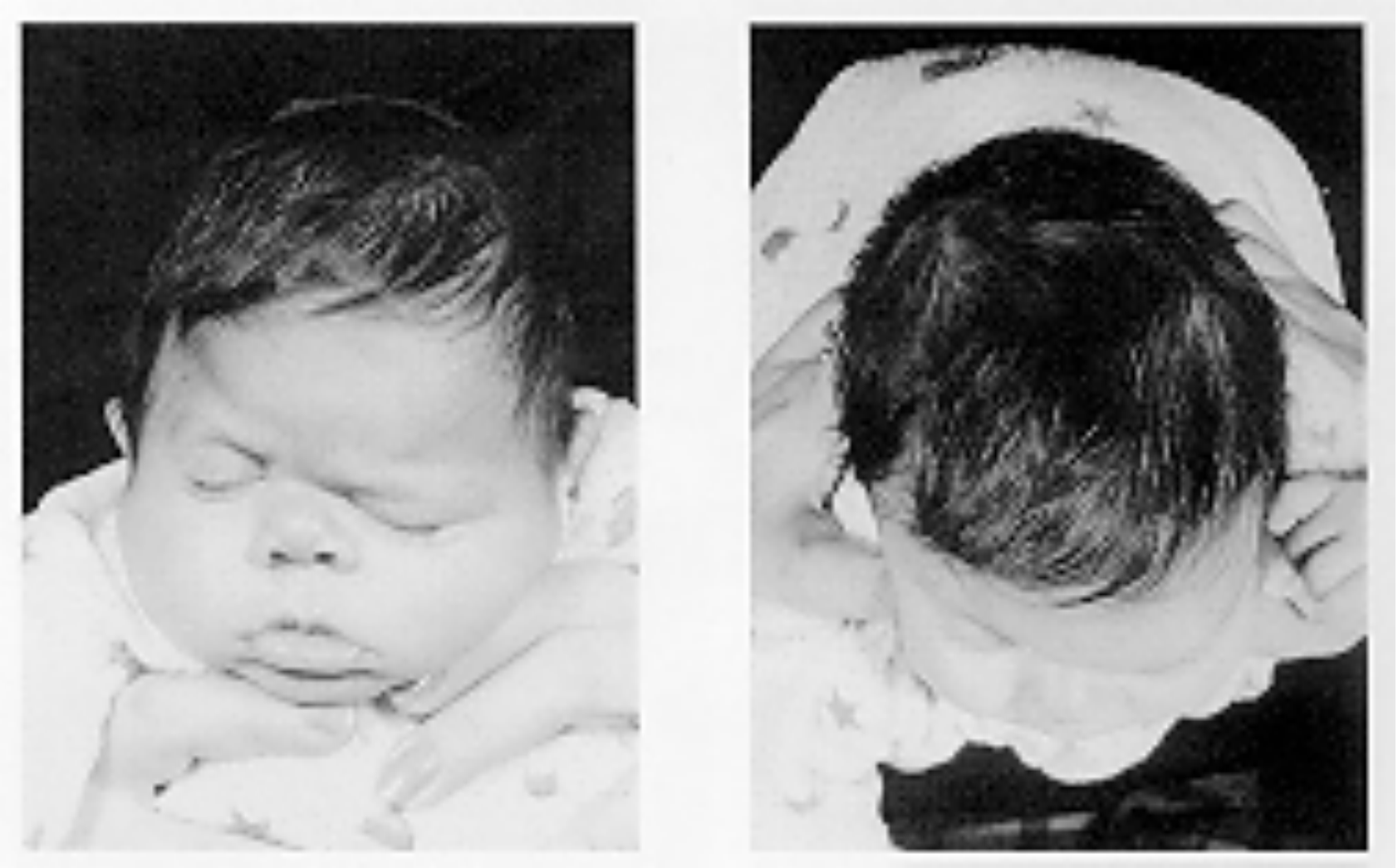


Coronal Synostosis

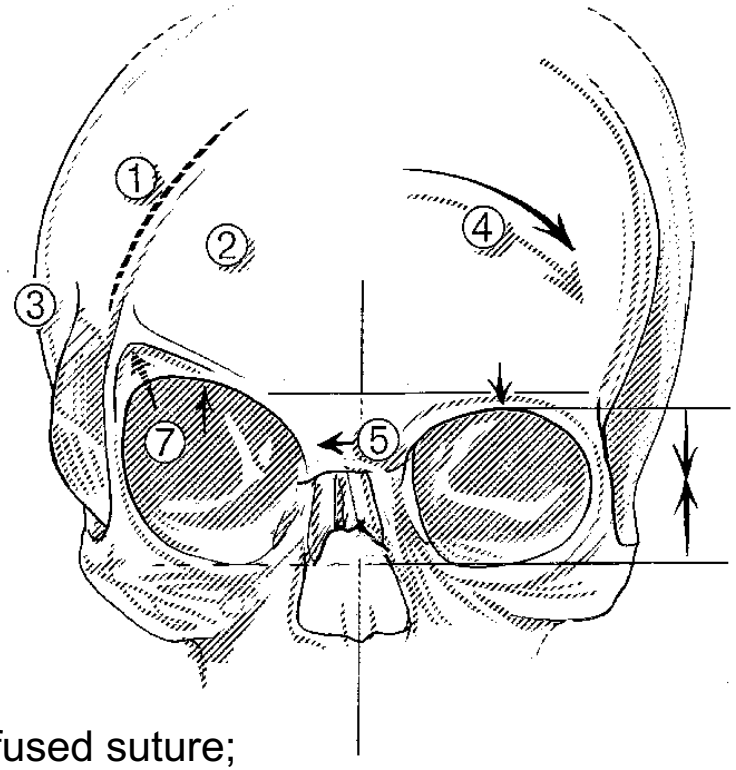


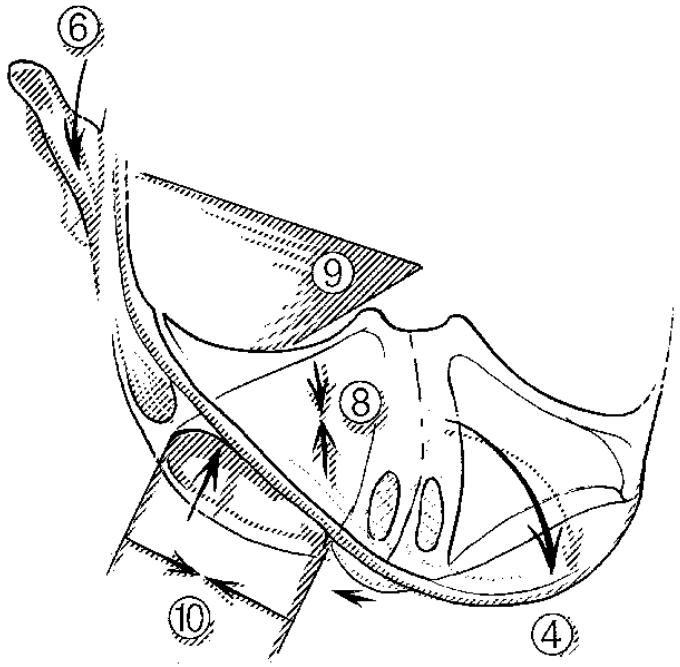
"Bent-Head"
(Plagiocepahly)

