Craniosynostosis

evaluation and treatment

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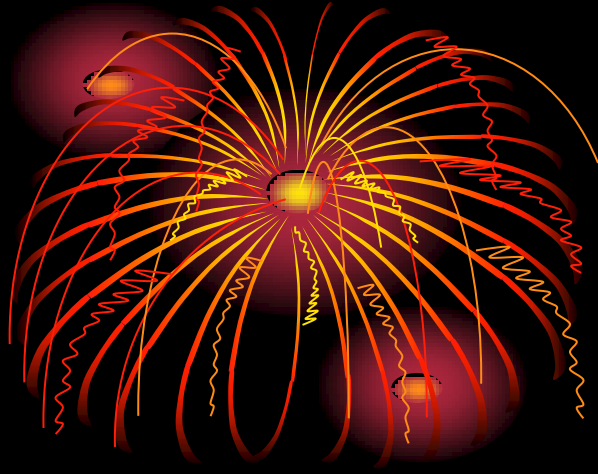
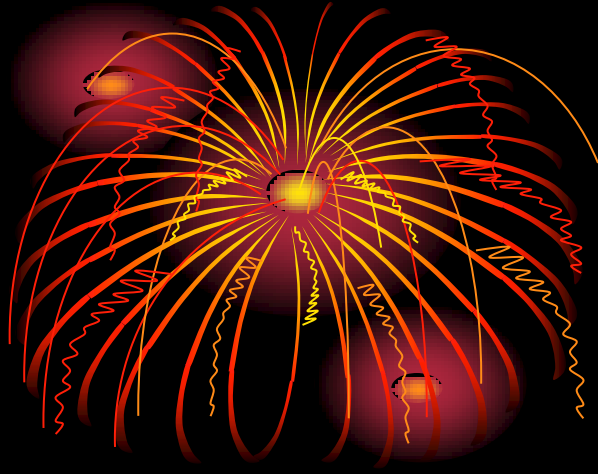
Definitions:

Craniosynostosis:

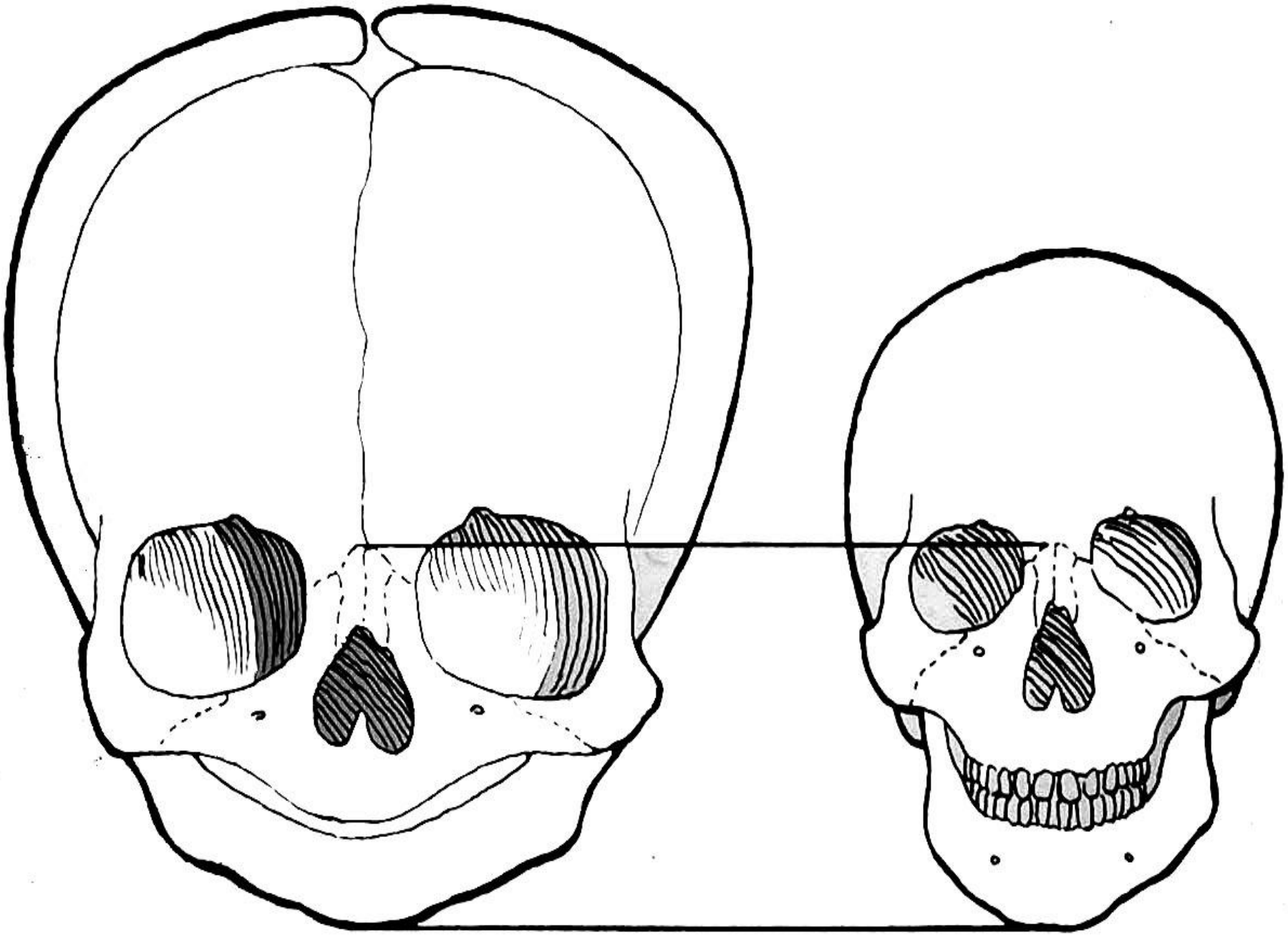
- A general term encompassing a variety of developmental disorders characterized by inadequate capacity of the cranium to accommodate the growing brain, which results in compensatory deformities. (Less accurate for the minor forms where there is deformity with out problems of capacity)

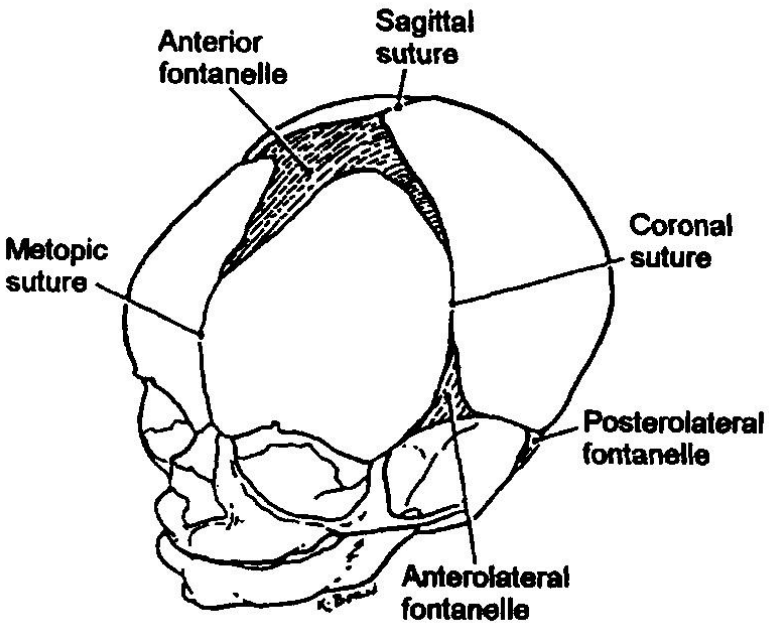


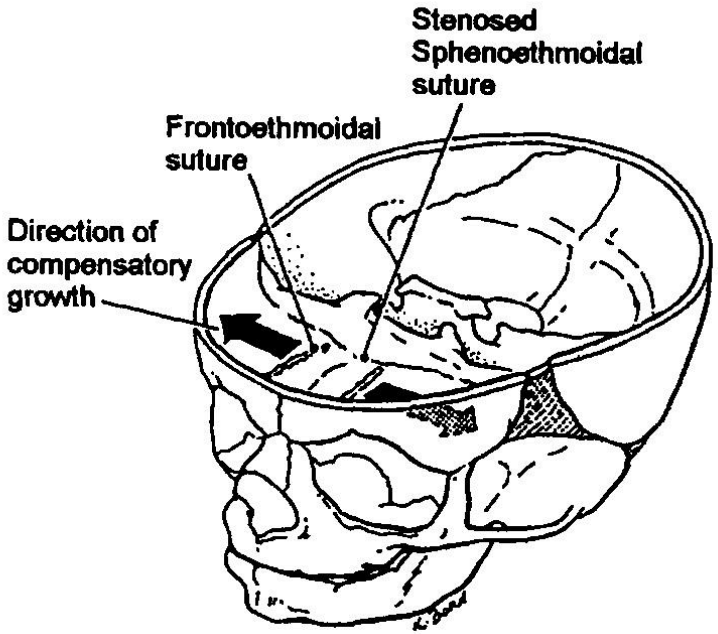
Simple: involves the cranial sutures, although there often is an extension into the base of the skull.

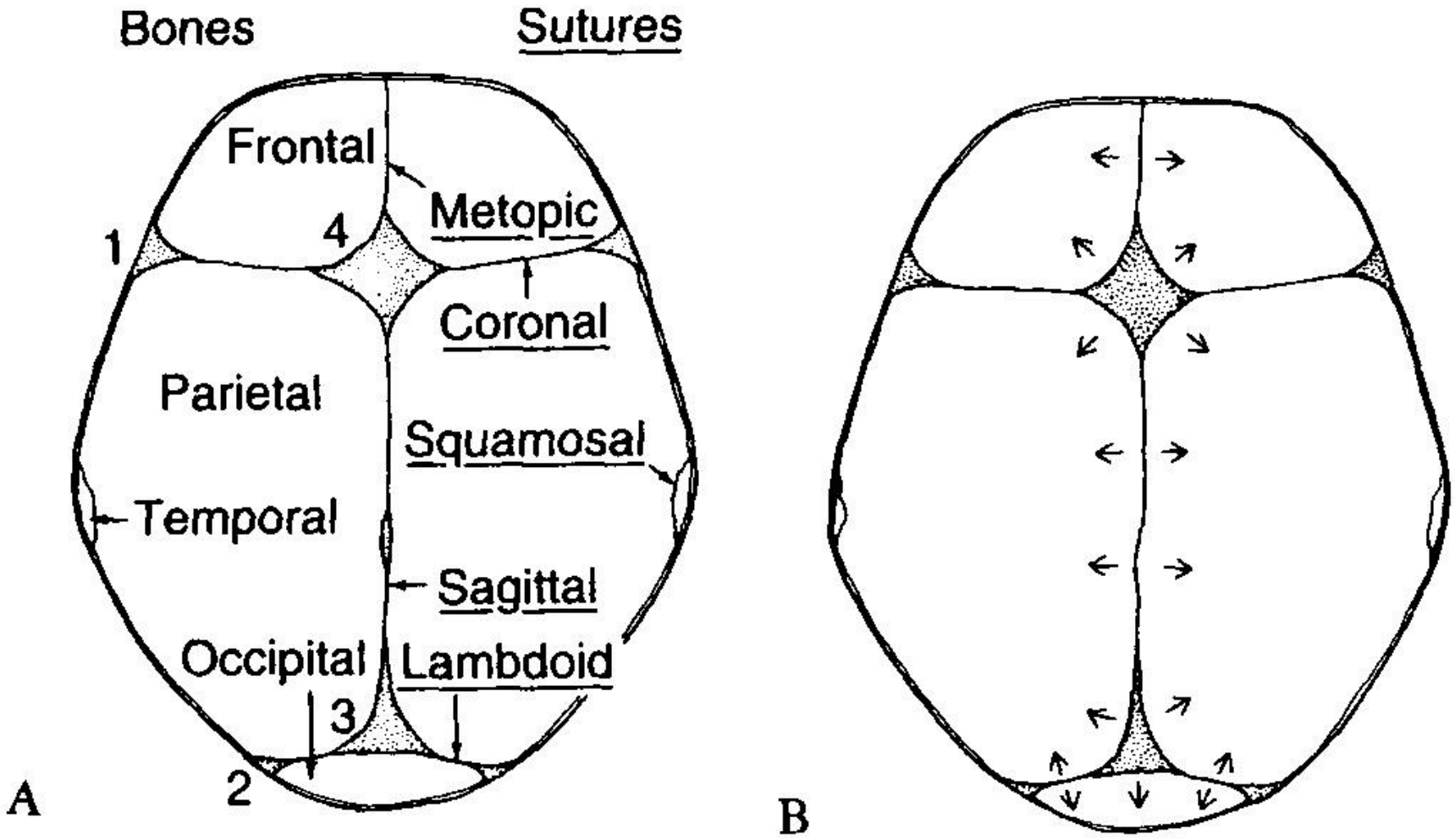


Syndromatic: involves also the face with possible involvement of the limbs.

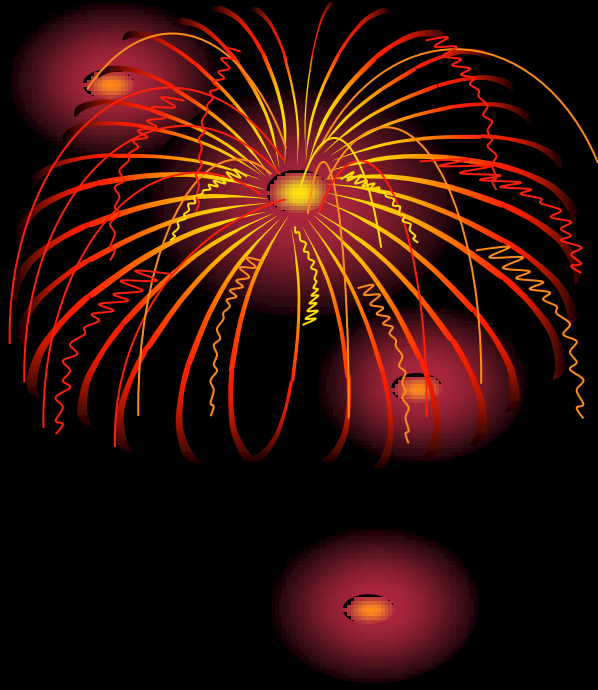



Anterior fontanelle
Sagittal suture
Coronal suture
Metopic suture
Posterolateral fontanelle
Anterolateral fontanelle

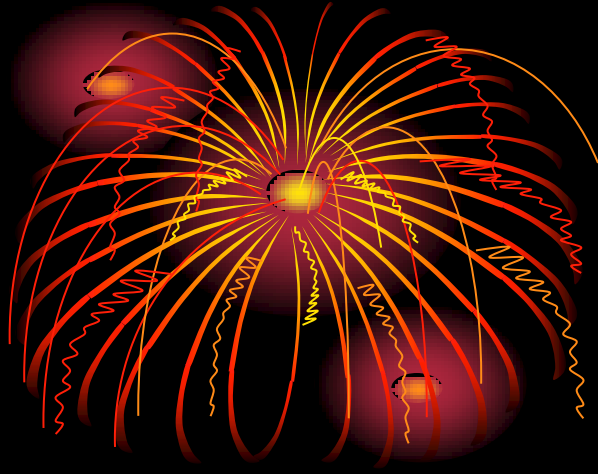

Stenosed Sphenoethmoidal suture
Frontoethmoidal suture
Direction of compensatory growth


Bones
Sutures
Frontal
Metopic
1
4
Coronal
Parietal
Squamosal
Temporal
Sagittal
Occipital
Lambdoid
3
2
A
B

Epidemiology



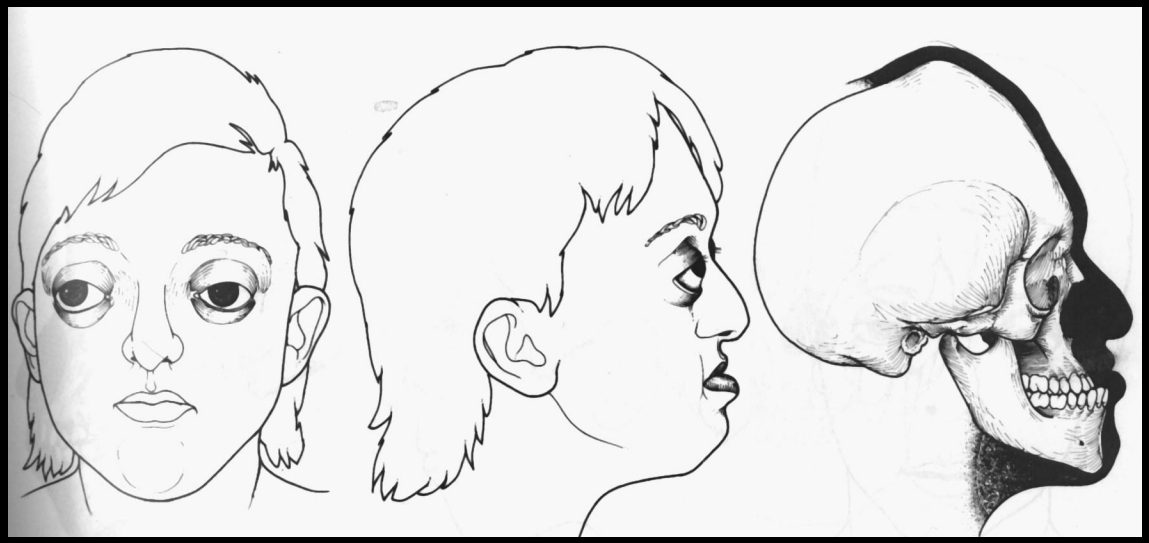
- Detecteble abnormalities due to craniosynostosis are seen in
  1:1700-1900 births.
- Most common types
- Sagital — schaphocephaly
- Metopic — trignocephaly
- Unilateral coronal — Plagiocephaly
  1:10,000
- Bilateral coronal — Brachicephaly

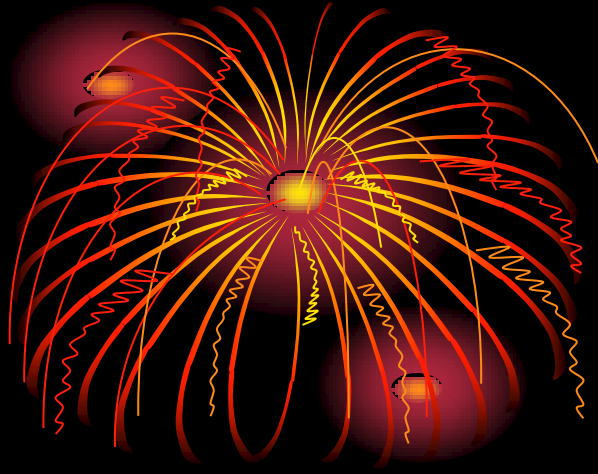


Impact

Aesthetic disfigurement

Functional disorders: Increased ICP, hydrocephalus, visual impairment.



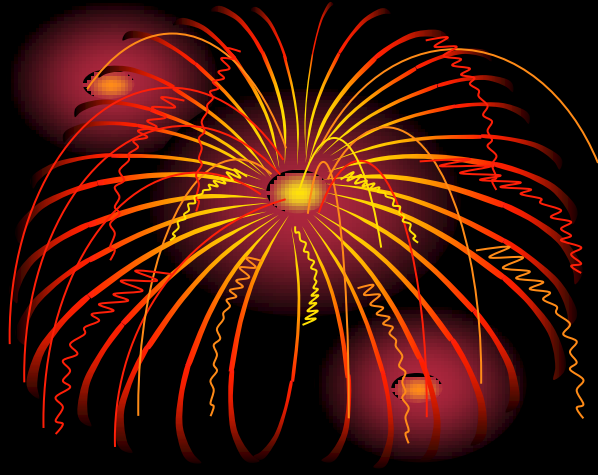


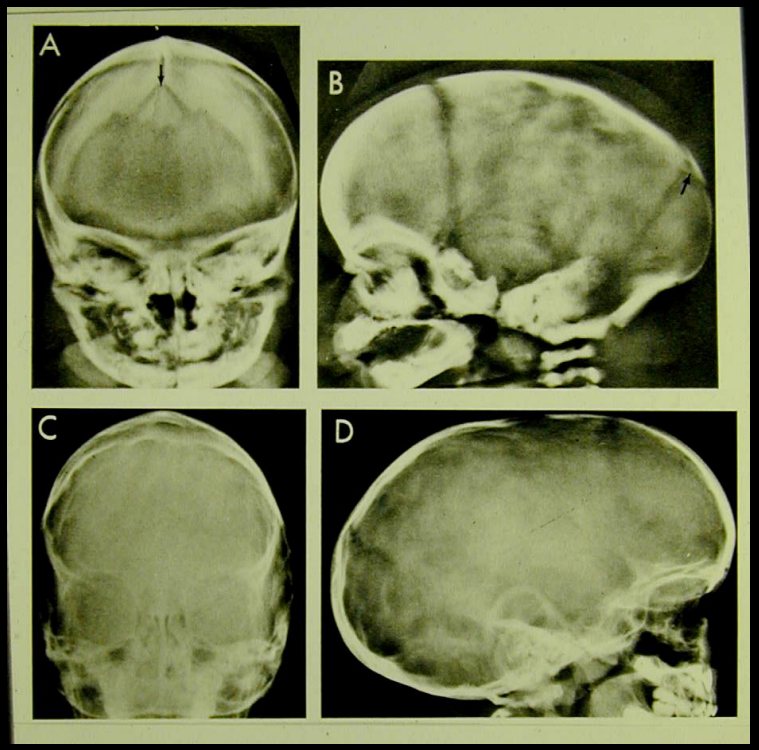
Impact

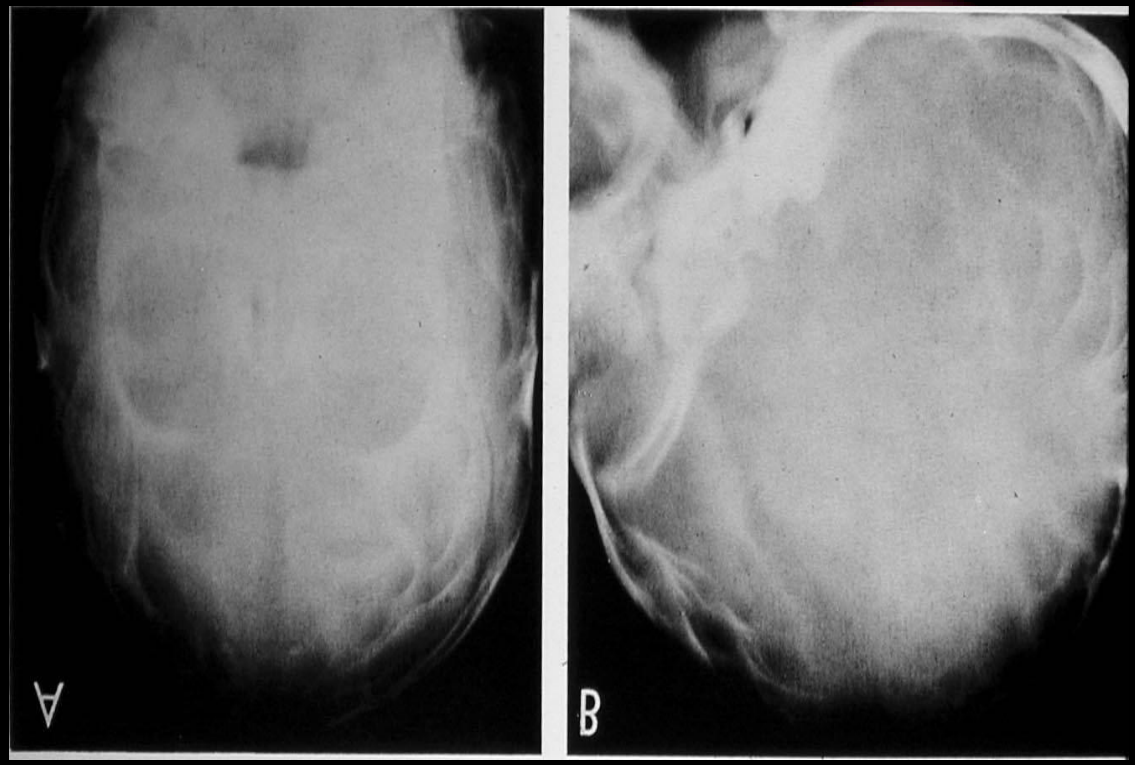
Functional disorders: Increased ICP, hydrocephalus, visual impairment.

