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CRANIOSYNOSTOSIS: A REVIEW OF 143 SURGICALLY-TREATED CASES*

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SUMMARY: Çolak A, Tahta K, Bertan V, Erbengi A, Sağlam S, Gürçay Ö, Özgen T, Benli K, Özcan OE. (Department of Neurosurgery, Hacettepe University Faculty of Medicine, Ankara, Turkey). Craniosynostosis: a review of 143 surgically-treated cases. Turk J Pediatr 34: 231-238, 1992.

In this study, 143 cases of craniosynostosis are presented. There were 109 males and 34 females. The major complaints were skull deformity (92 patients), proptosis (38 patients) and microcephalus (32 patients). Neurological examination revealed the presence of optic atrophy in 24 patients and papilledema in 20 patients. Seventy-four patients (53%) had three or more suture closures, with the sagittal suture being the most commonly involved (20% of patients). All patients underwent surgery. Suture removal was performed in 131 patients (91.7%), suture removal plus orbital decompression in 34 (23.8%), and linear craniectomy plus wrapping in 12 (8.3%). The reoperation rate was 6.2 percent. During the follow-up period, preoperative papilledema and proptosis improved in 88.2 and 78.9 percent of patients, respectively. Skull deformity disappeared in 46.9 percent of patients, but remained unchanged in 16.6 percent. *Key words: craniosynostosis, exophthalmos, hydrocephalus, suture removal, wrapping.*

With the use of advanced techniques and approaches, craniosynostosis is becoming one of the more popular entities[1-9]. Craniosynostosis is defined as the premature closure of one or more cranial sutures, producing a deformity of the skull[8-18]. Three classic theories have been advanced to explain craniosynostosis. Virchow[19] believed that craniosynostosis was a primary malformation and the cranial base deformity was secondary to it. Moss[20] postulated that the cranial base malformation was a primary abnormality, resulting in secondary premature fusion of the cranial sutures. The most recent theory proposes that a primary defect in mesenchymal blastema results in both craniosynostosis and an abnormal cranial base[11]. According to Virchow[19] compensatory growth of the skull occurs parallel with the plane of the fused suture. During the first several months of life, when the brain has reached 80 percent of its mature size, its development can be affected if two or more sutures are closed prematurely[6,9,10].

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In these cases, surgical intervention should be carried out as soon as a diagnosis is established. Several different surgical techniques have been used to correct this deformity[3,6,7,9-16].

In this report, we investigated patients according to age, sex, complaint, clinical features and neurological and neuroophthalmological findings. Surgical indications and results are discussed and compared with other reports in the literature.

Material and Methods

This retrospective study comprised 143 patients with craniosynostosis who were investigated and underwent surgery between 1965-1989 in the Department of Neurosurgery at Hacettepe University Faculty of Medicine.

Diagnosis was established according to clinical and neuroradiological findings.

The surgical techniques used in these patients were: suture removal carried out with a median width of two cm; parasagittal linear craniectomy about one cm in width, and the wrapping of the craniectomy edges with an inert film.

Results

Of the 143 patients, 109 were male and 34 female. The age distribution ranged from two weeks to nine years, with a median age of 23.4 months (Table I).

Table I: Age Distribution at Surgery

Age	No. of Patients	
0-6 months	42	(29.4%)
7-24 months	26	(18.2%)
2-5 years	56	(39.1%)
6-10 years	19	(13.3%)
Total	143	(100.0%)

The most frequently encountered complaints on admission were skull deformity (92 patients), proptosis (38 patients) and microcephalus (32 patients) (Table II).

Neurological examination revealed papilledema in 20 patients and optic atrophy in 24 patients. The other findings are summarized in Table III. Although 107 out of 143 patients had a normal head circumference, 135 patients had an abnormally shaped head (Table IV). We found that 74 patients had more than three suture closures, with the sagittal suture being the most commonly involved in 28 patients (Table V). Only the involved suture was removed in 23 patients while all were removed in 108 patients.

