ORIGINAL ARTICLE

CRANIOSYNOSTOSIS: EARLY RECOGNITION PREVENTS FATAL COMPLICATIONS

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Background: Craniosynostosis is the premature fusion of cranial vault sutures. The overall incidence is 3–5/10,000 live births. With multiple craniosynostoses, brain growth may be impeded by the unyielding skull. Most cases of single suture involvement can be treated with linear excision of suture. Involvement of multiple sutures or skull has usually required combined efforts of neurosurgeons and craniofacial surgeons. **Methods:** On the basis of visible skull deformity all patients were admitted in the Department of Neurosurgery, Liaquat University Hospital, Jamshoro, Pakistan. Patients were examined for signs of raised ICP and other congenital deformities. The records of patients were maintained till follow up. **Results:** Twenty-seven children were included in this study from 2002 to 2009. Age range was 1–6 years, boys were 18 (66.6%), and girls were 9 (33.3%). The common suture affected was coronal 12 (44.4%). Two children with craniostenosis belonged to same family, and all presented with suture involvement. Three (11.1%) deaths occurred due to hypothermia (1), and blood loss (2). **Conclusion:** Early diagnosis, expert surgical techniques and per- and postoperative care for bleeding and temperature regulation prevent mortality and morbidity.

Keywords: Craniosynostosis, children, skull defects, suture

INTRODUCTION

Craniosynostosis is a premature fusion of one or more cranial sutures. It can be present at birth but can be missed if mild. It usually manifests as an observable deformity within the first few months of life. The prevalence of craniosynostosis has been estimated as 3–5 per 10,000 live births.[1,2] It is a birth defect usually of unknown cause. Several potential risk factors have been identified in epidemiologic studies: male sex, advanced maternal age, maternal smoking, white race, nitro-stable drugs, and certain occupations. Genetic association has also been identified.[3–5] Many teratogenic agents play role in the development of craniosynostosis like diphenylhydantoin. Any agent that can cause ossification defects in the foetus including methotrexate and retinoic acid may cause craniosynostosis. Extrinsic forces *in utero* and abnormal position or early descent of foetal head in pelvis can result in craniosynostosis.[6,7]

Craniocynostosis can happen as an isolated defect (involvement of single suture). Saggittal suture is affected in 40–60% cases, coronal suture in 20–30%, and metopic in less than 10% cases.[8] Lambdoid synostosis is very rare.[9] Craniosynostosis is also seen in the context of various syndromes. The most common syndromes encountered in clinical practice are Crouzon, Apert, Saethre-Chotzen, and Pfeiffer.

Abnormal head shape produce because of restricted growth occurs perpendicular to the fused suture and compensatory growth under non-fused sutures. Growth of the calvarias is not the result of simple tissue growth at the level of suture, rather bones of skull grow secondary to the brain growth.[10,11]

Premature fusions of suture results in cosmetic deformity and compromised skull growth. Raised intracranial pressure has been reported in 50% of patients with multiple suture involvement and 10% in cases with single future involvement. Visual disturbances can occur in children with craniosynostosis secondary to derangement of facial and skull base bony structures. In case of multiple suture involvement blindness can occur because of both raise intracranial pressure (ICP) and facial skeletal deformity.[12] Vision outcome is poor in patients who develop optic atrophy.[13]

No medical treatment exists for craniosynostosis. Indications for surgical treatment in the form of cranial vault reconstruction in the early months of life include progressive facial and cranial deformity, intracranial hypertension, and progressive exophthalmos threatening the eyes.

Over three decades, various techniques for craniosynostosis have been tried. Otto[14] recognised the first premature closure of sutures as a discrete clinical entity in 1830 and coined the term craniosynostosis. The first reported procedure for correction of craniosynostosis was performed in 1890 by Lannelongue.[15] Later, Lane[16] described the first strip craniectomy. There have been many new developments such as distraction osteogenesis, biodegradable miniplate fixation, and development of minimally invasive endoscopic techniques. Though the techniques are standardised, all principles are universally accepted, the individual surgeon's

preferences, training and experience continue to modify the surgical correction of synostoses. Surgical technique start from strip carniectomy to the complete calvarial remodelling.[17,18] Goal of surgery should be correction of form and functions with minimisation of morbidity and mortality. We present our experience in 27 patients who underwent surgery and discussed their results.

