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ORIGINAL ARTICLE

Pediatric/Craniofacial

Craniosynostosis in an Indian Scenario: A Long-term Follow-up

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Background: Craniofacial surgery as a subspecialty has finally taken off in India. Young plastic surgeons in the early 1990s became fascinated by its enormous scope. The author was also among the lucky ones to be trained in this field with Dr. Ian Jackson at the Craniofacial Center at Southfield, Michigan, from 1993-1994. This facility for craniofacial surgery was established in the North Indian tertiary care hospital at the Postgraduate Institute of Medical Education and Research (PGIMER), Chandigarh, in 1995.

Methods: We have reviewed the outcomes of management of craniosynostosis patients and evaluated changes in these outcomes as the experience was gained. The study is a retrospective chart review of the 169 patients operated on for various craniosyntoses in the hospital, for the last 25 years from January 1995 to July 2019.

Results: The whole spectrum of craniosynostoses patients have been operated on and followed up for up to 20 years. All the patients underwent open surgery involving fronto-orbital advancement and remodelling of the deformed calvarium. It was noted that all the syndromic patients required more than one operation, whereas the non-syndromic patients had a stable result after a single surgery.

Conclusion: Craniofacial surgery has now truly arrived in India. Our experience with management of craniosynostosis at PGIMER, Chandigarh, has shown an acceptable morbidity and mortality which is at par with the global standards.
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INTRODUCTION

Craniofacial surgery is one of the newer disciplines of plastic surgery and has revolutionized the integration of patients with craniofacial deformities into the society as useful and confident members. The author had the great fortune of being able to spend 1 year with Dr. I. T. Jackson at the Craniofacial Institute at Southfield, Michigan, from 1993–1994. Thanks to the generous help from the hospital authorities at PGIMER, Chandigarh, we were able to start this specialty in 1995 when a 2-year-old child with brachycephaly underwent a fronto-orbital advancement and cranioplasty. Since then, we have managed a wide spectrum of craniofacial anomalies. We have been able to forge a core team comprising members from plastic surgery, neurosurgery, pediatrics, oral surgery, neuro-anesthesia, ophthalmology, genetics, and intensive care departments for a comprehensive management of these patients. The hospital caters to the neighboring 8 states of Punjab, Haryana, Himachal, Chandigarh, Jammu, Kashmir, Ladakh, and part of western UP with a population of about 110 million. The patients coming to the Institute are from varied social, economic, and ethnic backgrounds.

PATIENTS AND METHODS

The study is a retrospective chart review of the 169 patients operated upon for various craniosyntoses in the hospital for the last 25 years, from January 1995 to July 2019. In the early years (1995–2000), the patients were in older age group of 3–4 years, and some cases were even well past their teens. Thanks to the good word spread by print and electronic media, the patients soon started reporting in the first year of life. Table 1 depicts the changing trends in the age profile of these patients upon first visit to the unit.

Most of the patients reporting to us belong to nonsyndromic group. The syndromic patients constituted about 36% of all the cases 61/169 (Table 2).

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Table 1. Age at the Time of First Presentation to the Hospital

Years	Age < 1 year	Age 1–3 years	Age 4–6 years	Age >7 years	Total number
1995–2000	2	6	4	3	15
2001–2010	16	20	14	7	57
2011–2019	40	34	15	8	97
Total	58	60	33	18	169

Total patients: 169.

Multisutural involvement was seen in a sizable number of patients 36/169 (Table 3). The commonest presentation was coronal and sagittal.

Investigations

An accurate clinical examination was found to be quite reliable in the patients of craniosynostosis. However, a 3D CT scan was performed for confirmation and documentation as per the current suggestions.[1] An MRI was ordered only in suspected cases for any associated Chiari malformation or in suspected cases of microcephaly.[2]

All the patients had a detailed ophthalmic examination to document any changes in the fundus, associated squint, or exophthalmos.

The syndromic patients had associated midface hypoplasia leading to airway compromise. Complete choanal atresia was also seen in 2 patients with Apert syndrome. The threshold for postponing the intervention in such cases was kept low in case of a recent upper airway infection, as the chances of complications are very high.[3,4]

Operative Treatment

The majority of patients reported to the hospital for cosmetic improvement; however, some patients had associated symptoms of headache, proptosis, or subluxation of ocular globes. The deformed bones were remodeled to give normal shape to the calvaria. Any other associated deformities such as recessed supraorbital margins, hypotelorism, and frontal ridging/bossing were also addressed at the same time. A drainage procedure such as a ventriculoperitoneal shunt was performed before definitive surgery in cases with issues with the outflow of cerebrospinal fluid (CSF). About 10% of all the craniosynostosis patients needed drainage procedure before definitive cranial remodeling surgery.

