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Craniofacial Support



Non-syndromic Craniosynostosis

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What is Craniosynostosis?

Craniosynostosis may be defined as the premature closure or fusion of the calvarial sutures occurring in the uterus or shortly after birth. The calvarial sutures are lines of growth lying between the various bones of the skull. While there are a large number of sutures in the vault and base of the skull which can be involved, this discussion will be limited to the common ones on the top of the head.

These are six in number, namely the **sagittal suture** which runs longitudinally down the midline of the skull between the anterior and posterior fontanel, the **metopic suture** which runs longitudinally from the anterior fontanel to the area between the eyes, the two **coronal sutures** - one on each side running transversely from the anterior fontanel to the area just behind the orbits and the two **lambdoid sutures** which run obliquely downwards from the posterior fontanel to the areas behind the ears (see Fig 1).

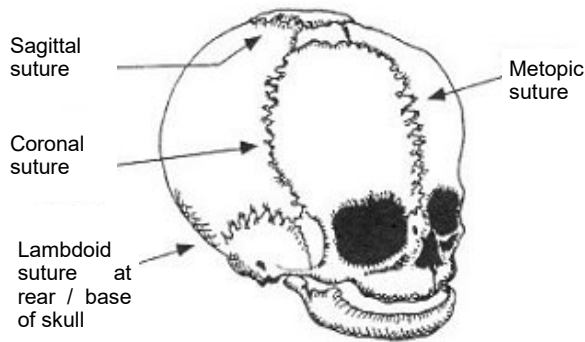
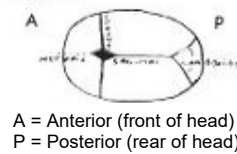


Fig 1



A = Anterior (front of head)
P = Posterior (rear of head)

The exact

cause of sutures fusing prematurely in isolated instances is as yet unknown. The overall incidence averages out at approximately 1 in 3,000 live births.

Under normal circumstances the growth of the individual skull bones occurs at right angles to the growing sutures. If a suture fuses prematurely the skull fails to grow at right angles to the involved suture(s). Importantly, normal adjacent sutures respond to this growth restriction by increasing their activity and there is thus generally a compensatory growth in a direction parallel to the involved suture.

It is the failure of normal growth at right angles to the suture, and the excessive compensatory growth at other sutures, which gives rise to the classical skull shapes associated with craniosynostoses. The typical shapes can be used clinically to predict the site of the abnormal suture.

forehead is reshaped from the existing or adjacent bone.

As mentioned, lambdoid sutures synostosis is uncommon and the surgery is aimed at preventing progressive disfigurement by releasing the suture and remodelling the posterior skull.

Irrelevant of the sutures involved, if there is evidence of raised intracranial pressure then more extensive skull vault surgery may be required to expand the volume of the skull and thus relieve the pressure.

It is important to note, once again, that while this is major surgery it is generally classed as safe if performed by a multidisciplinary team, i.e., a neurosurgeon with a plastic surgeon and/or a maxillofacial surgeon in an established unit. Further details of surgical procedures for treating single suture and syndromic craniosynostosis can be found in the Headlines - Craniofacial Support (previously CFSG) leaflet, No 4, **Craniofacial Surgery**.

Finally, it is also important to note that because the underlying defect in the growth centre is as yet uncertain, the surgery does not necessarily normalise the growth in all cases. There is a tendency in a certain percentage of cases for the condition to recur, or for progression to occur in other areas particularly the face, thus further operation or re-operations may be required in some cases. The predictive factors for this will depend on the sutures involved and the extent and rate of progression of the deformity. Once again it is impossible to give blanket guidelines for this and it should be discussed in detail with the treating surgeons.

Written by *Steven Wall, Consultant Plastic Surgeon, Oxford Craniofacial Unit, Radcliffe Infirmary*

These skull shapes have been given names which infer the involved suture and these will be covered at a later stage in this discussion.

Because skull growth is most rapid during the first two years of life and continues to adulthood, the presence of an abnormal or non-functioning suture gives rise to a progressive deformity. This is most rapidly progressive in infancy but has the potential to progress until growth is completed in adulthood. As mentioned there are sutures in the base of the skull as well which may be affected or, more frequently in the isolated synostoses, may grow compensatorially, causing distortion of the lower orbits and the face at a later stage.

What other effects does fusion of a suture have?

In the vast majority of cases of single-suture craniosynostosis the compensatory growth of the normal sutures is generally sufficient to allow the developing brain to grow without causing raised pressure. However, in a certain percentage of cases (somewhere between 10-15 %) the restriction is such that the pressure within the skull rises (so called *raised intracranial pressure*). This may cause functional problems in terms of development if left untreated. In addition there is evidence accumulating that there may be local pressure effects underlying the involved sutures which may be corrected by surgery.

