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Management of Craniosynostosis: A Clinical Study

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A B S T R A C T

Craniofacial anomalies are often complex. Affected individuals should be evaluated and treated by a craniofacial team. Examination of the patients for head shape, suture state, eye changes, deformity of face and other associated anomaly, X-ray skull and CT scan was done in almost all cases. Postoperatively skull x-rays were repeated after a week, every 3 months for first year and at yearly interval for comparison and for records. 40 patients (41.237%) underwent surgery in the form of front oorbital advancement with decompression of orbit and optic canal. Maxillary advancement was done in 2 cases. 3 patients were operated outside and 4 patients were reoperated as they had developed restenosis and signs of raised intracranial pressure.

Introduction

In craniosynostosis growth is arrested perpendicular to the fused suture and there is compensatory overgrowth parallel to the fused suture. Virchow believed that craniosynostosis was a primary malformation and that the associated cranial base deformity was secondary to craniosynostosis. Moss postulated the converse.

Park and Powers postulated a primary defect in the mesenchymal blastema that led to both craniosynostosis and an abnormal cranial base (Park and Powers, 1920). According to Moss theory, spatially malformed cribriform plate and crista galli in sagitally synostosis are viewed as primary abnormalities which at the points of dual attachment, transmit aberrant tensile forces upward through the dural fibre tracts, leading to the premature fusion of the overlying sutural tissues (Moss, 1959).

Craniofacial anomalies are often complex. Affected individuals should be evaluated and treated by a craniofacial team. It includes anaesthesiologist, audiologist, dentist, genestist, neurologist, neurosurgeon, otolaryngiologist, paediatrician, plastic surgeon, psychiatrist, psychologist, radiologist, speech pathologist and social worker. The ideal craniofacial center has

three important functions, service, research and education.

The above mentioned specialities provide care for patients. Research is another function of the team, new caraniofaical surgeries can be viewed as experiment, the patients serving their own control. Documentation yields valuable information. Education by media, disseminating information to community, updating general practitioners and paediatricians is necessary. Finally center should encourage clinical rotations of medical students interns and residents and consultants of relevant specialities.

Modern investigations such as axial and coronal 3-Dimensional CT, MR Scanning, genetic counseling combined with progress in instrument technology have made modern treatment of craniosynostosis safe and sometimes spectacular.

Methodology

Our study was based on clinical evaluation and management of the patient, which was evaluated on Outpatient Department basis and called for surgery electively, except for the patients with raised intracranial tension and for the patients who had presented with acute visual loss.

History emphasized on consanguinity and family history of similar deformity, along with social, physical and mental state of the patient. Most of the patients presented with abnormal head shape, delayed milestones, eye changes and with seizures.

Examination of the patients for head shape, suture state, eye changes, deformity of face and other associated anomaly, X-ray skull and CT scan was done in almost all cases. Postoperatively skull x-rays were repeated after a week, every 3 months for first year and at yearly interval for comparison and for records.

Results and Discussion

Complications

Haemorrhage was the commonest and frequently noted from dural venous sinuses because of deep grooves of the hypertrophied bony ridges enclosing the venous sinuses. Venous channels connecting the torcula and extracranial venous system bleed profusely. Dural tears were encountered in some cases, where the overlying bone was densely adherent to underlying dura and was difficult to separate, however there was no damage to the underlying brain.

Postoperative wound infection and fever was well controlled by antibiotics. Subgaleal collection was relieved by operation.

The coperitoneal shunt was required for CSF leak in one case. In one case V.A. shunt was changed to V.P. shunt following block, even this required to be revised as it was blocked (Two shunts were revised following block and one shunt was removed).

Mortality

Four died out of 40 operated patients (41.237%). First patient died on 7^{th} postoperative day after hypothermia and aspiration. Patient had became unconscious following seizures. Autopsy was performed in the above case which revealed, abnormal shaped brain olfactory nerves and bulbs were not seen. Marked herniation of cerebellar tonsil through foramen magnum upto C2-3. Pons was normal and medulla was flattened. Second patient died after one day of surgery following severe

hypothermia. Autopsy revealed intraparenchymal haemorrhages noted in lungs bilaterally. Brain showed frontal and occipital hypoplasia. Third patient expired on the same day following hypothermia and cardiac arrest. Autopsy performed showed, severe brain swelling of temporal lobe herniation with compression of the brainstem. Fourth patient died one year eight months after the surgery, death was at home. The details of which was not known. Patient's death was told to us when we had called the patient for follow up.