Right Coronal Synostosis

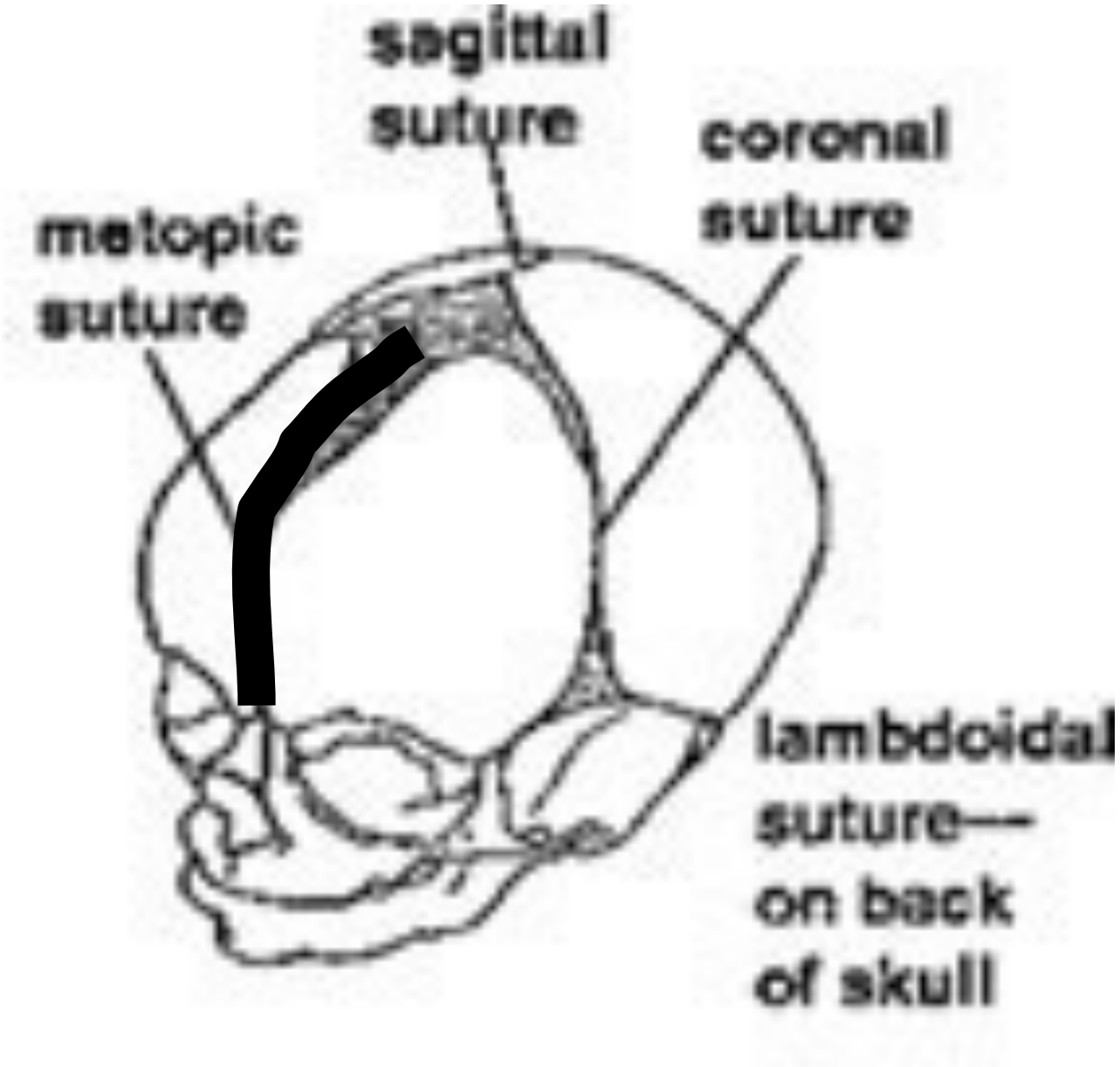


Unilateral Coronal Svnostosis

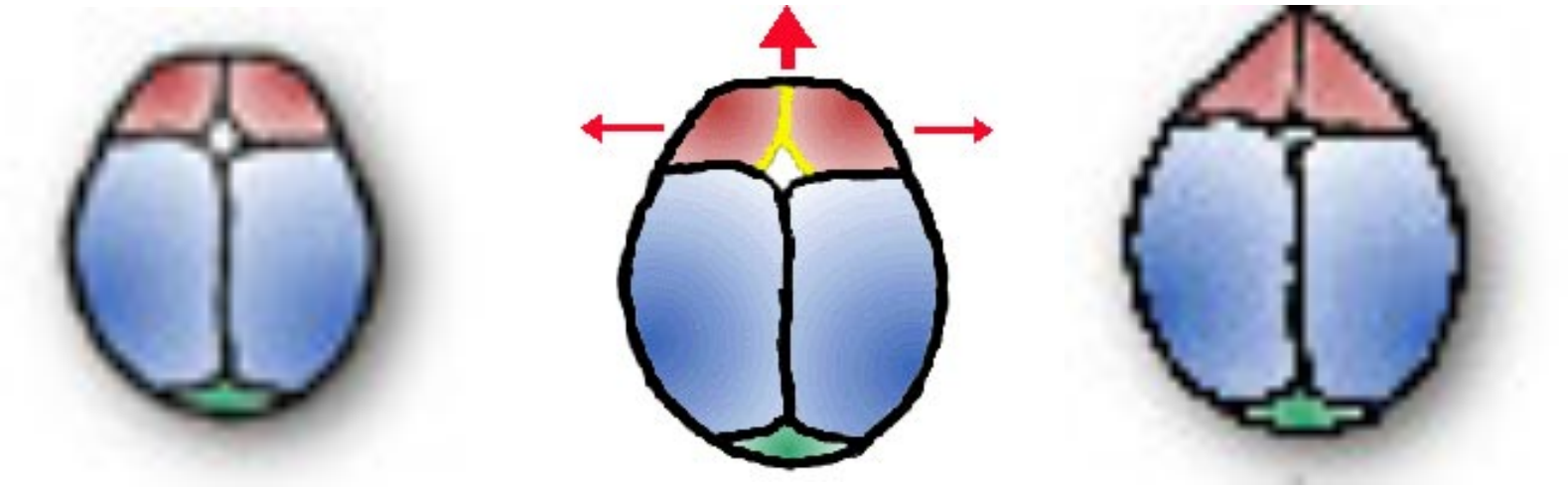




- 1, fused suture;
- 2, flattening of ipsilateral frontal bone;
- 3, bulging of right squamous temporal bone;
- 4, bulging of contralateral frontal bone;
- 5, nasal radix deviated to ipsilateral side;
- 6, ear ipsilateral to fused suture displaced anteriorly;
- 7, harlequin deformity, shown on radiograph, is superiorly displaced ipsilateral greater wing of sphenoid bone;
- 8, shortening of ipsilateral anterior cranial fossa;
- 9, narrowed ipsilateral sphenopetrosal angle; and
- 10, narrowing of ipsilateral mediolateral dimension of orbit.


sagittal suture
coronal suture
metopic suture
lambdoidal suture—on back of skull

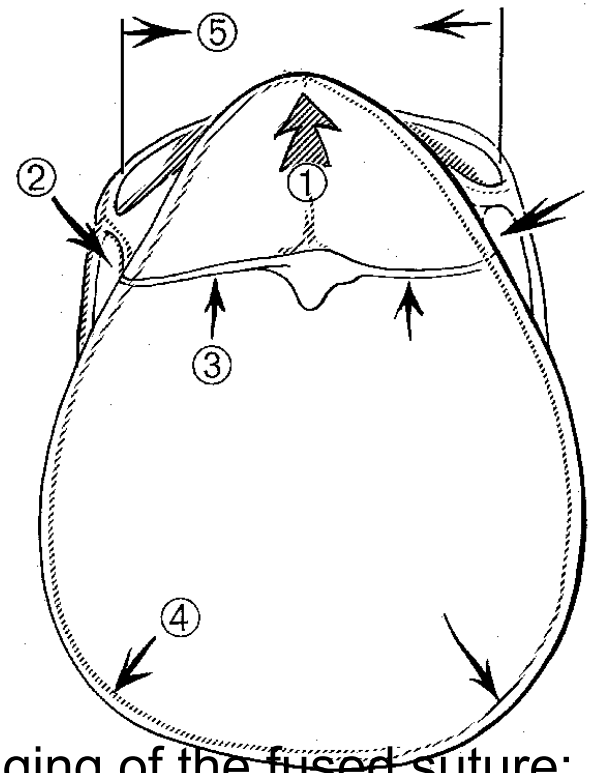
Metopic Synostosis

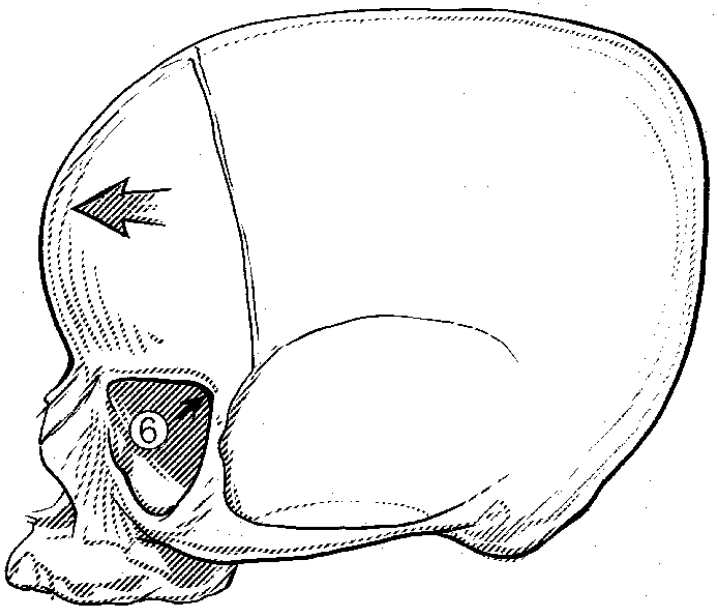


"Triangle-Head"
(Trigonocephaly)

