

Case Report

Sagittal and Partial Coronal Synostosis, Treated by Modified Pi Procedure with Barrel Stave Osteotomy : Report of One Case

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Abstract

Craniosynostosis consists of premature fusion of cranial suture(s) resulting in abnormal head shape. Sagittal craniosynostosis involves premature closure of the sagittal suture, producing an abnormally elongated skull called scaphocephaly. Multiple techniques and their modifications have been used for the correction of sagittal craniosynostosis. We present a case of sagittal and partial coronal craniosynostosis who was treated with Modified Pi procedure with barrel stave osteotomy.

Key Words: scaphocephaly, sagittal craniosynostosis, Modified Pi technique

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Introduction:

Premature fusion of a cranial suture is called craniosynostosis.

Nonsyndromic synostosis tends to comprise the majority of the cases of craniosynostosis (1 in 2,500),¹ more commonly involves one suture, and occurs sporadically in the population. Syndromic craniosynostosis occurs in conjunction with certain syndromic presentations in children.

The classification of the craniosynostosis is based on either the suture(s) involved or the shape of the head. Sagittal craniosynostosis is the most common type affecting a single suture. It usually results in dolichocephaly or scaphocephaly (boat shaped skull)² with frontal bossing, prominent occiput, palpable keel-like sagittal ridge. OFC remains close to normal, but the biparietal diameter is markedly reduced.

Multiple methods of surgical repair have been described, from strip craniectomies to extensive

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procedures involving calvarial vault remodeling. One of the more popular and well-described methods is the Pi repair, first described by Jane et al in 1976. [3] Our procedure involves a modified Pi craniectomy [4] with barrel stave osteotomy.

Case report

A 25 months old female baby got admitted to our department with complaint of head disfigurement since birth. The head of the baby was small in size and asymmetrical at birth which gradually increased to attain the present size. Parents of the baby went to local govt. hospital for this problem and they were assured that the disfigurement would subside with time. The mother also noticed developmental milestones of the baby were delayed. She learnt to control her neck at the age of 1 year and still can't sit without support.

Four months back, her mother noticed enlargement of head antero-posteriorly and bulging anteriorly while shaving the hair of head of her child. She also mentions that her baby had reluctance to feed, irritability and occasional vomiting. With above problems parents went to a private hospital and CT Scan of head was done which revealed hydrocephalus and left sided VP shunt was done there. No history of

prolonged fever, trauma to head, convulsion or unconsciousness. Her bowel and bladder habits were normal.

Mother did not take any antenatal checkup during her pregnancy and baby was delivered at full term by NVD at home. Her postnatal period was uneventful. She has a younger brother of 11 months of age who is healthy. There was no family history of craniosynostosis or craniofacial disorders. No history of consanguinity or marriage between parents.

On general examination, baby is conscious and playful. Systemic examination did not reveal any other congenital anomalies. Examination of head revealed occipito-frontal circumference was 44 cm. Anterior fontanelle was open but posterior fontanelle was closed. Frontal bossing was present. There was reduced biparietal diameter [Figure -1(a,b,c)]. Left sided VP shunt was present which was functioning. There was no palpable bony prominence over the sagittal suture. There was no relative movement of the bones on firm pressure on either side of the suture. Neurological examination revealed no abnormalities.

CT scan of brain with 3D reconstruction showed sagittal suture is completely fused. [Figure -2 (a,b,c)] Coronal suture is partially fused, but its impression is



Fig.-1 (a,b,c): Abnormally elongated skull (scaphocephaly) reduced biparietal diameter