Follow up

The cases were followed up according to the protocol described earlier in proforma (follow up) at regular intervals. Improvement in signs and symptoms, relief of raised ICT, to look for evidence of restenosis. Record of clinical features, radiological investigations and clinical photographs were maintained.

Treatment

Treatment of craniosynostosis is surgical. It involve team work with a craniofacial surgeon, trained anaesthesiologist. We had seen 97 patients from January 1990 to December 1994, 40 patients were operated. All our patients were admitted and operated electively after routine investigations. Anaesthesia fitness, blood donation, ophthalmology, ENT and plastic surgery opinion was sought before surgery. Fresh blood not less than five days old was arranged for transfusion. Patient was catheterized before surgery and the patient was wrapped completely with cotton wool to prevent hypothermia. Patient was operated under general anaesthesia with an endotracheal intubation.

Table showing the disease in male in comparison with David *et al.,* series (David Poswillo and simpson, 1986).

Disease	Montaut & Stricker Series	Our Series
Coronal	50	66.666
Sagittal	80	66.666
Metopic	75	91.666
Pansuture	37.5	46.428
Apert's	66	60.000
Clover Leaf	50	33.333
Crouzon's	56	62.500
Pfeiffer's	100	100.000

In our series all cases were of primary developmental error none of the cases were secondary to any metabolic error.

Clincial features

In our study, 95.876% of the patients had presented with deformed head, while in David *et al.,* series 70% had presented with deformed head. Other commonest presentation being in David *et al.,* series was mental retardation 40.029%, where as our study had 36.082%. Freeman and Brokowf had 44.117% of patients who had presented with mental retardation. Percentage of proptosis, being in our study was 48.453%, whereas in David *et al* series was 60%series (David Poswillo and simpson, 1986).

Raised intracranial pressure

Headache, papiloedema and mental retardation are the contributing factors for raised intracranial pressure.

In our study, 10.309% had presented with headache whilst, David *et al.,* reported

15.204% of papiloedema and 19.298% of optic atrophy cases, whilst our study has 7.216% of cases. Percentage of visual loss in Bertelsen's series is 34.502%, whilst our study is 17.525%. A survey of 776 blind children in Britain, only two children were found to have craniostenosisseries (David Poswillo and Simpson, 1986).

In our study of 7 patients, 6 had unilateral and one patient had bilateral blindness – papilloedema may result in optic atrophy and eventual blineness. We had a case of optic canal stenosis, some authors have suggested constriction in the optic canal or King king of the elongated optic never, however there is little evidence to support this hypothesisseries (David Poswillo and simpson, 1986).

Chronic papilloedema is common in multiple suture synostosis, and is uncommon in younger child because the split sutures and bulging fontanelle's help contain the pressure. Renier *et al.,* (Renier *et al.,* 1982) had recorded the raised ICP in 30%, whilst our study had 17.525% (more in multisuture than with single suture, which is true in our study). Hydrocephalus though uncommon may contribute to preexisting raised ICP secondary to crowding within the stenotic skull. We had 13 cases of hydrocephalus, of which 8 cases required venticulo-peritoneal shunt.

Epilepsy is more common multisuture involvement which is true in our series. bertelsen recorded 25% of his patients having epilepsy, however epilepsy is not a common symptom of raised pressure and may continue after surgery. Out study epilepsy recorded 13.402%.

Lateral rectus palsy is a nonspecific indicator of raised ICP. It resolves after decompressive surgery, however we had 5 cases of lateral rectus palsy, and in one case it persisted even after surgery. Bertelsen noted 10% of patients had deafness, whilst we had one case of hearing loss.

Table.1 Ocular Changes

Disease	Proptosis	Papilloedema	Optic Atrophy	Nystagmus	Keratitis	Squint
Coronal	3	1	-	-	-	-
Sagittal	1	1	1	-	-	-
Metopic	2	1	-	-	-	1
Plagiocephaly (R)	8	-	1	-	-	2
Plagiocephaly (L)	-	-	-	-	-	-
Lambdoid	-	-	-	-	-	-
Coronal + Metopic	-	-	-	-	-	-
Coronal + Sagittal	1	-	-	-	-	-
Microcephaly	-	-	-	-	-	-
Pansuture	21	4	3	1	1	1
Clover Leaf	3	-	-	-	-	-
Apert's	3	-	-	-	-	-
Crouzon's	5	-	2	-	-	1
Pfeiffer's	-	-	-	-	1	-
Secondary Synostosis	-	-	-	-	-	1
Total	47	7	7	1	2	5

Optic atrophy papilloedema was seen in 7 cases (7.216%) however, we had 17 cases (17.525%) who had presented with vision loss, six were blind in one eye and one bilaterally blind. 47 cases of proptosis are seen in our study. 5 cases were associated with squint and 2 cases of exposure keratitis, which required tarsorraphy.

