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CRANIOPLASTY
OUR EXPERIENCE IN LAST 5 YEARS

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ABSTRACT

Background: The incidence of craniofacial deformities in our country is not less than other countries. However the surgical treatment of these deformities is considered either difficult or out of the reach for most people. We present our experience at PNS Shifa in the last 5 yrs**.**

Subjects and methods: Five cases of craniofacial anomalies were operated at PNS Shifa, Karachi, from 2005 to 2010. Two had hypertelorism; one associated with frontonasal encephalocoele; one with plagiocephaly, one had Scaphocephaly and one with microcephaly **.**

Results: All the cases recovered well from surgery. Average age at surgery was 5 months. One case (20%) developed CSF leakage from nose and had to be re**-**explored. Cosmetic results were from good to very good.

Conclusion: With better facilities of anesthesia and better imaging techniques the surgery for craniofacial deformities is becoming safe and available in our country**.**

Keywords: Cranioplasty, hypertelorism, plagiocephaly, Scaphocephaly, microcephaly, encephalocoele.

INTRODUCTION

Craniofacial surgery is defined as surgery of craniofacial skeleton. Craniofacial deformities in our country are considered as a variant on the theme of normal and sufferers are often well settled in the close knit society albeit social isolation. They are often left untreated due to several religious beliefs and more importantly due to non-availability or the cost of such treatment. However with increasing awareness due to development of media and information technology more and more people are now agreeing to get the patients operated.

Now a days most congenital anomalies that involve the craniofacial skeleton are lumped together. The specialty has tumor surgery, orbital reconstruction, craniosynostosis and orbital hypertelorism[1]. Tessierin early 1970s is credited for the creation of the craniofacial surgery since he laid down the foundation of modern craniofacial surgery by analyzing the facial clefts and describing both intracranial and extracranial osteotomies for their correction[2]**.** Van der Meulen modified Ortiz monsterio module osteotomy, a wedge osteotomy between orbits to remove a portion of the ethmoids and the nasal bones to simultaneously correct the cortical hypertelorism[1].

The incidence of these abnormalities is estimated at 1.43 to 4.85 per 100,000 births[3]. Causes of the craniofacial defects appear to be multifactorial. They may be sporadic, non-hereditary or they may be a part of syndrome such as Treacher Collins syndrome or Apert syndrome in which case they are inherited. Considering all these variables a planned, staged and sequential approach is necessary to produce an ideal result at the completion of operation and at the completion of facial growth**.**

Orbital hypertelorism is increased distance between medial orbital walls and telecanthus is increased distance between medial canthi[4]. The normal distance between orbits is 15 to 23 mm while one of our patients had this distance of 55 mm**.** One of the patients with frontonasal encephalocoele had Telecanthus due to displacement of medial orbital walls and deformation of orbits[5,6] which automatically corrected after treatment for encephalocoele**.**

Microcephaly had resulted from early closure of sagittal and coronal sutures and lambdoid suture while plagiocephaly resulted from early metopic suture closure. Synostosis leads to elevated ICP and developmental delay.

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Optic nerve atrophy secondary to accompanying papilledema is common[2]. Other features include deafness secondary to CSOM, cleft palate strabismus and palpebral ptosis. Scaphocephaly results fromearlysagittal suture closure.

PATIENTS AND METHODS

This is a descriptive retrospective study conducted on patients presenting in PNS Shifa, Karachi from 2005 to 2010. These patients were essentially operated in the neurosurgery dept. with assistance from ENT colleagues as required. They were divided into 2 groups.

Group 1(2 cases): It included patients with midline defects. One was the case of Frontonasal encephalocoele with mild hypertelorism. Second was the case with large frontal midline defect and severe hypertelorism involving both soft tissues and bony defect. In both cases the defect did not involve lower third of nose or upper lip. CT and MRI of both cases revealed the defect in cribriform plate and frontal bone.

Group 2(3 cases): This group included cranial vault deformities without any herniation or facial component. Microcephaly due to premature closure of all the sutures. Plagiocephaly due to metopic suture foreclosure and Scaphocephaly in agirl of 4 years were the other cases.

Preoperative lab investigations, CT and MR scans and consultation with ophthalmologist and Otolaryngological clinics were done before planning for surgery.

Surgical Technique

Frontonasal encephalocoele:

Forehead of the patient was exposed with a bicoronal incision and anterior sub periosteal flap was raised. Frontal one sided osteoplastic flap was raised. Frontal lobe was retracted and the defect in the cribriform plate dissected out. Dural herniation with atretic brain tissue excised and dura was closed. Inner cortex of the frontal bone flap was used to close the bony defect in the cribriform plate.

Orbital Hypertelorism: Exposure was done by a bicoronal incision of the scalp extending down to the preauricular area on both sides. Sub periosteal elevation of the anterior scalp flap up to mid of the face was done. This was later split through a midline incision extending up to mid of nose.Laterally the incision was extended up to zygomatic arch. Anterior scalp flap was then everted to expose the supraorbital ridges and the whole lateral orbital walls bilaterally. Anterior cranial fossa was exposed by a bifrontal craniotomy using a craniotome and placing burr holes above the temporal crests and parasagittal posterior to the coronal suture. One cm strip of frontal bone with orbital margin was left as a strut. The anterior and lateral maxillary walls were exposed through intraoral route. Orbital dissection was completed. Osteotomy of the orbital cone was done 15 mm from the orbital margin after retracting the frontal lobes. Lateral orbital walls and malar bones osteotomy was then carried out. The widened interorbital regions were removed by vertical osteotomies and about 30 mm of midline defect containing ethmoid air cells were removed with care to save cribriform plate and olfactory nerves.The two orbits were then approximated medially using steel wires and then fixed to the frontal strut. Frontal craniotomies were closed. Soft tissue excision of intercanthal tissue was done and the incisions were closed using subcuticular stitches on the face.