Table 2. Types of Craniosynostoses Seen

Diagnosis	No. patients
Brachycephaly	31
Plagiocephaly	28
Trigonocephaly	26
Scaphocephaly	18
Clover leaf deformity	5
Apert syndrome	28
Crouzon syndrome	21
Other syndromes	12
Total	169

Table 3. Distribution of Multisutural Involvement

Sutures involved	No. cases
Coronal, metopic	5
Coronal, sagittal	16
Metopic, coronal, sagittal	7
Coronal, sagittal, lambdoid	1
Coronal, sagittal, metopic, sphenoparietal, lambdoid	7

The correction of the deformed skull was performed by remodeling techniques. These procedures involved open surgery, craniotomies, removal of deformed fronto-orbital segments, breaking down the calvaria into many segments, and then re-arranging these pieces to get an acceptable skull shape. These techniques allowed expansion of the constricted areas and, thereby, helped to improve the cerebral blood flow. They also addressed the regions with compensatory growth and permitted normalization of the skull in one operation. However, these procedures entail extensive dissection and major blood loss requiring blood transfusions.[5] A judicious mix of blood preserving techniques and the use of special agents such as tranexamic acid helped us minimize blood transfusion.[6,7]

The children who reported early in life were operated on around 1 year of age. Children at this age can withstand the surgical stress better and the calvarial bones are still soft and easily moldable.[8–10] Moreover, the secondary bony defects are likely to get ossified better if early surgery is performed. However, some patients reported late and were operated around the age of 2–5 years. Late surgical correction can lead to problems in social adjustment.[11]

Invasive monitoring involving use of a central venous line and an arterial line was undertaken in addition to conventional monitoring. This helped in early recognition of serious problems such as hypohydration, hypotension, and electrolyte disturbances.

Hyperflexion or rotation of the head was avoided to minimize venous congestion. Head-up position was used to reduce the blood loss and avoid venous air embolism. The eyes were protected, especially in children with proptosis or patients in prone position. A broad-spectrum antibiotic was given after induction of anesthesia. A sphinx position was used in correction of scaphocephaly, as it permitted access to the whole calvarial vault.

The open surgery was performed through a zig-zag coronal incision. Keeping the bevel of the blade parallel to hair follicles further avoided damage to hair follicles[12] and minimized scar alopecia. It is important to keep the incision behind the ear as the exposure is still excellent[13] and the scar is hidden unlike in the preauricular version of the coronal incision (Fig. 1). Infiltration of liberal amount of 1 in 500,000 adrenaline saline solution virtually made the elevation of this flap bloodless. Care was taken to avoid injury to the frontal branch of the facial nerve using a Farabeuf periosteal elevator to bluntly lift the nerves off in the temporal region. The