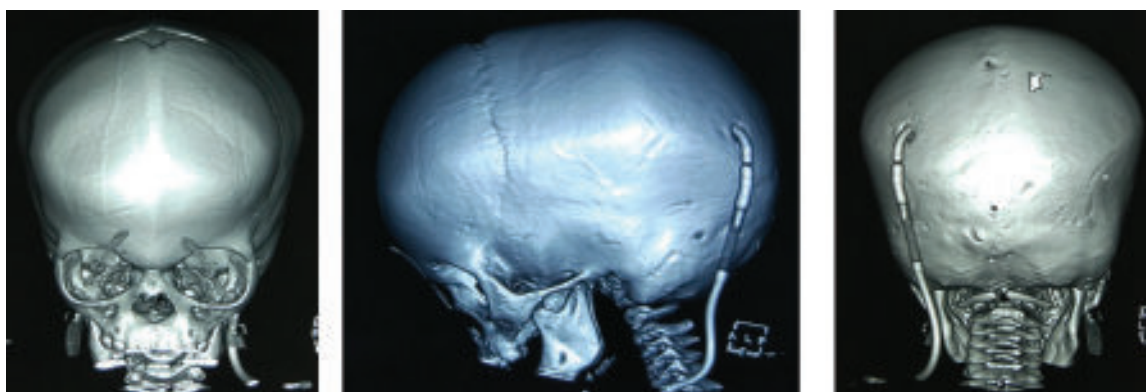


Fig.-2 (a,b,c): CT scan of brain with 3D reconstruction showed sagittal suture is completely fused.

still visualized. Anterior fontanelle is open, posterior fontanelle is closed. Frontal bossing is present. Left sided VP shunt is seen.

Operative technique

The baby was placed supine with head elevation to 20 -30 degree. Then she was wrapped with roll cotton to prevent hypothermia. Proper padding of pressure points was ensured. The zigzag incision was given halfway between the coronal and lambdoid suture. Zigzag skin incision decreases the visibility of the postoperative scar. Anterior and posterior flaps were raised in the subgaleal plane to supraorbital ridge anteriorly and lambdoid suture posteriorly [Figure – 3]. Pericranium was left on the skull during scalp reflection to minimize blood loss.

The targeted areas of craniectomy were marked [Figure – 4]. The horizontal bar of Pi was about 4 cm above

the supraorbital ridge bilaterally and two legs of Pi was made along both sides of the sagittal suture, and ends anterior to the lambdoid sutures [Figure – 5(a)]. Barrel stave osteotomies were then made [Figure – 5(b)]. A sub-galeal drain was placed and the scalp was closed in layers. Intermittent blood transfusion was done during surgery, about 100 ml of fresh blood was transfused peroperatively. Upon completion of the operation, the patient was extubated and were monitored in recovery room for 4 hours. Drain output was checked closely and postoperative hemoglobin level was checked on 1st POD and 50 ml blood was transfused. Next post-operative days were uneventful. Stitches were taken out on 7th post-operative day [Figure – 6]. A post-operative CT scan was done [Figure – 7(a,b)]. A hear gear was given to the baby [Figure – 8]. During discharge, parent were adviced to follow up after 2 months.



Fig.-3: (Anterior and posterior flaps were raised in the subgaleal plane)



Fig-4: (The targeted areas of craniectomy were marked.)

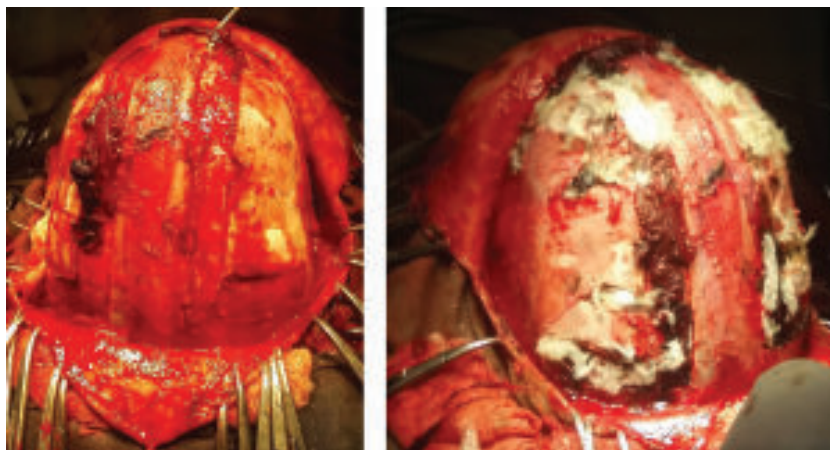


Fig.-5 (a,b): (Modified Pi procedure with barrel stave osteotomy)