ICP

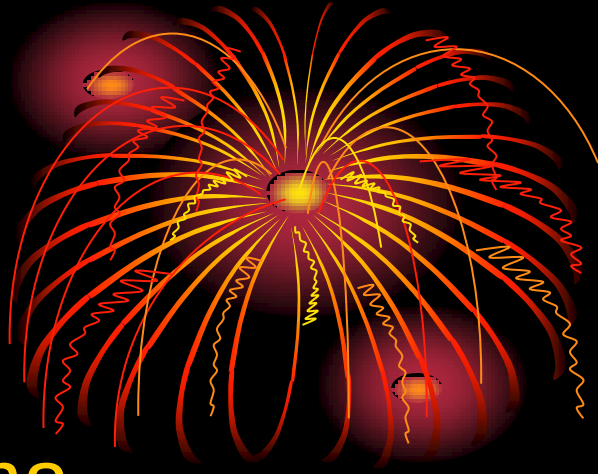
- Increased ICP is seen in cases of craniosynostosis more in cases where several sutures are involved.
- Surgery decreases ICP.
- A small skull doesn’t imply ICP elevation but ICP elevations usually seen in small skulls. Not all authors agree.
- Signs of increased ICP
- Papiledema, headaches, vomiting, lethargy, anorexia, beaten silver appearance on X-ray.




A
B
C
D


A
B

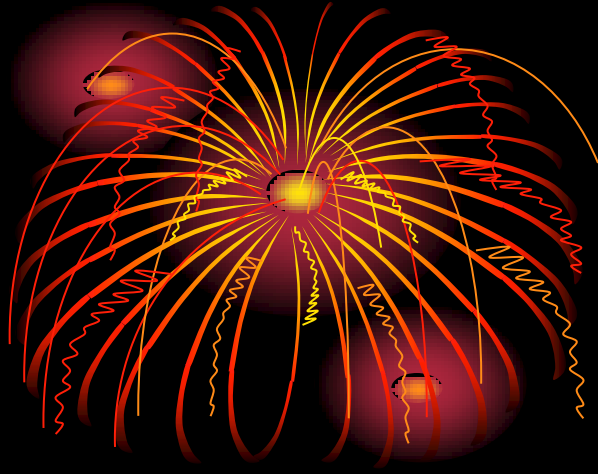
Visual abnormalities



- Optic atrophy and papilledema, stretching of optic nerve, pressure on the nerve. Damage from increased ICP.


A
B

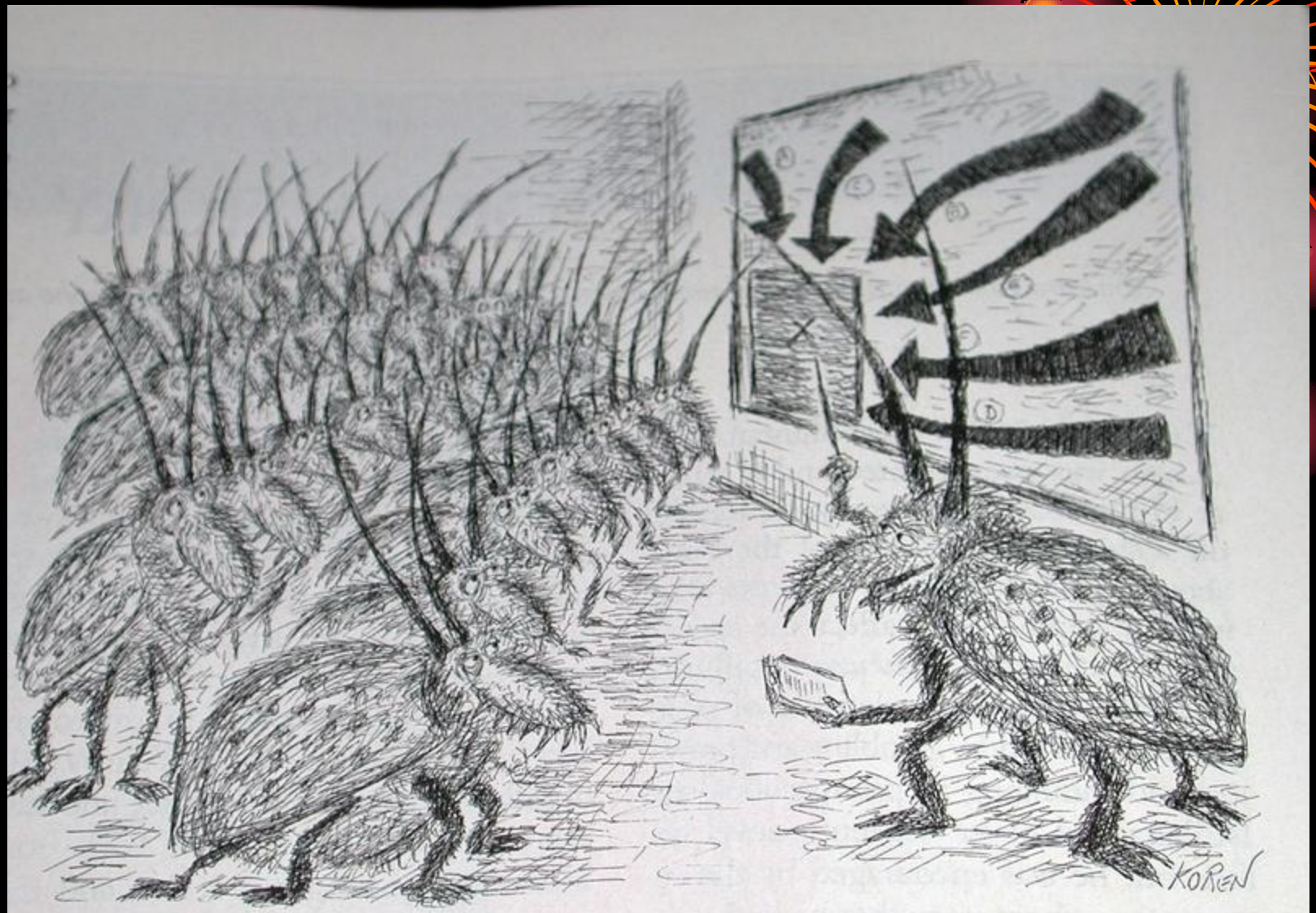
Hydrocephalus



- **Hydrocephalus**
- Rare, seen more in the syndromatic cases.
- Enlarged ventricles may be seen as part of the syndrome as in Apert's. Changes in ventricle size are more accurate for diagnosis.

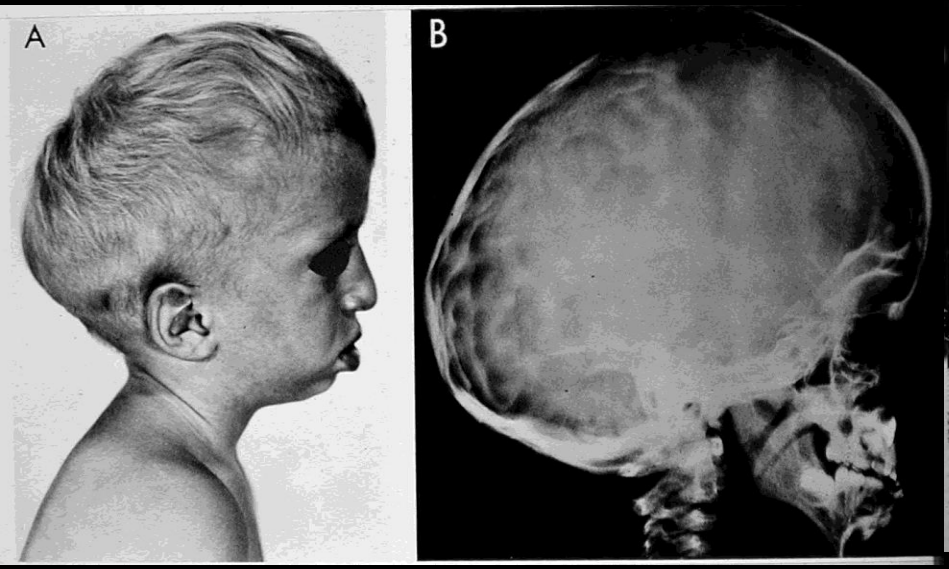
Mental retardation

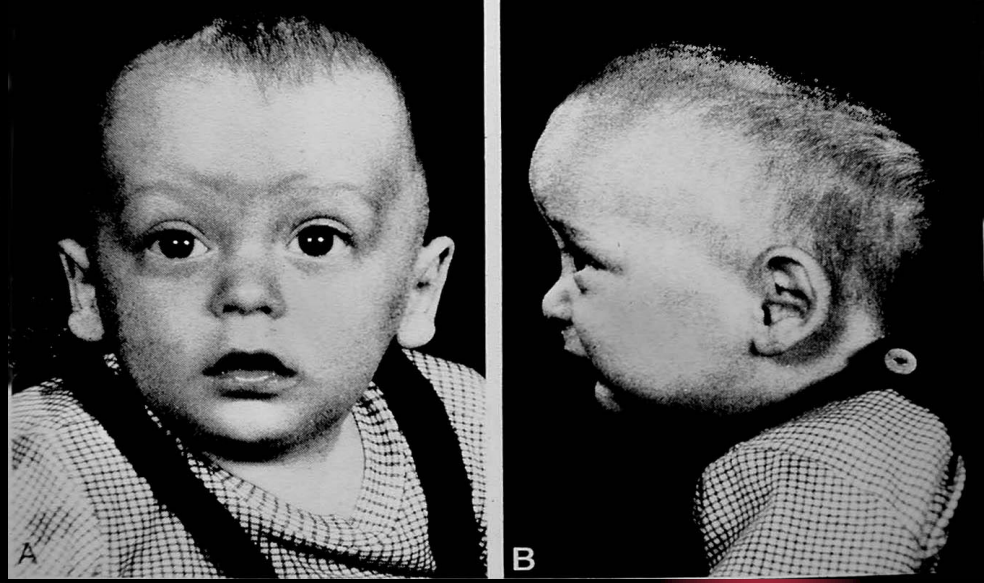
- It is not clear how many of these children really have mental retardation. It is not clear if it is related to the problem or independent
- Some just look retarded - environmental influences.
- In children with non syndromatic craniosynostosis there was no increase in IQ after surgery. Other studies claim there was an improvmentt.



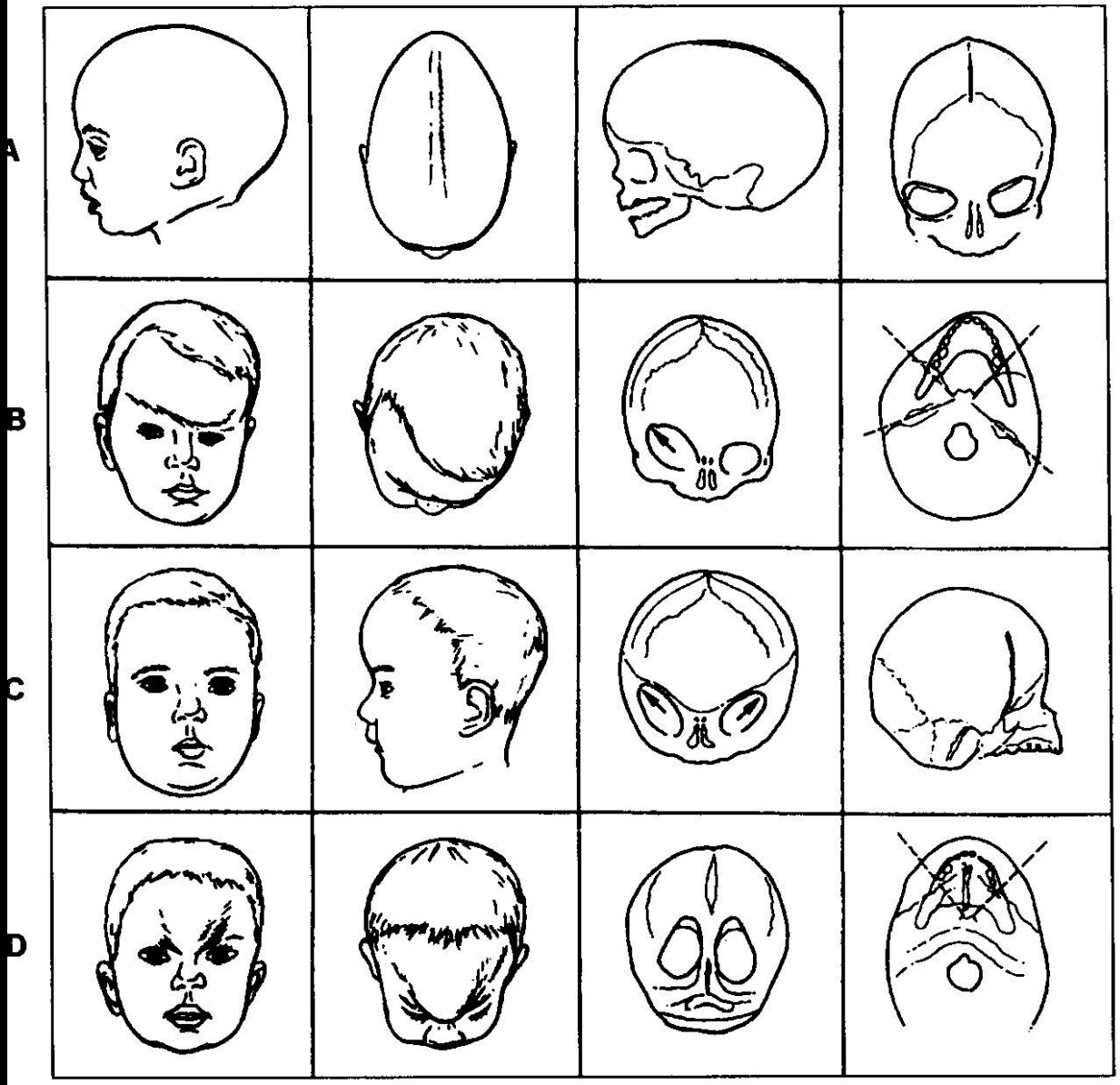
"Today's objective is the genetically modified corn in this quadrant."

Morphology


A
B

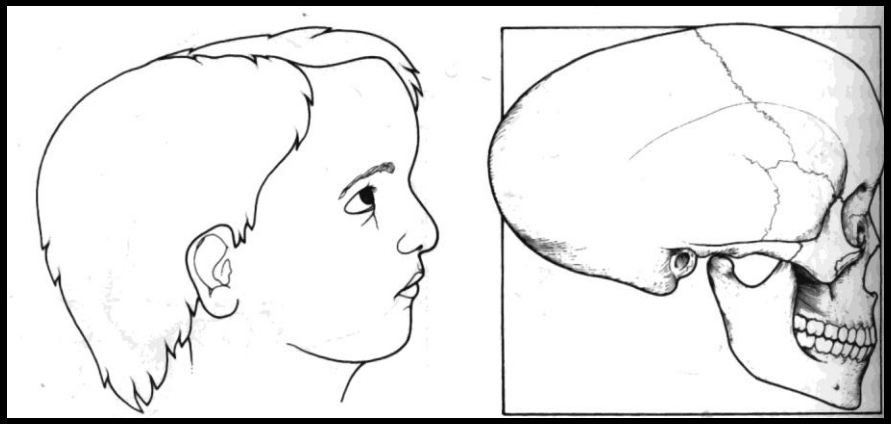

A
B

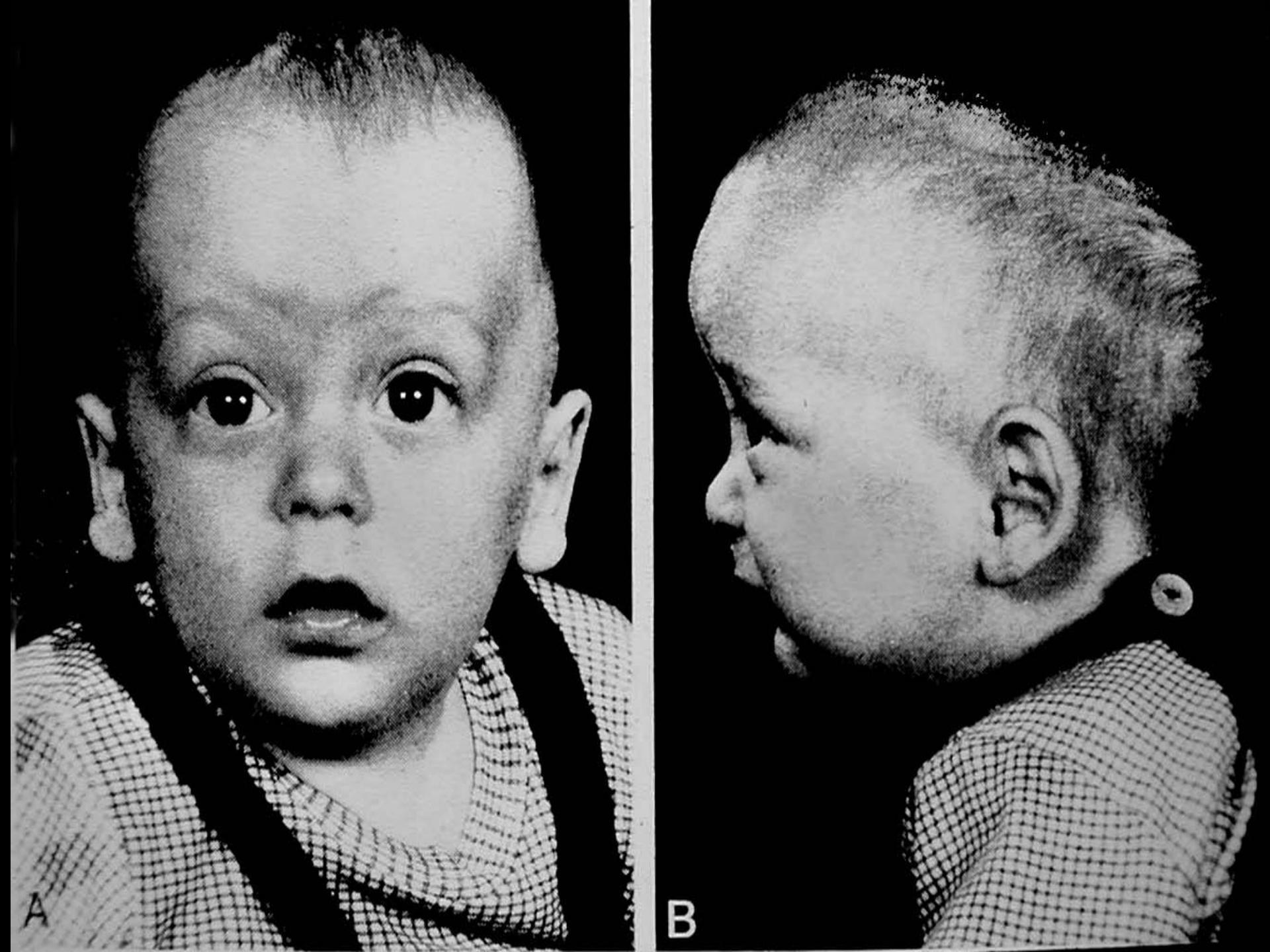

A
B


A
B
C
D

Scaphocephaly

Hull shaped cranium,reduced width with increased length, seen with premature sagital suture closure. The most common type. M:F 4:1