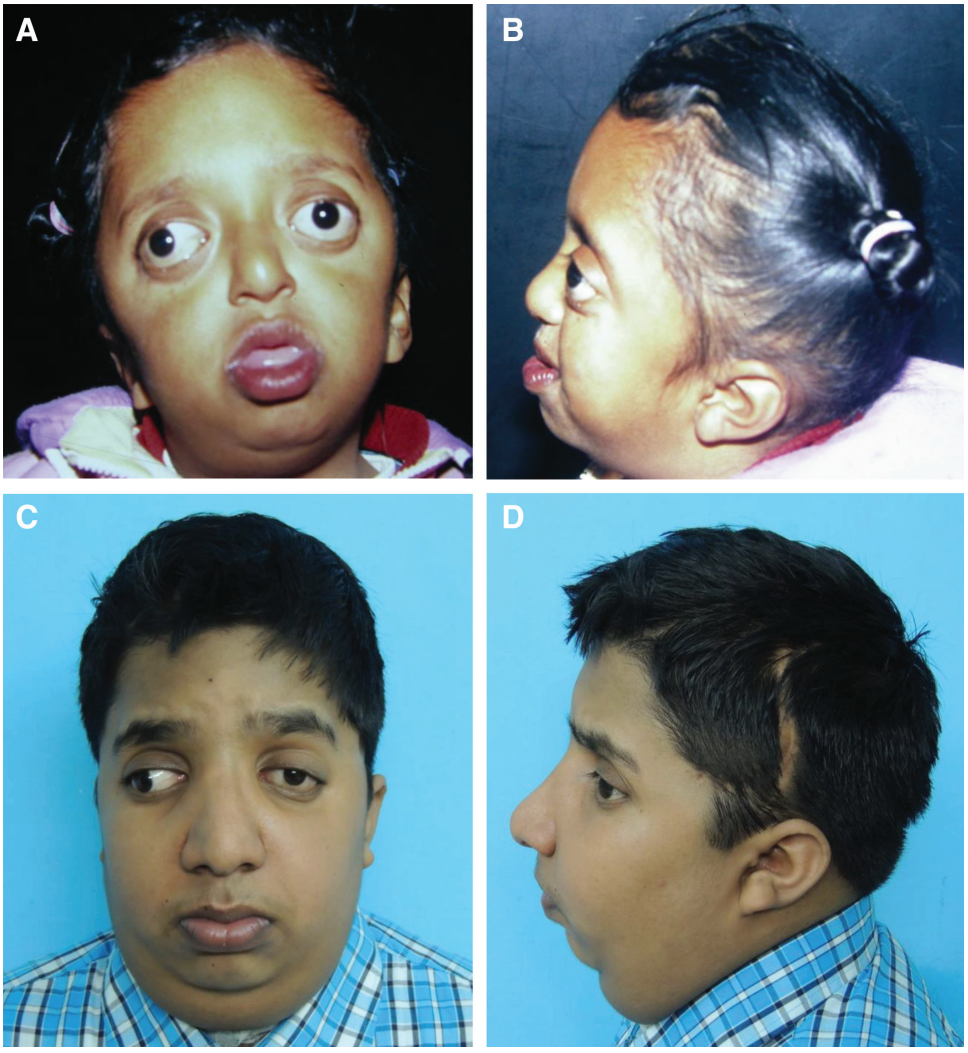

Fig. 1. A patient with Crouzan syndrome. A and B, The patient presented at the age 1.6 with years with a case of Crouzon syndrome with brachycephaly, marked proptosis and supraorbital recession. C and D, Appearance at the age of 16 years. The patient also underwent distraction at Le Fort 3 level at the age of 14 years.

dissection of the anterior and posterior scalp flaps was done in a subgaleal plane. The periosteum was incised about 2 cm above the supraorbital ridge and then rest of the dissection was done in the subperiosteal plane to expose the roof of the orbit and the lateral orbital margins and the lateral wall. The supraorbital nerves and the vessels were gently teased off the canals and if it was a foramen, then an osteotomy was performed to free the pedicles. The dissection was done on the medial orbital wall safeguarding the lacrimal sac and the lacrimal fossa. The temporalis muscle was partly cleared from the lateral wall. This is followed by a bifrontal craniotomy in all the cases. After proper retraction of the frontal lobes, the fronto-orbital segment consisting of supraorbital band and part of orbital roof and lateral orbital wall was also removed.

The fixation of the remodeled calvaria was done using screws and plates. The hospital received patients from the poor and lower middle-class strata of the society and we have always been struggling to cut the costs without unduly affecting the quality. In the earlier part of the journey, we were using stainless steel wires and later switched to use titanium plates. These days, we try to use the bioabsorbable plates whenever possible. These are particularly advisable in children below 1 year of age[14,15] In older children, metallic fixation may provide a better stability; however, there are concerns regarding their transcranial migration.[16]

All the patients were kept in the level 3 recovery room, ICU, for the initial 4–5 hours and once vital parameters stabilized, those patients were shifted to a level 2 care in high dependency unit facility. In cases with difficult airways, the endotracheal tube was kept overnight. A CT scan was performed if there was any deterioration in neurological status. After 48 hours, the patient was shifted to the general ward (level 1). The drains, central line, and urinary catheters were removed on the third day. The oral feeds were started by the second day and all intravenous antibiotics were stopped by the fifth day. The patient was then administered a broad-spectrum oral antibiotic for the next 5 days. The sutures were removed by the eighth day.

RESULTS

A variety of craniosynostosis patients were seen in our study (Table 2). In the initial part of our study, the cases were operated on at an older age, as these were reporting to us at a much older age. However, in the later part of the study, all the cases were operated on around 1 year of age (Table 1).

Coronal Synostosis

This was the most common variety of the craniosynostosis seen in our hospital. A total of 31 patients (18.3%) were seen. The patients underwent fronto-orbital advancement and cranioplasty at the age of around 1 year. The majority of the patients in the nonsyndromic group needed only one surgery. However, in syndromic cases the midface hypoplasia mandated an additional intervention in the later years. The most commonly performed procedure for correction of midface hypoplasia was a Le Fort 3 distraction. Figure 1 shows a case of Crouzon syndrome with gross brachycephaly who underwent fronto-orbital advancement and cranioplasty at the age of 1 year. Subsequently, he developed marked midface hypoplasia and airway narrowing. A midface distraction at Le Fort 3 level was performed at the age of 12 years. He continues to lead near normal life now at the age of 16 years.

