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IMPACT OF CRANIOSYNOSTOSIS: A REVIEW

Anatomy

Sebastain J Pengiparambil Professor/Principal, Little Flower Instituite of Medical Science and Research, Angamaly (LIMSAR)

ABSTRACT

Craniosynostosis is the premature fusion of sutures of the cranium, resulting inan abnormal skull shape and restricted brain growth. It usually occurs as an isolated condition, but may also be associated with other malformations as part of complex syndromes. Sutures are fibrous joints peculiar to skull and are immovable. In a neonatal skull these sutures corresponds to fontanelles or soft spots. These membranous gaps enable the infant's skull bones to over ride during vaginal delivery and also permitting brain growth. These sutures may synostose and get obliterated as age advances. When one or more of these sutures are prematurely closed there will be raised intracranial pressure, abnormally shaped skull and many other respiratory and neurological dysfunctions.

KEYWORDS

Craniosynostosis, syndromic, non syndromic, apert syndrome, crouzon syndrome fontanelles.

INTRODUCTION

The skull consists of several bones that are fused to form the cranium. These bones are fused by sutures. The frontal and parietal bone is fused by the coronal suture, the parietal and occipital by lambdoid suture, the two parietal bones by sagittal suture and in cases were frontal bone is paired the two bones are fused by metopic suture. Bregma is the meeting point between coronal and sagittal suture and in the foetal skull this is the site of membranous gap called anterior fontanelle, which closes at 18 months of age.

Lambda which is the meeting point between sagittal and lambdoid sutures and is the site for posterior fontanelle which closes at 2 to 3 months of age.

The metopic suture fusion is the earliest to occur between 9 months and 2 years of age and the sagittal suture fusion is the last to occur. The sutural side of each bone is covered by osteogenic layer which is continuous with the periosteum.

Skull growth occur by appositional growth and by the growing brain causing displacement of the overlying cranial bones which in presence of patent sutures drives bone deposition at the suture site in a direction perpendicular to the sutural line. Sutures normally fuse from back to front and lateral to medial except the metopic, which fuses from front to back.[1]

During infancy, there is an intial rapid calvarial growth, and the brain triples in volume to reach the two thirds of adult size over the first year of life. As the Monro-Kellie doctrine states, the intracranial volume is constant and is the sum of the volumes of brain, cerebrospinal fluid and blood; thus continued brain growth in a closed cavity leads to rise in intracranial pressure[2].

Thus the sutural gaps or soft spots in the foetal skull helps to maintain the intracranial pressure normal and helps in normal brain growth. There exists numerous complex mechanisms (genetic/acquired) that can disrupt this natural process and result in aberrant premature sutural fusion.

Craniosynostosis is an intricate heterogenous condition resulting in an abnormal skull shape due to aberrant premature fusion of one or more sutures of cranium. It may be primary or secondary, simple or complex and syndromic or non syndromic[2]. It is classified according to the frequency of sutural involvement as sagittal (60%) coronal (25%) metopic (15%) and lambdoid (2%).

Primary Craniosynostosis

It results in a primary deficit in the sutural ossification process. Complication increases according to the number of sutures involved. It is said to be simple craniosynostosis if only one of the sutures is involved and is complex when multiple sutures are involved [1,2]. Based on the sutural involvement the skull shows structural deformities as follows: Scaphocephaly, Brachycephaly, Trigonocephaly, Oxycephaly and Plagiocephaly.