MATERIAL AND METHODS

All case of craniosynostosis/craniostenosis admitted between 2002 and 2009 were included in this study. Patients were admitted either from OPD, as a referral, or direct in the Department of Neurosurgery, Liaquat University Hospital, Jamshoro. Patients were diagnosed on examination of suture involvement, and were investigated by simple x-ray and CT scan.

Following variable were analysed: Age, sex, type of suture involvement, and signs of raised ICP. Per-operative complications like blood loss, hypothermia and postoperative complications were recorded. SPSS was used for data analysis. Patients were reviewed at follow-up period of 6 months to 1 year.

RESULTS

Twenty-seven children were included in this study. Age range was 1–6 years. Children were divided into three groups according to their age. Most of the children (13, 48.1%) fell in 1–3 years age group (Table-1). Boys were 18 (66.6%) and girls were 9 (33.3%). Most common suture involved was coronal 12 (44.4%), followed by sagittal 10 (37.03). Strip craniectomies were performed in all cases. A bicoronal flap was made and Scalp flap turned to supraorbital region. One patient died on operating table because of hypothermia and two children expired on second day of surgery because of blood loss. Postoperative CSF leakage was found in 2 patients. Excellent results were seen in 2 patients with craniostenosis.

Table-1: Demography of the patients (n=27)

Variables		No. of Patients (%)
Age		
6 months to 1 year		9 (33.3%)
1 year to 3 years		13 (48.1%)
3 years to 6 years		5 (18.5%)
Gender		
Male		18 (66.6%)
Female		9 (33.3%)
Suture type	**Type of deformity**	**No**
Coronal	Plageocephally	12
Saggittal	Scaphocephally	10
Metopic	Triagonocephally	2
Lambdoid	Posterior Plageiocephally	0
All sutures	Craniostenosis (non-syndromic)	3

DISCUSSION

Craniosynostosis can be either primary from a problem with the involved suture or it can be secondary to position that children spend most of their time. Not all the children need surgery especially those with mild deformity while children with obvious deformity benefit from early surgery. Our catchments area was the rural area of Pakistan and people due to illiteracy go to quake and are initially mishandled. People in rural area believe that children who are born with small heads are God's special creatures and source of pray, and they are compelled to live at shrines for their ill means. It is awful meditation of the people that they make them their earning channels for their livelihood. Many children became mishandled because of lack of knowledge about these skull problems in community and less number of neurologist and neurosurgeons in rural areas of Pakistan. Cultural and socioeconomic reasons of mishandling are also involved.

Surgical technique can be suture release, strip craniectomy, craniofacial remodelling and orthognathic surgery in adolescence.[19] In our series, linear strip craniectomies only were used (Figure-3). Strip craniectomy releases pressure from brain and eyes, and when needed, craniofacial reconstruction should be used. The timing of surgery for isolated non-syndromic craniosynostosis is controversial. We tried to avoid surgery in younger children less than 6 months age because of severe peroperative complications like blood loss and hypothermia.

In our study, maximum number of patients were under 3 years of age and above 6 months which is compatible with other studies.[2] Male gender was predominant in our study compatible to other international studies.[19–21] Common suture involved was coronal followed by sagittal in our series which is contrary to the literature.[19,22,23] We did suturectomies in all cases with extension into temporal and supraorbital regions.

During surgery, dura was found attached to the skull bone and torn during suturectomy in many cases. This showed underlying raised ICP. No significant postoperative complications were found except CSF leakage from wound in two patients who were managed by repeated lumbar puncture.

Death occurred in 3 patients, 2 were male and 1 was female child. Female child expired at operating table because of hypothermia. Same mortality rate was also observed by others.[19]

CONCLUSION

Diagnosis and management of craniosynostosis at an early stage gives good results. Expert surgical techniques and per- and postoperative care for bleeding and temperature regulation prevent mortality and morbidity in children with craniosynostosis.

