Craniosynostosis

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Craniosynostosis

- Definition Characteristic skull deformities that occur as a result of premature fusion of various cranial vault sutures.
- Craniosynostosis are frequently associated with impaired central
 - nervous system functions due to raised intra-cranial pressure, hydrocephalus, and brain anomalies.
- Incedence-1 / 2000-2500 live birth.
- Males sagittal and metopic stenosis
- Females coronal

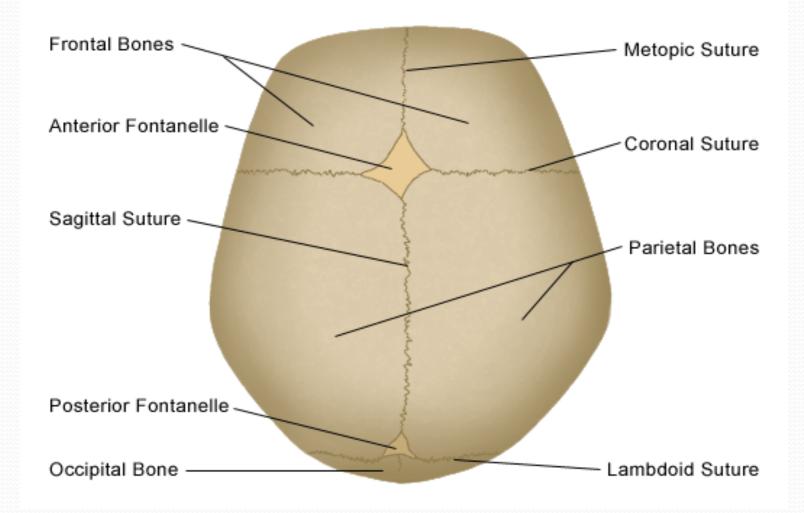
History of craniosynostosis

- Otto (1830) coined the term craniosynostosis.
- **Virchow**: Classify the different types of skull deformity.
- L.C. Lane:
 - 1st surgical procedure to release stenosed suture
- Lannelogue (1890) :
 - performed B/L strip craniectomies
- Paul Tessier father of modern craniofacial Surgery.
 - First to attempt major surgical procedures on the craniofacial skeleton.

Pathophysiology

- Normal Infant skull-flexible enough to get through vagina and expansile enough to accommodate rapidly growing brain.
- The calvarial sutures serve 2 important functions (a) maintenance of head malleability,(b) growth sites
- The cranial base was proposed as the primary locus of the abnormality in children with multi sutural craniosynostosis and may not be the primary anomaly in bicoronal or sagittal craniosynostosis.

Normal Skull of the Newborn



- Brain Growth
 At term nearly 40% of adult brain volume, and
 This increases to 80% by 3 years of age and continues to grow until the age of 12 yrs.
- The cranium
 At term 40% of adult size and
 by 7 years of age increases to 90%.

Principles and practice of pediatric neurosurgery. New York: Thieme Medical, 1999:219-42.

Timing of closer of sutures & Fontanelles

Sutures & Fontanelles	Timing of closer
Metopic Suture	9 months - 2 year
Coronal , Sagittal and Lamdoid suture	40 years
Anteior fonanelles	18-24 months
Posterior fontanelles	3 - 6 months
Mastoid fontanelles	1 year
Sphenoid fontaneles	2-3 months

Rodger E, Hall CM. Craniosynostosis. Clin Radiol 2002;57:94 Craniosynostosis

Etiology of Craniosynostosis

- Exact etiology is not known
- Sporadic in most instances
- Potential risk factors :-
 - white maternal race,
 - -advanced maternal age,
 - -male infant sex,
 - -maternal smoking,
 - -maternal residence at high altitude,
 - -use of amine containing drugs (e.g., nitrofurantoin, chlordiazepoxide, chlorpheniramine)
 - -certain paternal occupations (e.g., agriculture and forestry, mechanics)
 - -fertility treatments.

- Familial Nonsyndromic craniosynostosis,
 - Transmitted as an Autosomal Dominant disorder
 - Affect 2-6 % with sagittal synostosis & 8-14% with coronal synostosis
- Fibroblast growth factor and fibroblast growth factor receptor (FGFR): regulate fetal osteogenic growth and possibly influence fetal suture patency.
- Mutation in gene coding for

FGFR1 -Pfeiffer's syndrome

FGFR2 -Apert's syndrome and Crouzon's syndrome

TWIST - Saethre chotzen syndrome

Theories of Cranoisynostosis

- Sommering(1839) –noted that bone growth in skull primarily occurs at suture line and if it prematurely fused, an abnormal skull shape developed and skull growth restricted.
- Virchow(1821) and Otto(1830)- similar observation were made and they noted restriction of growth adjacent to suture and compensatory growth occurred at elsewhere in skull to accommodate growing brain.
- Jane JA: the major cause of the global cranial deformity was compensatory overgrowth at adjacent sutures.