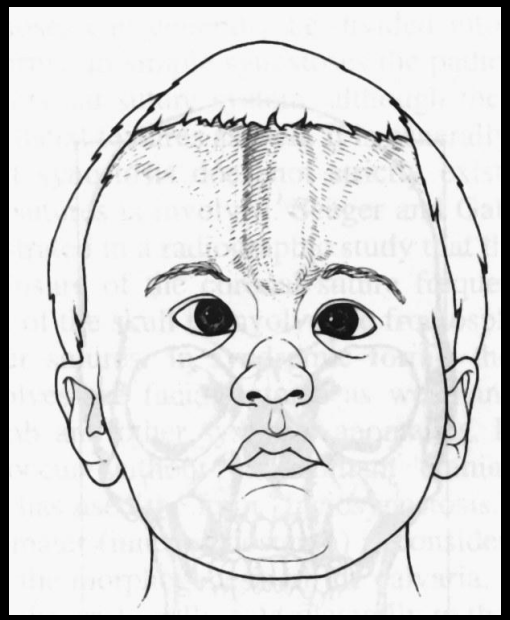


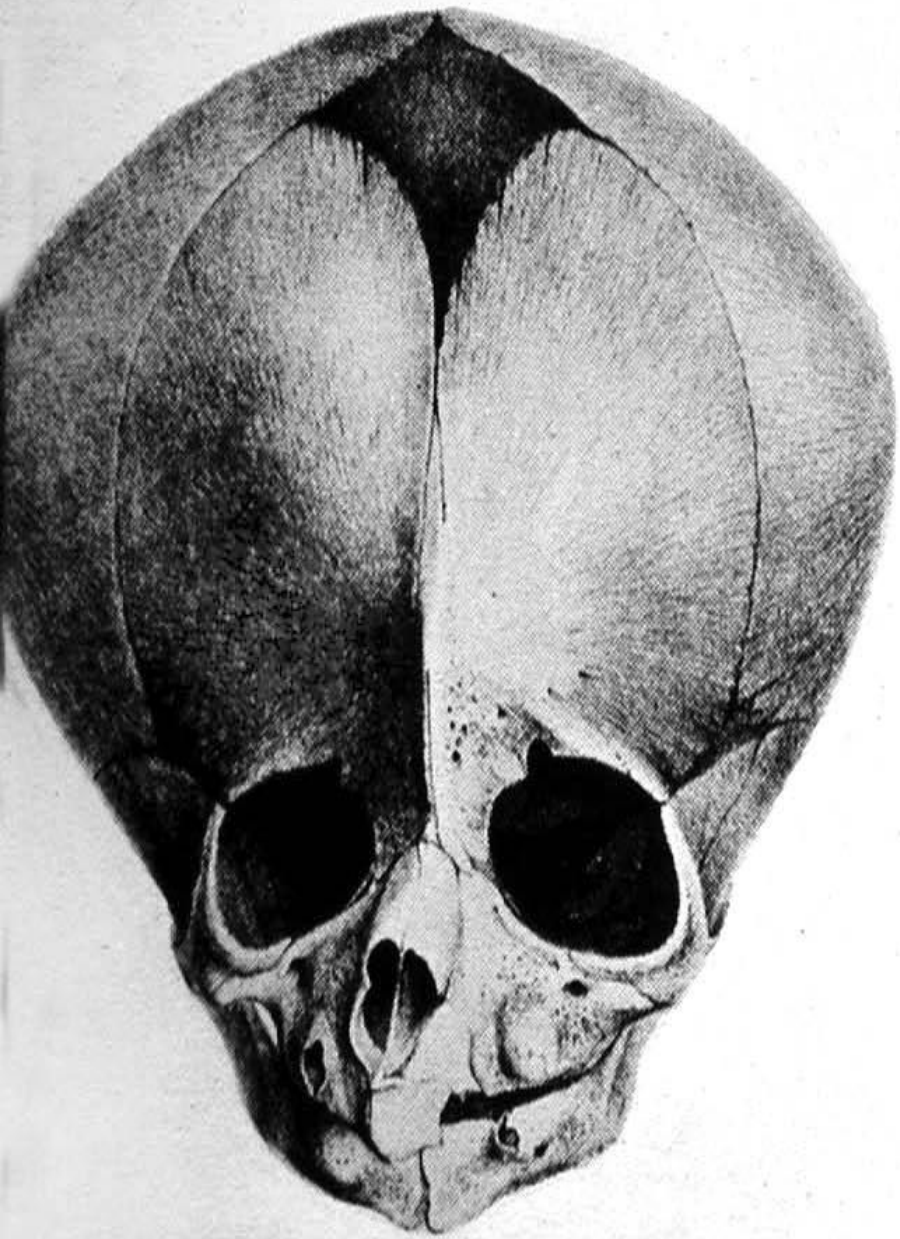

A
B

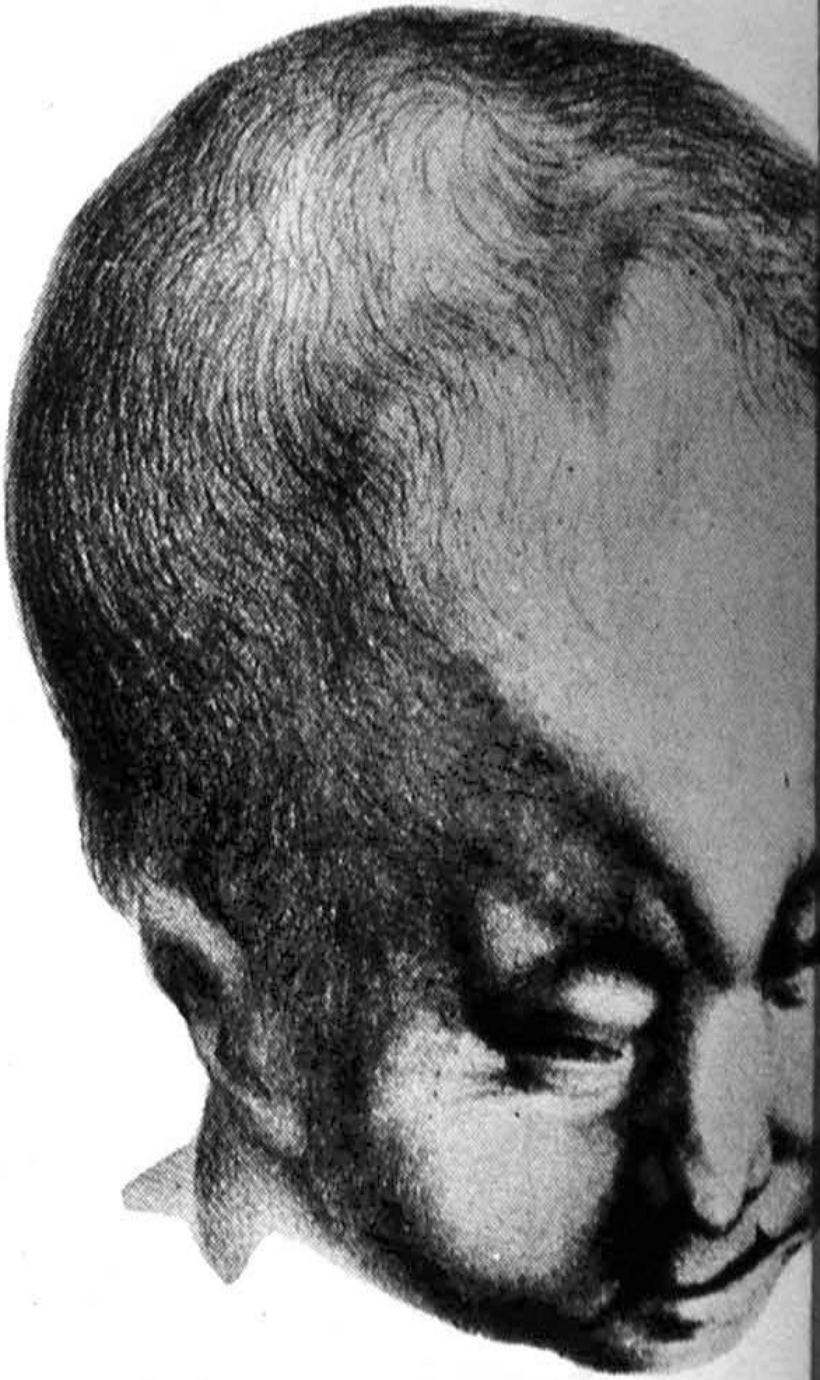
Trigonocephaly

Narrow and sharp forehead caused by premature closure of

metopic suture. May be accompanied by hypotelorism.





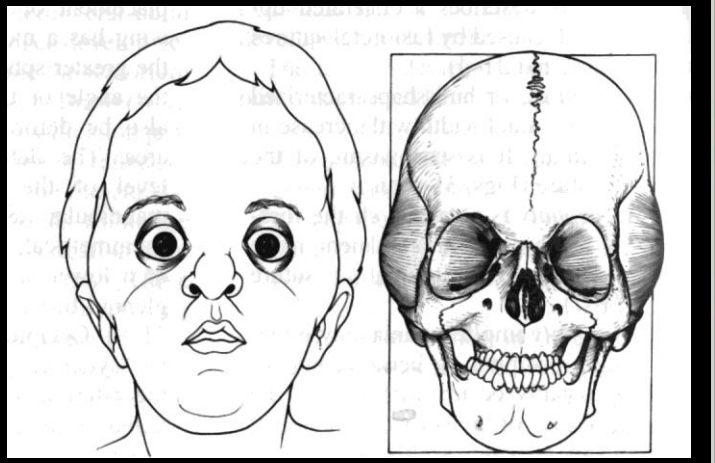


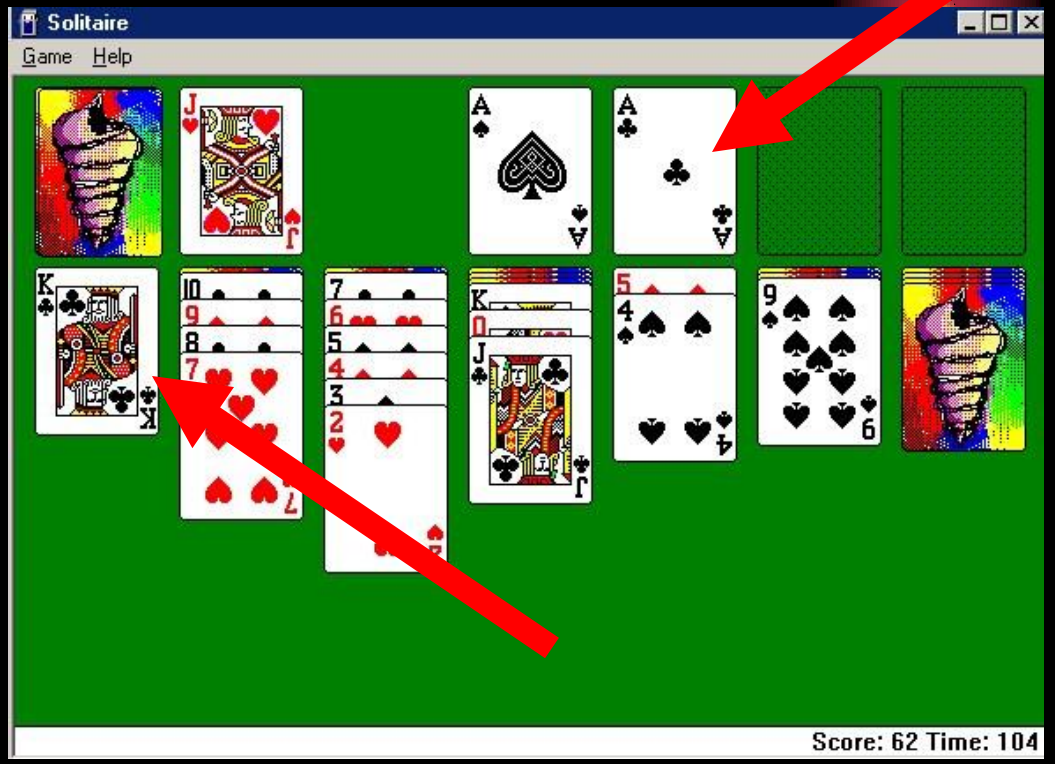

A


B

Clover leaf (Kleebattscaädel)

Multiple sutural fusions, constriction ring in the lamboid-squamosal zone, bulging of frontal and temporal lobes

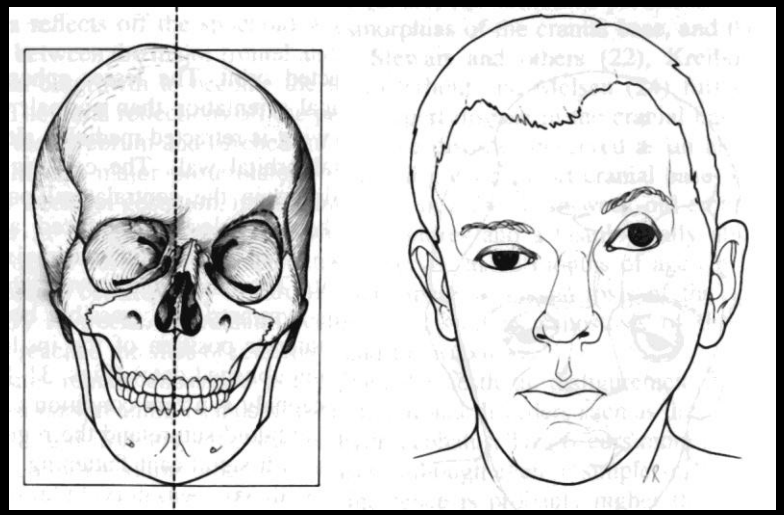


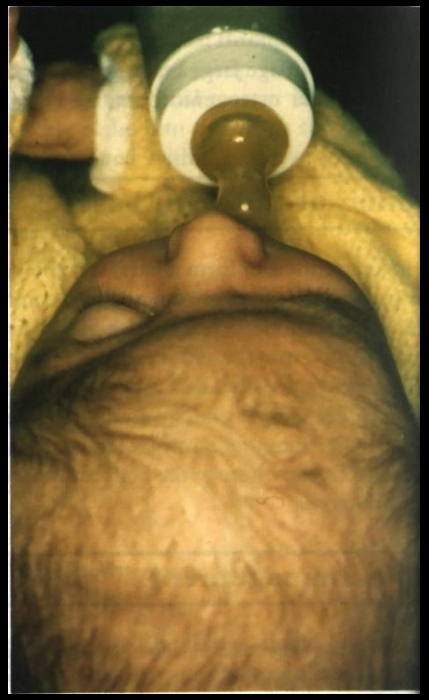


Plagiocephaly

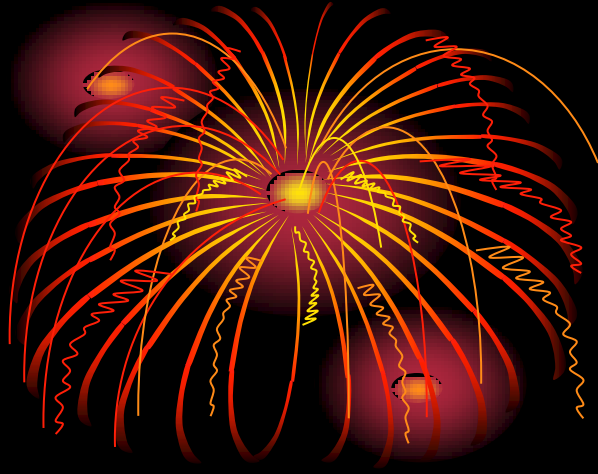
(various sutures)

- Cranial asymmetry



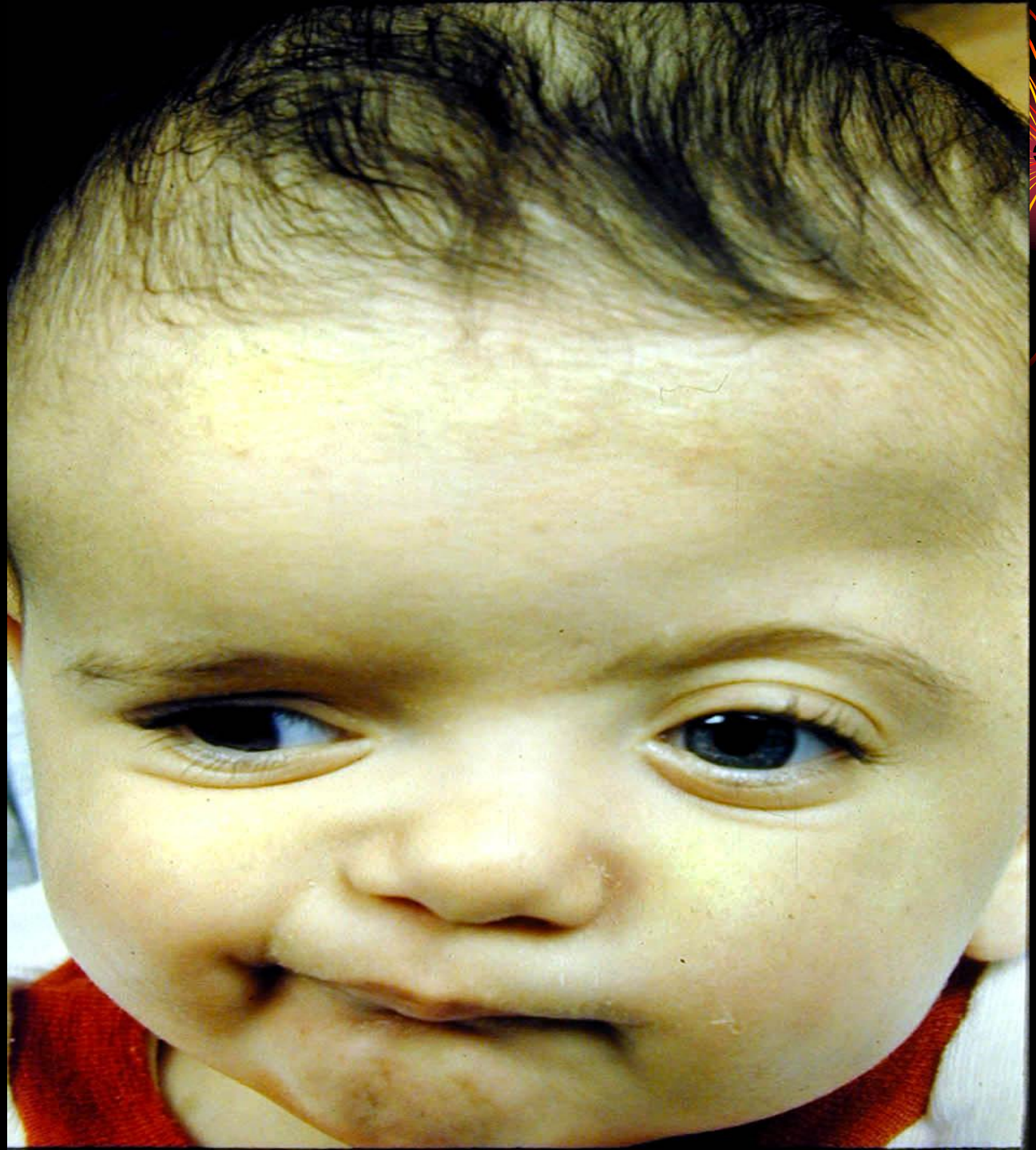


Plagiocephaly

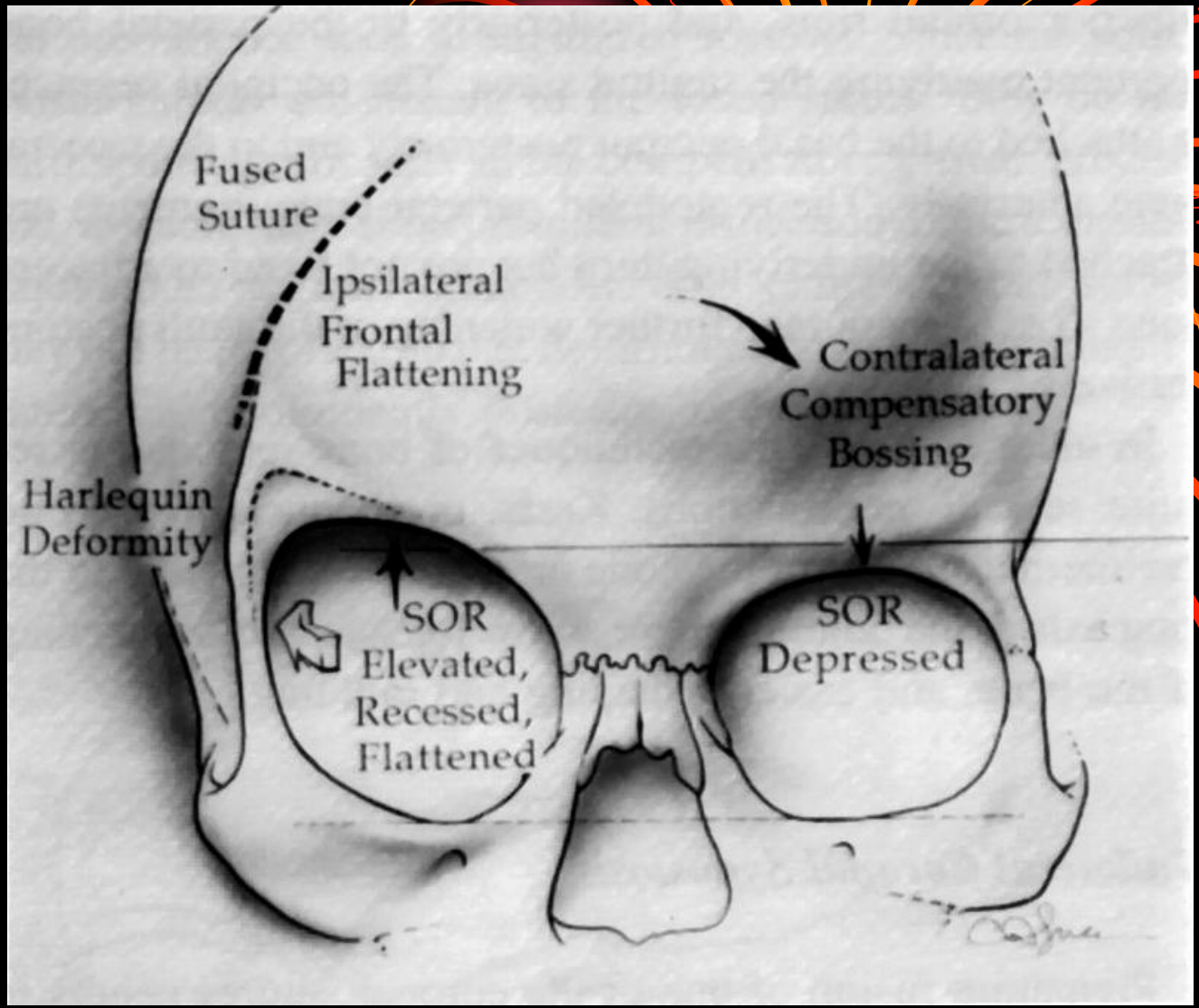


- Frontal
  - Ipsilateral coronal and frontosphenoidal sutures, flat forehead, backward displacement of orbit, bulging of contarlateral parietal area. Mandibular asymmetry.”harlequin” orbit.

