- Pathological condition that results from premature fusion of one or more sutures in the cranial vault;

- Associated with a deformity of the vault and cranial base.
Development

Bones of the cranium
- the skull base and the calvarial vault
- growth of skull bones
  expanding growth of the brain.
Brain Growth

- At term has nearly 40 percent of his or her adult brain volume,
- and this increases to 80 percent by three years of age.
- Continues to grow until the age of 12 yrs
The cranium

- at term is 40 percent of adult size;
- by seven years, this increases to 90 percent.

Mature suture closure occurs by 12 years of age, but completion continues into the third decade of life and beyond.

<table>
<thead>
<tr>
<th>Type of suture/fontanelle</th>
<th>Time to closure</th>
</tr>
</thead>
<tbody>
<tr>
<td>Metopic suture</td>
<td>Nine months to two years (may persist into adulthood)</td>
</tr>
<tr>
<td>Coronal, sagittal, lambdoid sutures</td>
<td>40 years</td>
</tr>
<tr>
<td>Anterior fontanelle</td>
<td>Nine to 18 months</td>
</tr>
<tr>
<td>Posterior fontanelle</td>
<td>Three to six months</td>
</tr>
<tr>
<td>Anterolateral fontanelle</td>
<td>Three months</td>
</tr>
<tr>
<td>Posterolateral fontanelle</td>
<td>Two years</td>
</tr>
</tbody>
</table>

Adapted with permission from Aviv RI, Rodger E, Hall CM. Craniosynostosis. Clin Radiol 2002;57:94.
HISTORY AND PATHOGENESIS

- **Otto** (1830) coined the term craniosynostosis.

- **Stahl** and **Hyrtl** noticed that premature closure of the cranial vault sutures leads to an abnormal skull shape.

- In **1851**, **Virchow** described how skull growth is restricted to a plane perpendicular to the affected, prematurely fused suture and is enhanced in a plane parallel to it.
Suture Growth

- Sutures allow growth perpendicular to them
  - Growth at suture lines related to brain growth
Early Closure Causes Growth Parallel to the Suture
• Virchow - classify the different types of skull deformity
• Van der Klaauw, in 1946, and Moss, in 1959,

cranial base source of abnormal physical stress leading to dural abnormalities that yielded premature sutural fusion
animal studies

the cranial vault abnormalities typical of synostosis can be produced with experimental fusion of developing cranial vault sutures.

Marsh and Vannier – following cranioplasty in patients with individual suture craniosynostosis in which surgery altered only the cranial vault structure, previously developed cranial base abnormalities were ameliorated.

- **L.C Lane** first surgical procedure to release stenosed suture
- **Lannelogue-1890** performed b/l strip craniectomies
- **Tessier** - father of modern craniofacial Surgery.

First to attempt major surgical procedures on the craniofacial skeleton.
Incidence

- one per 1,800 to 2,200 live births
- Males - sagittal and metopic stenosis
- Females - coronal

Familial Non syndromic Craniosynostosis

- Affects 2 to 6 percent with sagittal synostosis
- 8 to 14 percent of infants with coronal synostosis,
- Autosomal dominant disorder.

Syndromic craniosynostosis

- is less common (20 percent)
- more than 150 syndromes with craniosynostosis have been identified.
- multiple sutures are involved.
- Autosomal dominant

Etiology

- Unknown
- sporadic in most instances
Risk factors

- white maternal race
- advanced maternal age
- male infant sex
- maternal smoking
- residence at high altitude
- nitrosatable drugs (e.g., nitrofurantoin, chlordiazepoxide, chlorpheniramine),
- Certain paternal occupations (e.g., agriculture and forestry, mechanics, repairmen)
- fertility treatments.

Pathophysicsology

- Cranial sutures - fibrous joints
- Abnormal osteoblastic activity – observed in cultures of synostotic bone
- Decreased growth rate
- Decreased alkaline phosphatase production
- Increased levels of osteocalcin
  platelet-derived growth factor
  epidermal growth factor.
Fibroblast growth factor and fibroblast growth factor receptor (FGFR) regulate fetal osteogenic growth expressed in cranial sutures in early fetal life.