Plagiocephaly

This was the second most common variety of craniosynostosis (28/169; 16.5%). The aims of the treatment here included correction of the forehead flattening on the affected side, correction of the vertically oriented bony orbit, and the deviated root of the nose. We performed bilateral frontal craniotomy in all patients, as it afforded a better reconstruction of the deformed structures. Figure 2 shows a case of nonsyndromic plagiocephaly operated on at the age of 1 year. She had just one surgery and has an acceptable result at 20 years follow-up. However, in syndromic cases a secondary surgery was often needed; these included correction of the persistent depression of the forehead by an onlay bone grafting and correction of the deviated nose.

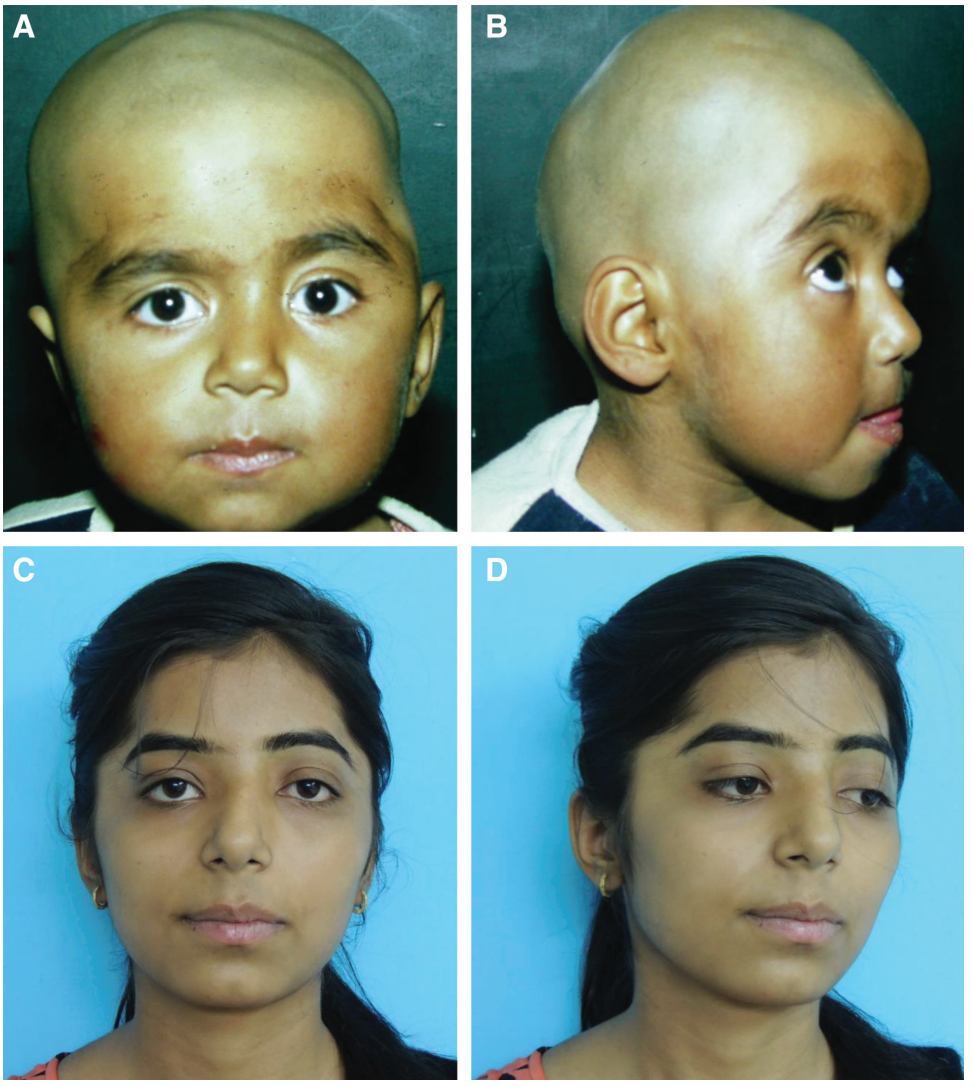

Fig. 2. A patient with unilateral coronal craniosynostosis. A and B, The patient presented with plagiocephaly aged 1 year. C and D, Appearance at 20 years postoperatively, at the age of 21 years.

About 50% of all the plagiocephaly patients needed correction of the associated exophorias.