Metopic Synostosis

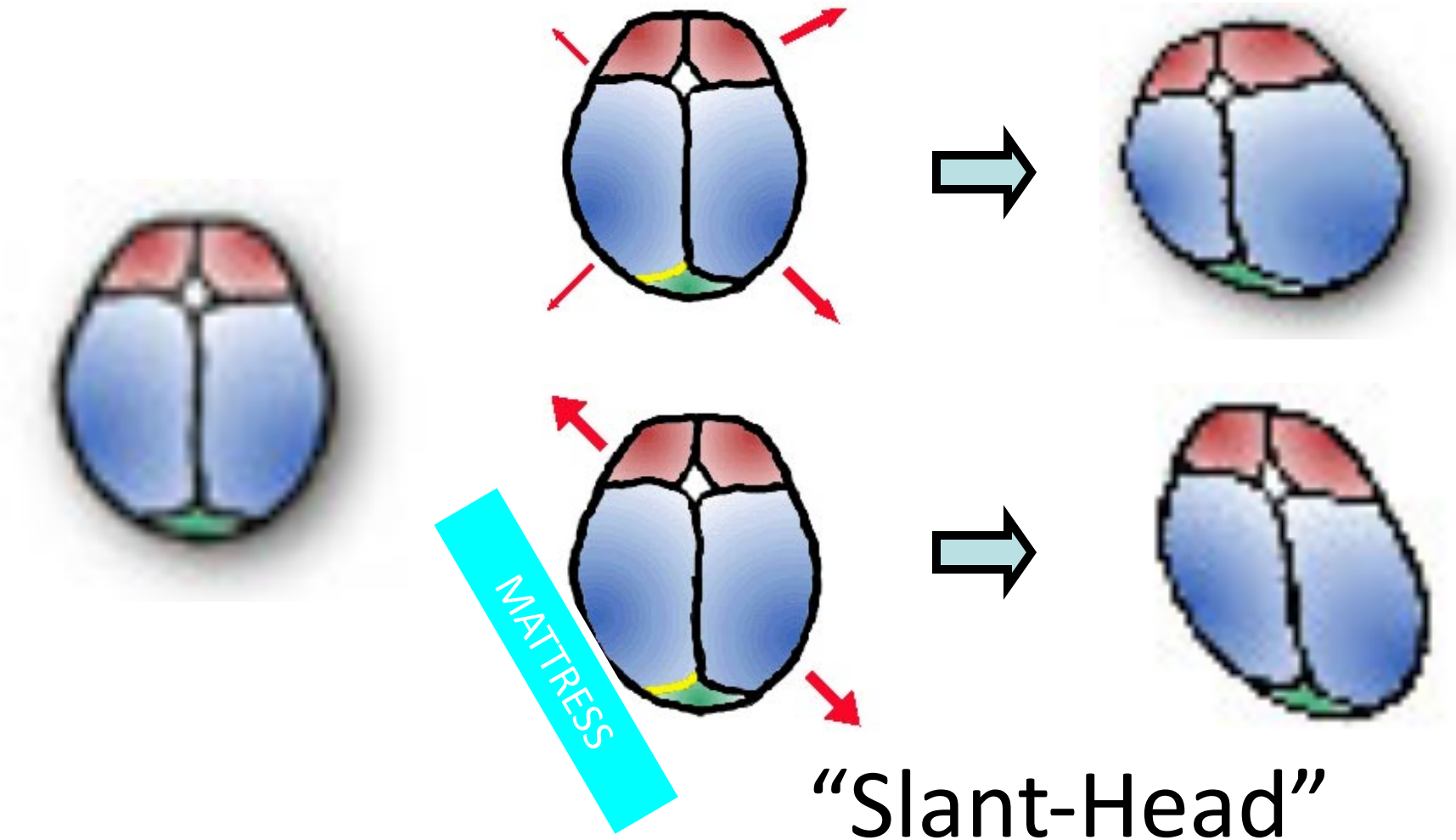
Metopic Synostosis





- 1, ridging of the fused suture;
- 2, temporal narrowing;
- 3, patent coronal suture displaced anteriorly;
- 4, compensatory bulging of the parieto-occipital region, contributing to the pear-shaped appearance of skull;
- 5, narrowed bizygomatic dimension; and
- 6, posterior displacement of superolateral orbital rim.

Lamboid Synostosis



"Slant-Head"
(Occipital Plagiocephaly)

Clinical Exam

- Head circumference
- Head shape (from above, side)
- Ear and facial symmetry
- Palpate suture lines & fontanelles
  - Look for ridging
- Look for associated anomalies
- Skull X-ray or CT

Craniosynostosis

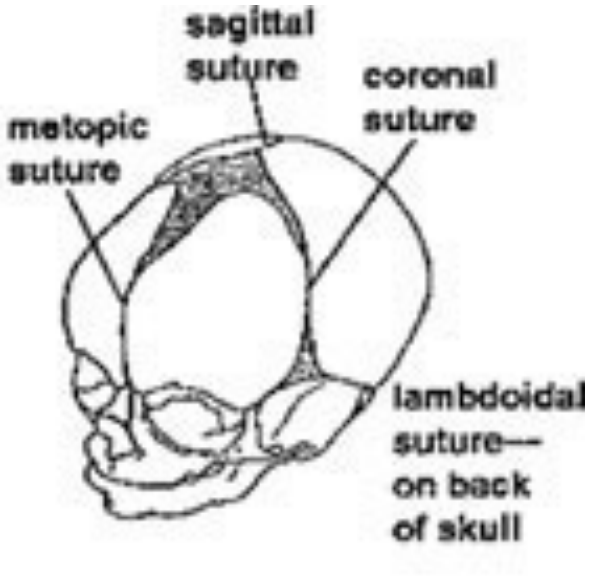
Secondary

Microcephaly

Prematurity

VP Shunting

Positioning

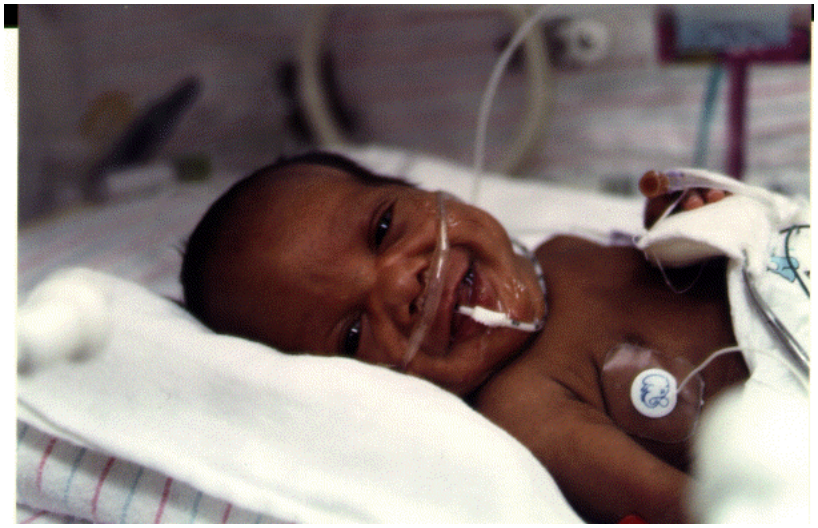


Primary

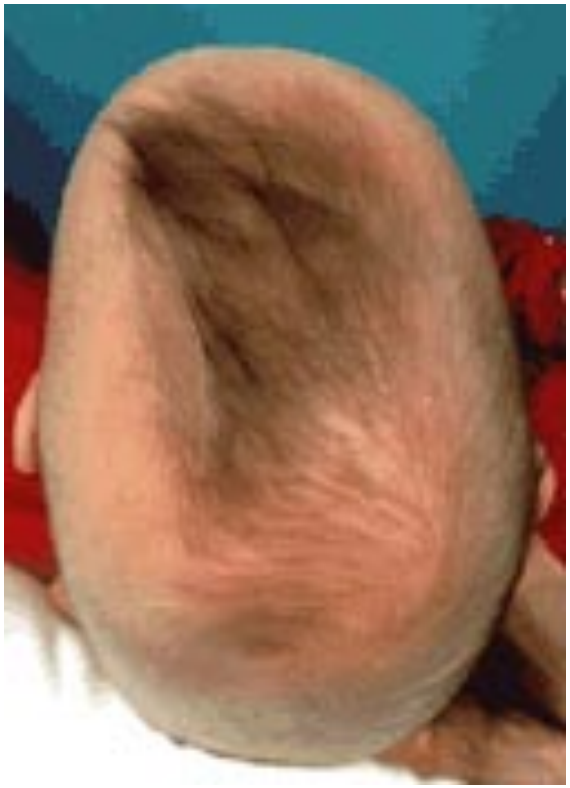
Isolated Abnormal Suture

Syndromic

Prematurity



- Deformational Scaphocephaly
- Impaired mobility & prolonged positioning
- Persists until adulthood
- Prevention:
  - Donut-shaped head supports
  - waterbed mattresses
- Does not warrant intervention