Mutations in the gene coding for
- FGFR1 → Pfeiffer’s disease,
- FGFR2 → Apert’s syndrome and Crouzon’s disease.
| TABLE 2 |
| Classification of Craniosynostosis |

**Primary**

**Simple**
- Nonsyndromic: sagittal, coronal, metopic, lambdoid

**Compound**
- Nonsyndromic: biconoral
- Syndromic: Crouzon’s disease, Apert’s syndrome, Pfeiffer’s disease, Saethre-Chotzen syndrome

**Secondary**

**Metabolic disorders (e.g., hyperthyroidism)**

**Malformations (e.g., holoprosencephaly, microcephaly, shunted hydrocephalus, encephalocele)**

**Exposure of fetus (e.g., valproic acid, phenytoin)**

**Mucopolysaccharidosis (e.g., Hurler’s syndrome, Morquio’s syndrome)**

Types

- Scaphocephaly (Sagittal synostosis)

  derived from the Greek words *scaphos*, meaning boat, and *kephali*, meaning head.

  Most common 45-50%
FIGURE 5. (Left) Sagittal synostosis (superior view) with a ridged, fused sagittal suture, bitemporal narrowing, and (right) frontal and occipital bossing.
Superior view of a child with sagittal craniosynostosis demonstrating frontal bossing bilaterally
Lateral 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.
Plagiocephaly

- Greek word plagios, meaning oblique or sloping, and corresponds to unilateral coronal synostosis.
- 10–20%

- posterior plagiocephaly corresponds to lambdoid synostosis.
- 1.3%
Left coronal synostosis
asymmetry of the orbits
widened palpebral fissure on the left
superiorly displaced left eyebrow.
Right coronal craniosynostosis
Lamboid Synostosis

“Slant-Head” (Occipital Plagiocephaly)
left posterior plagiocephaly.
Trigonocephaly

- derived from the Greek word *trigonos*, meaning *triangular*

- *metopic synostosis.*

- 5 → 10% incidence
hypotelorism
Brachycephaly

- Greek word *brachys*, meaning *short*.
- Both coronal sutures
- 10–20%
FIGURE 4. Bilateral coronal synostosis results in a prominent frontal bone, flattened occiput, and anterior displacement of the skull vertex.
Child with bicoronal synostosis with exorbitism and recessed frontoorbital bar.
exorbitism and recessed frontoorbital bar
Oxycephaly

- oxys, meaning sharp, and is a high, conical head with sharp bossing in the region of the anterior fontanelle.

- coronal and sagittal sutures
  results in an abnormally high conical head shape

- encountered in syndromic types.
Cloverleaf skull deformity, (Triphylocecephaly)

- (derived from the Greek word *triphyllos*, meaning *trefoil, with 3 leaves*),
- multiple suture synostosis

  head shaped like a cloverleaf

  three bulges-two temporal and top

  pronounced constrictions in both sylvian fissures
Frequency

sagittal $\rightarrow 45\%-50\%$

unilateral coronal $\rightarrow 15\%$

metopic synostosis $\rightarrow 5\%$

lambdoid $\rightarrow 1.3\%$
CRANIOSYNOSTOSIS SYNDROMES

- 10-20% of cases

- Autosomal Dominant
  - Linked to Chromosome 10
  - Multi-sutural, complex case

- If a suture is fused, check hands, feet, big toe and thumb
Crouzon’s

- Autosomal-dominant pattern.
- one of every 25,000 live births
- 5 percent of cases of craniosynostosis.

Clinical findings

- Brachycephaly,
- significant hypertelorism, proptosis, maxillary hypoplasia, beaked nose
- Intracranial anomalies
  - Hydrocephalus, Chiari 1 malformation, and hindbrain herniation (70 percent).
• Normal intellect
• Normal extremities
• 5% have acanthosis nigricans
• 30% have progressive hydrocephalus
Apert’s

“Crouzon’s with Hand Involvement”

• 1 in 55,000
• Varying intellect (50 % with MR)
• **Syndactyly**
• Cervical vertebral anomalies
• Rare hydrocephalus
Apert Extremity Findings
PFEIFFER SYNDROME

- 1 in 2 lakhs
- Clover leaf skull in 20%
- Broad thumbs, broad great toes
- Intelligence is reported to be normal
PFEIFFER SYNDROME
CARPENTER SYNDROME

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

- Clinical history
- physical examination
- radiographic studies
- Passage of the head through the birth canal deforms the head. This shape is retained for 2-3 weeks postnatally.