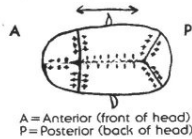
So, there are a number of reasons why surgery is generally indicated in craniosynostosis

- (1) For the treatment of an established disfigurement.
- (2) To attempt to prevent the significant progression of a developing disfigurement.
- (3) To relieve established raised intracranial pressure.
- (4) To decrease the risk of developing raised intracranial pressure or other functional pressure-related effects.

Types of single suture craniosynostosis

The type of surgery required is frequently determined by the degree of the disfigurement and the underlying sutures involved. Therefore we will briefly review the different synostoses and their main features before proceeding to the discussion of surgery.

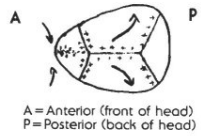
Sagittal synostosis



Sagittal synostosis

Fusion of the midline sagittal suture. Growth fails to occur across the head and excessive growth occurs from front to back. The resultant disfigurement is a narrow head which is excessively long. This is traditionally known by the term “*scaphocephaly*” which means “boat-shaped head”.

Metopic synostosis

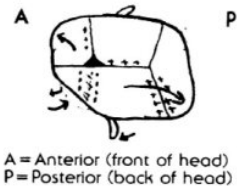


Metopic synostosis

Fusion of the anterior midline suture with failure of adequate transverse growth in the forehead and compensatory growth posteriorly and laterally. The end result is a central ridge of the forehead with a pinched look above the brows. The eyes tend to be fairly close together (*hypotelorism*). The overall skull shape (particularly anteriorly) is triangular in nature and this is classically known as “*trigonocephaly*” or “triangle skull”.

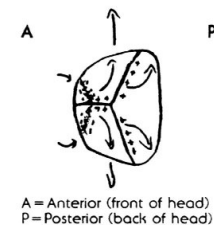
Coronal synostosis

This may be divided into cases where only one coronal suture is involved or those where both coronal sutures are involved:



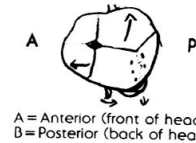
Unicoronal synostosis is where one suture is involved and there is a failure of adequate anterior posterior growth on the side of the involved suture. There is flattening of the brow and elevation of the upper part of the orbit and eyebrow area. The ear on the involved side tends to be pulled forward. The opposite forehead bulges significantly and there is increased growth in areas of the posterior

skull. In addition there is frequently compensatory growth in the cheek area of the involved side, which in untreated cases may potentially cause facial distortion. The overall impression is of a skull which has been twisted skew. This condition is classically known as “*plagiocephaly*” or “oblique skull”.



Bicoronal synostosis is when both coronal sutures are involved. There is failure of adequate anterior, posterior growth on both sides of the skull. The skull becomes excessively wide and short from front to back. There may be excessive growth at the upper part of the forehead and both eyebrows and orbital rims are pulled up and are flattened. This short front to back appearance of the skull has given rise to the name “*brachycephaly*” or “short skull”.

Lambdoid suture synostosis



This is rare, with an incidence of approximately 1 in 10,000 live births. This presents with flattening of the skull around the involved suture with compensatory bulging occurring at the forehead on the involved side. Marked bulging of the mastoid bone behind the ear and a significant bulge on the parietal bone (which is the area above the ear) on the opposite side.

Surgical treatment

As already indicated there are a variety of reasons for proceeding to surgery in craniosynostosis. Each case needs to be individually assessed in terms of functional indications and established or progressive disfigurement. Currently craniosynostosis surgery is one of the supra-regionally designated services. There are four nominated centres in England: The Radcliffe Infirmary, Oxford; Children's Hospital, Birmingham; Great Ormond Street, London and Alder Hey Hospital, Liverpool.

Whilst the surgery required is fairly extensive, if performed in an established unit it is regarded as safe, with acceptable risk in terms of the benefits achieved. These “benefits versus risks” are best discussed by the individual surgeons based on the individual indications on each child. It is not appropriate to give an analysis of all the various combinations in a pamphlet of this nature.

In general, as mentioned above, the surgery is aimed at preventing progressive disfigurement, correcting established disfigurement and reducing any functional risk in terms of raised intracranial pressure.

It has been established that it is safe to remove segments of bone in the area of the involved sutures, and to reshape segments of bone and change their position with predictable survival of bone fragments and reliable healing of the bone and soft tissue.

Sagittal suture synostosis is traditionally treated in the early phases by removal of some of the bone overlying the abnormal midline suture, the so called “*strip craniectomy*”. This may be combined with various other ancillary procedures such as “*plication*” or tightening of the bones laterally to encourage the development of a broader, shorter head with growth. In established cases in older children more extensive reshaping of the skull may be required.

In the anterior synostoses (namely metopic, bicoronal and unicoronal synostosis) the aim is to recreate a symmetrical forehead and orbital rim. To “release” the area of the involved suture and thereby allow more normal growth of the skull. The mainstay of this type of surgery is the “*fronto orbital advancement and remodelling*” procedure whereby the upper aspects of the orbits are freed and advanced unilaterally or bilaterally, as appropriate, and a more symmetrical