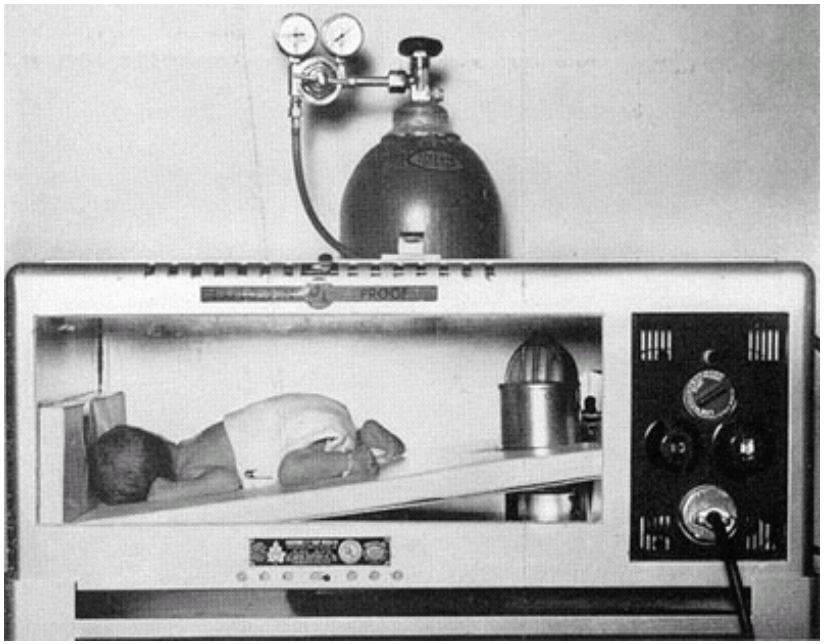
VP Shunting

- Scaphocephaly
- Chronic hydrocephalus thickens the skull
- Once decompression with shunt, the suture fuses
- Surgery Indications:
  - HC > 50 cm (4-5+ STDs)
  - When VPS performed during when VLBW

Microcephaly

- Surgical correction not indicated
- Abnormal HC
  - in primary craniosynostosis, HC remains normal yet oddly shaped
- Rare cases of multisutural craniosynostosis restricting head growth, but manifests with increased ICP

Positional Deformation



- Most common cause
- Usually forehead asymmetry
- Sometimes associated with torticollis
- Usually acts on coronal or lamboidal suture
- 40% of newborns

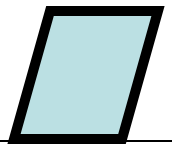
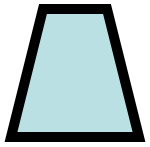
An Epidemic of Lamboidal Plagiocephaly

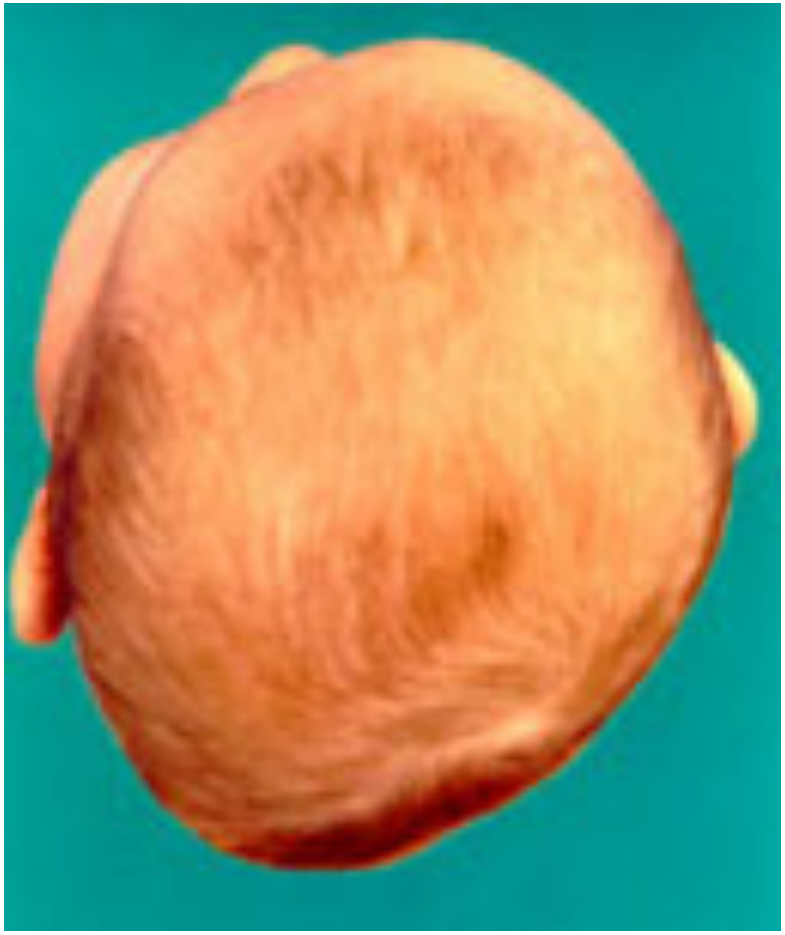
- 1992: Back to Sleep Campaign

- 1996: Tertiary Care Centers report rise in lamboidal plagiocephaly from 3% to 20%

Sorting out the “Epidemic”

- 102 Patients with occipital plagiocephaly over 4 year period
- Only 4 (3%) had true lamboidal synostosis
- The rest were deformational
  - Only 3 were progressive (required surgery)
  - Other responded to positioning or helmets

	Deformational	True
Incidence	Common	Rare
Occipital Flattening	Yes	Yes
Suture	Open	Closed/Ridged
Fronto-parietal Bossing	Ipsilateral	Contralateral
Ipsilateral Ear	Forward / Down	Back / Down
Head Shape		



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Occipital Flattening	Yes	Yes
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Fronto-parietal Bossing	Ipsilateral	Contralateral
Ipsilateral Ear	Forward / Down	Back / Down
Head Shape		

Syndromic Craniosynostosis

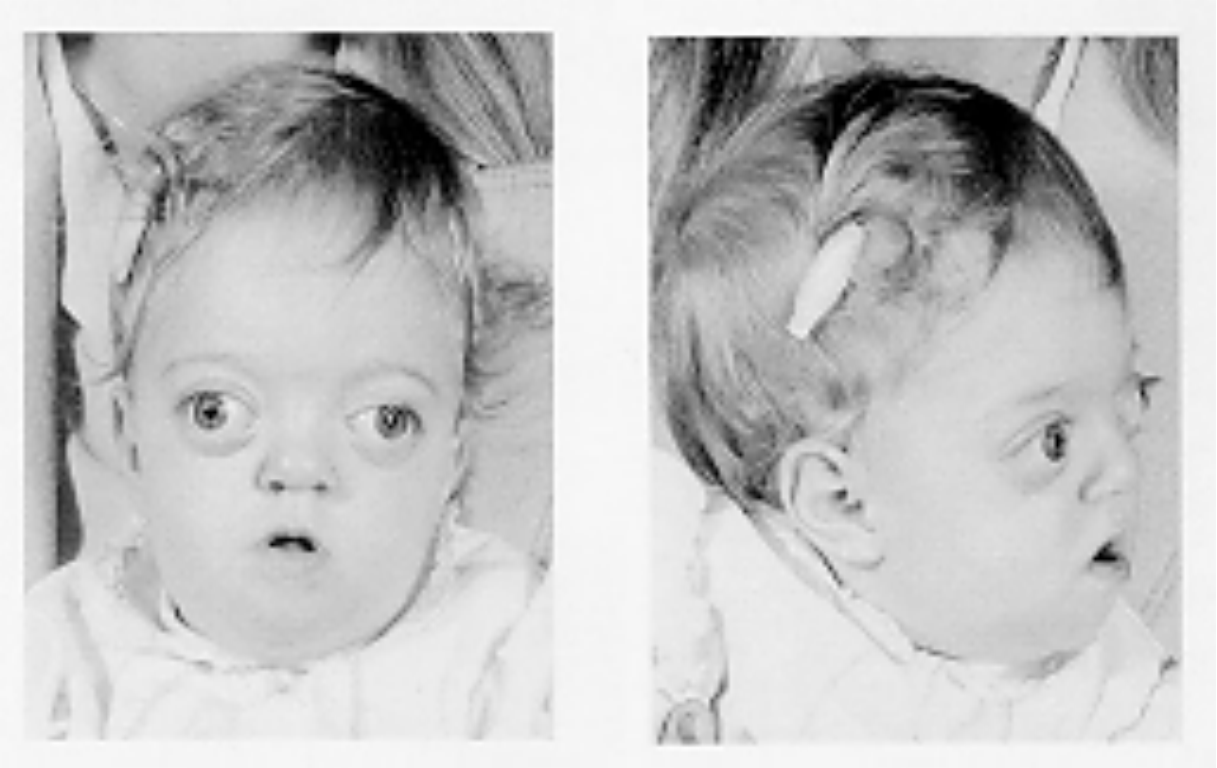
- 10-20 % of cases
- Autosomal Dominant
  - Linked to Chromosome 10q
- Multi-sutural, complex cases