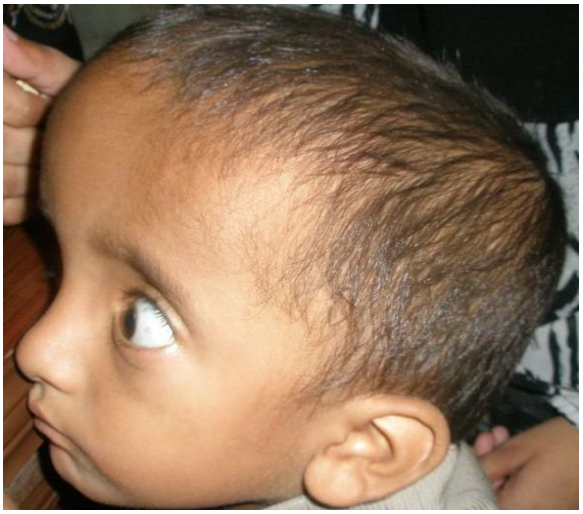

Figure-1: Photograph showing anterior plagiocephaly

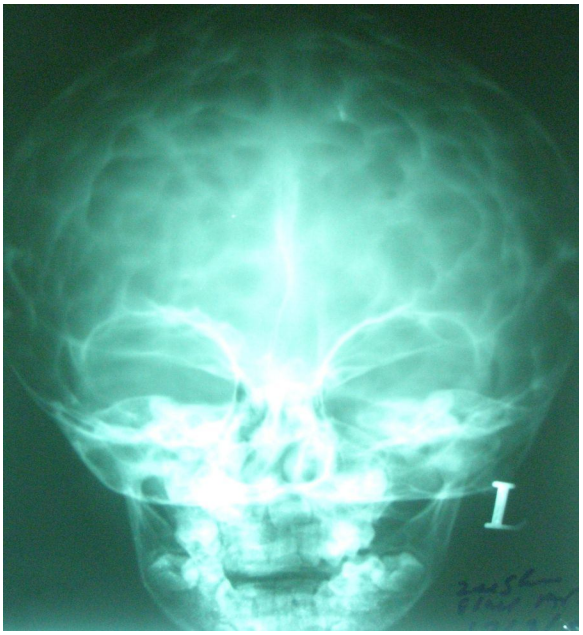

Figure-2: X-ray skull with showing copper beaten sign

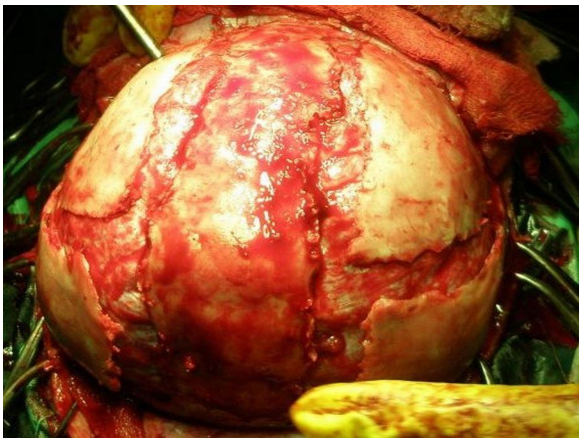

Figure-3: Peroperative photograph showing strip craniectomy

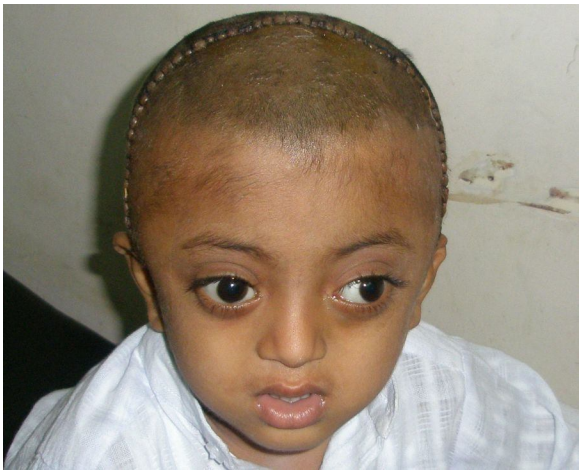

Figure-4: Postoperative photograph of 4 years old boy. Note scar of flap and ocular findings

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