The Metopic Synostosis

Almost all the patients were nonsyndromic here. It was seen in 15% patients (26/169). In half of these patients, the metopic suture involvement was associated with other suture involvements (Table 2). These children invariably had an associated hypotelorism. The fronto-orbital segment was wedge shaped and constricted in horizontal orientation. Figure 3 shows a picture of child with marked trigonocephaly. The child underwent cranial vault remodeling and expansion of the remodeled fronto-orbital segment at the age of 8 months. A bone graft was also wedged into the frontonasal junction to increase the intercanthal distance. The postoperative picture at the age of 15 years in Figure 3C,D shows long-term correction of the forehead contour and near normal intercanthal distance.

Scaphocephaly

This constituted about 10.7% of all the cases (18/169). The aim of the treatment was expansion of the calvaria in the transverse dimension. Most of the children reported before 2 years of age. Barring two patients, all the patients underwent correction in one stage. In some patients, the cranial and caudal folds of the calvaria resulted in difficulty lying prone or supine. The surgery was performed between 9 months and 1 year and included removal of the frontal, parietal, and occipital bone plates, which were molded, reshaped, and repositioned. This was done by performing zig-zag osteotomies in such a way that the calvarial width can be increased by moving the segments laterally on either side. The resultant bony defects were

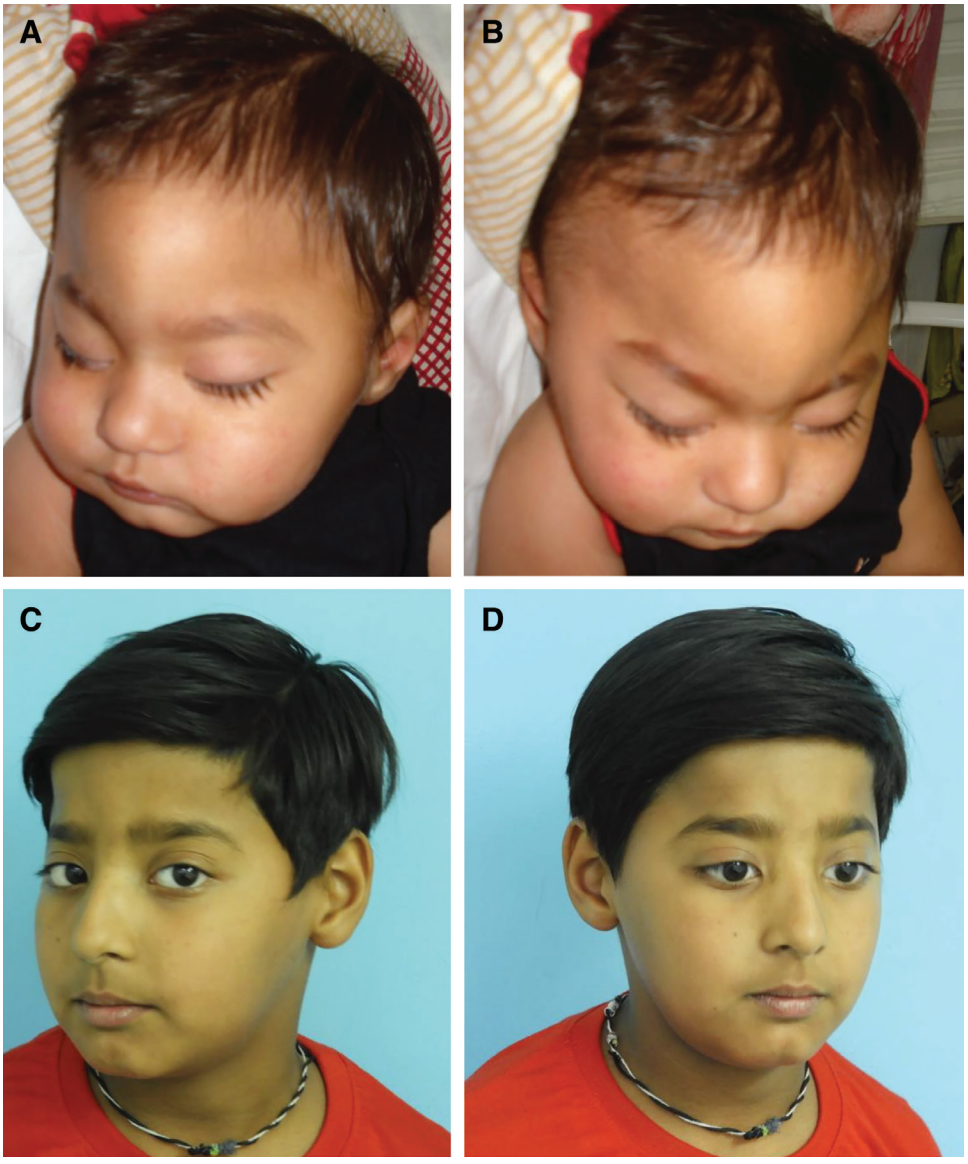

Fig. 3. Metopic synostosis. A and B, A marked trigonocephaly in an 8-month-old child. C and D, Appearance at 14 years postoperatively.