- Occipital
  - Flattening Lamboid suture must be distinguished from positional plagiocephaly.

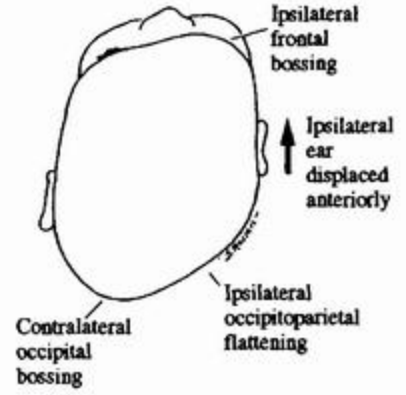


Plagiocephaly



Plagiocephaly

- Positional caused by extrinsic forces
- Positional plagiocephaly does not require surgical treatment.



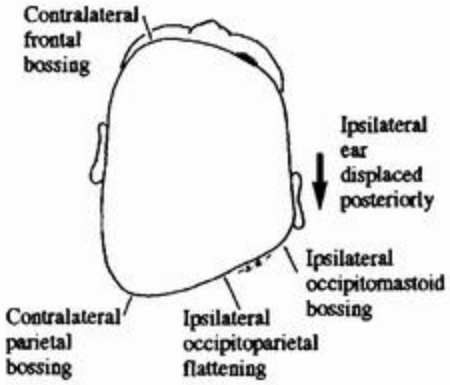


Fig 5. Some differences between positional molding *(top)* and unilambdoid synostosis *(bottom)*. *(Reprinted with permission from Huang MHS, Gruss JS, Clarren SK, et al: The differential diagnosis of posterior plagiocephaly: true lambdoid synostosis versus positional molding. Plast Reconstr Surg 98:765, 1996.)*

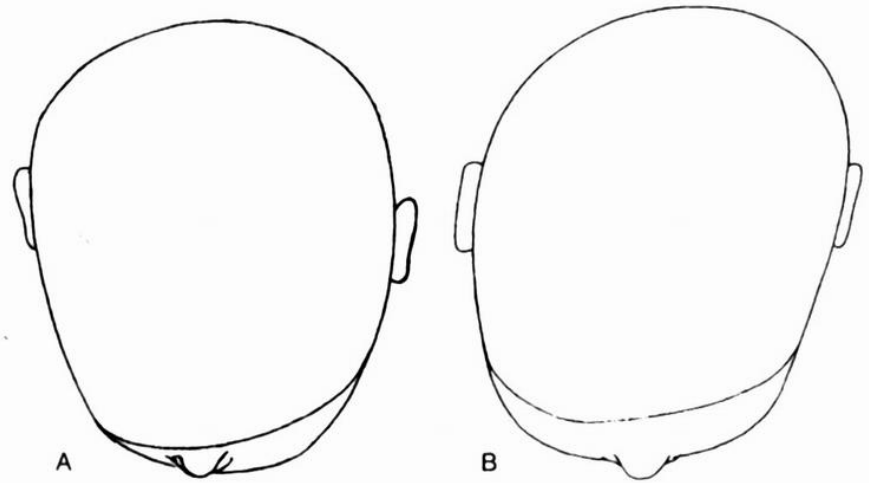
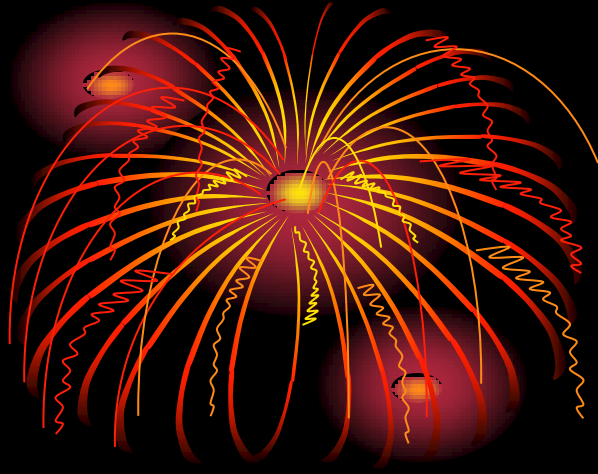
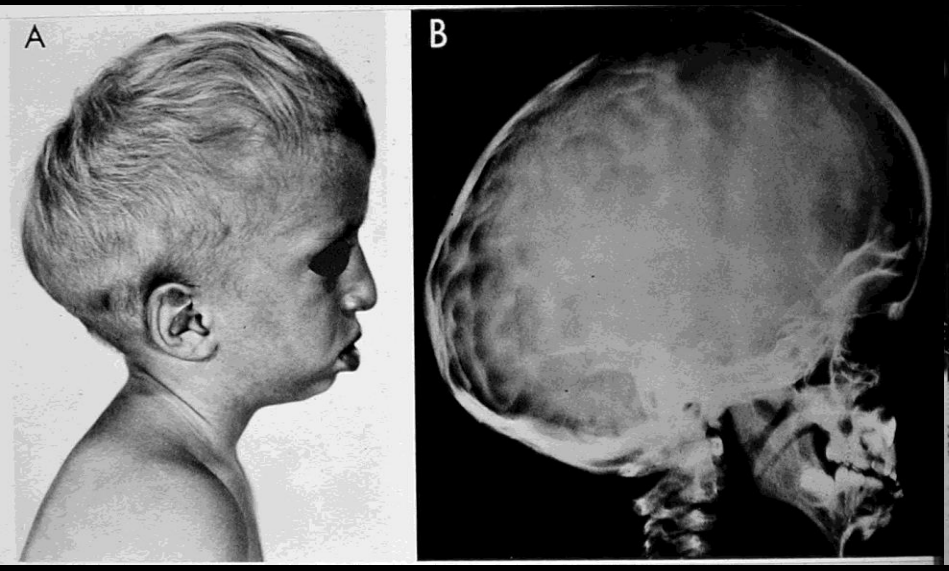


FIG. 15. Illustration of differences between left synostotic **(A)** and left deformational **(B)** frontal plagiocephaly. Note opposite configuration of left malar eminence and opposite position of ears. (*From* Hansen, M., and Mulliken, J. E. Frontal plagiocephaly: Diagnosis and treatment. In *Clin. Plast. Surg.* 21(4): 547, 1995.

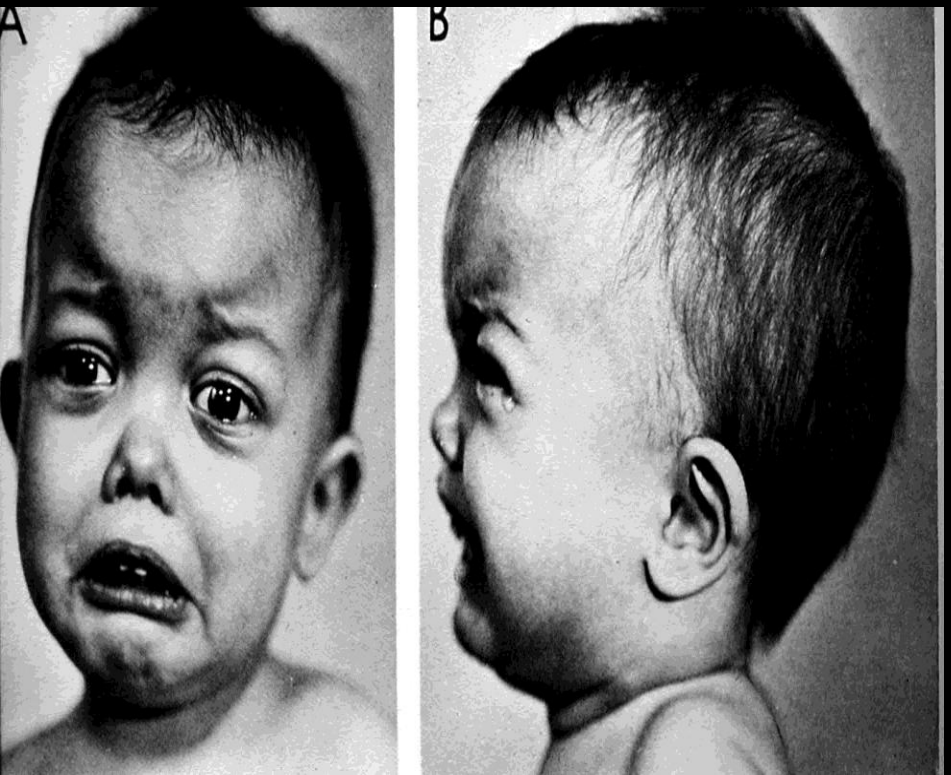


Syndromes:

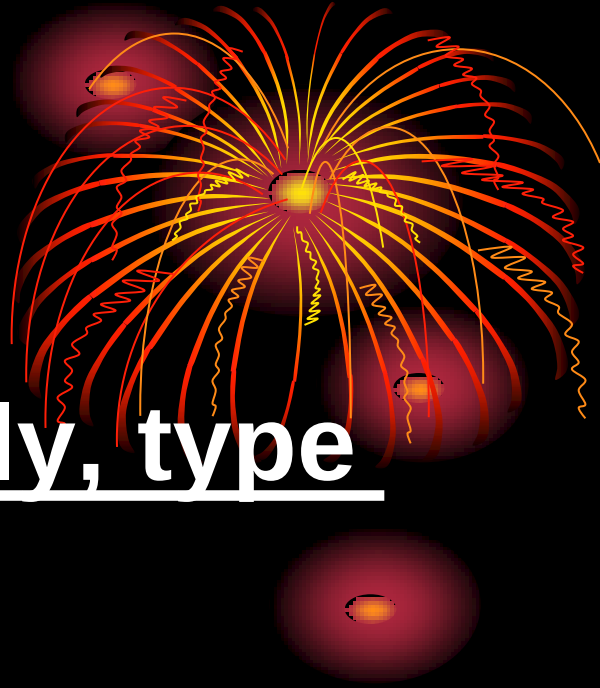
- Different sutures may be involved in different patients with the same syndrome.
- The difference may be in other parts of body.
- Mental retardation found to some extent in all syndromes.


A
B


A
B

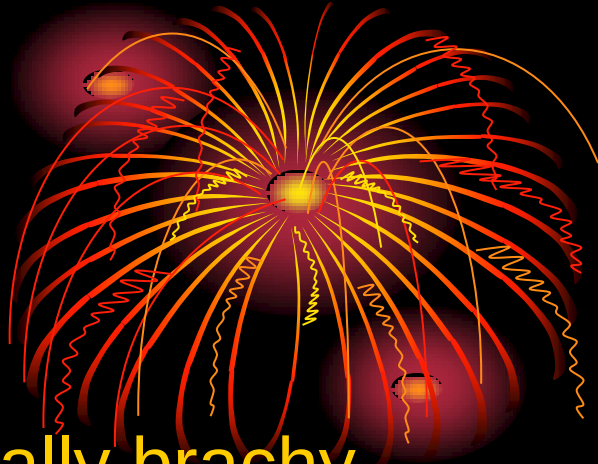

A
B

Crouzon (Craniofacial dysostosis) (Acrocephalosyndactyly, type II)

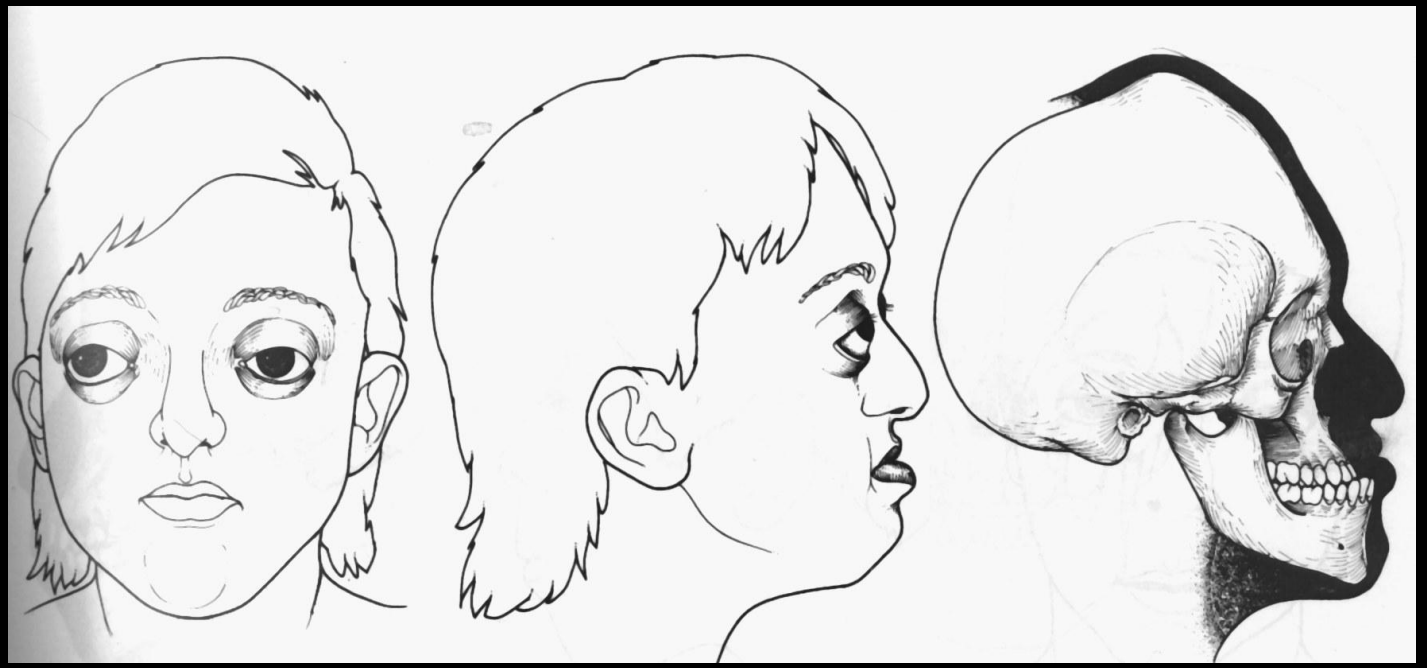


- 1:25,000
- Autosomal dominant (Most cases sporadic) Chromosome 10
- Craniosynostosis, midfacial hypoplasia, exopthalmos.

Crouzon cont



- Various dformations of skull- usually brachy.
- Midfacial retrusion – class III bite and shallow orbits. May cause damage to eyes. Beaked nose, High arched palate
- Conductive hearing loss is common.
- Synostosis develops during 1st year of life usually complete by 3rd.
- Increased ICP common.

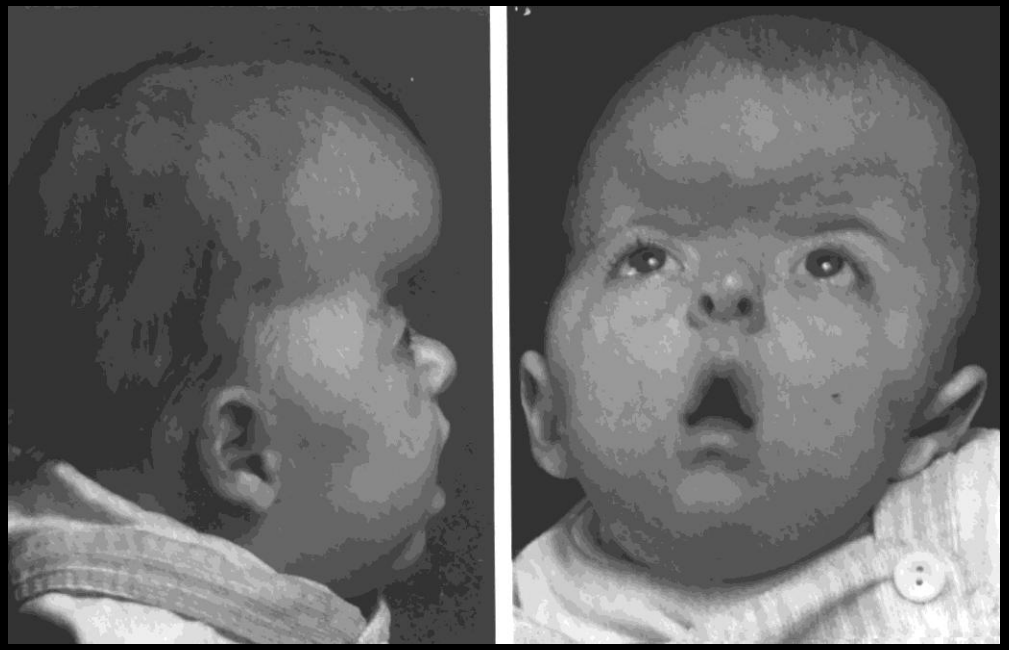


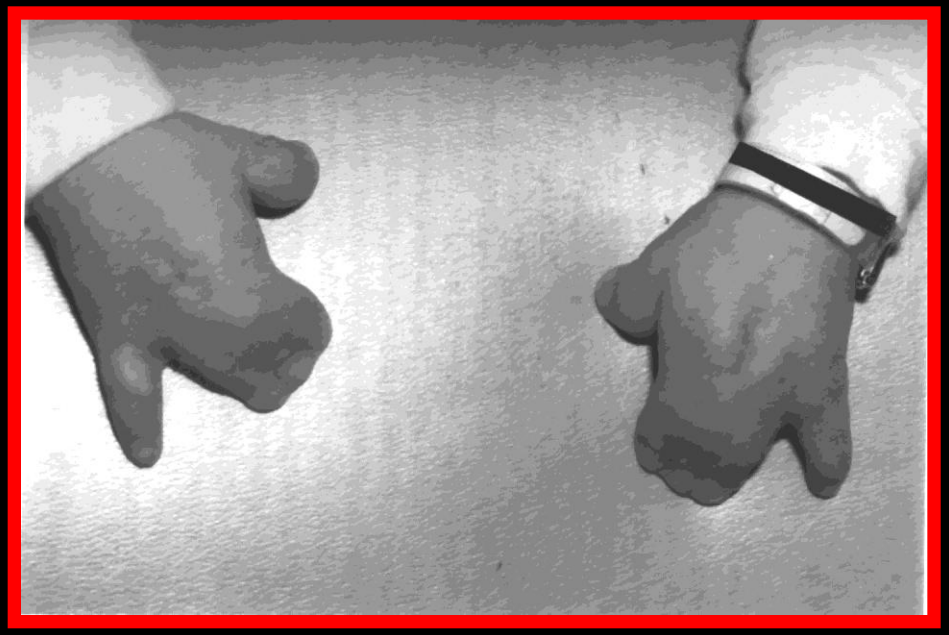
Apert (Acrocephalosyndactyly, type I)

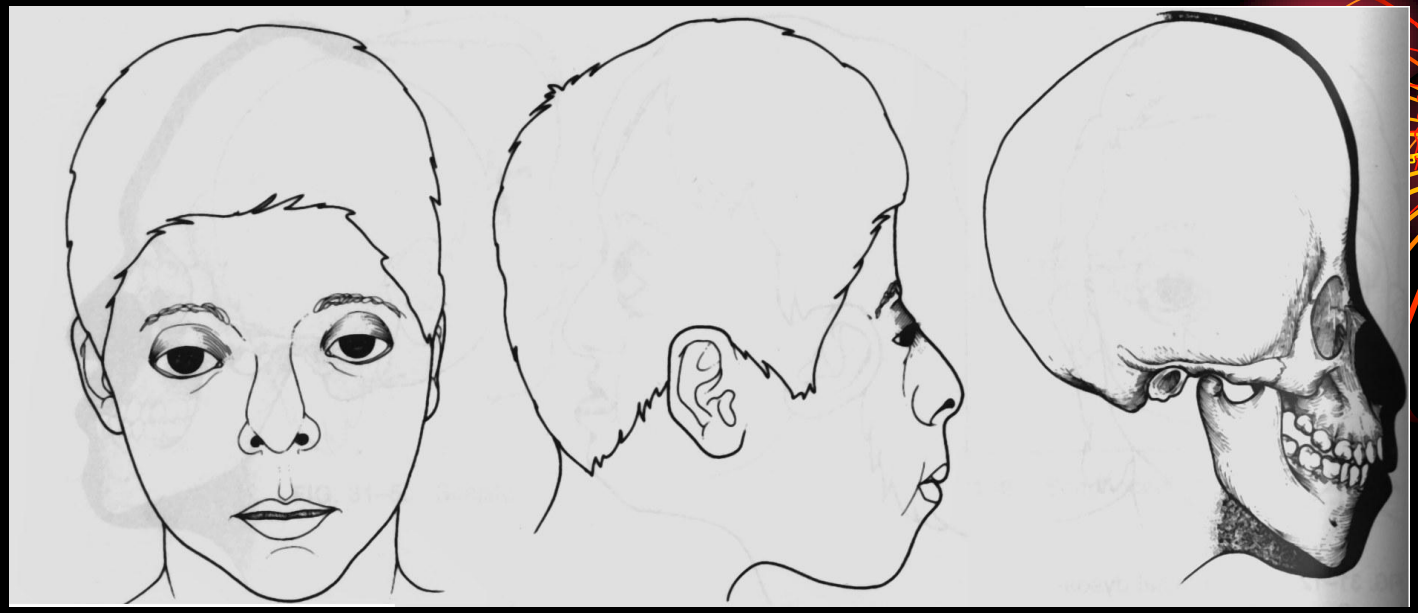
- 1:160,000
- Autosomal dominant (Most cases sporadic)
- High cranial vault, flat posteriorly while bulging in the front. Brachycephalic, Turribracephalic.
- Syndactyly of all four extremities- symmetrical middle three digits or more.