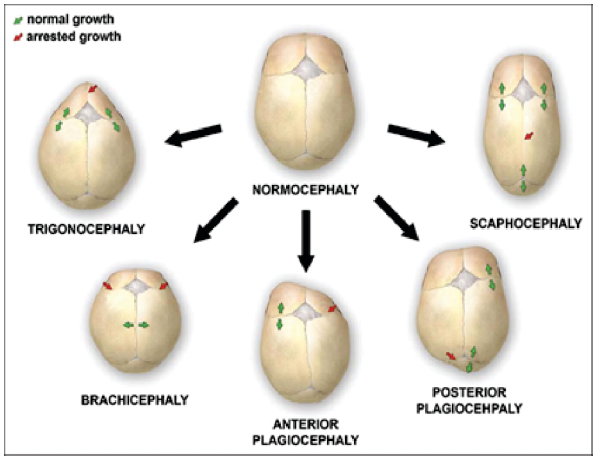

Figure1 shows the structural deformities of skull in craniosynostosis

Scaphocephaly

Most common craniosynostosis in which there is premature fusion of sagittal suture. Male predominance seen and elongated head in the antero-posterior direction and frontal bossing present.

Brachycephaly

Mostly seen in syndromic forms in which there is bilateral coronal suture fusion resulting in short and wide skull. Here in this case, hypertelorism present.

Trigonocephaly

The shape of the head will be triangular and premature fusion of metopic suture is seen. More over, occipital part of the head is broad but the forehead is narrow and pointed.

Oxycephaly

There is multiple sutural fusion.

Plagiocephaly

There is unilateral coronal sutural fusion with subsequent facial asymmetry leads to a flattened forehead on the side of the skull, raised ipsilateral supraorbital margins and characteristic Harlequin sign on the radiograph of the skull. If the lambdoid suture closes too soon the infants head may be depressed on the back and it is referred to as posterior plagiocephaly.

Secondary Craniosynostosis

Secondary craniosynostosis can result from underlying systemic diseases like sickle cell anaemia, rickets, hypothyroidism etc. Or it can develop in newborn with microcephaly due to failure in brain growth or in children with hydrocephalus[3,4].

Although most cases of craniosynostosis are nonsyndromic, craniosynostosis is known to occur in conjunction with other

anomalies in well-defined patterns that make up clinically recognized syndromes.

Syndromic Craniosynostosis

Craniosynostosis is caused by a complex interaction between genetic, epigenetic and environmental factors. Genetics contribute to approximately 20% cases, autosomal dominant mode of inheritance or single gene or polygenic mode of inheritance pattern. Syndromic craniosynostosis has a genetic basis and mostly associated with FGFR2 (Fibroblast Growth Factor Receptor gene) mutations. FGFR2 gene codes for the morphogenesis and migration of mesenchymal cells in craniofacial development. Most commonly seen craniosynostosis associated syndromes include Crouzon syndrome, Apert syndrome, Pfeiffer syndrome etc.

Crouzon syndrome is the most common form of syndromic craniosynostosis exhibiting earlier closure of lambdoid and sagittal sutures. Bicoronalsynostosis is the most common feature with brachycephalic head. A cloverleaf skull deformity has also been described. Fusion of cranial base sutures causes shallow orbits with ocular proptosis. Midface hypoplasia is also seen. In most of the cases children are with normal intelligence [5,6].

Apert syndrome is a rare condition characterized by craniosynostosis with acrobrachycephaly (tower skull). The occiput is flattened and a tall appearance to the forehead is noted. Ocular proptosis is a characteristic finding along with hypertelorism and downward slanting palpebral fissures noted. Midface hypoplasia with mandibular prognathism seen. Syndactyly of second, third and fourth digits of hands and feet is observed. This characteristic limb defects help to distinguish Apert syndrome from other craniosynostosis syndromes [5,7,8]. The physician may recommend genetic counseling to evaluate the child's parents for any disorders that may run in families in cases of syndromic craniosynostosis.

CONCLUSION

Synostosis of a particular suture alters the skull shape in a recognizable manner. An abnormal skull shape at birth is not always craniosynostosis and may be related to fetal head position or birth trauma. Craniosynostosis can affect a child's brain and its development. The degree of the problems depends on the severity of the craniosynostosis, the number of sutures that are fused, and the presence of brain or other organ system problems that could affect the child[9]. Typically, surgical procedures are undergone within the first year of life. Infants with extremely minor craniosynostosis may not require surgery. The aim of the surgical treatment is to enable the normal brain development and to achieve an acceptable cosmetic effect[10].

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