Table.2 Facial Anomalies

Disease	Maxillary Hypoplasia	Telorism		H.A.P.	C.A.	C.P.
		Hypo	Hyper			
Coronal	4	-	5	2	-	-
Sagittal	1	-	-	1	-	-
Metopic	3	8	1	2	1	-
Plagiocephaly (R)	8	2	1	1	1	1
Plagiocephaly (L)	1	-	-	-	-	-
Lambdoid	1	-	-	-	-	-
Coronal + Metopic	-	-	-	-	-	-
Coronal + Sagittal	1	1	-	-	-	1
Microcephaly	-	-	-	-	-	-
Pansuture	10	1	6	1	-	1
Clover Leaf	3	-	3	2	2	-
Apert's	5	2	2	1	-	2
Crouzon's	5	1	2	4	1	1
Pfeiffer's	-	-	-	-	-	-
Secondary Synostosis	-	-	-	-	-	-
Total	42	15	20	14	5	6

H.A.P. = High Arched Palate.
C.A. = Choanal Atresia.
C.P. = Cleft Palate.

Maxillary hypoplasia was seen in 42 cases (43.298%), Hypertelorism in 20 cases (20.618%), Hypotelorism in 15 cases (15.463%), High Arched Palate in 14 cases (14.432%), Cleft Palate in 5 cases (5.154%) and Choanal Atresia in 5 cases (5.154%).

Table.3 Other Anomalies

Disease	AAD	Syndactyly	CTEV	Torticolitis	Spinabifida	CHD	CVS
Coronal	1	1	1	-	-	-	-
Sagittal	-	-	-	-	-	-	-
Metopic	-	-	1	3	1	1	-
Plagiocephaly (R)	-	-	1	-	1	-	-
Plagiocephaly (L)	-	-	-	-	-	-	-
Lambdoid	-	-	-	-	-	-	-
Coronal + Metopic	-	-	-	-	-	-	-
Coronal + Sagittal	-	-	-	-	-	-	-
Microcephaly	-	-	-	-	-	-	-
Pansuture	1	1	-	-	-	1	1
Clover Leaf	-	1	-	-	-	-	-
Apert's	-	5	-	1	-	-	-
Crouzon's	-	-	1	-	-	-	-
Pfeiffer's	-	1	-	-	-	-	-
Secondary Synostosis	-	-	-	-	-	1	1
Total	2	9	3	4	2	3	2

CTEV = Congenital Talipus Equino Varus.
CHD = Congenital Hip Dislocation.
CVS = Cardiovascular System.

We had 2 cases (2.061%) of ADD, one each in coronal and pansuture, they were offered surgery. Only 1 case with pansuture was operated (C1 arch excision was done). Other refused surgery. Both of them were reducible.

9 cases of Syndactyly was recorded (9.278%) of which 5 in Apert's and 1 each in Clover Leaf, Pfeiffer's pansuture and in Coronal synostosis. One case of Polydactyly associated with syndactyly was seen in all 4 limbs in Apert's syndrome. CTEV (Congenital Talipus Equinovarus) deformity was found in 3 cases (3.092%). One each Coronal, Metopic and Plagiocephaly (R).

Torticollis was noted in 4 cases (4.123%) of which 3 were in Plagiocephaly (R) and 1 in Apert's syndrome.
Congenital hip dislocation was seen in 3 cases (3.092%), 1 in each of Metopic, Pansuture and Secondary Synostosis.
Cardiac abnormality was seen in 2 cases. One in Pansuture in the form of ASD and other in Crouzon's syndrome.
We had a case of Plagiocephaly with 4 arms [(R) 3 arms, (L) 1 arm].

In 69 cases, CT Scan was done of which 13 were found to have hydrocephalus, 8 cases were severe, which contributed to raised intracranial pressure and required ventriculoperitoneal shunt surgery. Cerebral anomalies were found in 3 cases of which absent corpus callosum and septum pellucidum was found in microcephaly and coronal synostosis. Poorly formed ventricle in coronal synostosis, hypoplastic corpus callosum was found in coronal synostosis.