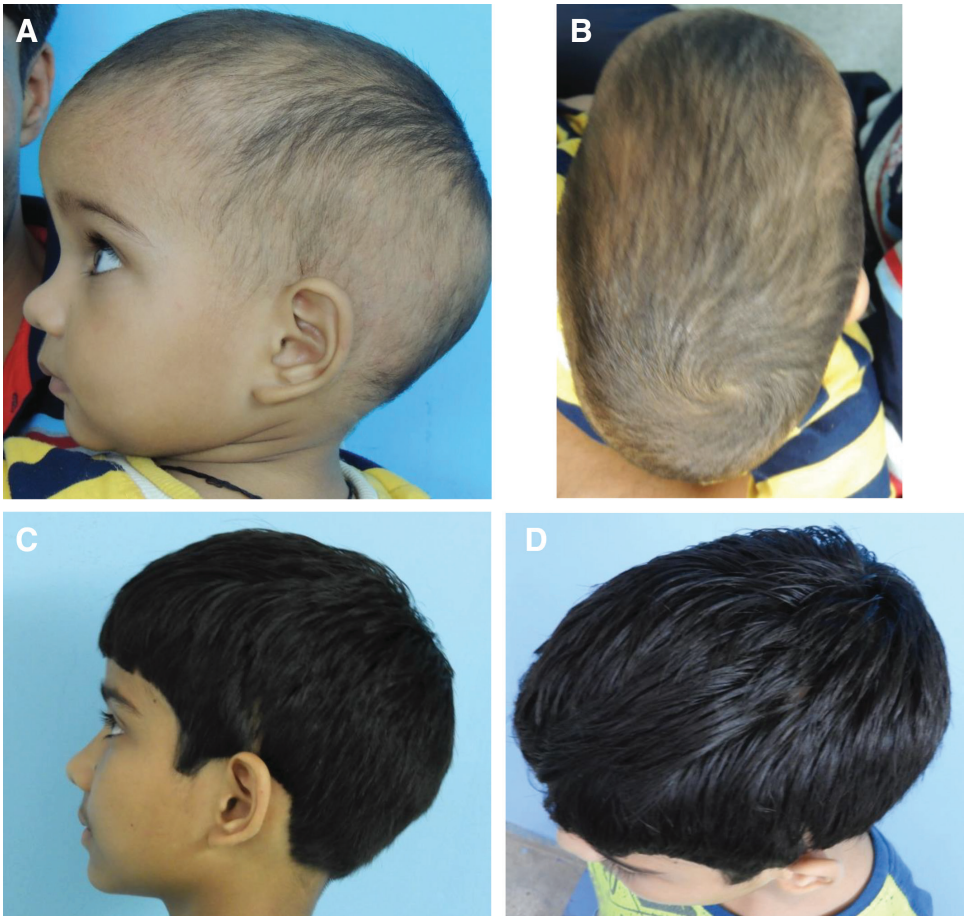

Fig. 4. A case of sagittal craniosynostosis. A and B, A 1.5-year-old child with scaphocephaly. C and D, Appearance at the age of 12 years following cranioplasty and cranial remodeling.

grafted using bones from the calvaria itself. Barrel staving was performed in the temporal regions to match the expanded calvaria. Figure 4 shows a 10-year follow-up of the child who was operated on at the age of 1 year.

Clover Leaf Deformity

We had 5/169 (3%) patients with this deformity. All the cases had features suggestive of raised intracranial pressure as evidenced by gross thumb printing and thinning of the calvaria. These patients required preliminary shunt CSF drainage procedure. It was very difficult to remodel the calvaria as the bones were paper thin and did not hold screws firmly. PDS sutures were used along with plates to maintain the reconstruction. Figure 5 shows long-term result in a 17-year-old child who underwent cranial remodeling at the age of 7 months

Multisutural Craniosynostosis

Twenty-one percent cases of our series (36 out of 169 cases) had involvement of more than one suture. The most common combination was of coronal and sagittal synostoses. (Table 2) The majority of patients with multisutural involvement required shunt procedures to control the raised intracranial pressure before calvarial remodeling operation. **Supplemental Digital Content 1** shows a case of coronal and sagittal synostoses in a patient who reported with abnormal shape of skull and headache at the age of 3 years. (see **figure, Supplemental Digital Content 1,** which displays marked (A and B) turri-brachycephaly in a 2.5-year-old child. C and D show appearance at the age of 21 years; http://links.lww.com/PRSGO/B325). The skull was tower-shaped because the calvarial growth occurred along the spheno-parietal sutures. He underwent cranial vault remodeling and fronto-orbital advancement. He had a long-term stable result 18 years after the surgery.