Fig.6: (well healed scar along the zigzag incision mark)

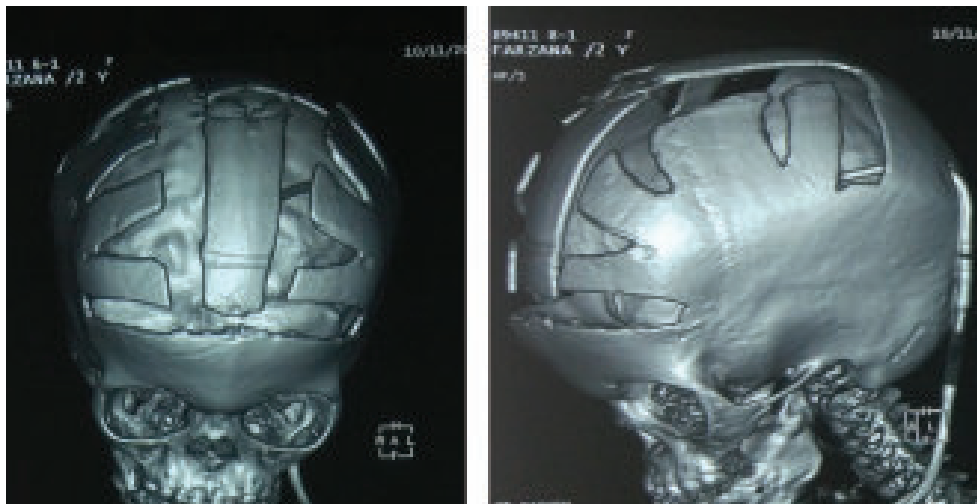


Fig.-7 (a,b): (post operative image shows osteotomy bone gaps)



Fig.-8: A head gear was given to the baby (Reproduced with consent of patient)

Discussion:

The premature synostosis of some of the cranial vault sutures causes expectable skull deformation and, in some cases, produces increased intracranial pressure. [5] Severe psychological effects of having of a disfiguring head is an important issue. [6] Different configurations of the skull are observed depending on the engaged suture. The premature synostosis of the sagittal suture or the sagittal craniosynostosis is the most frequent reason for a calvarial dysmorphism, requiring operative correction.

The premature synostosis of the sagittal (interparietal) suture produces the typical scaphocephalic configuration with frontal and occipital bossing and small biparietal distance (reduction of the normal calvarial wide and a compensatory anterior-posterior extension). Ordinary, these deformations get worse with time.

That is why the purpose of the applied treatment is "normalization" of the head shape. The management of sagittal synostosis (SS) is surgical, the 2 main aims being to normalize the aesthetic appearance of the skull, and to relieve increased intracranial pressure, thus providing a cranium of appropriate geometry and volume to allow for the normal development and growth of the child's brain. [7] As our understanding of the disease process has evolved, and with the advent of surgical innovations, the operative approaches for managing patients with SS have considerably changed, ranging from the initial suturectomy as described by Lannelongue in 1890 and Lane

in 1892 to the more extensive calvarial vault remodeling and variants thereof. More recently, the use of spring-assisted surgery and endoscopic-assisted craniectomies, with and without helmets as postoperative adjuncts, has been described. [8]

In comparison with the other techniques in the literature, the advantages of our technique are the fast and easy technique, the small risks of bleeding [9] and operative shock at its both stages, as well as the retaining of the bone protection of the sagittal sinus.

Conclusion:

If normalization of the head shape is the primary goal of the performed operative treatment, we consider that the proposed operative correction of the scaphocephaly is an part of the management of the sagittal craniosynostosis.

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