Theories of Craniosynostosis

- Moss(1959) described functional matrix theory. According to this theory cranial base abnormality was the primary pathological process and cranial vault suture abnormality was secondary as cranial base mature embryologically before cranial vault.
- Persson (1979) cranial vault suture pathology may be primary in the development of synostosis leading to cranial base and facial deformity.
- Marsh and Vannier(1986) –following cranioplasty in patients with individual suture craniosynostosis, surgery altered only the cranial vault structure, the previously developed cranial base abnormalities were not ameliorated.

Classification of Craniosynostosis

Primary

- Simple(single suture) : Nonsyndromic: sagittal, coronal, metopic, lambdoid
- Compound (≥2 suture): -Nonsyndromic: bicoronal
 Syndromic: Crouzon's disease, Apert's syndrome,
 Pfeiffer's disease, Saethre-Chotzen syndrome

Secondary

- Metabolic disorders: e.g., hyperthyroidism (bone mature faster)
- Malformations : e.g. holoprosencephaly, shunted hydrocephalus, microcephaly, encephalocele.
- Exposure of fetus: e.g. valproic acid, phenytoin.
- Mucopolysaccharidosis : e.g., Hurler's syndrome, Morquio's syndrome.

Primary vs secondary craniosynostosis

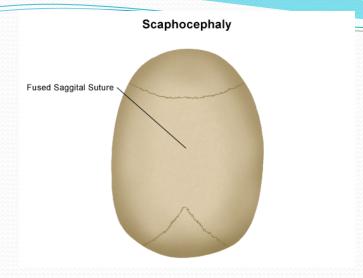
Primary

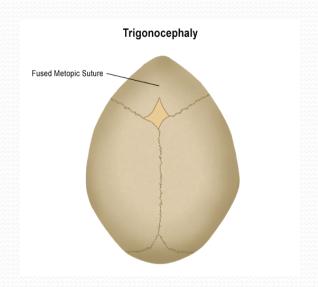
- Primary defect of ossification
- suture growth is altered
- head frequently asymmetric
- the brain continues to grow in areas where sutures are open
- most individuals are normal neurologically
- benefit from surgery

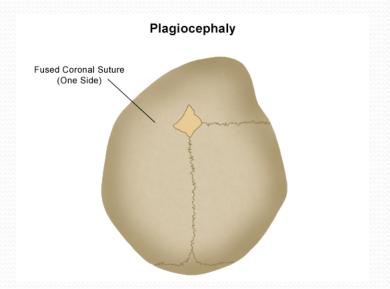
Secondary

- Secondary to brain malformation
- Head symetric
- growth of brain is impaired
- neurologically abnormal usually
- No benifit of surgery

TYPE OF CRANIOSYNOSTOSIS	CHARACTERISTIC HEAD SHAPE
Single-Suture Synostosis	
Sagittal	Scaphocephaly(Dolichocephaly)
Coronal	Anterior plagiocephaly,
Metopic	Trigonocephaly
Lambdoid	Posterior plagiocephaly
Double-Suture Synostosis	
Bicoronal	Anterior brachycephaly
Bilambdoid	Posterior brachycephaly
Saggittal plus metopic	Scaphocephaly
Complex Multisuture Synostosis	
Bicoronal, sagittal,metopic	Turribrachycephaly
Multisuture, pansynstosis	cloverleaf (Kleeblattschadel)





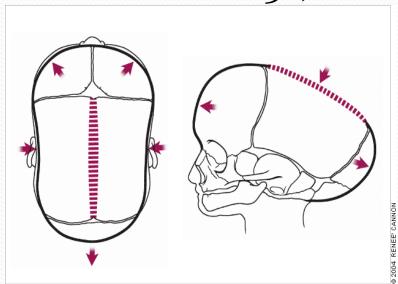


Types of craniosynostosis

(A) Sagittal craniosynostosis (Scaphocephaly, dolichocephaly)-

Most common – 40-55%, 1 in 5000 live birth

Male : Female = 3:1,



Skull shape characterized by biparietal narrowing, ridging of the sagittal suture, and bilateral bulging of the frontal and occipital region or both.

Sagittal craniosynostasis





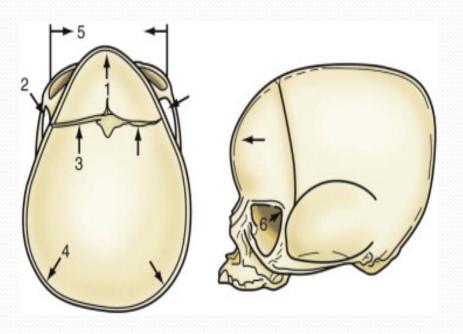
Clinical photograph-Lateral and Superior view of a child with sagittal craniosynostosis demonstrating frontal and occipital bossing.