If a suture is fused,
check hands, feet, big toe and thumb

Distinguishing Clinical Features in the Craniosynostosis Syndromes

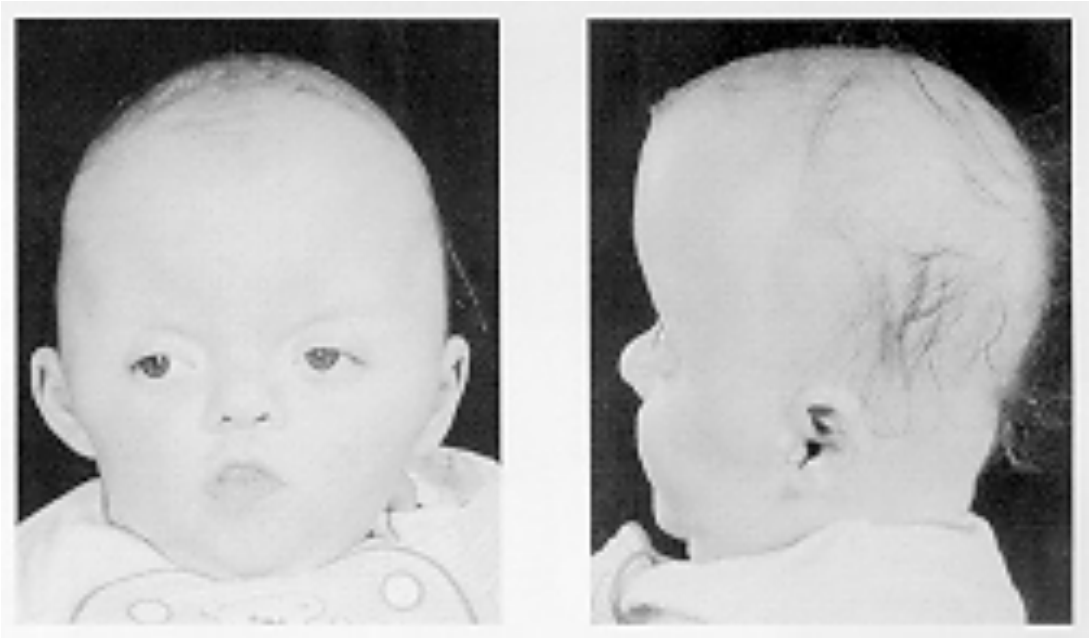
	Muenke	Crouzon	Jackson-Weiss	Apert	Pfeiffer	Bear-Stevenson
Thumbs	Normal	Normal		Fused to fingers	Broad, deviated	Normal
Hands	± Carpal fusion	Normal	Variable	Bone syndactyly	Variable brachydactyly	Normal
Great Toes	± Broad	Normal	Broad, deviated	Fused to toes	Broad, deviated	Normal
Feet	± Tarsal fusion	Normal	Abnormal tarsals	Bone syndactyly	Variable brachydactyly	Normal

Crouzon's



- Normal intellect
- Normal extremities
- 5 % have acanthosis nigricans
- 30 % have progressive hydrocephalus

Apert's
"Crouzon's with Hand Involvement"



- Varying intellect (50 % with MR)
- Mitten Glove Syndactyly
- Cervical vertebral anomalies
- Rare hydrocephalus

Apert Extremity Findings

True Craniosynostosis & Surgery

- Single Suture Synostosis: Confirm by exam and skull x-rays
- Complex cases: CT or 3D CT
- X-Ray: Fused sutures have a broad ridge of overgrowth of solid bone along a previous suture, or suture is completely obliterated
- Ridge is especially characteristic of fused sagittal suture

Management

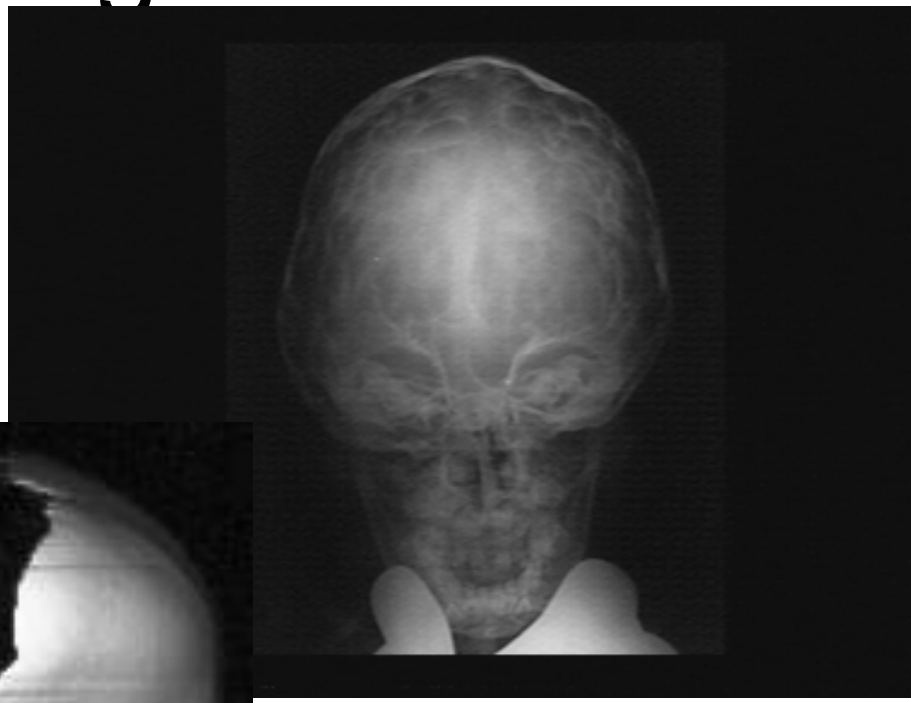
- Surgery vs. Conservative Management

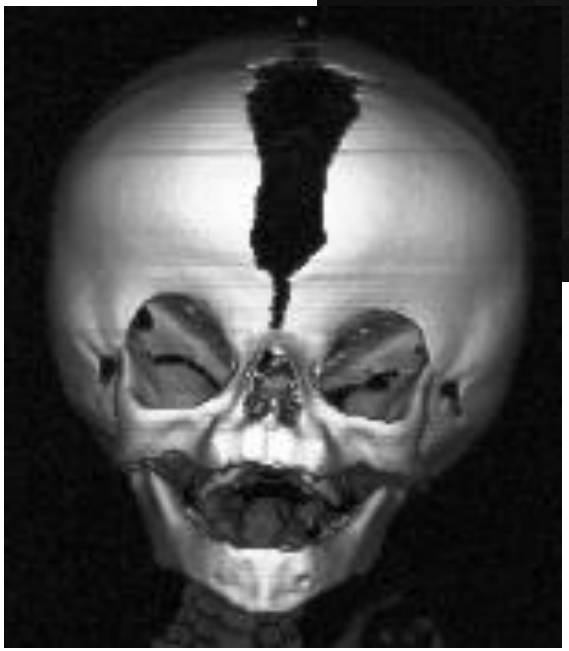
The Decision to Operate

- Raised ICP in 1/3 of cases, but no neuro impairment
- Cosmetic considerations usually most important
  - affects peer acceptance, parent-child bonding, self-image and coping