Syndromic Craniosynostosis

The Apert and Crouzon syndromes were commonly seen in syndromic patients. These cases required multiple surgeries that included skull remodeling, release of syndactyly (Apert syndrome), and midface distraction at Le Fort 3 level. **Supplemental Digital Content 2** shows a case of Apert syndrome who underwent similar management for an acceptable result 15 years postoperatively. (**See figure, Supplemental Digital Content 2**, which displays (A and B) a 1-year-old child with Apert syndrome. C and D show appearance at the age of 14 years. She also had midface distraction at Le Fort 3 level at the age of 12 years; http://links.lww.com/PRSGO/B326.)

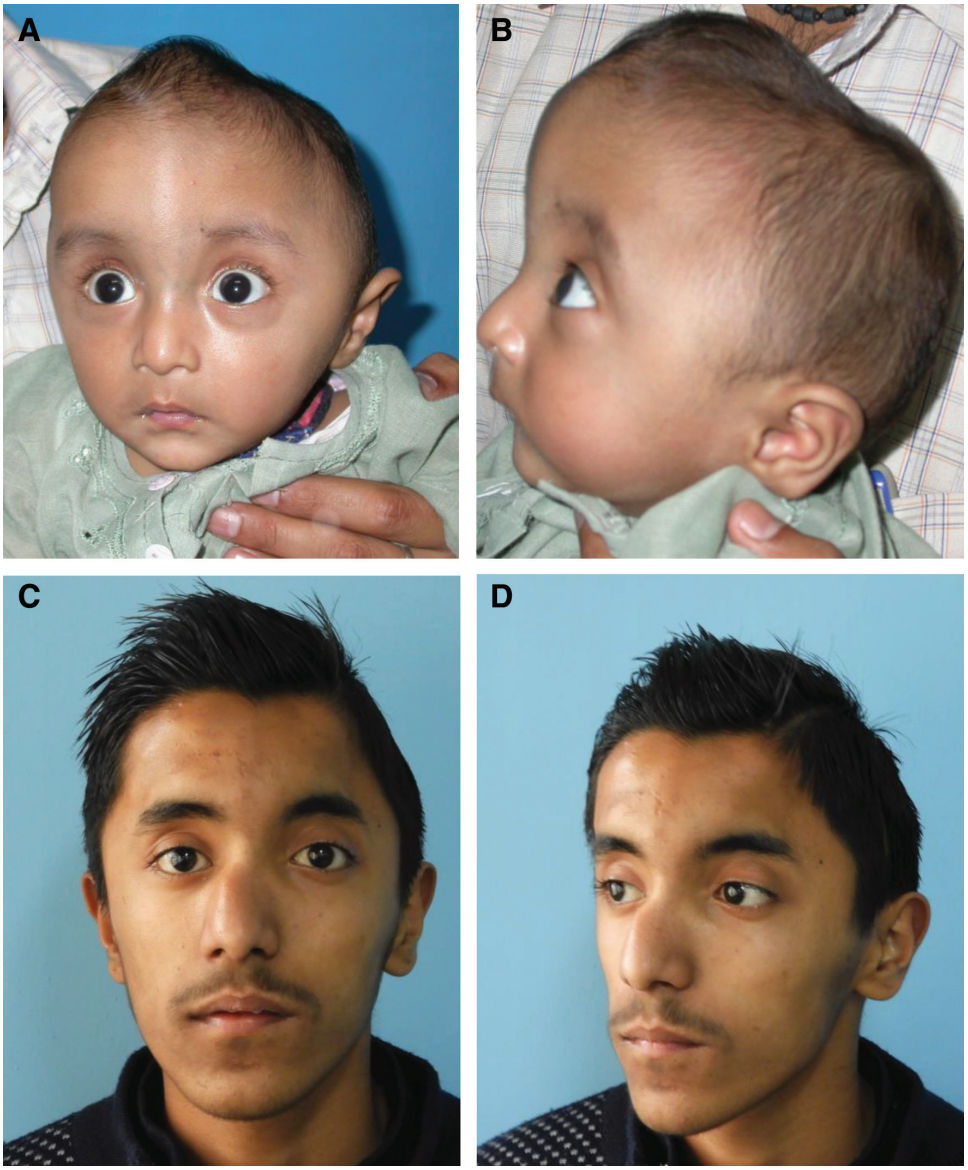

Fig. 5. A case of multisutural craniosynostosis. A and B, Preoperative appearance of a child with clover leaf deformity aged about 7 months. C and D, Appearance at 17 years later.