Superior view of a 3-dimensional CT scan demonstrating a fused sagittal suture with frontal bossing and patent coronal sutures.

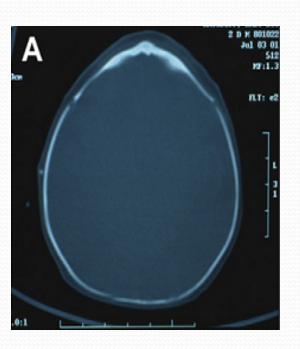
Metopic synostosis

- Also known as trigonocephaly,
- Incidence 4-10 %, M > F
- The distance between the medial canthi of the eyes is reduced.



metopic synostosis. 1, Ridging of metopic suture. 2, Temporal narrowing. 3, Patent coronal suture displaced anteriorly. 4, Compensatory bulging of the parieto-occipital region. 5, Narrowed bizygomatic dimension. 6, Posterior displacement of the superolateral orbital rim.

Metopic synostosis





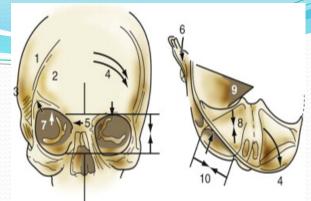


Coronal craniosynostosis

- Second most common type (20 -25 %)
- 1/10,000 live births
- female > male with ration of 2:1
- Two types :
 - i) Bilateral coronal cranosynostosis- Brachycephaly
 - ii) unilateral coronal cranosynostos- Anterior plegiocephaly

Unilateral coronal synostosis

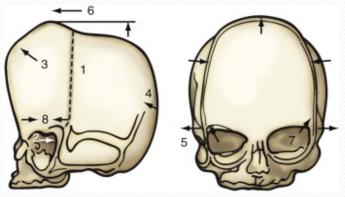
- •prematurely fused one coronal suture,
- flattening of the ipsilateral frontal and parietal bones,
- •bulging of the contralateral frontal and parietal bones
- •bulging of the ipsilateral squamous portion of the temporal bone,
- •ipsilateral ear displaced anteriorly compared with the contralateral ear.
- •radiographic findings include the "harlequin" orbit deformity (elevation of supra orbital margin)due to elevation of the greater and lesser wings of the sphenoid







Bilateral coronal synostosis







- 1, Fused bilateral coronal suture.
- 2, Recessed superior orbital rim.
- 3, Prominent frontal bone.
- 4, Flattening of occiput.
- 5, Anteriorly displaced skull vertex.
- 6, Shortened anterior cranial fossa.
- 7, Harlequin deformity of greater wing of sphenoid.
- 8, Protrusion of squamous portion of temporal bone.

Lambdoid synostosis

- Represents 2- 4% ,
- prevalence of 1 / 150,000 live births
- unilateral fusion Posterior plegiocephaly
- bilateral fusion Posterior brachycephaly

 Unilateral posterior plegiocephaly should be differentiated from positional plagiocephaly

Oxycephaly

Turricephaly (high head syndriome)

 Fused coronal and sagittal suture results in a an abnormally high conical head shape with bossing in

the region of the anterior fontanelle

Seen in syndromic craniosynostosis



Cloverleaf skull deformity,

- Pansynostosis/multiple suture synostosis
- head shaped like a cloverleaf (Kleeblattschadel)
- three bulges-two temporal and top
- pronounced constrictions in both sylvian fissures





Craniosynostosis syndrome

- 10-20 % of cases
- Autosomal Dominant
- Linked to Chromosome 10
- Multi-sutural, complex craniosynostosis
- If a suture is fused, check hands, feet, big toe and thumb

Craniosynostosis syndrome

Crouzon's syndrome

- one in 25,000 live births
- 5 % of cases of craniosynostosis
- Autosomal-dominant pattern. paternal age older than 35 years.
- Normal intellect
- Normal extremities
- 5 % have acanthosis nigricans
- 30 % have progressive hydrocephalus
- Mutation of gene coding FGFR2, FGFR3

Corde Mason A, Bentz ML, Losken W. Craniofacial syndromes. . Atlas of pediatric physical diagnosis. 4thed. St. Louis: Mosby, 2002:803-17

Clinical Feature

- Brachycephaly,
- significant hypertelorism, proptosis, maxillary hypoplasia, beaked nose
- Intracranial anomalies:

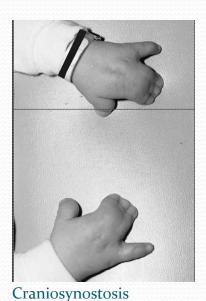
 Hydrocephalus, Chiari 1
 malformation, and
 hindbrain herniation (70
 percent).