Imaging

- Skull X-ray
- CT
- 3-D CT



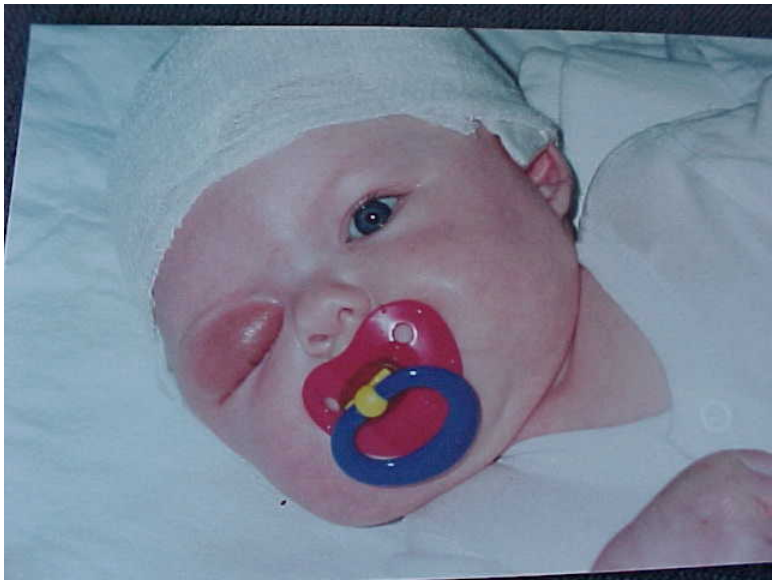


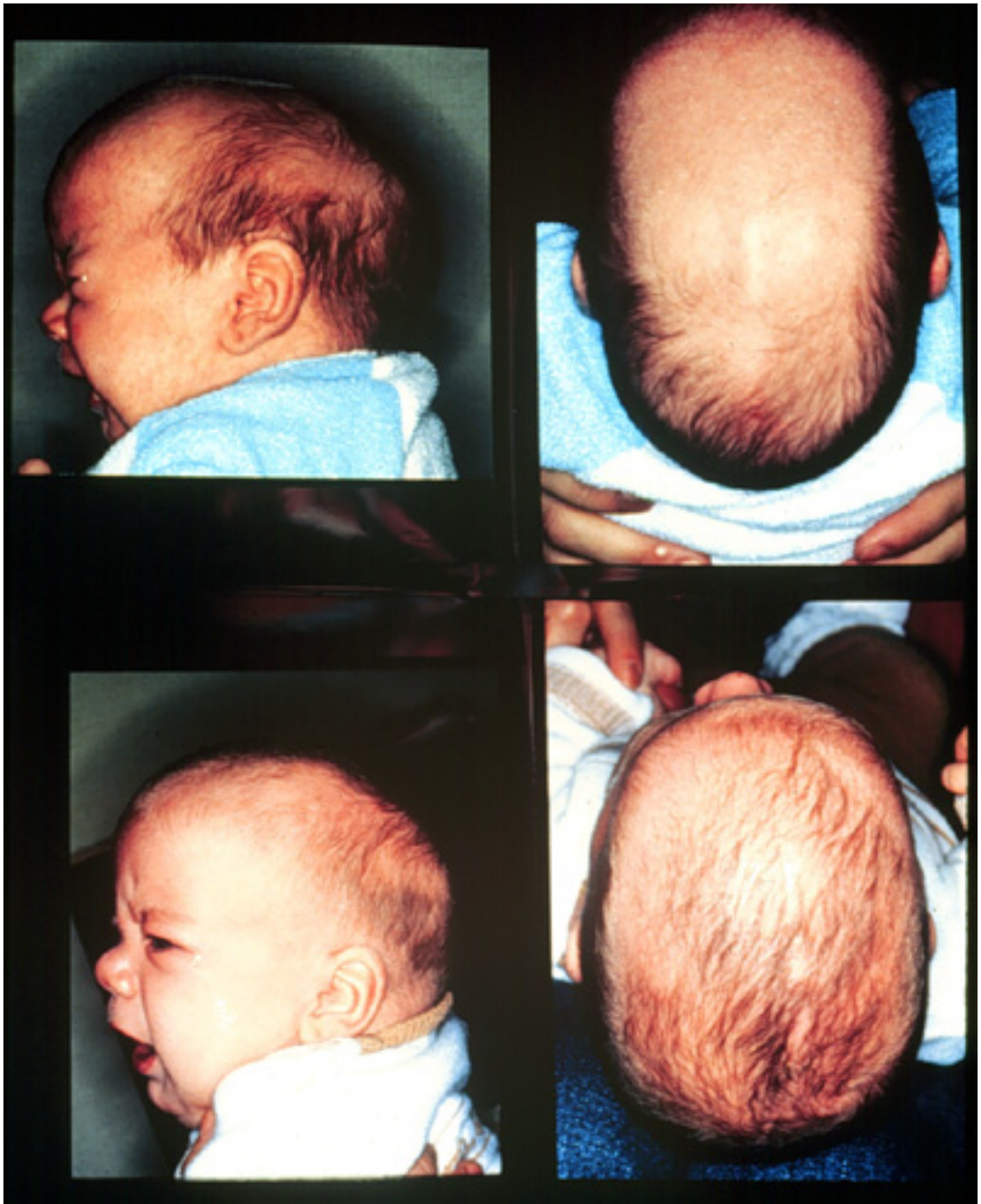
Surgery

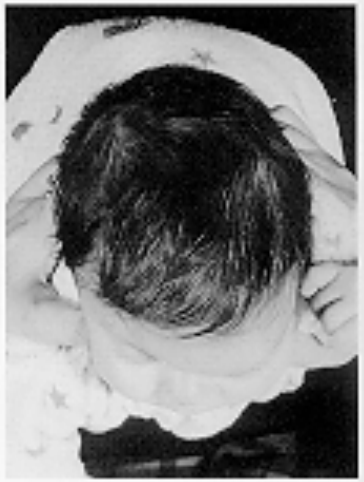
- If not part of syndrome, the earlier the operation the better
  - At the latest 6-12 months (by 12 months, skull is 85% of adult size)
  - For coronal suture, operate before 2 months because of facial symmetry and visual system development
  - Procedure depends on continuing skull growth

