Craniosynostosis

Premature closure of cranial suture originally called craniostenosis, incidence 3-5/1000 live birth. The growth of the skull normally is perpendicular to suture line.

Primarily a prenatal deformity, postnatal craniosynostosis (CSO) occur uncommonly (postnatal causes consist primarily of positional alterations which may not represent true synostosis). (CSO) rarely associated with hydrocephalus (HCP)

Treatment is usually surgical. In most instances, the indication for surgery is for cosmetics and to prevent the severe psychological effects of having a disfiguring deformity. However with multiple (CSO), brain growth may be impeded by the unyielding skull. Also, ICP may be pathologically elevated, and although this is more common in multiple (CSO), elevated ICP occur in 11% of cases with a single stenotic suture. Most cases of single suture involvement can be treated with linear excision of the suture, the best timing of surgery between (3-6) months of age .Involvements of multiple sutures or the skull base usually required the combined efforts of a neurosurgeon and craniofacial surgeon, and may need to be staged in some cases.

Risks of surgery include: blood loss, seizures, and strokes.

Types of the cranial sutures

1. Sagital: between anterior and posterior fontanels or bet (bregma and lambda) which separate tow parietal bone.

2. Coronal: Run along the top of the skull from side to side in front of the ear which separate frontal from parietal bones.

3. Metopic: which separate frontal bone in to two halves.

4. Lambdoid: which separate occipital bone from parietal.

Causes

The causes of premature fusion is unknown the craniosynostosis is either primary or secondary.

1.Primary (CSO):multiple theories have been proposed for the etiology of primary (CSO),but the most widely accepted is the primary defect in the ossification of the mesenchymal layer of the cranial bones.intrauterine factors may also play a role.

2. Secondary (CSO): more frequent than the primary type and is typically due to the underlying systemic disorder e.g. sickle cell disease, thalassemia, rickets and all causes of microcephaly causing primary failure of brain growth and is associated with premature fusion of all skull sutures.2nd (CSO) can also developed after placement of shunt in a child with hydrocephalus .the (ICP) not elevated but there is often associated with neurodevelopment delay.

Types of craniosynostosis

1. Sagittal synostosis.

The most common (CSO) affecting a single suture. Results in **(Scaphocephaly)** boat shaped skull with frontal bossing, prominent occiput, and palpable keel like sagittal ridge. (OFC) remain close to normal, but the biparietal diameter is markedly reduced.

2. Coronal synostosis.

Bilateral coronal (CSO) **(Brachycephaly)** with broad, flattened forehead

Unilateral coronal (CSO) **(Plagiocephaly)** with forehead on the affected side flattened or concave above eye (normal side falsely appears to bulge abnormally), supra-orbital margin higher than normal side. The orbit rotates out on the abnormal side, and can produce amblyopia.

3. Metobic synostosis.
Result :**(trigonocephaly)** pointed forehead with midline ridge

4. Lambdoid synostosis.
Bilateral lambdoid (CSO) produce **(Brachycephaly)** with both ears displaced anteriorly and inferiorly
Unilateral produce lambdoid **(plagiocephaly)** result in bulging of ipsilateral forehead (rhomboid skull) with ipsilateral ear locater anterior and inferior to contralateral ear. The contralateral orbit and Forehead may also be flattened.

5. Multiple synostoses.
Fusion of many or all cranial suture **(oxycephaly)** tower skull with undeveloped sinuses and shallow orbit, this patient has elevated (ICP)