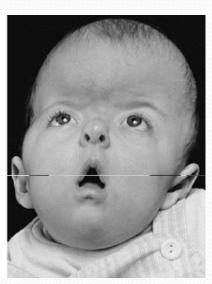


Apert's syndrome

- Crouzon's with Hand Involvement
- 1 in 100,000 to 160,000 live births, mutation FGFR2
- Varying intellect (50 % with MR)
- Cervical vertebral anomalies
- Rare HCP
- Syndactaly







10/10/2012

Pfeiffer's syndrome

- 1 in 2,00,000
- Clover leaf skull in 20%
- Intelligence is reported to be normal
- broad thumbs, broad great toes
- Mutation of gene coding for FGFR₁, FGFR₂







10/10/2012

Craniosynostosis

Carpenter's syndrome

- autosomal recessive.
- syndactyly of feet
- Sagittal and lambdoid suture closes first coronal last
- Cardiac abnormalities

Consequence of craniosynostosis

Intracranial hypertension

- 4-20%: single suture craniosynostosis
- 42-60 %: multiple suture/syndromic craniosynostosis
- Neurologic symptoms of elevated ICP

 (Headaches, vomiting, sleep disturbances, feeding difficulties, behavioral changes, and diminished cognitive function).
- If ICP: 10 15 mmhg were considered borderline,
 - > 15 mmhg abnormal
- Causes- abnormal venous drainage
 - respiratory obstruction
 - -chiari malformations

Consequence of craniosynostosis

Hydrocephalus

- 4% to 18%
- multiple-suture craniosynostosis >> nonsyndromic single suture craniosynostosis.
- Communicating
- causes
 - Cerebral maldovelopment
 - Brain atrophy
 - Abnormal csf circulation
 - Venous outflow obstruction
 - Hind brain herniation
 - Aqueductal stenosis

Consequence of craniosynostosis

Ophthalmologic Effects

- Papilloedema, optic nerve atrophy, and even loss of vision may occur with prolonged, untreated elevated ICP.
- Visual function and eye motility are often impaired.
- orbital dystopia secondary to unilateral coronal synostosis result in disturbances of extra-ocular muscle movement (ie, strabismus), upper eyelid ptosis, and poor binocular vision.
- Decreased orbital volume (in coronal craniosynostosis) causes proptosis, corneal exposure, and increased risk of direct ocular trauma.

Diagnosis

- (A)Detailed history
- (B)Clinical examination
- (C)Radiological evaluation

Diagnosis

(A) Detailed history

- Birth history, sleeping position.
- Head tilt, torticollis (deformational plagiocephaly)
- Delayed developmental mile stone
- family history
 abnormal head shape or multiple systemic
 problems (eg,cardiac, genitourinary, musculoskeletal)

Diagnosis

(B)Clinical Examination

- HC(micro/macrocephaly)
- Head shape (from above, side)
- Palpate suture lines & fontanelles (Look for ridging)
- Ear and facial symmetry, neck, spine, digits, and toes
- Look for associated anomalies (cvs,genitourinary,musculoskeletal)
- Fundus examination

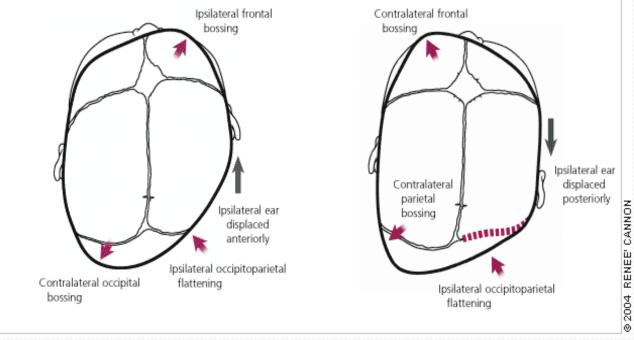
Diagnosis

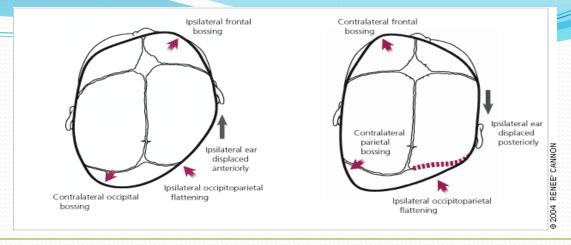
(C) Radiological Evaluation

- Plain radiography-AP and lateral
 views of the skull -bony bridging across the
 suture ,sclerosis, straightening and narrowing of the
 suture and loss of suture clarity
- CT scan Head -more accurate . structural abnormalities (e.g., hydrocephalus, agenesis of the corpus callosum).
 3D CT scanning accurately delineate the craniofacial deformity and plan surgical reconstruction.