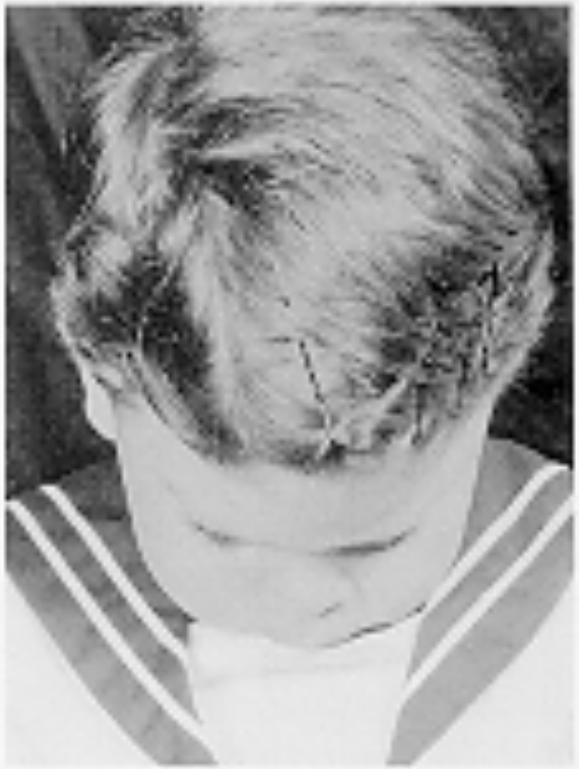
Surgery

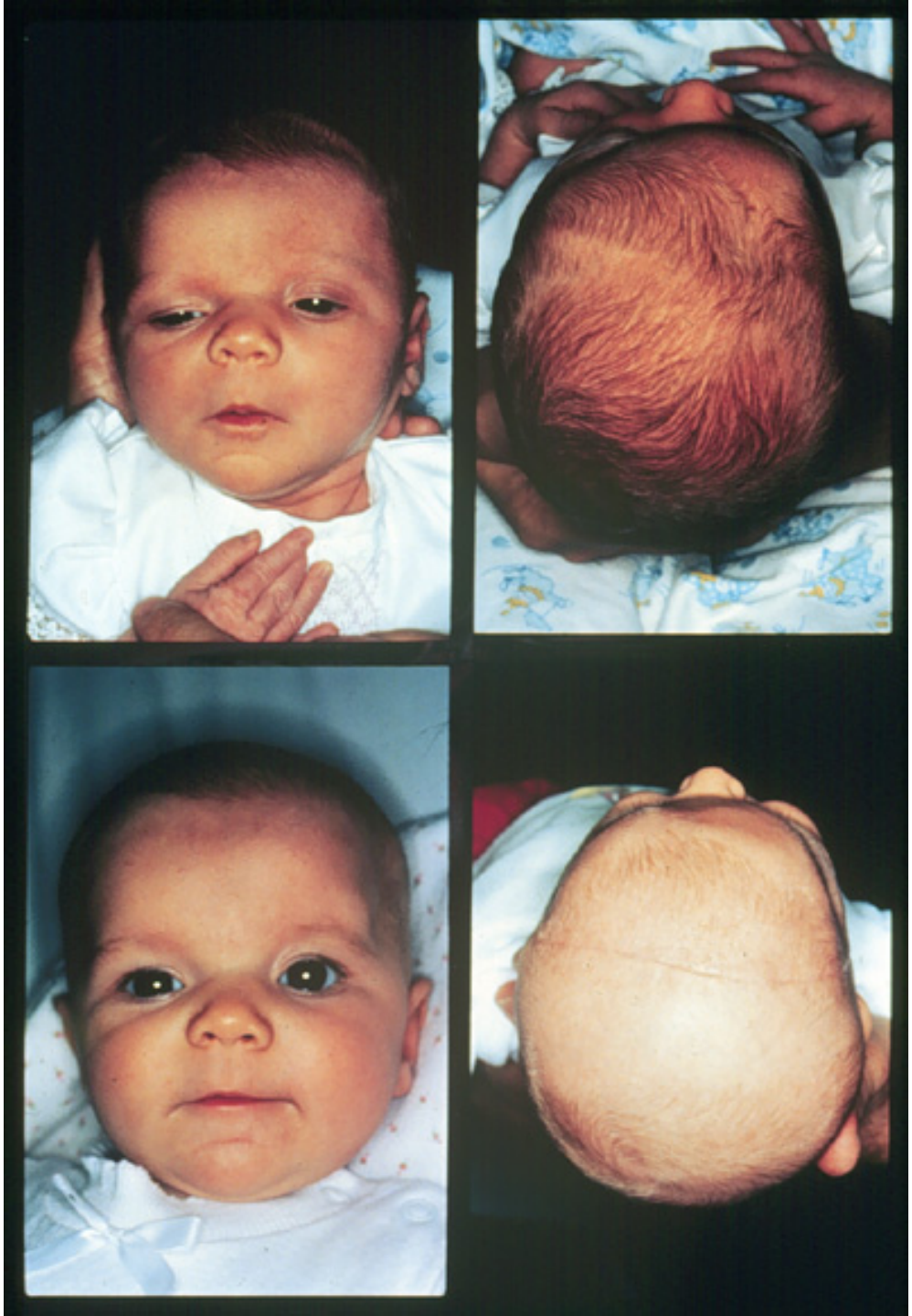
- Syndromic cases may need special airway support
- Blood loss significant due to scalp vascularity
  - transfusion rates 20-500 % of infant estimated blood volume
- May require PICU stay (facial edema)



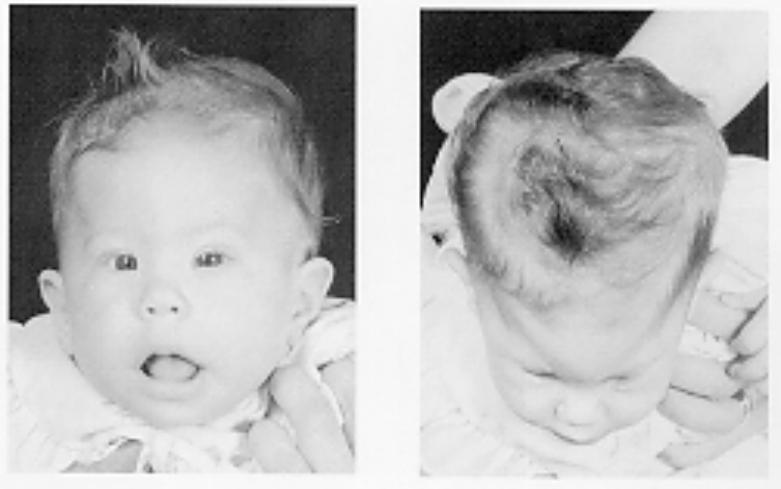








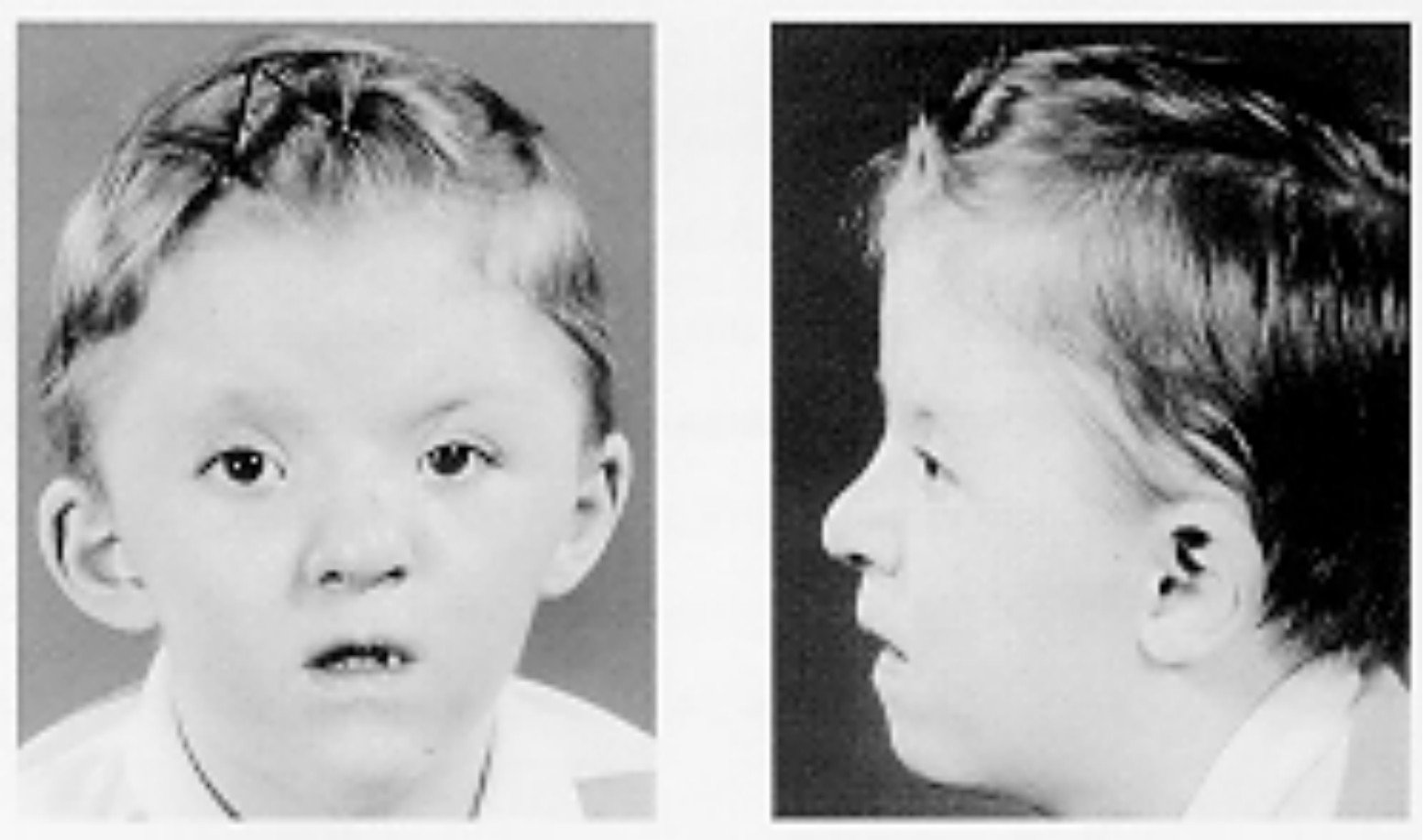
Metopic Synostosis



Surgery

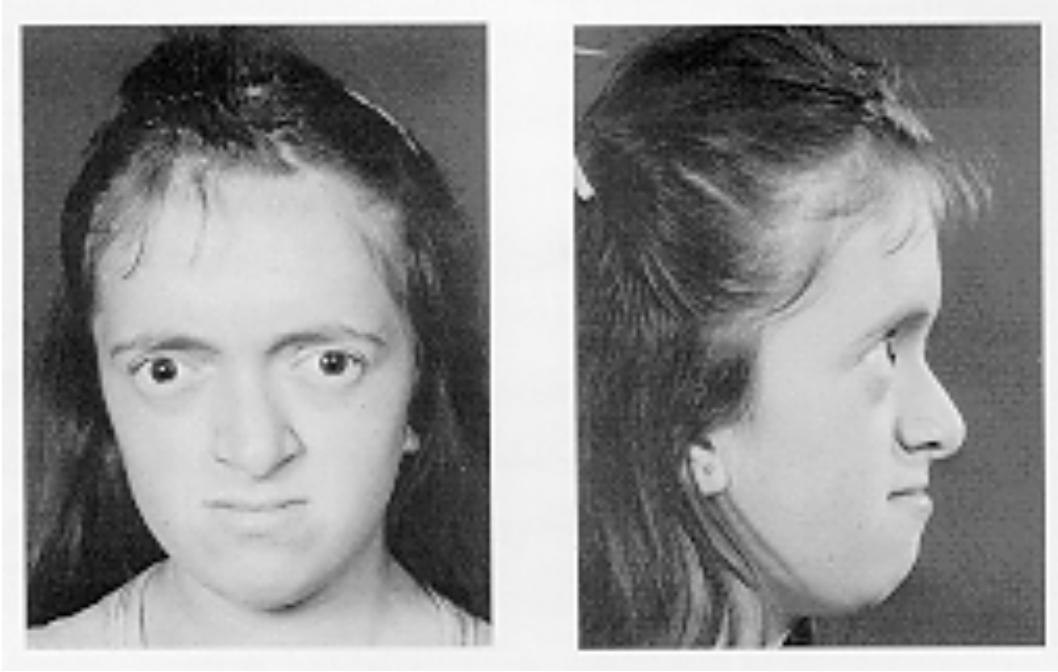
- Unilateral coronal suture: difficult. Orbital relocation as well.
- Syndromic or multi-suture cases: staged repairs.