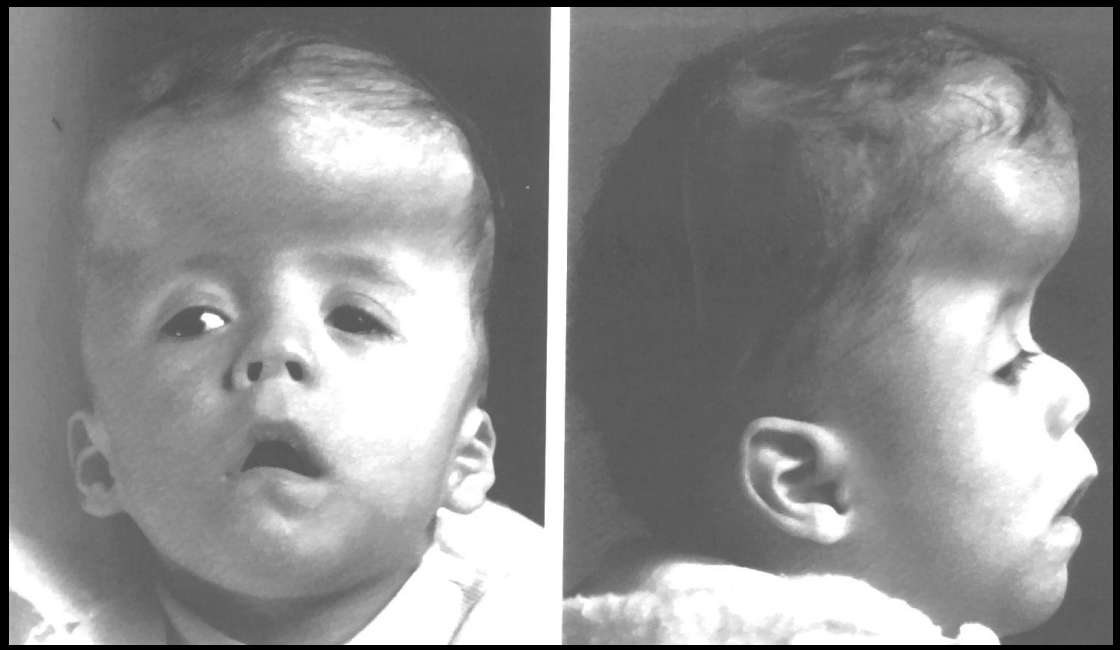
Apert cont

- Mild telorbitism, midfacial retrusion, deficient maxilla, abnormal dentition, Cleft palate 11-30%, low hair line, hypertrichosis of the eye brows, ptosis (mainly lateral), well developed protruding tongue.
- Mental reardation seen in most.

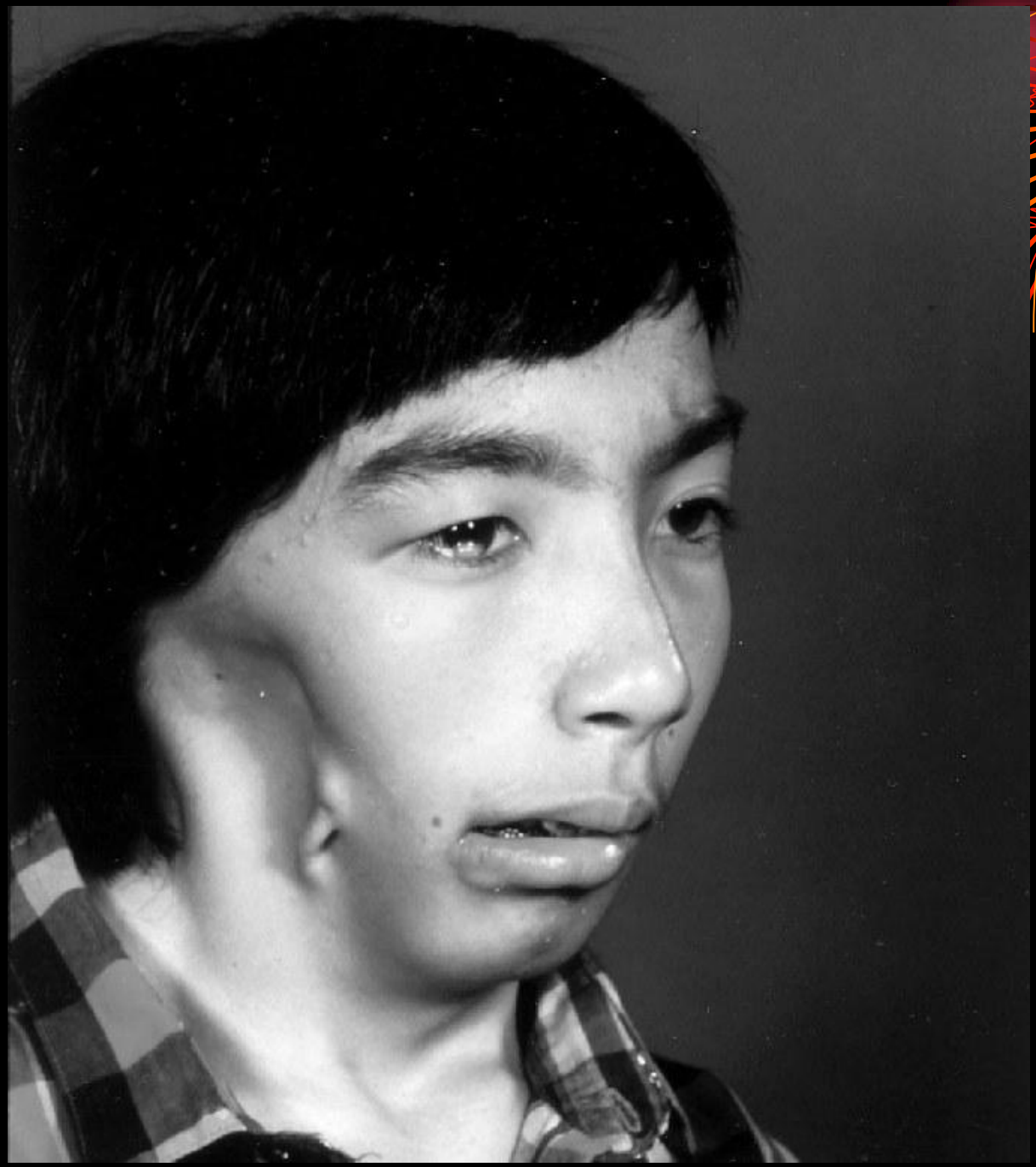








Hemifacial microsomia
First and second branchiall arch syndrome
Otomandibular dystosis
Craniofacial microsomia
Lateral facial dysplasia
Otomandibular syndrome



Craniofacial surgery

- surgery of the cephalic extremity
  - skull, face, Orbit
  - Congenital
  - Post traumatic
  - Tumors

Surgical repair

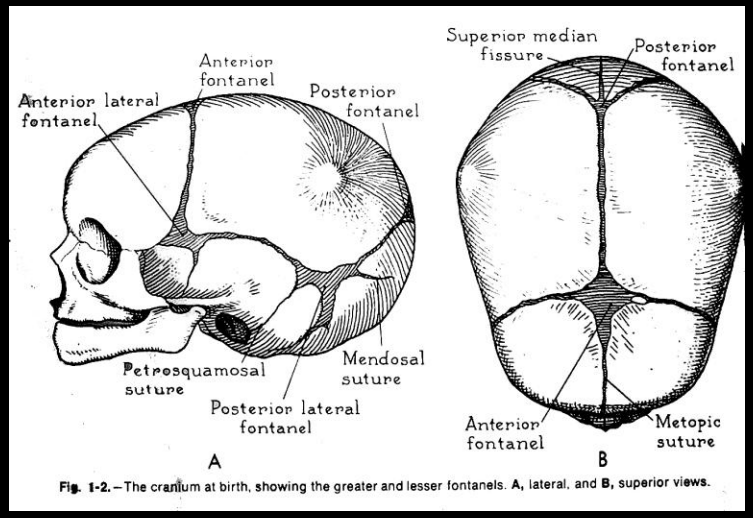
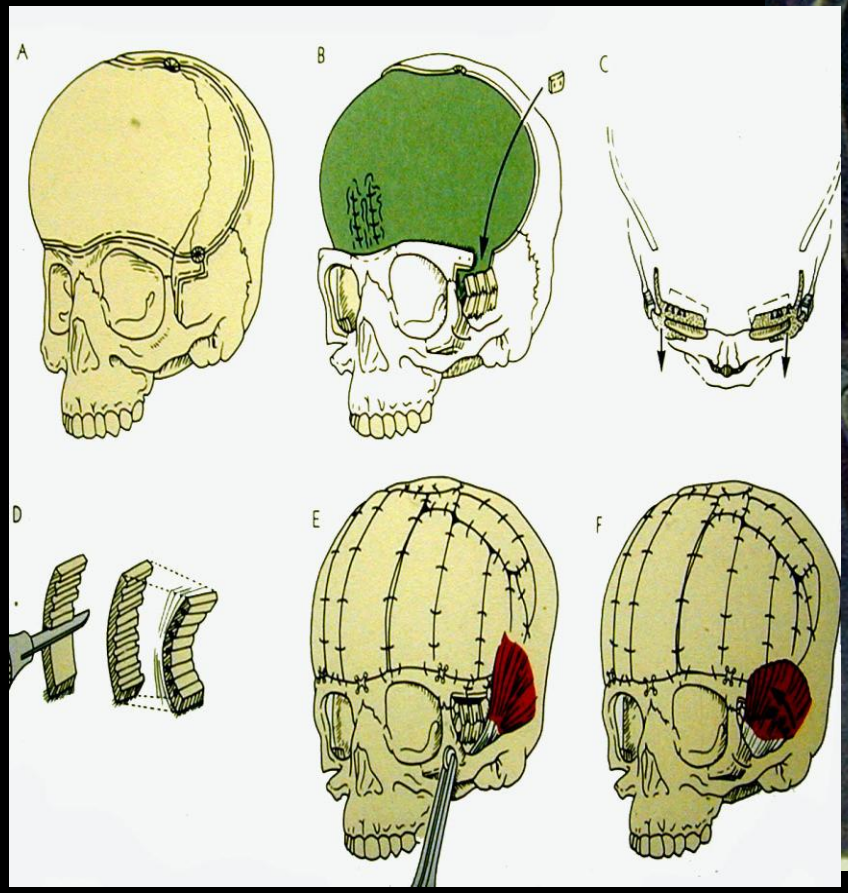
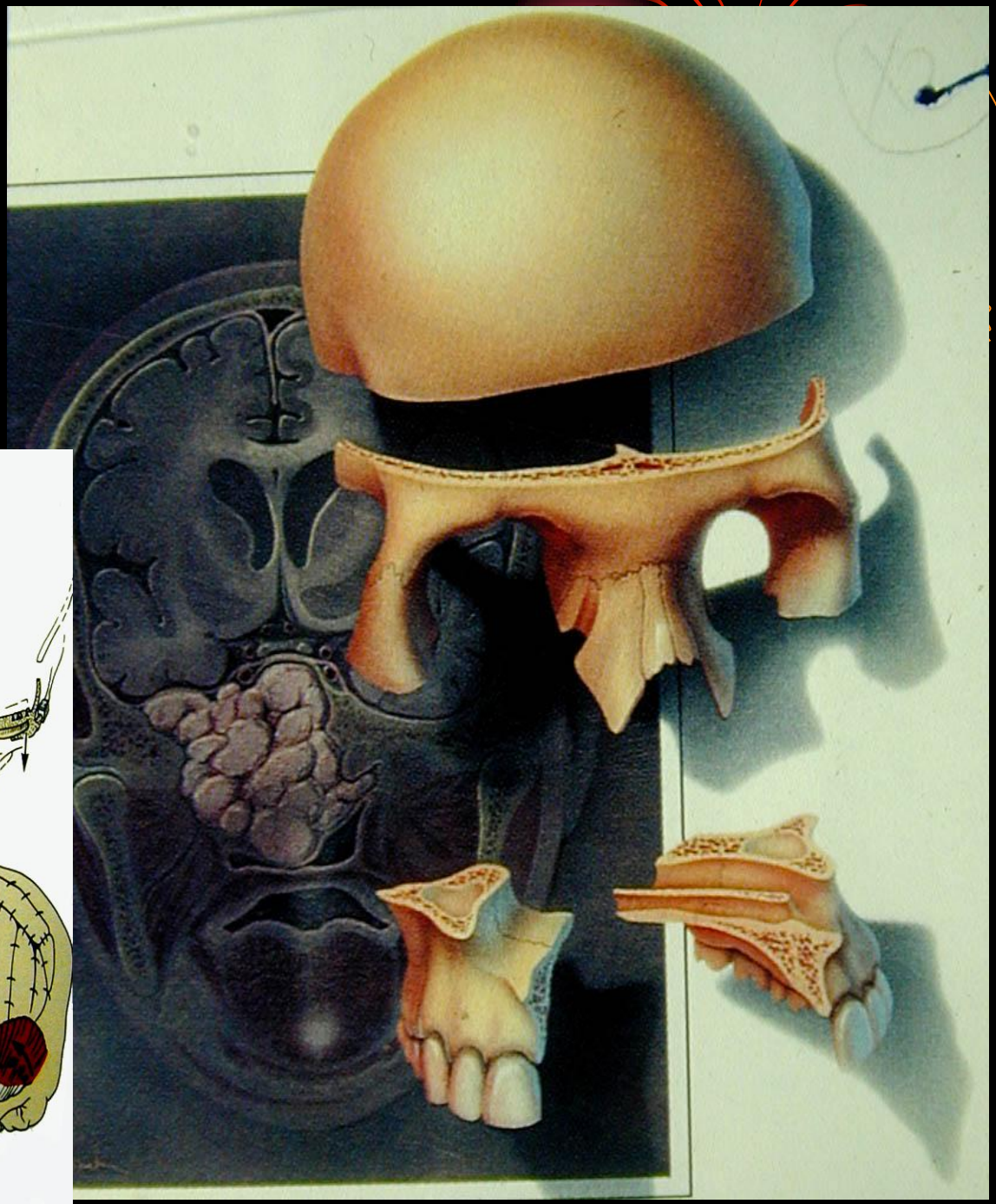


Fig. 1-2.—The cranium at birth, showing the greater and lesser fontanels. A, lateral, and B, superior views.