Positional plagiocephaly, the diagnosis can be made based upon the history and physical examination. Positional plagiocephaly seen in : 1 /300







Anatomic features	Positional plegiocephaly	Unilambdoid Synostosis
Frontal bossing	Ipsilateral	Contralateral
Displacement of ipsilateral ear	Anterior	Posterior and inferior
Ridging of limbdoid sututre	Absent	Present
Head shape: vertex view	Parallelogram	Trapezium
Head shape: posterior view	Normal	Parallelogram
Status of lambdoid suture	No fusion or fibrous union	Bony union
Contralateral posterior bossing 10/10/2012 Craniosynostos	Occipital	Parietal

Treatment

- primary objectives in nonsyndromic craniosynostosis are release of the involved (fused) suture and reconstruction of all dysmorphic skeletal components
- Older surgical techniques: removal of sutures via a strip craniectomy. Allow only cerebral decompression but a dysmorphic skull does not reshape itself. residual bony deformity persist.
- Modern surgical management involves release of the involved sutures and reconstruction by removal, dismantling and reassembly of all dysmorphic skeletal components into a more appropriate anatomic position.

- most popular procedures
 - wide-strip craniectomy with bilateral wedge parietal craniectomy,
 - extended vertex craniectomy,
- complete calvarial remodeling :
 - The pi procedure for advanced sagittal synostosis
 - orbitofrontal advancement for metopic, unicoronal, or bicoronal synostosis

Surgery indication

- In single suture- cosmetic / esthetics
- Optimizing brain growth
- Early treatment of intracranial hypertension
 (improvement in IQ, behavior and resolution of papilledema after cranial vault reconstruction with proven elevated ICP)
- When Severe proptosis and impending corneal damage

Timing of surgery

Early operation(3-6 months)

- Better compliance of brain ,dura and scalp
- Calvarium is much more malleable, easier to shape and providing a better outcome
- Rapid brain growth reshape the bone

YN Anantheswar, NK Venkataramana, Pediatric craniofacial surgery for craniosynostosis. Journal of pediatric neurosciences, 2009, 4; 86-99

• Prefers operating within 3-6 months time frame to take advantage of the ability of the rapidly expanding brain and skull to grow more normally and so that the skull can be remodeled more readily.

John A. Persing, M.D.New Haven, Conn .Management Considerations in the Treatment of Craniosynostosis. Plastic and Reconstructive Surgery • April 2008

- Early frontocranial remodelling is performed between 2 and 4 months for brachycephalies
- other operated on between 6 and 12 months of age.
- For syndromal craniofacial synostosis, two-step operation:

forehead advancement first, facial advancement later, to avoid the risk of frontal osteitis

Marchac D, Renier D. Timing of treatment for craniosynostosis and faciocraniosynostosis: a 20-year experience, Br J Plast Surg. 1994 Jun;47(4):211-22

Positioning

Supine on a padded horseshoe head rest for correction of metopic and unicoronal craniosynostoses



Prone on a horseshoe head rest allowing access to the posterior half of the skull.



Modified prone position with chin support in a padded "bean bag" to allow simultaneous access to the anterior and posterior skull. sagittal and bicoronal craniosynostoses



Incision

- Zigzag bicoronal incision(Stealth incision)
 - to minimize incisional scap alopecia
 - scar tends to spread less redistribution of the forces.
- Incision begins slightly anterior and superior to the helix of the ear.

Electrocautery is used cautiously.

Sagittal synostosis

- Goals of surgery:
 - i) eliminate the biparietal constriction,
 - ii) increase the width of the skull,
 - iii) decrease the anterior-posterior (AP) length
- mild scaphocephaly: strip or extended craniectomies to remove the sagittal suture using either open or endoscopic techniques, and then relying on future cranial vault remodeling with brain growth.
- marked scaphocephaly: extending the craniectomy along the lambdoid sutures and bilateral parietal bone or total cranial vault remodeling

Scaphocephaly.

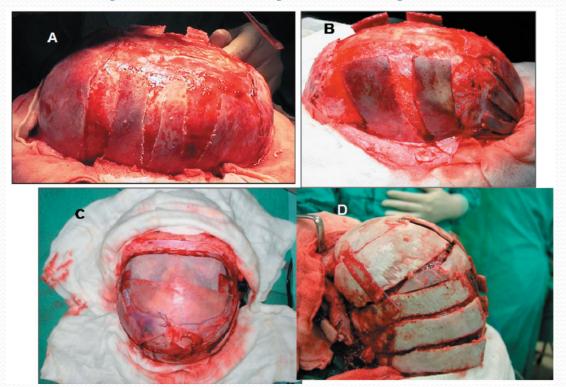
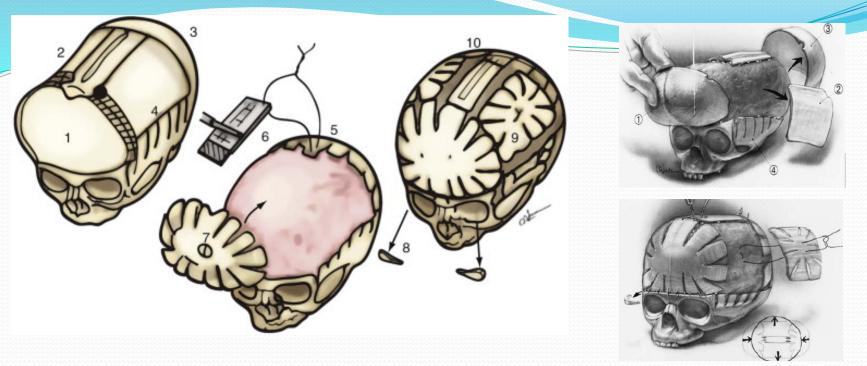