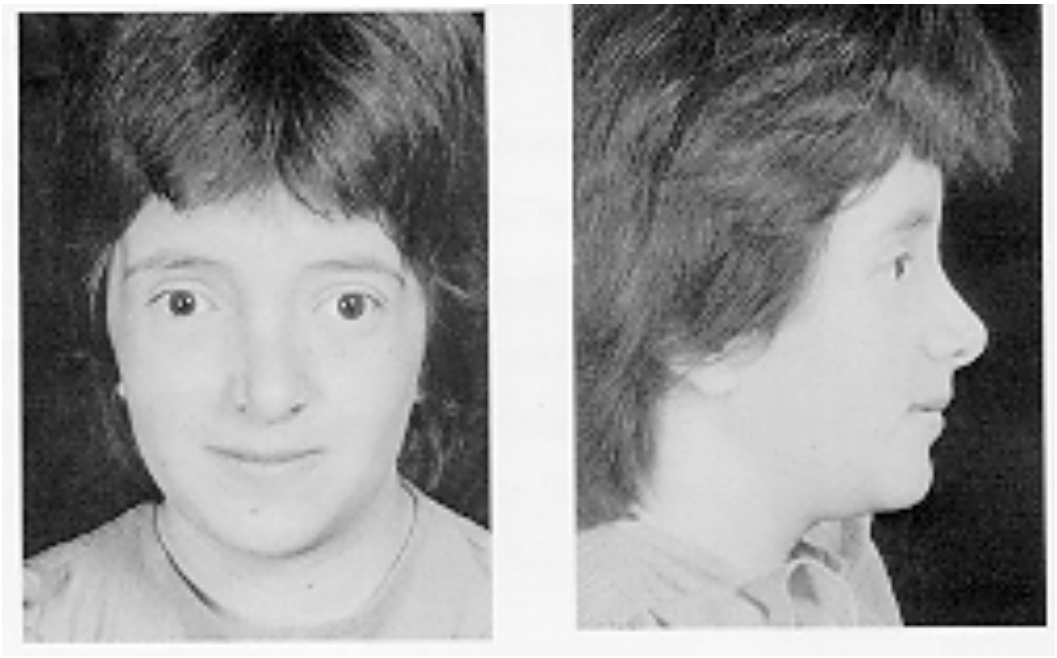
Apert: Post-Op



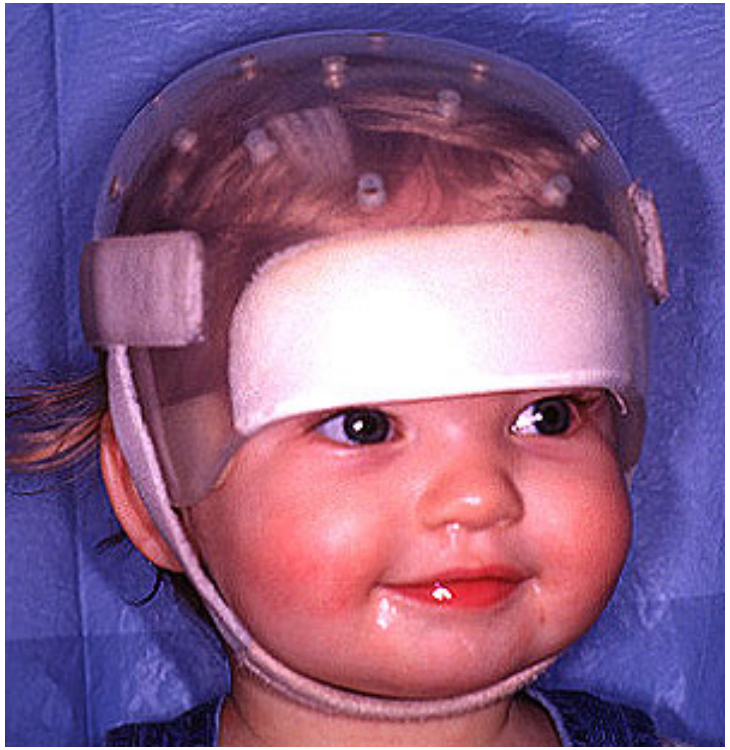
Crouzon

Surgical Pics

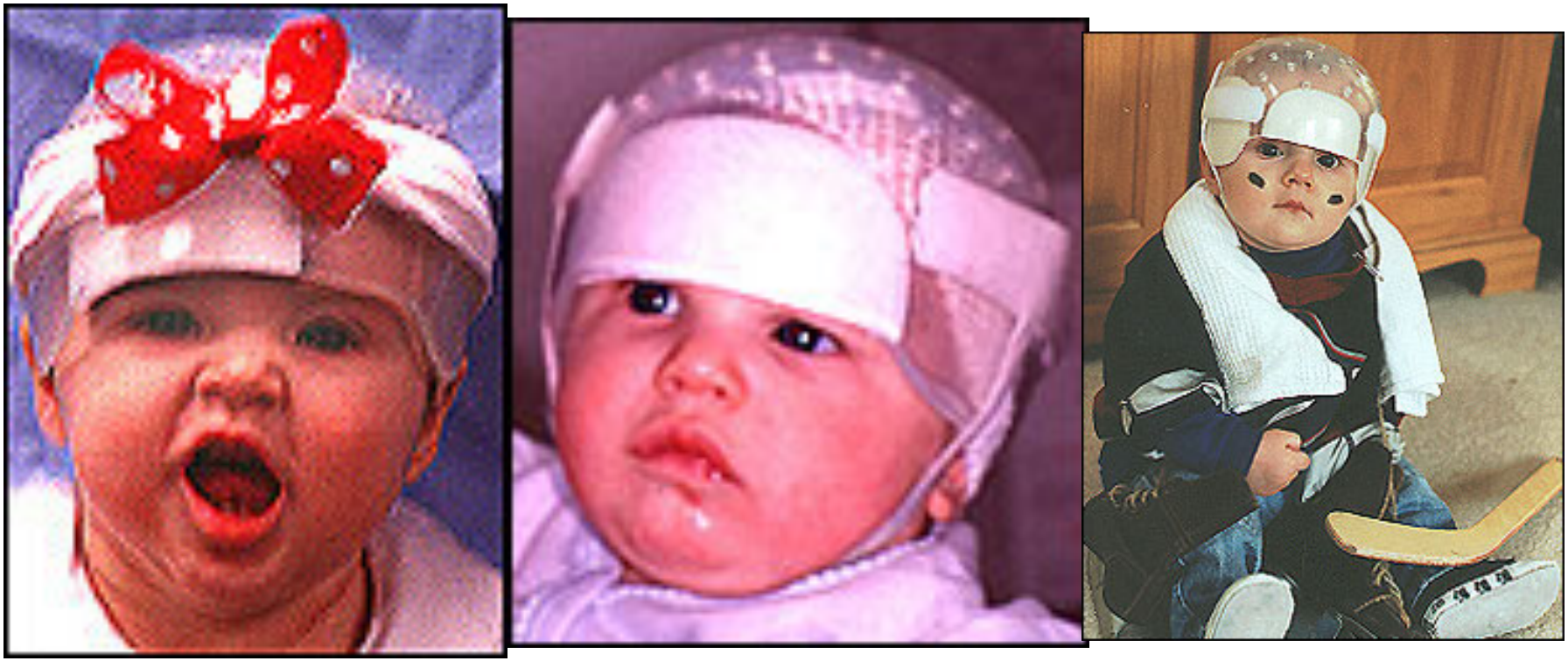




Conservative Therapy for Deformational Plagiocephaly



- Re-positioning
- If no improvement by 6 months….
  - Helmet Molding



Custom Made for each head

24/7 wear for 4 months