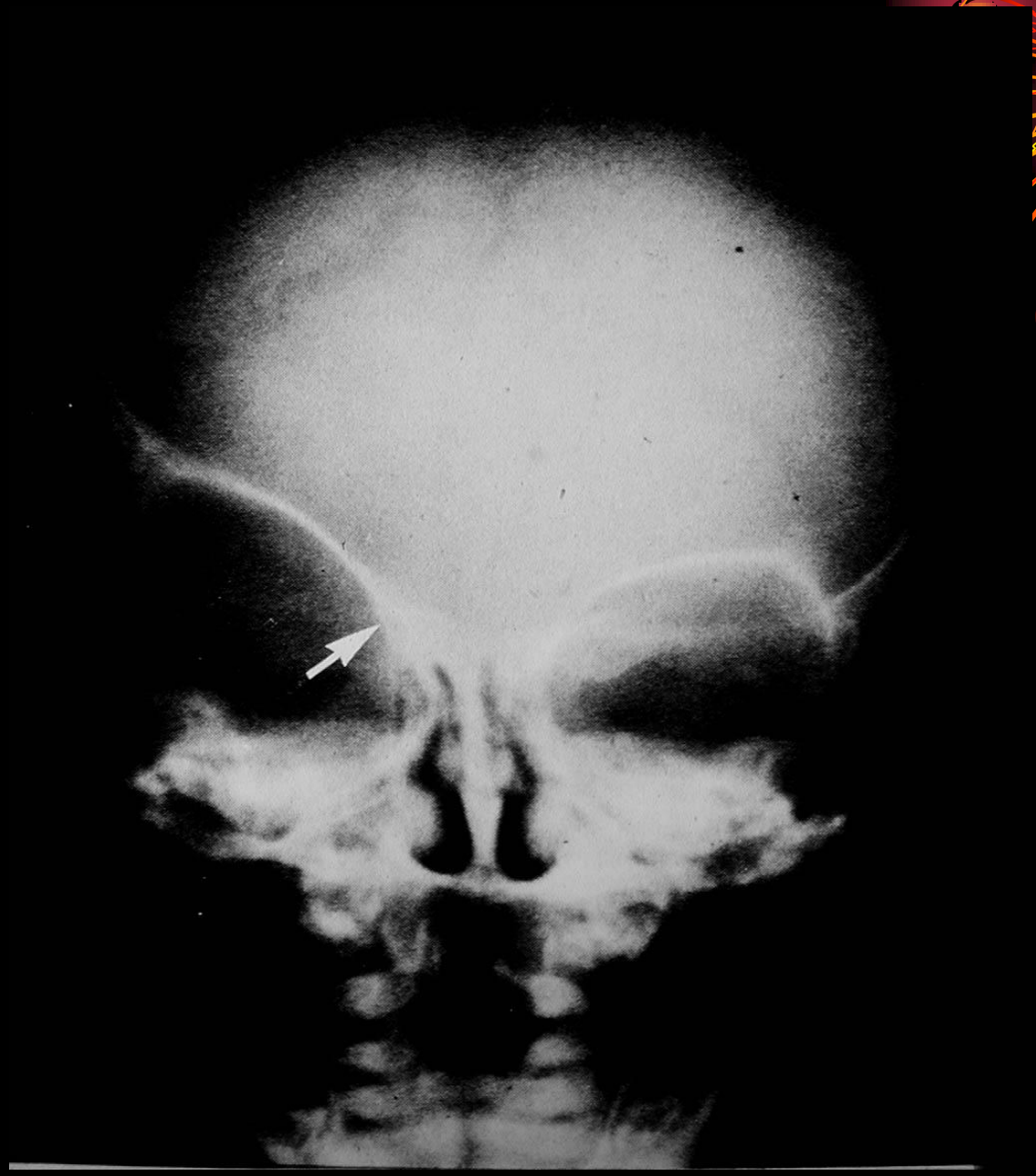


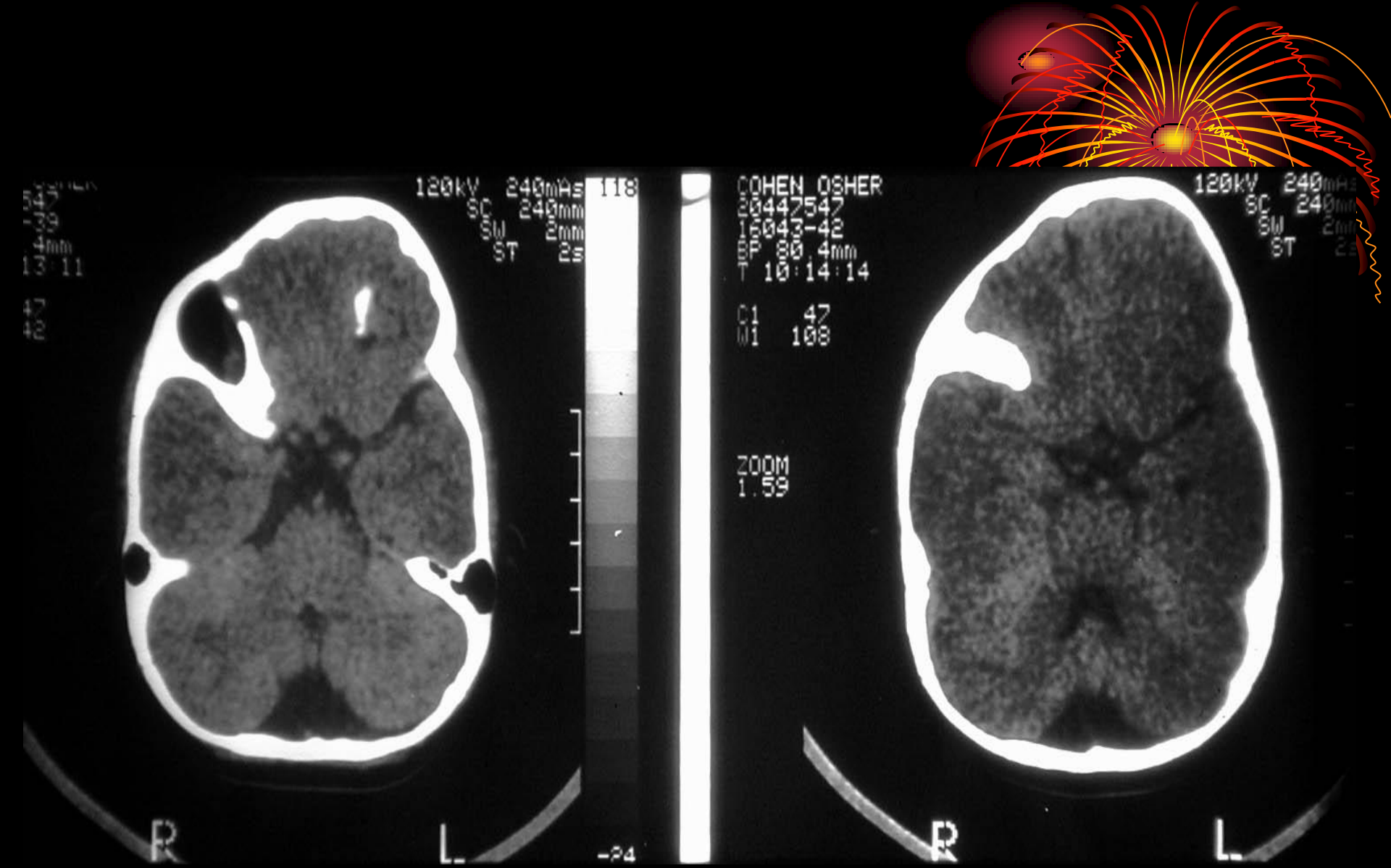
Pre operative evaluation

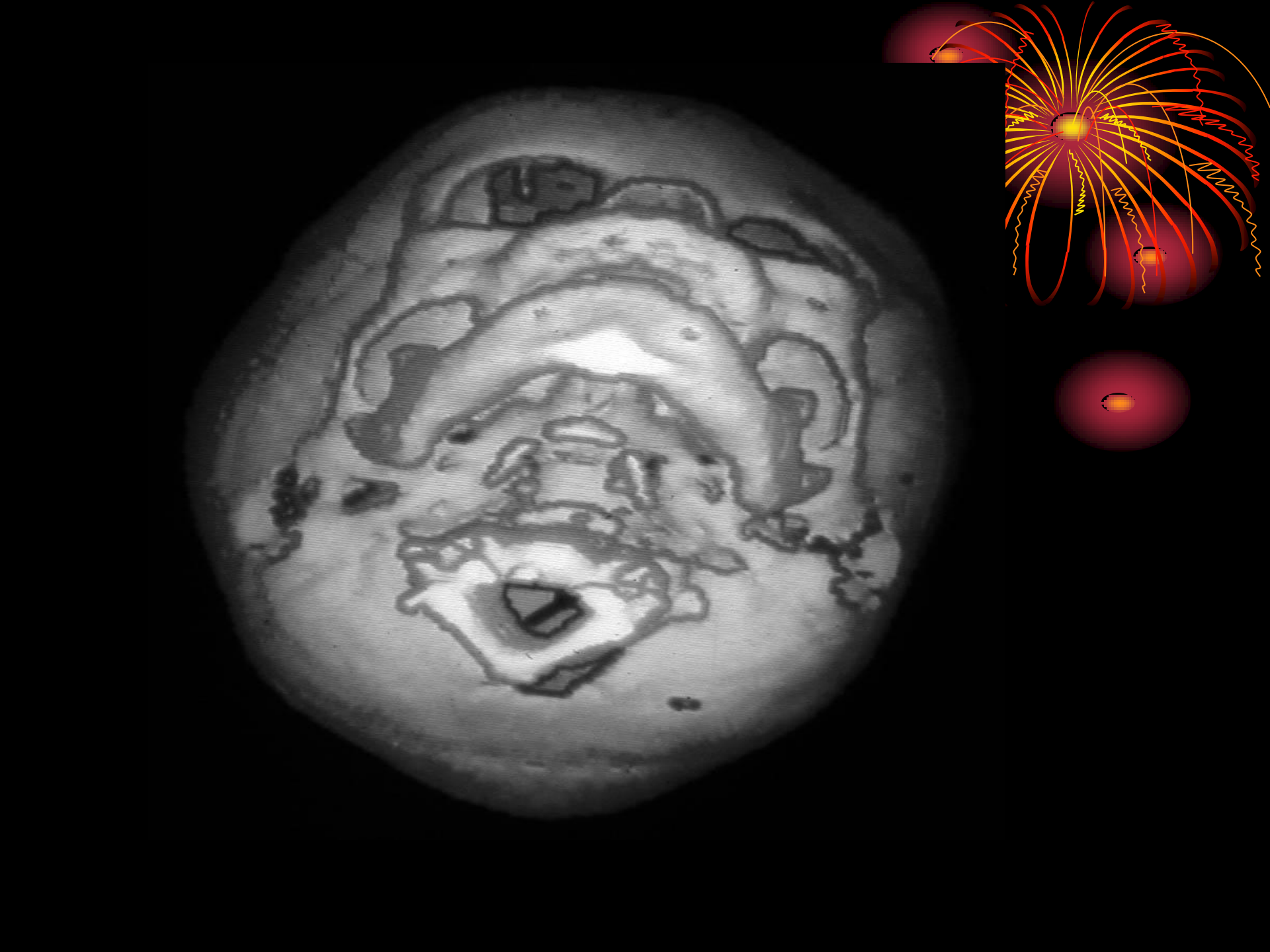
- History
  - Family, pregnancy..
- Physical
  - Observation, palpation of sutures, fontanels, neurological exam.
- Other abnormalities
  - search for other abnormalities

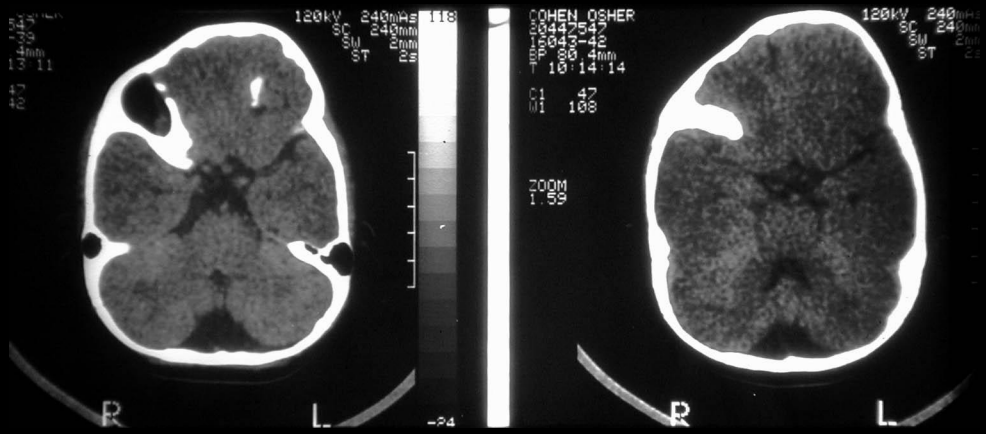
Pre operative evaluation

- Imaging
  - X-ray (including C-spine)
  - Cephalometris
  - CT 3DCT
  - MRI
- Imaging has a major importance in the management of these cases with the possibility of performing mock surgery




120kV 240mAs 118
SC 240mm
SW 2mm
ST 2s
COHEN OSHER
20447547
16043-42
BP 80.4mm
T 10:14:14
C1 47
W1 108
ZOOM
1.59
R
L
120kV 240mAs
SC 240mm
SW 2mm
ST 2s
R
L




120kV 240mAs
SC 240mm
SW 2mm
ST 2s
COHEN OSHER
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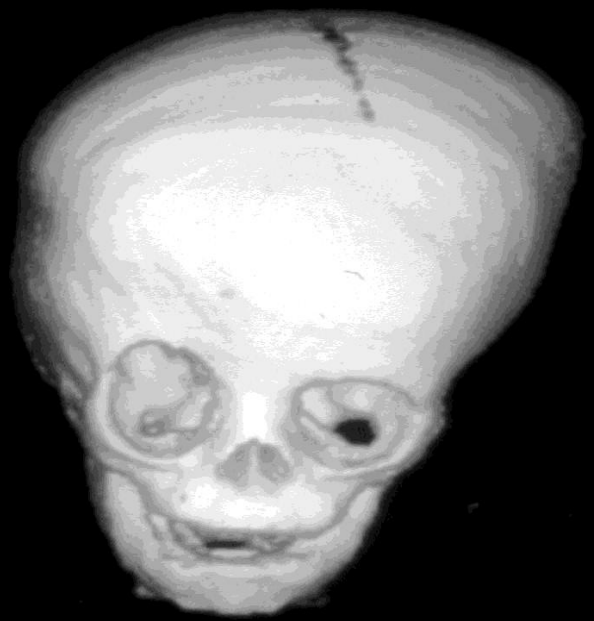
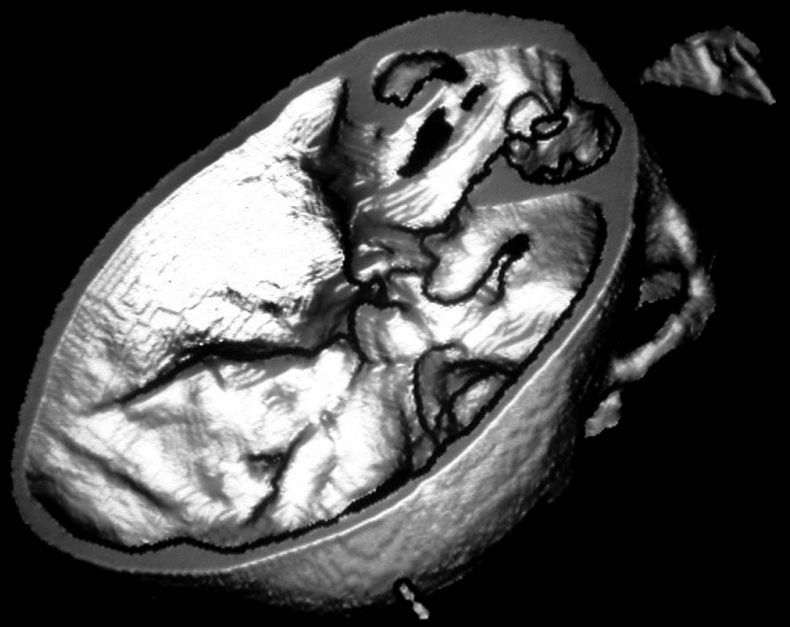
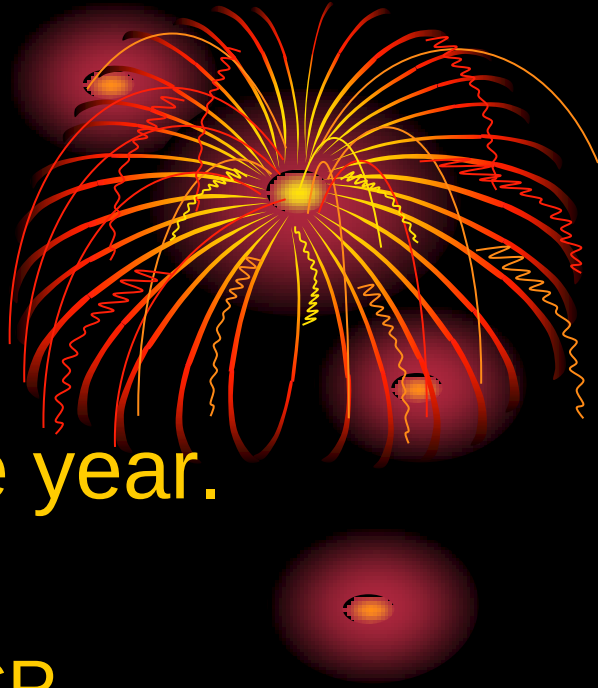
ZOOM
1.59



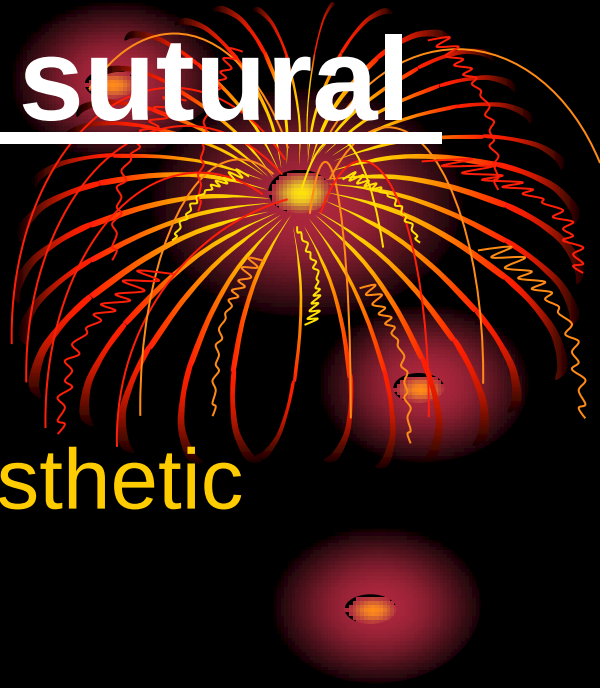

5 cm

Timing of surgery



- The consensus is before one year.
- Earlier
  - Better growth, early relief of ICP.
- Later
  - Child stands surgery better, nearer to adult size.
- Facial osteotomies
  - are performed at a later age.

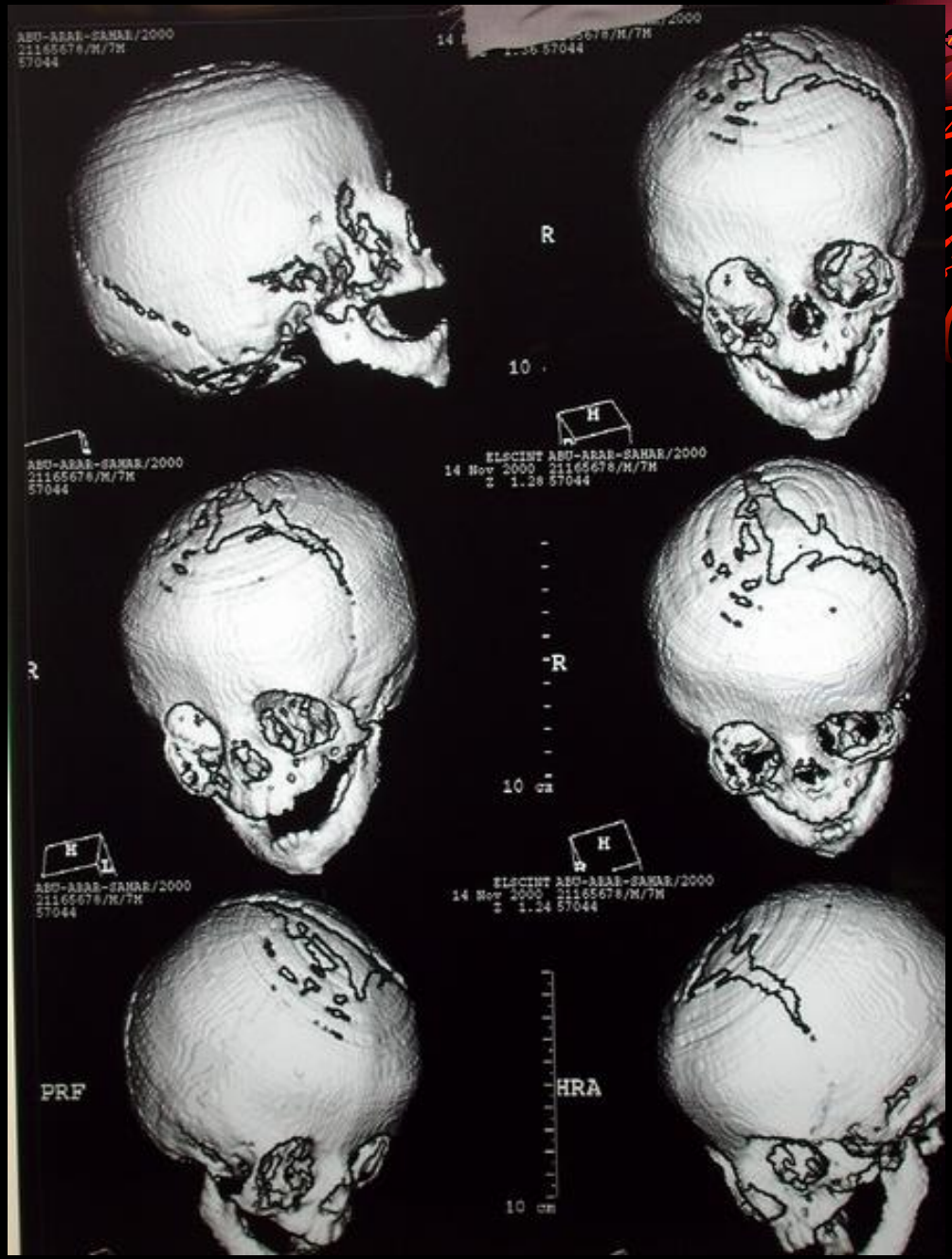
Correction of simple sutural synostosis

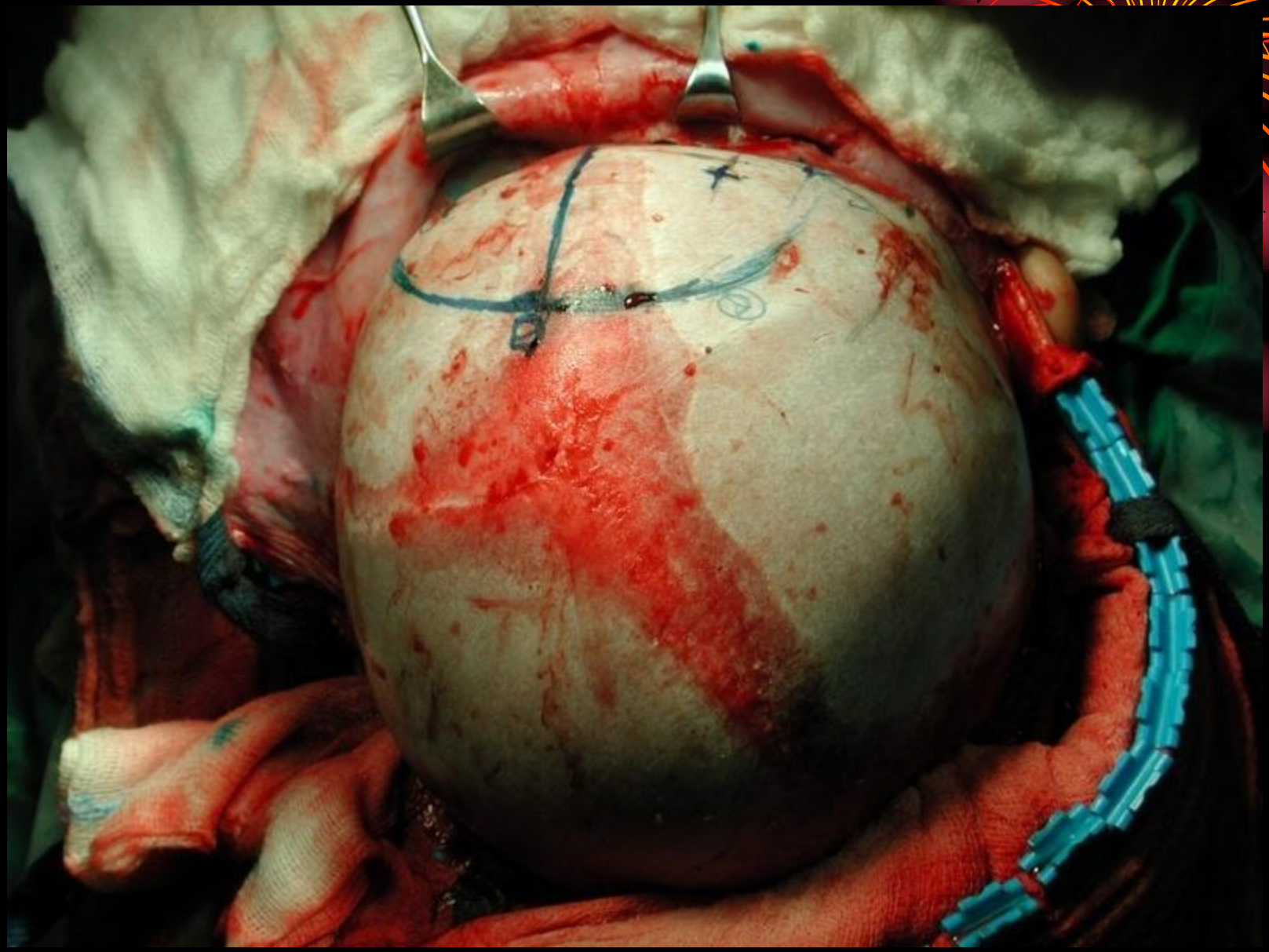


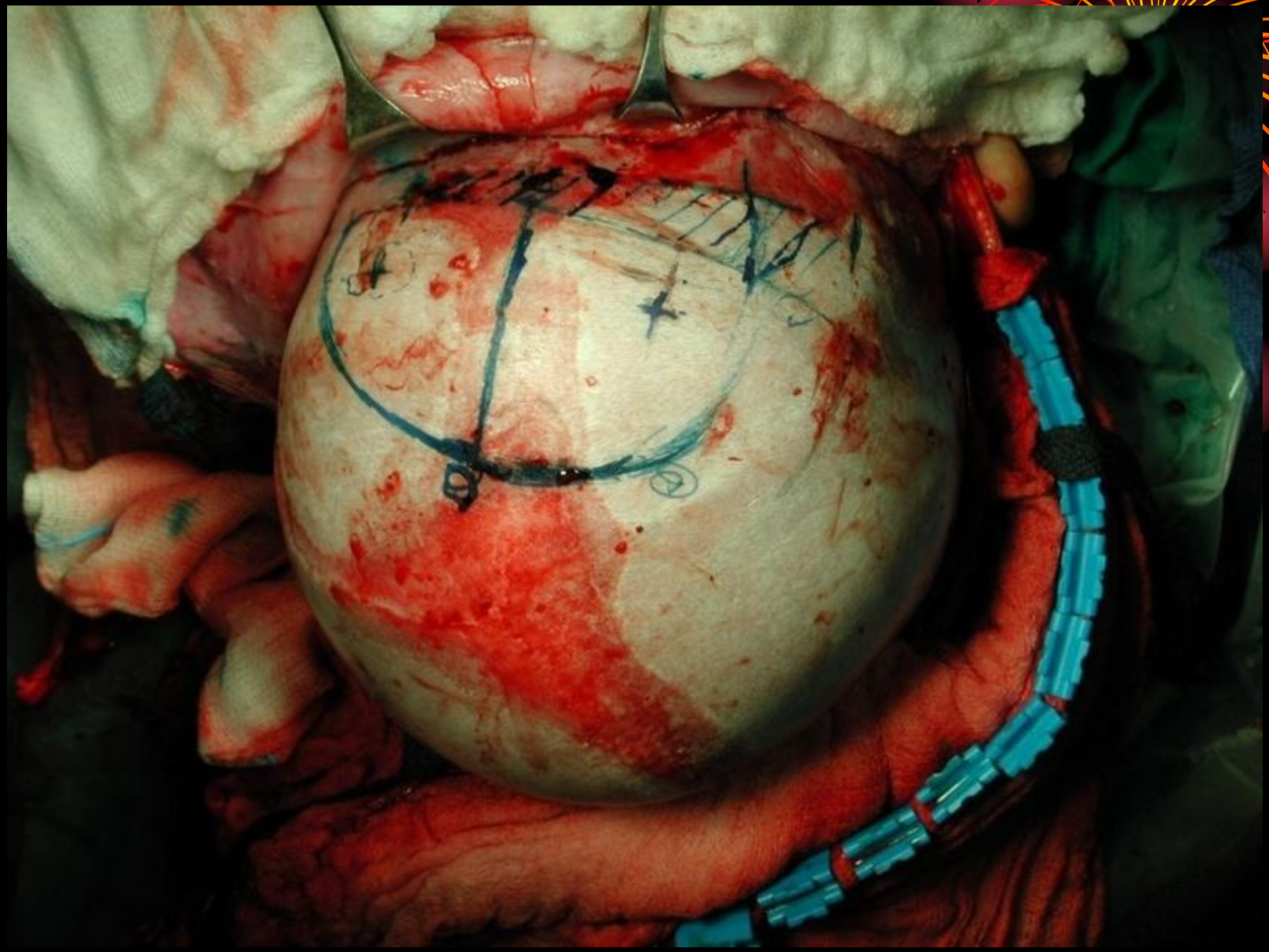
- Aim: correct functional and esthetic deficits
- Strip craniotomies
  - Not suficent for an esthablished deformity but may be used in infancy, Bone growth and regeneration fill the defects.