Fig A: Suturectomy and expanding osteotomies. Standard technique.

Fig B and C: Scaphocephaly. Suturectomy and expanding osteotomies including frontal bone remodelling.

Fig D: total cranial vault remodel-ling (holocranial dismantling).



Sagittal synotosis, operative approach. 1, Bifrontal craniotomy. 2, Separate parietal craniotomies bilaterally. 3, Biparietal -occipital craniotomies. 4, Lateral barrel stave osteotomies. 5, Recontour of occiput and replacement of bone. 6, Reduction in skull length by attachment of shortened midline bone. 7, Contour of projecting sagittal ridge, with reduction in length. 8, Bifrontal bone graft has undergone radial osteotomy. 9, Removal of infralateral frontal bone to allow posterior tilting of frontal bone on visor. 10, Parietal bone grafts attached to underlying dura.

sagittal synostosis

- In older children, the bone is more difficult to reshape, and a more extensive calvarial reconstructive procedure is performed with bifrontal, biparietal, and occipital bone removal and reconstruction.
- With the advent of endoscopes in neurosurgery, extended-strip craniectomy is performed and the patient is placed in a custom-made molding helmet to correct the frontal bossing and bathrocephaly.

Pi procedure

- For isolated sagittal synostosis—Jane and colleagues developed the pi procedure in 1978.
- In this technique, the sagittal, bilateral coronal, and lambdoid sutures are first removed and the parietal bones are out fractured to increase the skull width.
- The sagittal suture used as a strut to maintain the outward position of the parietal bones. Finally, the frontal and occipital bones are secured to the parietal bones with adjustments of anterior-posterior dimension and frontal bossing.
- Advantages -addressing the primary suture fusion and also the global cranial deformity and providing immediate correction without the need for a postoperative helmet

Pi procedure

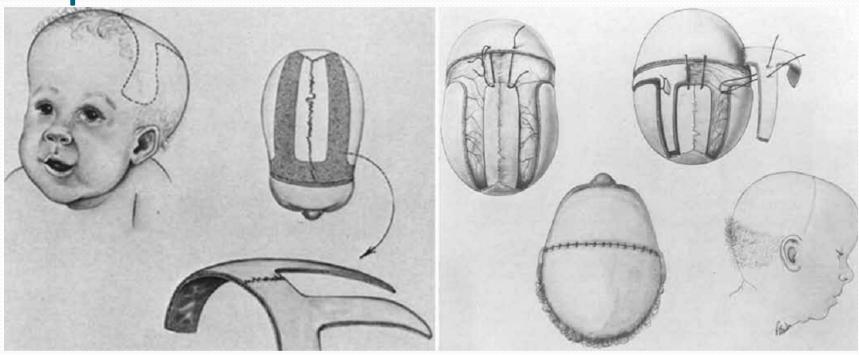
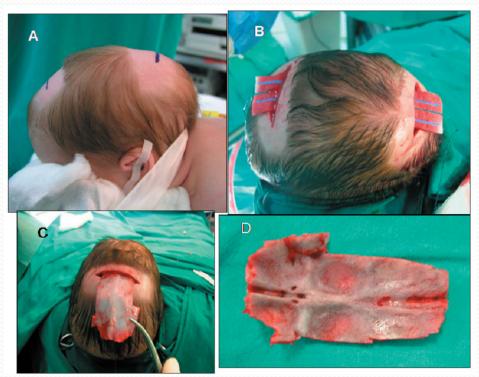


Fig. Pi procedure. bilateral parasagittal strip craniectomies with outfracturing of the temporal bones, removal of the vertex bone and AP shortening of the calvarial vault with bifrontal osteotomies.

From Jane JA et al: J Neurosurg 49:705–710, 1978.

Modern Endoscopic Strip Craniectomy: Scaphocephaly.



Endoscopic-assisted suturectomy and osteotomies.

A: patient resting in modified prone position.