- **Early diagnosis is important**
  
  the brain grows rapidly during this period delay only worsens the deformity of the head shape.
Detailed history

- birth, sleeping position
- Head tilt, torticollis
dehismental plagiocephaly
- family history
  abnormal head shape or multiple systemic problems (eg, cardiac, genitourinary, musculoskeletal)
Clinical Exam

- Head shape (from above, side)
- Ear and facial symmetry
- Palpate suture lines & fontanelles
- Look for ridging
- Look for associated anomalies
- If a suture is fused, check hands, feet, big toe and thumb
FUNCTIONAL CONSEQUENCES

**Intra cranial hypertension**

- Complicates one third of cases
- Principal indication of surgery
- Icp monitoring
- Syndromic forms → 30%
- 17% single suture
- **Causes** abnormal venous drainage
  respiratory obstruction
  chiari malformations
Hydrocephalus

- 4% to 18%
- Communicating
- ?causes
- Cerebral maldvelopment
- Brain atrophy
- Abnormal csf circulation
- Venous outflow obstruction
- Hind brain herniation
- Aqueductal stenosis
Respiratory Abnormalities

- Syndromic craniosynostosis
- Manifest during sleep
- Maxillary hypoplasia, choanal stenosis, tonsillar hypertrophy
- Nasal stents, tonsillectomy or tracheostomy
- Nocturnal CPAP
- Surgical correction of midfacial hypoplasia
Feeding

- Abnormalities of palatal shape and movement
- Disordered dentition
- Dental malocclusion
- Nasogastric tube or gastrostomy
Vision

- Chronically raised icp----papilloedema----optic atrophy(crouzon syndrome)
- Shallow orbits ---------exposure
- Primary optic atrophy :compression,traction
- Early craniectomy
Plain Films

- simple and inexpensive,
- Absent or line of increased density
- Harlequin appearance → coronal
- Cannot differentiate lambdoid synostosis and deformational plagiocephaly (plagiocephaly without synostosis).

- To visualize all the sutures, special Waters views must be taken.
Ultrasound

- Noninvasive
  more effective than plain skull radiographs in detecting fused sutures
- accuracy depends on a reliable and experienced operator.
CT Scan

- standard for the complete visualization of the skull and cranial sutures.
- detailed anatomy of the calvaria and the brain parenchyma
- document effect of corrective surgery
MRI

- Complex craniosynostosis
- Improved definition of intracranial soft tissue structures
- Hindbrain herniation
- Identify sites of respiratory obstruction
Radio isotope scanning

- Diminished uptake → complete fusion
ICP monitoring

- Clinically occult – majority
- Radiological signs inconclusive
- Deciding nature and timing of surgery
- Features of ↑ ict
  - Mean pressure > 15 mm Hg
  - Raised base line value
  - Prolonged plateau wave
Management

- Surgery vs. Conservative Management
Goal

- Normalization of deviated appearance, growth and function of skull
- Keep the suture open till brain growth is complete
- Rarely achieved
Indications

- Correction of cosmetic abnormality
- Early treatment of intracranial hypertension
- Optimizing brain growth
- Severe proptosis and impending corneal damage
Timing of surgery

Early operation (3-6 months)

- Rapid brain growth reshape bone
- Better compliance of brain dura and scalp
- Calvarium in an infant aged 3-9 months is much more malleable, easier to shape and providing a better outcome.
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.

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

- Surgical intervention should be performed during infancy, preferably in the **first 6 months** of postnatal life, to prevent the further progression of the deformity and possible complications associated with increased intracranial pressure.

*Plast Reconstr Surg. 2004 May;114(6):2032-48; quiz 2049. Management of craniosynostosis. Panchal J, Uttchin V. Oklahoma University Health Science Center, Oklahoma*
• 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.


Late intervention

- closer the cranium is to the adult size, the less overcorrection for reconstruction and the better the ultimate skull shape.
- Higher risk of recurrent deformity
- Surgical correction more complex
Positioning

Supine on a padded horseshoe head rest.

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.
Basic mechanisms

- Passive reshaping: generous removal of bone
- Strip craniectomy
- Morcellation

- Active reshaping:
  - Fronto orbital advancement
  - Cranial vault reshaping
Incision

- zigzag bicoronal incision
- prevents parting of the hair along a straight line
- 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 craniosynostosis

- objectives
  correction of scaphocephaly
  frontal bossing and occipital protrusion
- Initial surgical procedures included a narrow-strip craniectomy→higher restenosis.
- wider and more extensive craniectomy
  do not address the frontal bossing and occipital bathrocephaly relied on the growing brain to correct these deformities.
• more extensive cranial vault remodeling with barrel-stave osteotomy

• Spring assisted cranioplasty

• 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.