Management in Older Children

Many patients reported to us quite late with well-established deformities. In the initial part of our study (1995–2010), 39% patients reporting to us were above the age of 4 years; however, in the later part of the study (2010–2019), the number dropped to just 23%. There were both aesthetic and functional concerns in this group. In older children, the bones were thicker and less moldable and required extensive bench surgery for cranioplasty. About 15% of these patients had a CSF drainage procedure performed previously to manage the raised intracranial pressure. In patients presenting after 2 years of age, large bony spurs were seen arising from the frontal bone and indenting the frontal lobes. These spurs were likely to cause dural tears and bleeding from the sagittal sinus during craniotomy. (See **figure, Supplemental Digital Content 3**, which displays spurs arising from the frontal bone indenting into the frontal lobes in a 3-year-old child with brachycephaly, http://links.lww.com/PRSGO/B327.)

Some of the older children had an intervention in the childhood elsewhere and calvarial craniectomies had been performed to relieve the pressure on the underlying brain. The calvaria had re-ossified leading to bizarre shapes leaving some areas of bone defects. All such patients required a reoperative cranioplasty and fronto-orbital advancement.

DISCUSSION

The creation of a craniofacial facility can be quite challenging in a resource crunch scenario prevalent in a public hospital in a developing country. One must be able to create a team of dedicated individuals who can work under one roof to manage such cases. The number of patients in

India is enormous and one needs a lot of resources to meet the demand of such patients. It is possible to cut down the costs by utilizing strategies such as the use of absorbable sutures such as PDS along with the absorbable plates. We have also been using only the minimum number of plates to further cut down the costs. (See **figure, Supplemental Digital Content 4**, which displays the use of PDS suture along absorbable plates to create a reasonably stable construct, http://links.lww.com/PRSGO/B328.)

Craniosynostosis surgery caught up in India in the late 1990s and is now being performed in many centers on a regular basis.[17]As the public awareness of these deformities is growing, the patients report to the hospital in early life (before 6 months) and the standard line of treatment is carried out early in life. In the initial phase of our experience, the average age for surgery was about 2 years and this has come down to between 9 and 12 months currently. However, still there may be situations where the patients report later in life, primarily for cosmetic corrections. The treatment in such patients is primarily tailored to meet the aesthetic needs and may include some camouflage procedures also. Another group of patients are the ones where the surgeons, mainly the neurosurgeons, had performed craniectomies for the treatment of synostosis in the first few months of life to allow for the brain to grow normally. Such patients will have calvarial deformities or large bony gaps that need to be addressed on an individual basis.

During the initial few years of the experience with theses patients, the average time taken for a cranioplasty was about 8 hours. This has come down to an average of 5.5 hours, thanks to the better coordination between the team members. This has also resulted in shorter hospitalization after the surgery. The average postoperative stay currently is about 5 days after the surgery as compared with about 8 days in the earlier part of our experience.

Likewise, there has been a steady improvement in the morbidity and mortality in these patients. In the earlier phase our study (1995–2010), the infection rate was about 4% and this has now stabilized to <1%. The incidence of infection has been reported to be around 3%–6%.[18]

The mortality has been reported to be around 2.2%–0. 1% and this falls with experience.[19] In our center, the mortality has been about 2% cases.[20,21] This has now been <1% for the last 10 years. This can primarily be attributed to better intraoperative and postoperative monitoring in these patients.

CONCLUSIONS

Craniofacial surgery has now truly arrived in India. Our experience with management of craniosynostosis at PGIMER, Chandigarh, has shown an acceptable morbidity and mortality which is at par with the global standards.

However, we still have a large number of cases which do not get the benefit of surgical correction. There is an urgent need to develop more tertiary care centers that will take up such cases on priority basis. The funding of the treatment is another thorny issue, as majority of the patients are poor and can ill afford the expensive treatment. However, with the ambitious launching of "Ayushman Bharat" insurance scheme by the federal government of India, this major obstacle may be resolved to some extent.

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