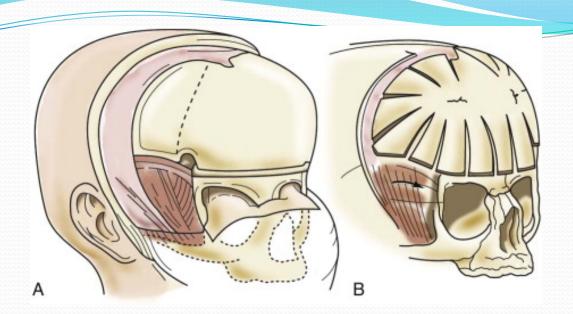
B: surgical patties showing subcutaneous dissection.

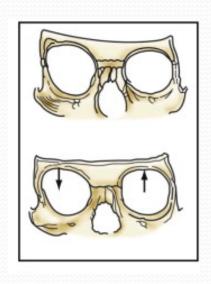
C: sagittal suture excision complete.

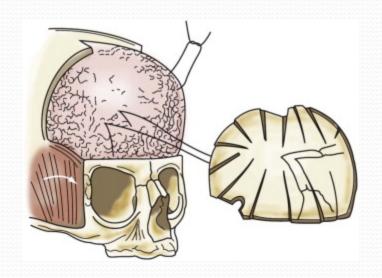
D: excised sagittal suture showing characteristic grooving of the superior sagittal sinus

Coronal Synostosis (Unilateral)

- goal of surgery: to achieve forehead and orbital symmetry.
- If contralateral compensatory deformity, a bifrontal craniotomy with a bilateral or extended unilateral orbital roof osteotomy, with lateral cuts at the frono-zygomatic sutures.
- The orbital rims are contoured, advanced and secured.
- The frontal bone reconstructed to reduce the projection of the bulging side and increase the projection on the flattened side, and then attached to the advanced orbital rim.
- In older children, the frontal bone reconstruction requires a series of osteotomies to achieve appropriate contouring, and bilateral orbital advancement, sometimes extending into the body of the zygoma

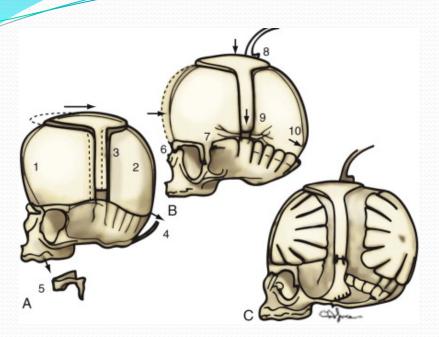


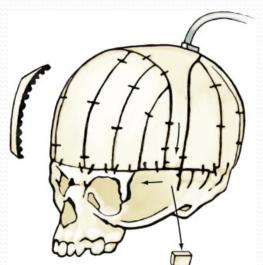




Bilateral coronal synostosis

- Challenge to treat, owing to the difficulty of reducing the height of the cranial vault.
- In younger children, bifrontal and bioccipital craniotomie, leaving behind a strut of bone from the vertex to the temporal bone.
- The skull's height is reduced by dividing and shortening the strut and fixing it to the temporal bone bilaterally.
- Bilateral orbital advancement is performed, and both the frontal and occipital bones are reshaped with osteotomies. The bones are then replaced, with the frontal bone attached to the newly advanced orbit.



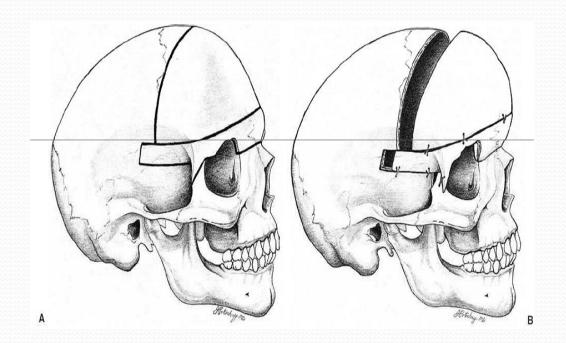


bilateral coronal synostosis technique. **A**, Osteotomies: bifrontal craniotomy (1); biparietal occipital craniotomy (2); struts severed inferiorly, with bone removed (3); barrel stave osteotomies in occiput (4); bilateral supraorbital rim elevation (5).

B, Height reduction: bilateral supraorbital rim advancement and fixation at nasion (6); composite myo-osseous temporalis flap advanced and fixed to rim (7); intracranial pressure monitoring as height of skull is reduced (8); height reduction achieved by slowly cinching wire struts (9); greater prominence achieved in parietal occiput(10).

C, Completed remodeling of skull.