Microcephaly: Bicoronal scalp incision and anterior and posterior sub periosteal flaps were elevated. Excision of bony strips of 1.5 cm lateral to sagittal suture and along the coronal sutures and lambdoid suture were achieved. Bone edges were waxed and the incisions were closed.

Scaphocephaly: In this case we used another technique called morcellation. Exposure was through same bicoronal incision and anterior and posterior flaps were raised. Three cm bone over the closed sagittal suture was removed. Bone over the temporal sides bilaterally was morcellated into small tile like pieces attached to dura and covered with periosteum.

Plagiocephaly: In this case exposure was with bicoronal incision and anterior flap was raised. One sided craniotomy and advancement of the superior orbital margin including orbit and frontal bony flap was achieved.

RESULTS

The results of the surgical correction were very satisfactory and gratifying as regards the cosmetic achievement and the functional outcome was also very good. The results of surgery are shown in figures before and after surgery (Fig 1, 2 and 3). Despite the magnitude of surgery, early age of the patient and long duration of operation we did not encounter any mortality. This could be attributed to lack of anesthetic complications and good control of bleeding. One of the cases developed CSF leakage which required early re-exploration and the site of dural rent was promptly repaired. There were no cases of cutaneous sinuses, wire exposure or persistent CSF fistulae.

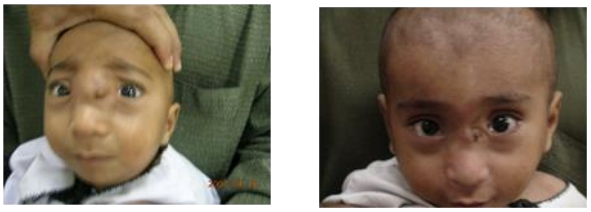

Figure 1: Frontonasal Encephalocoele before and after surgery

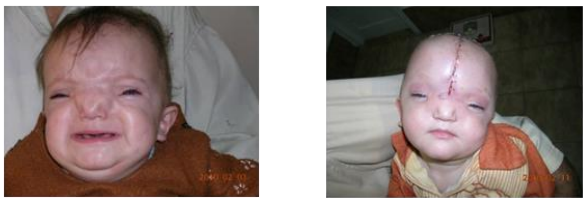

Fig.2: Hypertelorism before and after surgery

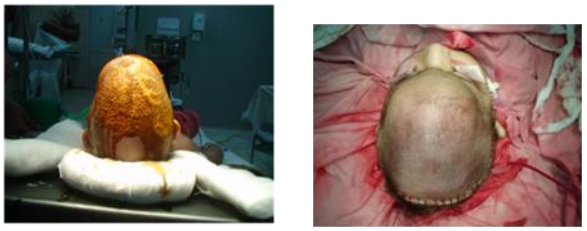

Fig.3 : Scaphocephaly before and after surgery

DISCUSSION

Craniofacial anomalies are grouped into two categories, those which involve failure of fusion or disruption of the embryological units leading to facial clefting, or those which involve premature closure of cranial sutures leading to craniostenosis. These children suffer the severest degree of psychological and educational drawbacks unrelated to mental retardation but due to their inability to interact with their community, or being ridiculed by the fellows due to apparent deformity. Education of the parents and primary care physicians and the possibility of improvement with surgery at an early stage need to be stressed. Early surgery is crucial to prevent psychological problems later in life.

The orbital hypertelorism is corrected by the operation of facial bipartition which was originally described by Tessier in 2 stages[2]. Later Converse and his associates developed this procedure in a single stage which preserves the cribriform plates and olfactory nerves.

Orbit rim advancement for correction of plagicephaly was done initially on the effected side only but the results in long term were less than satisfactory. So now the procedure was done on both orbits simultaneously[7,8,9].

Different procedure have been used to correct Scaphocephaly. Morcellation is an established procedure whichgives excellent results[10]. Other procedures being done in Centers of Excellance since 1997 are the use of spring assisted carniotomy whose results are compareable to results of our procedures except for short hospital stay and less morbidity[10,11].

Frontal carniotomy for the correction of nasal ensephalocoele; unilateral or bilateral; with excision of the sac and closure of the dura and reconstruction of the frontal fossa floor (nasal roof) is being practiced with minor variations in the finer details of surgery (the use of auto and allograft or synthetic bone or dural replacement materials, use of small plates, fibrin glue sealant etc.) has become the standard procedure for this kind of defects.

The timing of this procedure is also very controversial as some surgeons delay this procedure till the age of 5 or 6 years claiming that the bones are very fragile at young age and secondly that operation at young age may impair mid face development. This issue, however, has been addressed by Monasterio and his colleagues who have found normal mid face development in patients with early operation. To qualify as a craniofacial operation at least one orbit must be entirely stripped of soft tissues except for attachment of the

nasolacrimal apparatus at the point of attachment of the entry of the optic nerve[1].

We have used steel wires and prolene suture in our surgeries and did not have any problem with the fixation. Minimal amount of hardware was used to prevent restriction of growth of the facial skeleton and migration of the hardware with growth. Thin facial bones were actually easier to handle and osteotomy and molding and greenstick fracturing was easier in the patients at younger age.

CONCLUSION

With increasing awareness of the parents, better education and developing media, more and more people are approaching the doctors for the reconstructive surgery of the craniofacial defects. On our end now we have easier availability of CT and MR scans with 3 D reconstructions to better evaluate and plan for the surgery. Helped by the development of better and safe anesthetic techniques we are now able to offer safe and corrective surgery at a much younger age with acceptable to very good results.

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