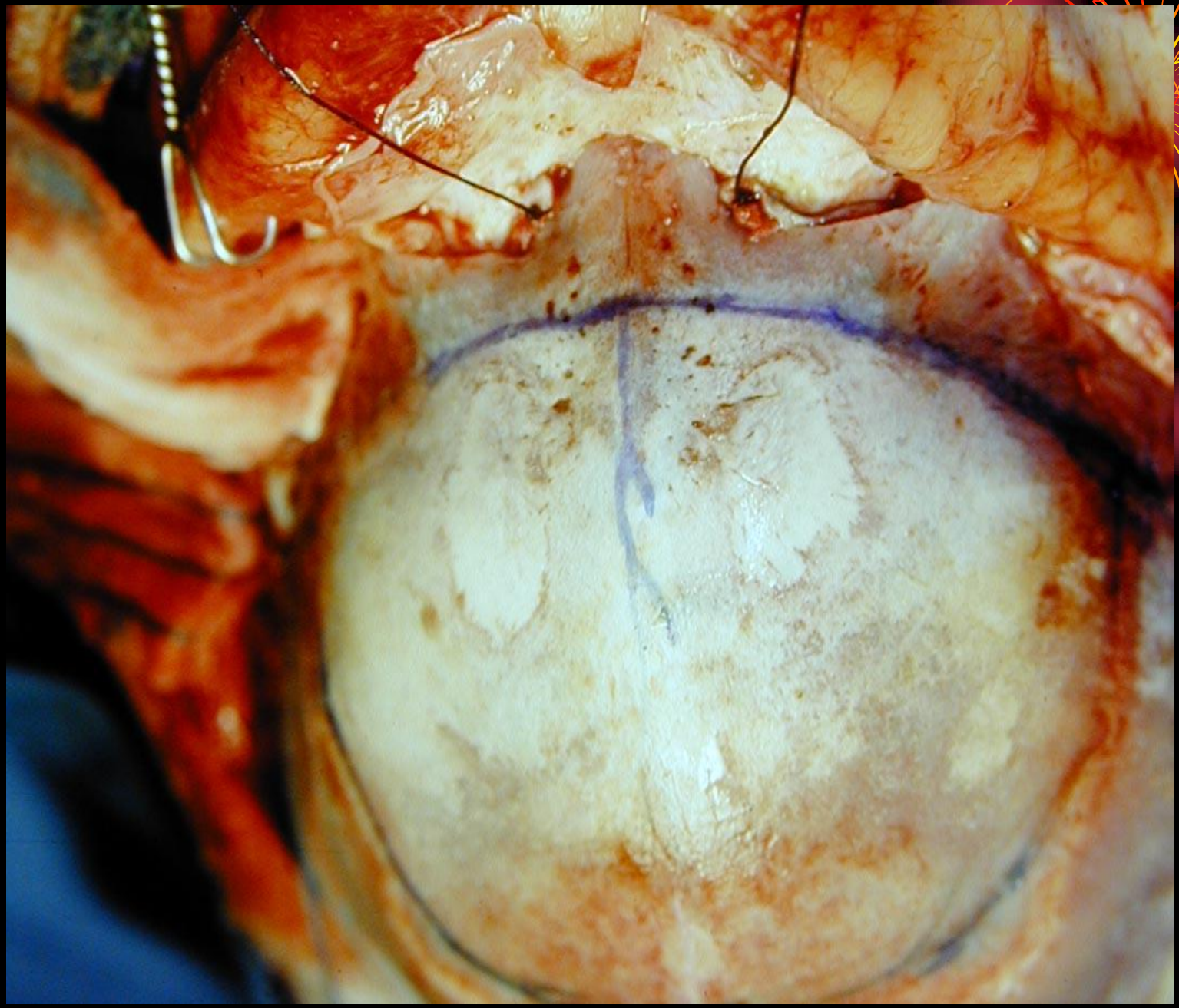
Correction of simple sutural synostosis

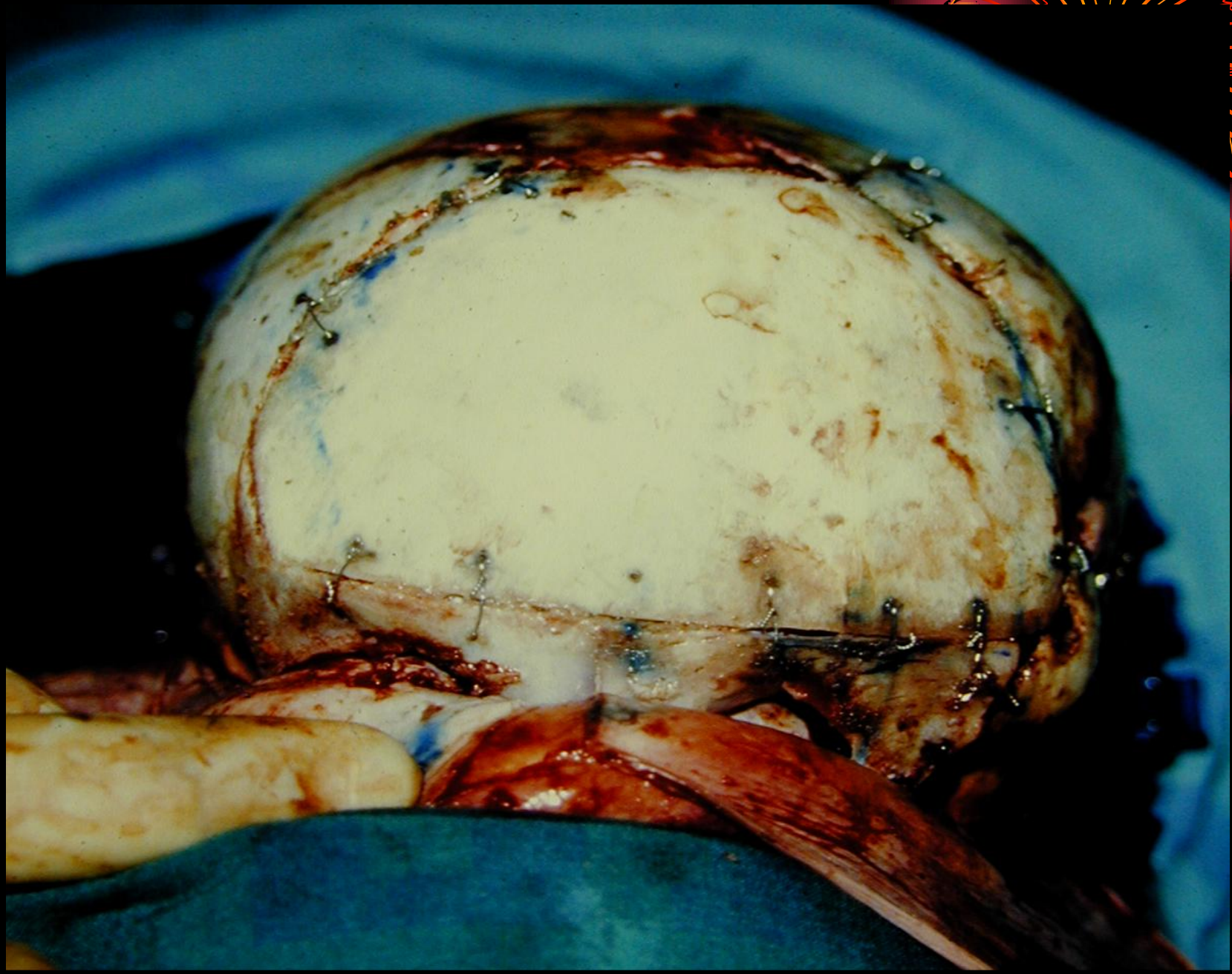
- Plagiocephaly
  - Frontoorbital remodeling
- Brachycephaly
  - Frontoorbital remodeling
  - The orbital bar is remodeled and repositioned in an appropriate location
- Metopic
  - Deformed frontal bone Remodeled or replaced by parietal bone

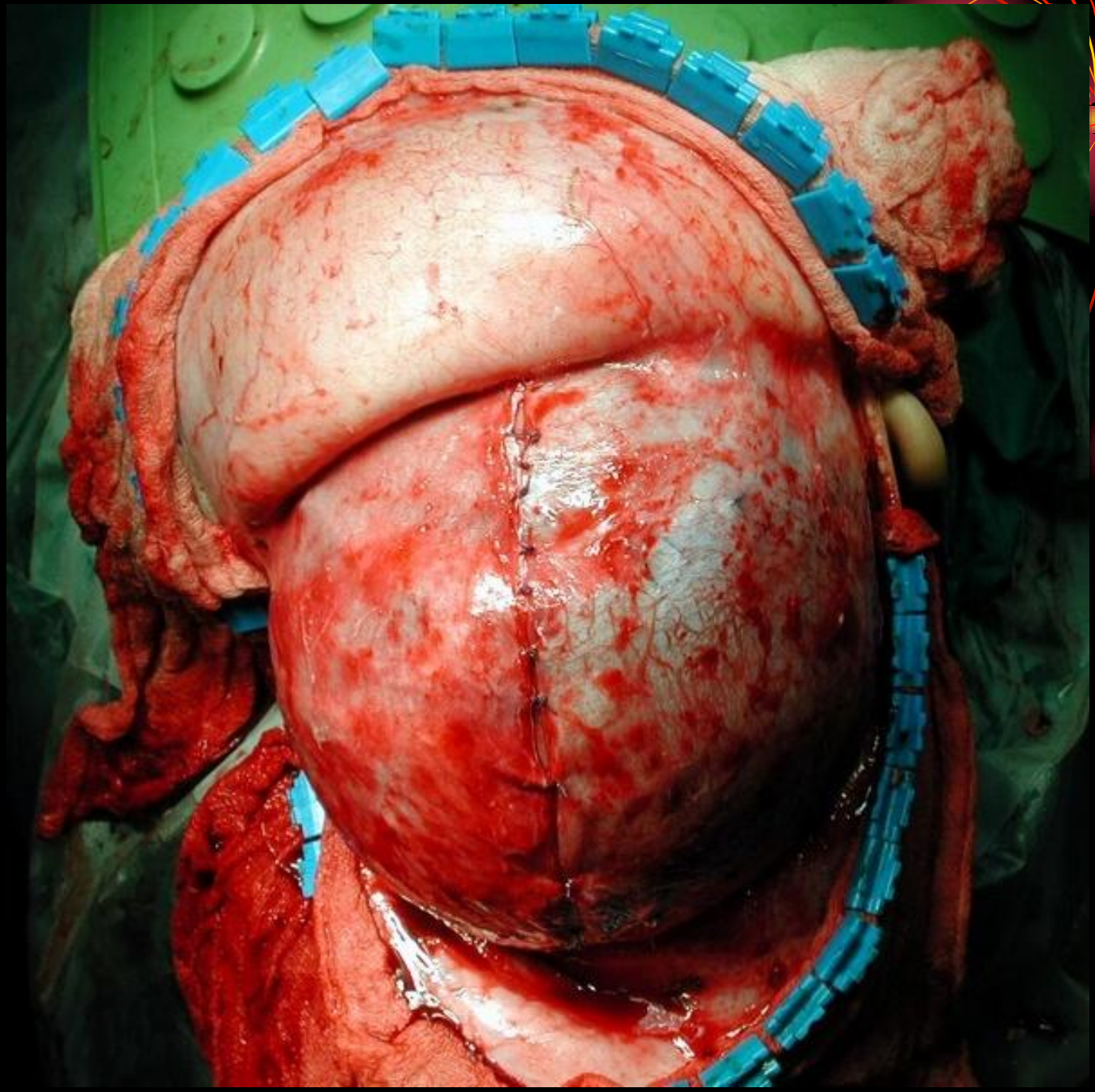

ABU-ARAR-SAHAR/2000
21165678/M/7M
57044
R
10
ELSCINT ABU-ARAR-SAHAR/2000
14 Nov 2000 21165678/M/7M
Z 1.28 57044
R
10 cm
ELSCINT ABU-ARAR-SAHAR/2000
14 Nov 2000 21165678/M/7M
Z 1.24 57044
PRF
HRA
10 cm





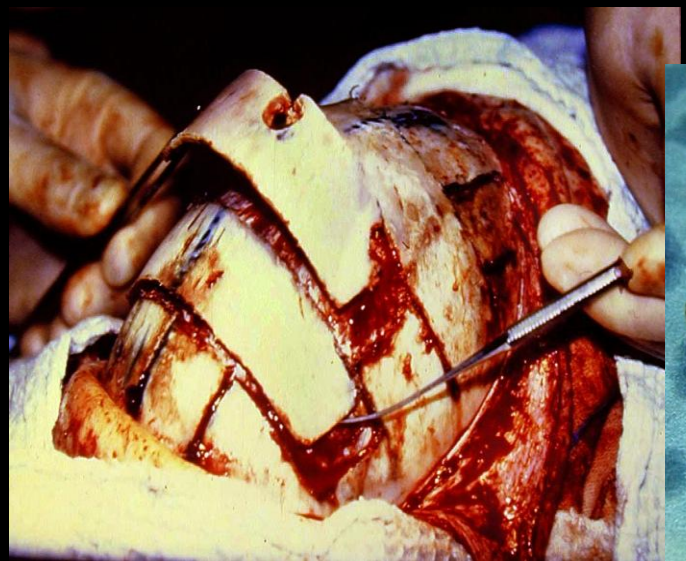







POD 7




Continuous soaking prevents edema & swelling and washes out bacteria & debris
D 5


2 Years


Father…

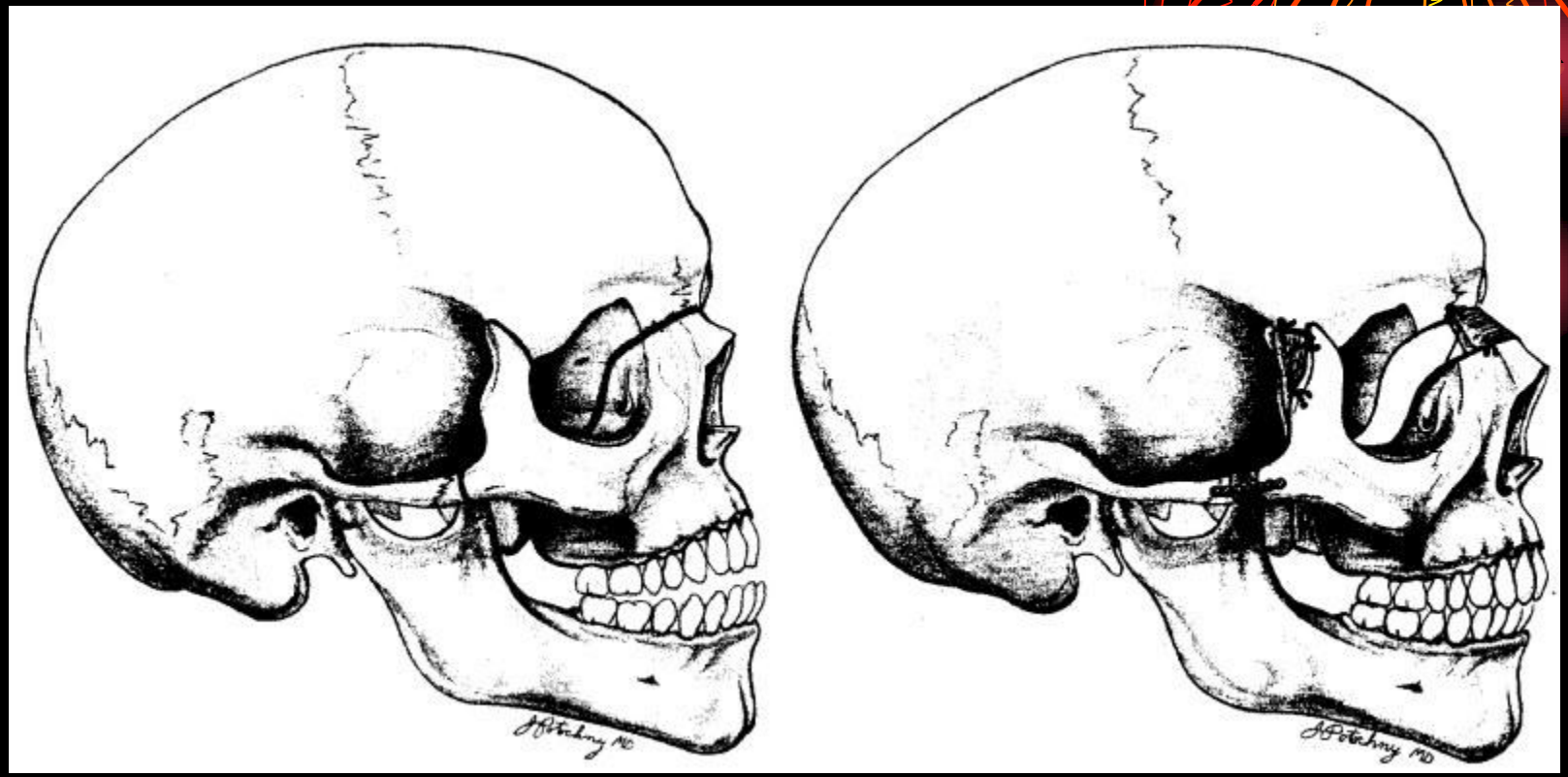


5 Years

Correction of syndromatic sutural synostosis

- Advancement of forehead and midface with monoblock frontal osteotomy.
- Le fort osteotomies.
- Facial bipartition for telorbitism.
- Mid face deformities
- Le Fort III, Le Fort III+I, Monoblock advancment