Table.4 Surgical state of the patients

Disease	Shunt Surgery	Definitive Surgery	Re-Surgery	Not operated	Operated Outside
Coronal	1	3	-	6	-
Sagittal	-	1	-	2	-
Metopic	-	3	-	9	-
Plagiocephaly (R)	-	8	-	13	-
Plagiocephaly (L)	-	-	-	1	-
Lambdoid	-	-	-	1	-
Coronal + Metopic	-	-	-	1	-
Coronal + Sagittal	-	1	-	-	-
Microcephaly	-	-	-	1	-
Pansuture	2	13	1	15	1
Clover Leaf	1	2	1	1	-
Apert's	-	3	1	2	-
Crouzon's	2 (TPS-1)	5	-	3	2
Pfeiffer's	-	1	1	-	-
Secondary Synostosis	2	-	-	2	-
Total	8	40	4	57	3

TPS -1 Thecoperitoneal shunt.

All cases which were operated electively after investigations and anaesthesia fitness was obtained.

In 8 patients (8.247%) ventriculoperitoneal shunt wad done. We had reoperated upon 4 patients.

In 2 patients, large skull defect had reossified with raised ICP. In third case, reoperation was done because the bone at the outer canthus had grown and it was sharp enough to penetrate outside with reossification of the defect was also seen. Fourth patient had developed pseudomeningocele at Frontotemporal region with reossification of the defective skull. All the patients were given antibiotics, anticonvulsants were given postoperatively. In selected patients steroids were given and tapered off over 3-4 days, where there was evidence of raised intracranial pressure preoperatively. Patients were discharged 8-10 days after surgery with x-rays and their clinical condition noted. Patients were asked to follow up at regular intervals.

Table.5 Type of surgery

DISEASE	Advancement of Head & Orbit	Advancement of Maxilla	Para Sagittal Craniectomy	Decompression Orbital Roof & Optic Canal	Lower of Height of Vault	Previous Craniectomy
Coronal	3	-	-	-	-	-
Sagittal	1	-	1	-	-	-
Metopic	3	-	-	-	-	-
Plagiocephaly (R)	8	-	-	-	-	-
Plagiocephaly (L)	-	-	-	-	-	-
Lambdoid	-	-	-	-	-	-
Coronal + Metopic	1	-	-	-	-	-
Coronal + Sagittal	-	-	-	-	-	-
Microcephaly	13	-	-	-	-	1
Pansuture	2	-	-	-	-	1
Clover Leaf	3	-	-	-	-	1
Apert's	5	1	-	1	1	-
Crouzon's	1	1	-	-	-	1
Pfeiffer's Secondary Synostosis	-	-	-	-	-	-
Total	40	2	1	1	1	4

40 patients (41.237%) underwent surgery in the form of frontoorbital advancement with decompression of orbit and optic canal. Maxillary advancement was done in 2 cases. 3 patients were operated outside and 4 patients were reoperated as they had developed restenosis and signs of raised intracranial pressure.

Hypothalamic dysfunction due to distortion of the diencephalon has been noted by Bertelsen and Montaut & Stricker. We had a case of short stature however, patient at 5 years 2 months of age, the height was not much effected, despite reminder, patient did not turn up for followup. The exact growth of the patient after that is not know. Anosmia is a rare finding. Bertelsen noted tringeminal neuralgia in 5 patients. We did not have any such case (Bertelsen, 1951).

Faciostenosis

Maxillary hypoplasia is commonly seen in Crouzon's and Apert's syndrome. We had 5 cases of Crouzon's (of 8 cases), all cases in Apert's Pansuture 10 cases, and in Plagiocephaly 9 cases.

Maxillary hypoplasia effects the midface causing high arched palate and thereby reducing the nasopharyngeal space, nasal cavity along with paransal sinuses. We had 5 cases of choanal atresia, as the patients had difficulty in breathing. One patient required choanal dilation.

Speech

Midface deformity with cleft uvula, cleft palate and malocclusion of teeth aggravates the speech problem. Mental retardation may also contributes to impairment of speech.

Conclusion

Modern management of craniosynostosis has to be a team effort with Neuosurgeon, Maxillofacial surgeon, ENT surgeon, Ophthalmologist, paediatrician, clinical psychologist, anaesthesiologist, resuscitation specialist and social worker. The aim of modern surgery in craniosynostosis is to operate as early as possible to correct the deformity as completely as possible to

ensure the best results as rapidly expanding brain can mould the elastic skull effectively. Maxillary advancement is best done after the age of 10 years to allow the roots of teeth to descend.

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