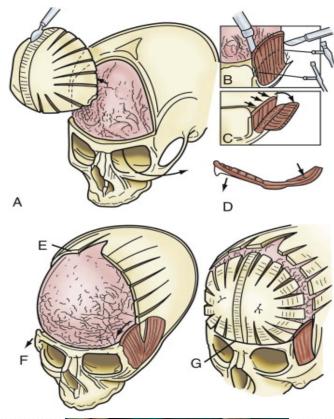
Fronto - orbital advancements

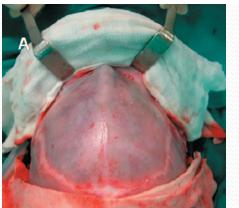


- older children, height reduction is carried out more slowly.
- Barrel stave osteotomies of-frontal and occipital bones
- bilateral orbital and in some cases occipital advancement
- Some surgeons prefer to perform staged anterior and posterior procedures, rather than a total calvarial reconstruction

Metopic Synostosis

- Goal: to eliminate the frontal keel, and to advance and widen the forehead.
- In mild cases, a frontal keel present without significant temporal narrowing, and this can be treated by burring down the keel only.
- bifrontal craniotomy is performed, followed by bilateral orbital osteotomies with remodeling of the supraorbital rim. The frontal bone is remodeled to eliminate the keel, and to widen it bilaterally.
- Lastly, the bilateral squamous temporal bones are out fractured.
- In older children, the frontal bone remodeled by radial osteotomies or splitting into vertical slats.





A,Bifrontal craniotomy is performed with burring of the central ridge, peripheral radial osteotomy, and bilateral removal of the visor.

B, A composite flap consisting of vascularized temporalis muscle and squamous temporal bone is developed

C, The composite flap is reflected laterally, and squamous temporal bone is split vertically into "barrel staves" and out- or infractured (*arrows*).

D, The supraorbital rim is recontoured with internal kerfs, and the rims are bent to achieve a rounded contour.

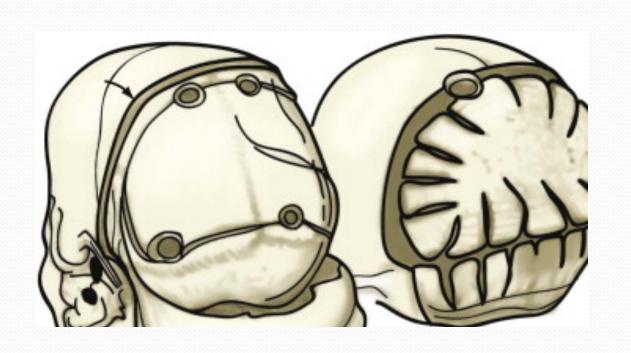
E, Anterior parietal bone undergoes parallel-oriented osteotomies to expand the parietal region laterally.

F, The visor is reattached at the nasion, and the composite myo-osseous flap is advanced forward (*arrows*) to the orbital rims.

G, Reshaped bifrontal bone is attached to the supraorbital rim and underlying dura.

Lambdoid suture Synostosis

- the goal : to restore the natural contour to the occipital bone.
- An ipsilateral perisutural osteotomy to address the primary sutural abnormality,
- compensatory contralateral deformity addressed by a bi parietooccipital craniotomy to achieve adequate cosmetic correction.
- the occiput is outfractured on the flattened side and infractured on the bulging side.
- supplemented with the use of barrel stave osteotomies.



Unilateral lambdoid technique.

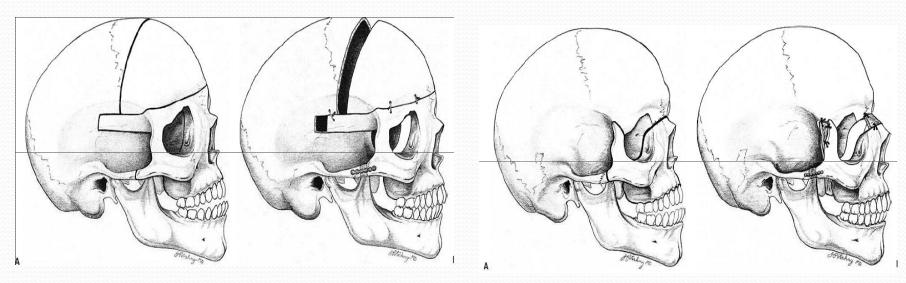
A, Biparieto-occipital bone flaps.

B, Biparietal asymmetry recontoured.

Syndromic craniosynostosis

current surgical treatment approach

- Initial fronto-orbital and cranial vault remodeling,
- midface advancement procedure with or without distraction (Le Fort III or monobloc)



Conservative Therapy for Deformational Plagiocephaly

- •Educate parent to keep head off the flat surface
- Helmet Molding
- Re-positioning if no improvement by 6 months.
- •Rarely surgery may be indicaated



complications

Uncorrected craniosynostosis

increased intracranial pressure, asymmetry of the face, and malocclusion. Asymmetry of the orbits leads to strabismus.

Complication of surgery

- Acute –blood loss, air embolism, dural tear & csf leak, meningitis, acute air way obstruction.
- Late –cranial defect and growing skull fracture,plate migration, relapse and restenosis

Long Term Follow-Up

- Speech
- Genetic Counseling
- Feeding / Swallowing
- Ophthalmology evaluation