Le Fort III



Facial Bipartition

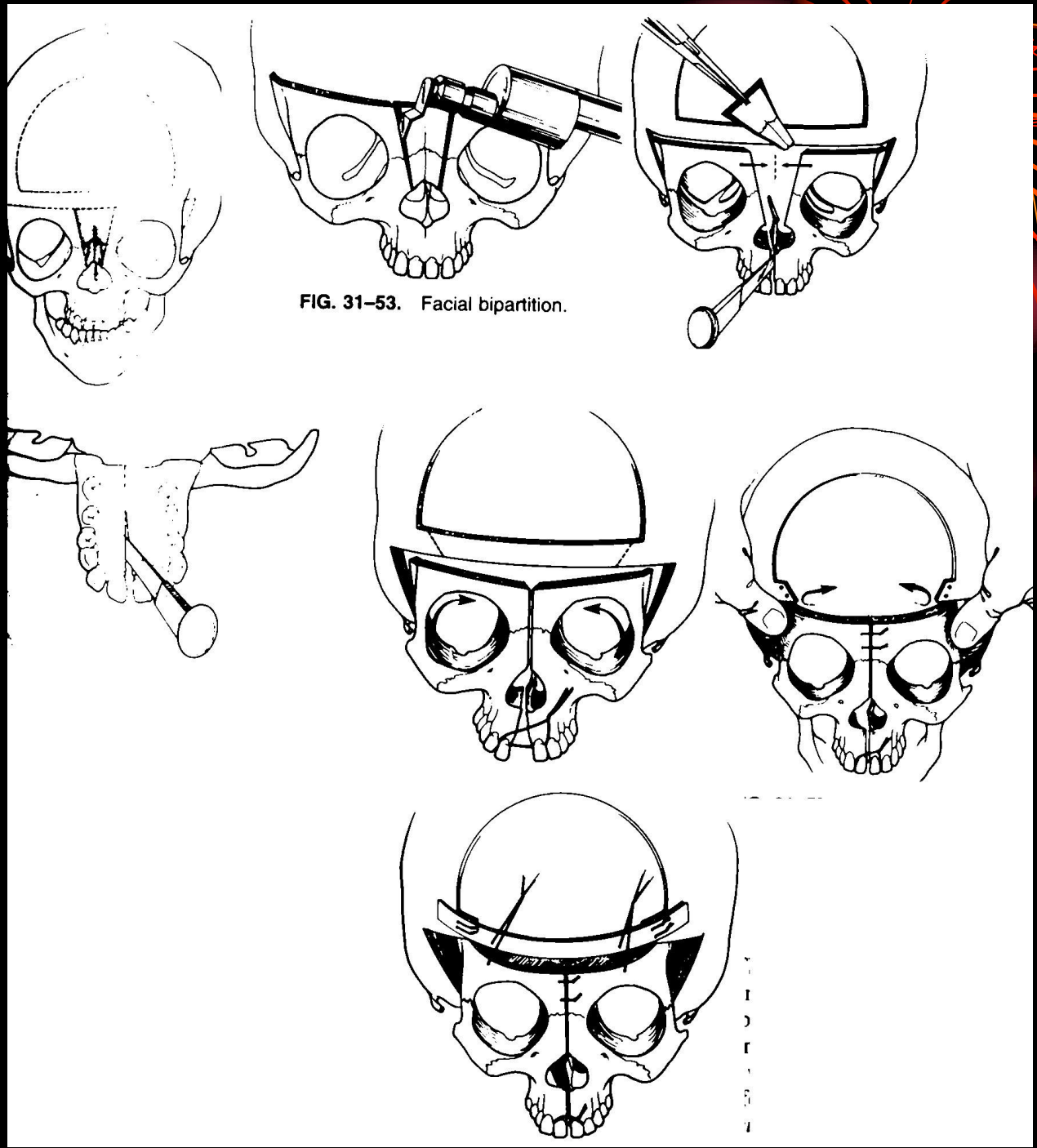
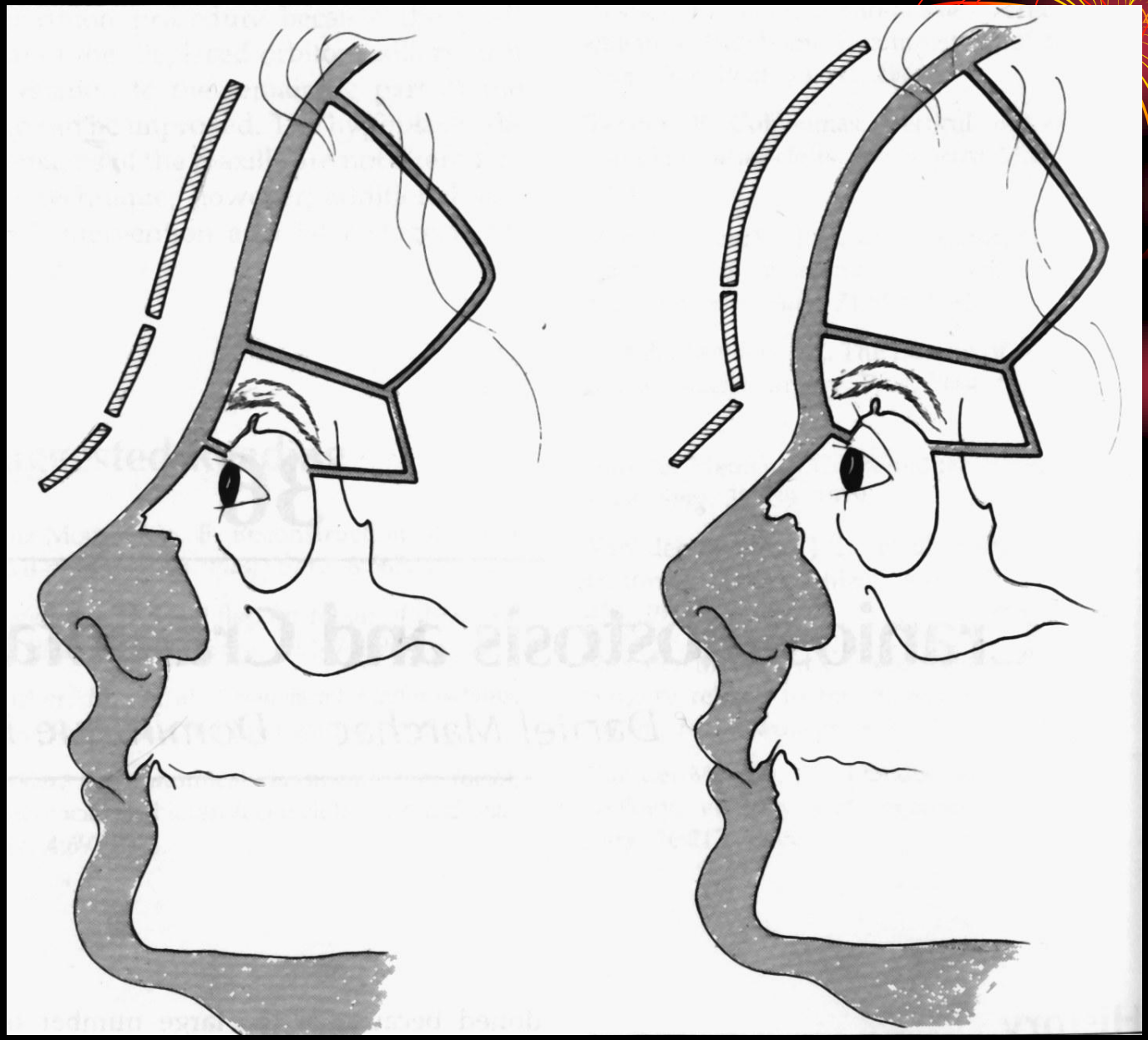
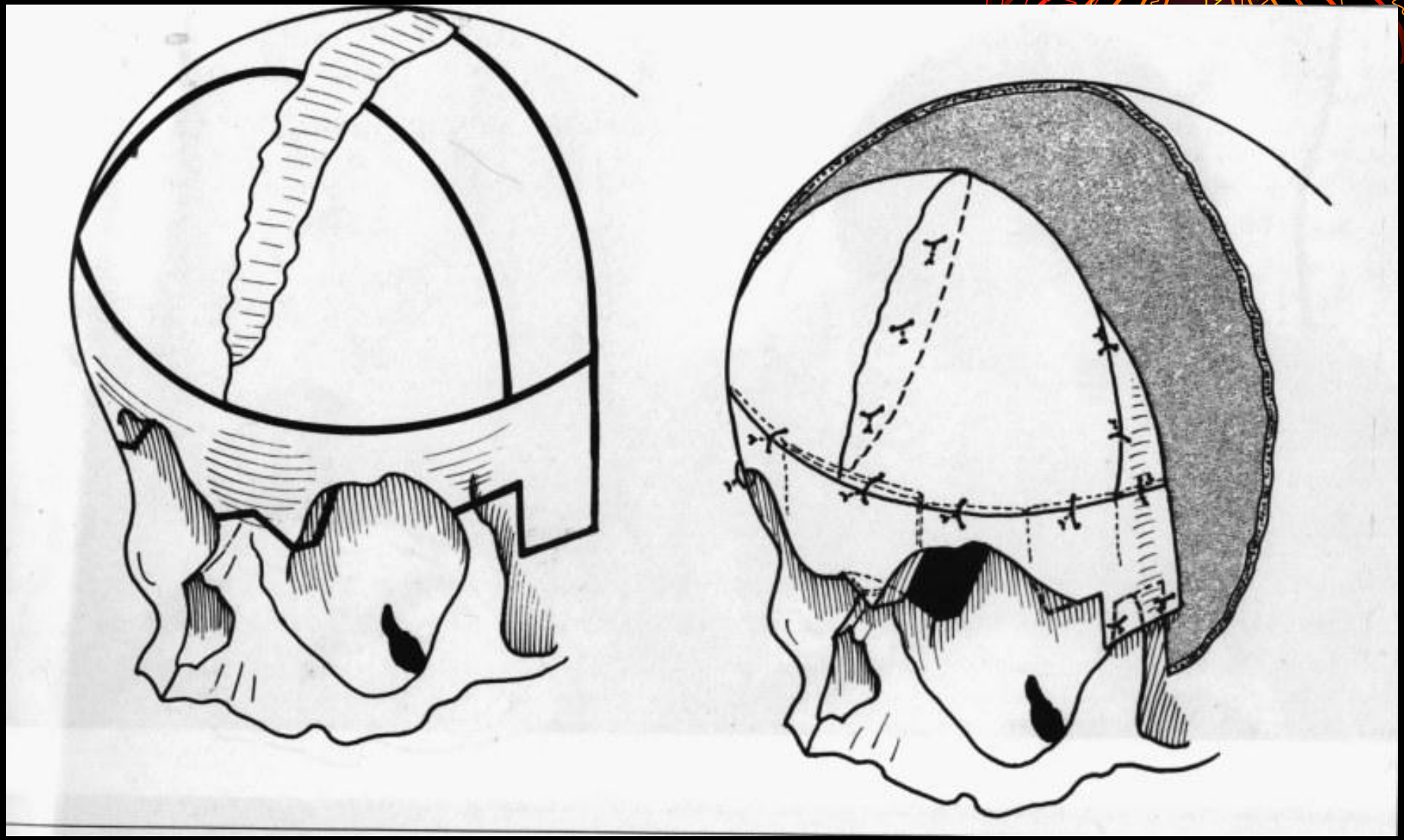


FIG. 31–53. Facial bipartition.

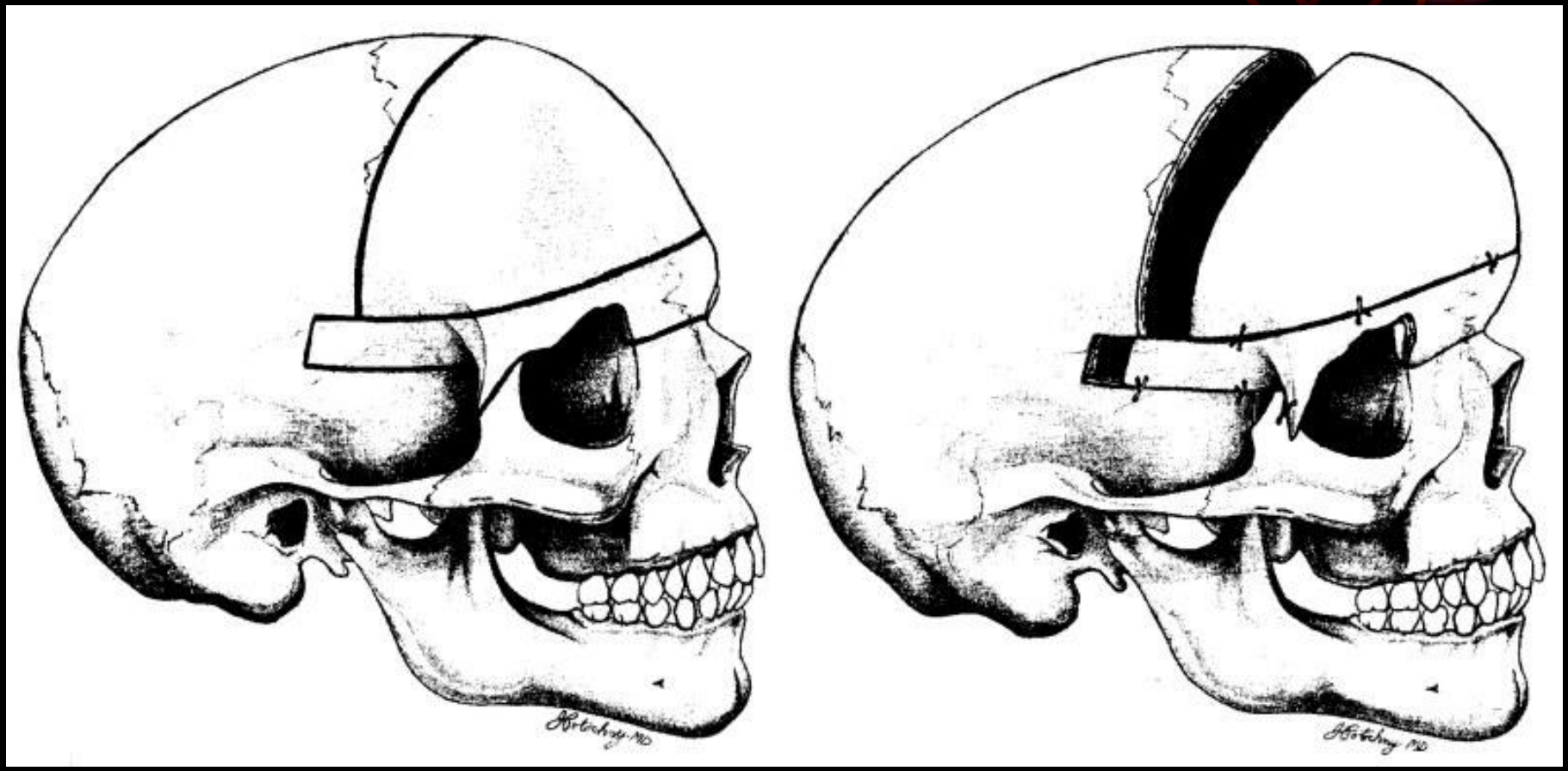
Forehead Advancement

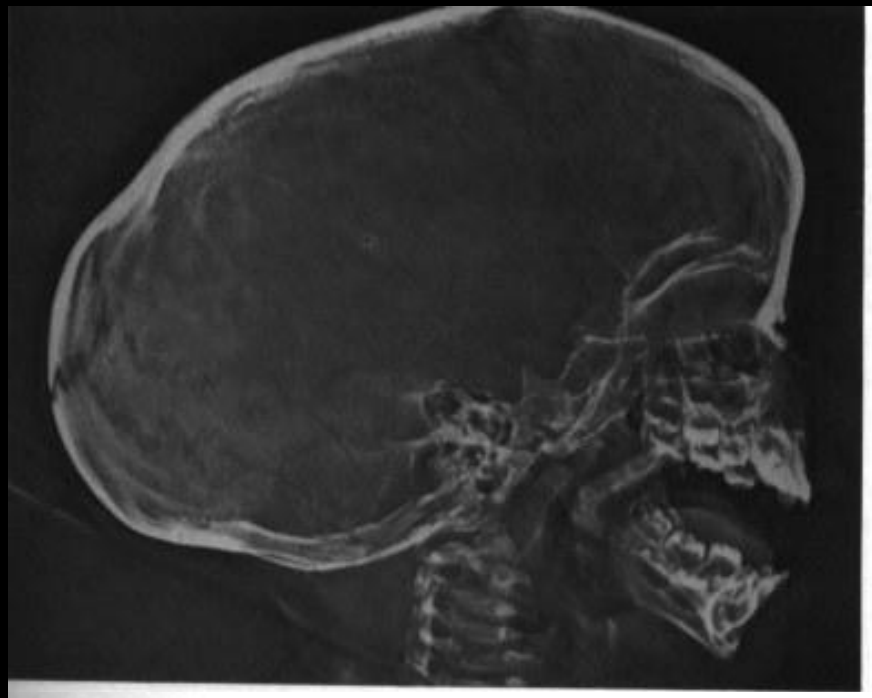


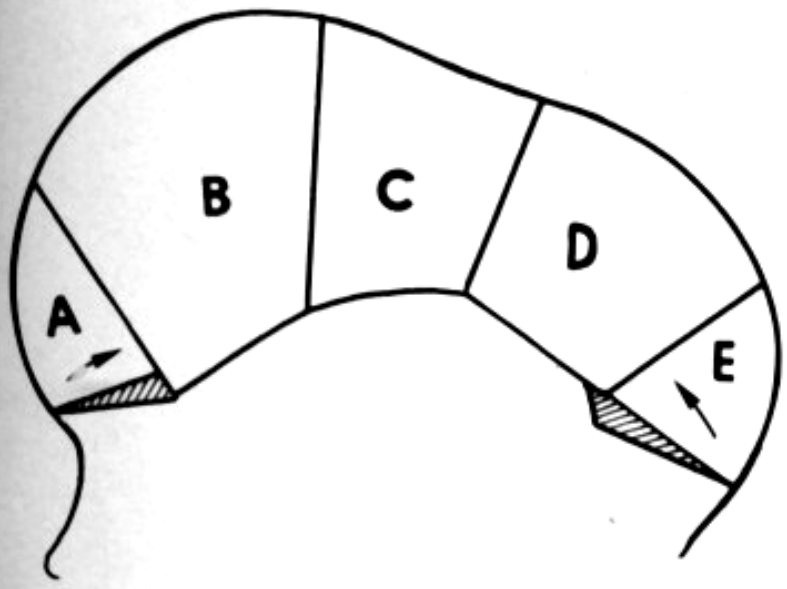
Forehead Advancement Floating

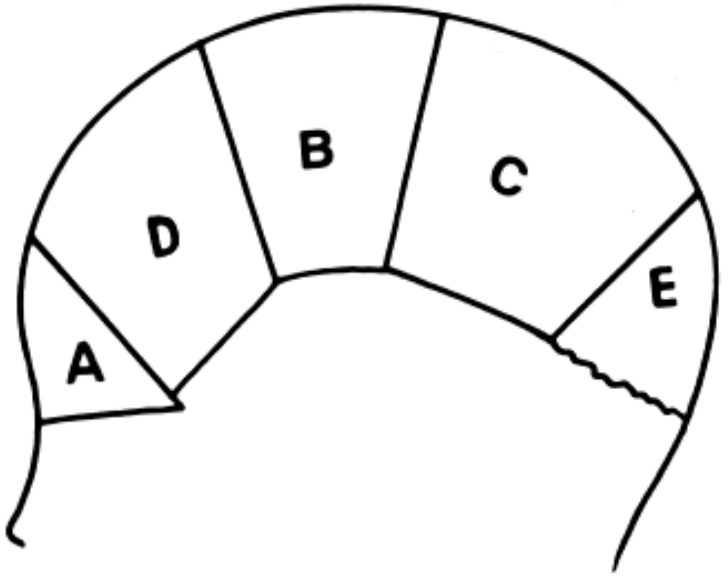


Forehead Advancement

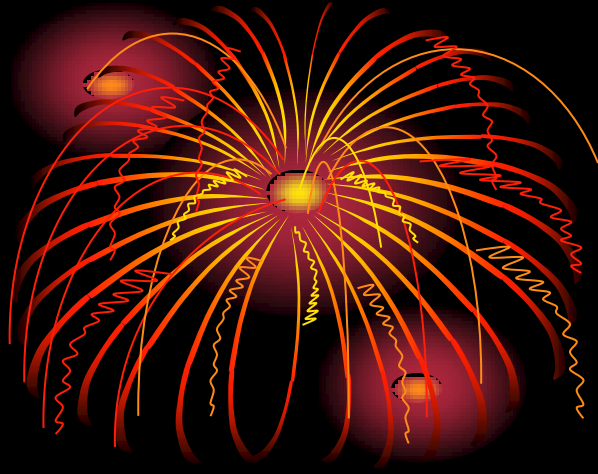





A
B
C
D
E


A
D
B
C
E

Complications



- Death 1.5-2%
- Hemorhagic complications. There is significantand continous blood loss in these operations. Replacment required
- Air emboli.

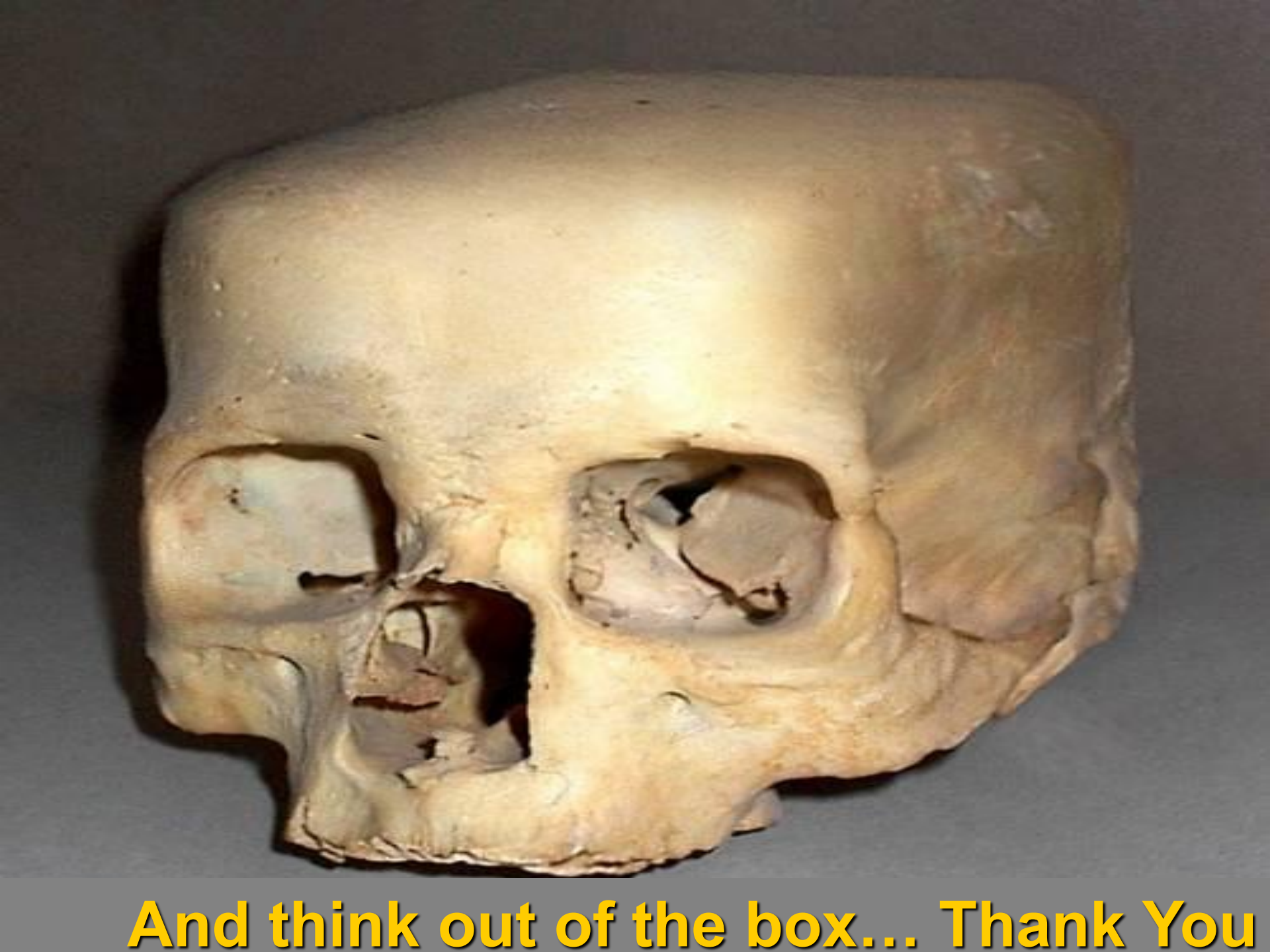
Complications

- Infection of soft tissues and bones more when there is a communication of bone with the respiratory system. Related to the length of surgery. Prophylactic antibiotics are given.
- Infection (meningitis)
- CSF leak
- Ophthalmic complications- blindness.
- Cerebral edema
- Respiratory obstruction
- Sagital sinus thrombosis
- Seizures

Thanks

When will you fix me ?


хуЯndex.net


And think out of the box… Thank You