



### Craniosynostosis:

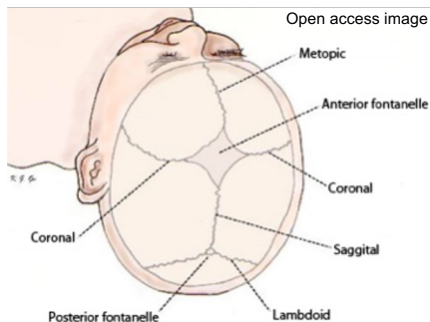
premature fusion of 1+ cranial sutures; this is *always abnormal*

### Functional sequelae:

craniofacial deformity, restricted brain growth, elevated ICP, ophthalmic anomaly, Chiari malformation

## PATHOPHYSIOLOGY

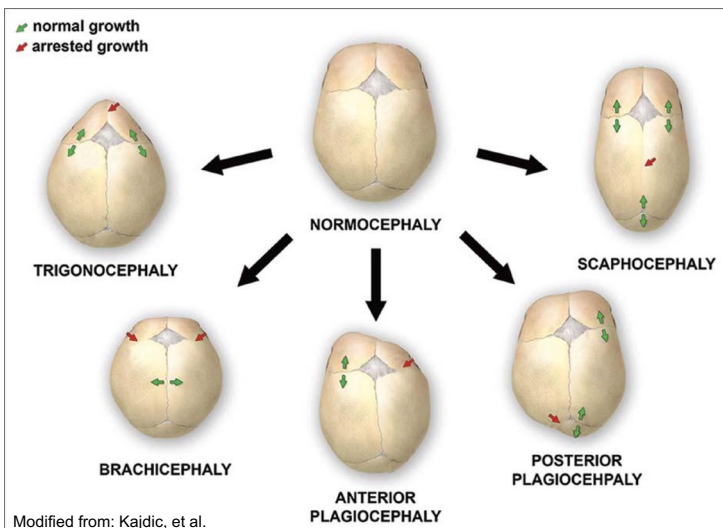
- **Cranium growth:** occurs through *sutural growth* (perpendicular to cranial sutures) and *appositional growth* (resorption of bone from the inner cranial surface & deposition on the outer surface)
- **Virchow's Law:** premature suture fusion results in cranial growth predominantly parallel to fused sutures
- **Causes:** intrauterine compression, syndromic, endocrinopathy, cranial base anomaly, abnormal dural attachment, medications



The resulting craniosynostosis is dependent on suture(s) involved

## PRESENTATION

HISTORY	PHYSICAL EXAM
<p><b>Onset &amp; evolution</b></p> <ul style="list-style-type: none"> <li>• At birth or early acquired</li> </ul> <p><b>Positional preference</b></p> <p><b>Torticollis</b></p> <p><b>Signs of elevated ICP</b></p> <ul style="list-style-type: none"> <li>• Lethargy, irritability, poor feeding, nausea, vomiting</li> <li>• visual changes, altered mentation, discoordination</li> </ul>	<p><b>Inspection</b></p> <ul style="list-style-type: none"> <li>• Abnormal cranial morphology or facies</li> <li>• Syndromic features</li> </ul> <p><b>Palpation</b></p> <ul style="list-style-type: none"> <li>• Ridge or lack of movement at suture, bulging fontanelles</li> </ul> <p><b>Head Circumference</b></p>



## DIFFERENTIAL DIAGNOSIS

(abnormal head shapes not caused by suture fusion)

### Deformational plagiocephaly:

parallelogram skull shape with anterior displacement of ipsilateral occiput, ear, forehead

**Deformational brachycephaly:** shortened AP distance and flattened occiput with normal forehead and orbits

**Dolichocephaly:** elongated AP distance without biparietal narrowing or frontal/occipital bossing; common in NICU

- *Distinguish by positional preference, movement at suture, lack of palpable sutural ridge, no functional sequelae, imaging*

## MANAGEMENT

**Consult:** to pediatric craniofacial surgeon. If signs of elevated ICP, consult neurosurgery (do not delay!)

**Imaging:** radiographs of skull (AP, lateral, Towne's projection) and C-spine (for associated abnormalities)

- Discuss with craniofacial surgeon if low dose CT indicated for complex diagnosis and/or surgical planning

**Surgery:** ~1st year; technique depends on timing & specific features; +/- helmet therapy

**Multidisciplinary:** neurosurgery, ophthalmology, speech language pathology, occupational therapy

**Prognosis:** variable based on craniosynostosis +/- associated syndrome

October 2023