• rapid recovery of the child and diminished need for blood transfusion,
FIGURE 24.8. Operative technique in sagittal synostosis. Reduction in anteroposterior skull length is achieved frontally by removing a segment of midline bone. Triangular wedges of frontal bone are removed just cephalad to the supraorbital margins to allow for posterior inclination of the forehead. The frontal bone laterally is remodeled to create a neocoronal suture. As the wires are cinched down frontally, bulging occurs in the parietal region, for which the parietal bone is reshaped to add increased contour laterally. (From Persing JA, Jane JA, Edgerton MT. Surgical treatment of craniosynostosis. In: Persing JA, Edgerton MT, Jane JA, eds. Scientific Foundations and Surgical Treatment of Craniosynostosis. Baltimore: Williams & Wilkins; 1989:190, with permission.)
Bilateral coronal stenosis

- extended bicoronal craniectomies with reconstruction of the forehead.
- The supraorbital bar or brow is reshaped and advanced forward with the forehead.
- The reconstructed forehead and brow are rigidly fixed to the nose and lateral orbits with microplates.
Metopic stenosis

- Objectives: increase in width of the bifrontal diameter, an increase in volume of the anterior cranial fossa, and normalization of frontal bone shape.

- The frontal bones and the frontoorbital bones are excised and transferred to the side assembly. The frontoorbital bar is advanced to create an appropriate brow position.

- The interdacrion distance is increased by placing a bone graft between the 2 halves of the frontoorbital bars.
FIGURE 24.17. Barrel staves are performed in the occipital bone to increase projection ipsilateral to the fused suture. A retractor is used to protect the transverse and sagittal sinuses during the osteotomy (insert). (From Persing JA, Jane JA, Edgerton MT. Surgical treatment of craniosynostosis. In: Persing JA, Edgerton MT, Jane JA, eds. Scientific Foundations and Surgical Treatment of Craniosynostosis. Baltimore: Williams & Wilkins; 1989:201, with permission.)
Syndromic craniosynostosis

current surgical treatment approach

- initial fronto-orbital and cranial vault remodeling,
- a midface advancement procedure with or without distraction (Le Fort III or monobloc)
- secondary orthognathic surgery to correct any dentofacial deformities (Le Fort I, mandibular osteotomies)
### SYNDROMIC CRANIOSYNOSTOSIS TREATMENT OPTIONS

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Timing (age)</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Craniectomy, fronto-orbital advancement</td>
<td>4–12 mo</td>
<td>Repeat procedure may be indicated in childhood or adolescence for continued growth restriction or abnormal growth of the skull.</td>
</tr>
<tr>
<td>LeFort III osteotomy and advancement—conventional or by distraction osteogenesis</td>
<td>4–8 y</td>
<td>If performed in this age group, a secondary LeFort III may be required in teenage years.</td>
</tr>
<tr>
<td>LeFort III osteotomy and advancement—conventional or by distraction osteogenesis</td>
<td>9–12 y</td>
<td>Delaying to this age in less-severe cases may obviate the need for a second major midface advancement.</td>
</tr>
<tr>
<td>LeFort I ± mandibular osteotomy</td>
<td>14–18 y</td>
<td>Required to establish neutral dental occlusion after facial growth has ceased.</td>
</tr>
<tr>
<td>Monobloc frontofacial advancement—conventional or by distraction</td>
<td>4–12 y</td>
<td>Simultaneously improves forehead, orbital, and midface aesthetics. Suitable for a patient whose deformity allows simultaneous advancement.</td>
</tr>
<tr>
<td>Contouring via reduction, onlay bone grafts, bone substitutes, or alloplasts</td>
<td>15–19 y</td>
<td>Performed as the final procedure to enhance aesthetics after all growth has ceased.</td>
</tr>
</tbody>
</table>
Conservative Therapy for Deformational Plagiocephaly

- Re-positioning
- If no improvement by 6 months….
  - Helmet Molding
Long Term Follow-Up

- Speech
- Genetic Counseling
- Feeding / Swallowing
- Ophtho
Thank you