Single-Suture Craniosynostosis

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This pamphlet was written to provide information and guidance for parents of a child with premature closure of a single cranial suture system. These disorders are called the single-suture craniosynostose – the most common types are scaphocephaly, synostotic frontal plagiocephaly (also called "unilateral coronal synostosis"); and trigonocephaly. This presentation includes a description of these disorders, the cause (what is known), and the treatment. A glossary of selected medical terms is also included. This pamphlet is intended as an aid, not a substitute, for discussion between parents and the various medical specialists. Parents are encouraged to ask all their questions and to be certain they are given answers by members of the craniofacial team.

Introduction

The term non-syndromic craniosynostosis denotes that these disorders are usually isolated cranial abnormalities, without problems in other areas. These are spontaneous anomalies that are rarely the result of a genetic mutation; nor are they familial or hereditary. The best known in order of frequency are: *scaphocephaly* (premature closure of the sagittal suture); *synostotic frontal plagiocephaly* (premature closure of the coronal suture); *trigonocephaly* (premature closure of the metopic suture). Rarely is simple,

non-syndromic craniosynostosis associated with increased intracranial pressure or developmental delay.

Clinical Features

Scaphocephaly: Children with scaphocephaly have a long head that is narrowed from side to side. The forehead and back of the head (occiput) may be very prominent. There is little evidence that scaphocephaly causes increased pressure on the brain.

Synostotic Frontal Plagiocephaly (Unilateral Coronal Synostosis): Children with synostotic plagiocephaly have an abnormal flattening on one side of the forehead often, with a compensatory bulging of the normal side of the forehead. In addition to flattening of the forehead, there is elevation and posterior retrusion of the eye, giving the eye a wide appearance. The root of the nose is deviated to the involved side. On the involved side the ear is forward as is the cheek bone. The chin is often deviated to the side opposite of the flattened forehead.

Synostotic frontal plagiocephaly (secondary to premature closure of one half of the coronal suture) must be differentiated from the very common abnormality, that results from intrauterine deformational forces called *deformational anterior plagiocephaly.* The features of this benign condition include, flattening of the forehead, and a downward position of the supraorbital rim, giving the eye a small appearance. The cheek is often flattened on the same side and the ear is always posteriorly displaced on the same side. The importance of making this distinction is that the treatment is

entirely different for deformational frontal anterior plagiocephaly. Usually it is only necessary to use physical therapy to loosen the neck (torticollis), which is often tight on the same side.

Far more common than anterior positional flattening is *deformational posterior plagiocephaly*. This is likely related to keeping babies on their back to sleep to prevent SIDS. Often, a molding helmet is recommended. This posterior cranial flattening must be differentiated from the very uncommon *synostotic posterior plagiocephaly* caused by unilateral fusion of a lambdoid suture.

Trigonocephaly

Trigonocephaly is a triangular or keel-shaped forehead, caused by abnormal closure between the two frontal bones. Usually the distance between the eyes is narrow and the lateral corner of the eyes are angled upward.

Cause

Scaphocephaly

The cause of premature closure of the sagittal suture is unknown. This abnormality can occur in association with fusion of the coronal suture in Crouzon syndrome. Very rarely is isolated scaphocephaly familial.

Synostotic Frontal Plagiocephaly

The cause of most cases of most cases is also unknown; it is almost always a sporadic condition and not familial. Nevertheless, synostotic frontal plagiocephaly can occur in certain genetic disorders such as Saethre-Chotzen

syndrome (chromosome 8), craniofrontonasal syndrome (X-linked), and a mutation in FGFR3 (chromosome 4) or FGFR2 (chromosome 10).

Trigonocephaly

Like the other non-syndromic craniosynostoses, this usually occurs without any family history. It can, however, be associated with chromosomal deletions involving chromosomes 9, 11, and 13 and other very rare disorders. The metopic (interfrontal) suture is the first of the cranial sutures to close, variably between six months and age 2 years. In some families, premature closure occurs but with growth, there is a normal flattening of the vertical ridge of the forehead.

Treatment

Scaphocephaly: If the infant is seen promptly, i.e., during the first few months of life, the corrective surgical procedure is called a "strip craniectomy." This involves removing a midline plate of bone including the fused suture. If the child is seen late in the first year of life or thereafter, a more complex surgical release and remodeling of the posterior, and sometimes anterior cranium is necessary.

Synostotic Frontal Plagiocephaly: Surgical correction is done around 8-10 months of age. It involves bilateral repositioning of the forehead and superorbital area. The nose is also straightened during the same procedure. The majority of children with this disorder need only one operation.

Trigonocephaly: Some minor forms of trigonocephaly can be simply watched and will improve in time. Others can be treated by burring down the

abnormal central keel. If an operation is necessary, it is scheduled between eight and ten months of age. The procedure involves removing the abnormal suture along with expansion and modeling of the forehead.

Glossary

Craniectomy

Removal of cranial (skull) bone.

Deformational Plagiocephaly

The cranium is flat because of external forces, but the sutures are open. This condition is often designated as either anterior (frontal) or posterior (occipital).

Mutation

A change in a gene. The human genome (46 chromosomes) is made up of about 30,000 gemes.

Non-syndromic

Refers to a condition in which there is a single or isolated abnormality.

Scaphocephaly

Boat-shaped head, the anterior-posterior dimension is overly long and the width of the cranium is narrow. It is caused by premature closure of the sagittal suture.

Syndromic

A group of findings or symptoms that occur together.

Synostostic Plagiocephaly

Fusion of a suture, either anterior (coronal suture) or posterior (lambdoid suture).

Torticollis

Twisted neck, caused by a tightness or shortening of the muscle that originates behind the ear (mastoid process) and inserts on the sternum and clavicle (collar bone).

Trigonocephaly

Triangular-shaped forehead due to premature fusion of the metopic suture (between the two forehead bones).