Table II: Complaints on Admission

Complaint	No. of Patients	
Skull deformity	98	(68.6%)
Proptosis	38	(26.6%)
Microcephalus	32	(22.4%)
Visual disturbance	23	(16.1%)
Headache	16	(11.2%)
Seizure	6	(4.2%)

Table III: Neuroophthalmological Findings

	No. of Patients		
Vision	143		
Normal		115	(80.5%)
Decreased		28	(19.5%)
Fundoscopic Findings	143		
Papilledema		20	(14.0%)
Atrophy		24	(16.7%)
Normal		99	(69.3%)
Exophthalmos	63		
Slight		30	
Severe		33	

Table IV: Head Circumference and Shape

	No. of Patients		
Head Circumference	143		
Normal		107	(74.8%)
Increased		25	(17.5%)
Decreased		11	(7.7%)
Head Shape	143		
Normal		8	(5.6%)
Abnormal		135	(94.4%)

Table V: Suture Involvement

Involved Suture	No. of Patients
Three or more	76 (53.1%)
Only sagittal	28 (19.6%)
Bilateral coronal	11 (7.7%)
Coronal and sagittal	10 (7.0%)
Coronal and metopic	9 (6.3%)
Sagittal and metopic	6 (4.2%)
Only coronal	3 (2.1%)
Total	143 (100.0%)

The surgical techniques used are shown in Table VI. The most frequent complications were: wound infection (five patients), cerebrospinal fluid fistula (three patients), and leptomeningeal cyst formation (one patient). There were ten postoperative deaths due to hemodynamic imbalance (eight patients), sepsis (one patient), and bleeding diathesis (one patient).

Table VI: Surgical Procedures

	No. of Patients	
Suture Removal	131	(91.7%)
Only involved suture	23	(16.1%)
All sutures	108	(75.6%)
Suture removal + Orbital Decompression	34	(23.6%)
Linear craniectomy + Wrapping	12	(8.3%)

Primary craniosynostosis was observed in 142 patients and secondary craniosynostosis due to subdural hematoma in one.

Follow-up examinations revealed normal fundoscopic findings in 17 patients with preoperative papilledema. Although exophthalmos remained unchanged in six out of 52 patients, it disappeared in 43. Table VII shows the preoperative and follow-up findings of all patients.

A repeat operation was performed in nine patients because of resynostosis of the sutures. In eight of them, the sutures were removed, and a linear craniectomy was performed in one. One patient was operated on three times and the others twice.

Table VII: Findings at Follow-up

Preoperative Findings	No. of Patients*	Postoperative Findings	No. of Patients
Papilledema	17	Normal	15 (88.2%)
		Optic atrophy	2 (11.8%)
Proptosis	52	Decreased	5 (9.6%)
		Unchanged	6 (11.5%)
		Normal	41 (78.9%)
Skull Deformity	115	Complete Improvement	54 (46.9%)
		Marked Improvement	33 (28.7%)
		Slight Improvement	9 (7.8%)
		Unchanged	19 (16.6%)

* Number of cases whose follow-up examination was possible.

Findings of raised intracranial pressure were found in 52 patients (36.5%). Associated congenital anomalies consisted of corpus callosum agenesis (one patient), low posterior hair line and phimosis (one patient), clavicula abnormality (one patient), and encephalocele (one patient). One patient with cystic fibrosis also had craniosynostosis and patent ductus arteriosus. One patient had puberty precox. Four patients had hypertelorism and this condition was corrected in two of them by a combined surgical procedure performed by a team of neurosurgeons and plastic and reconstructive surgeons. One patient had a family history of hyperthyroidism. The mother of this patient had received propicyl therapy during her pregnancy. Hyperthyroidism could cause premature fusion of cranial sutures[11].

Discussion

Craniosynostosis occurs in approximately 1 per 1900-2000 live births[6,9-11,13,17], and there is a male preponderance accounting for 76 percent of patients, even though some authors report a slight female preponderance for brachycephaly[7,9,13,17]. In our series, the male/female ratio was 3.5/1.

Radiographic and/or clinical evidence of raised intracranial pressure was found in most of the reported cases[4-6,9,10,13]. It is documented that this pressure drops to normal after surgical treatment[3,4,6,10,13]. In our series, the incidence of increased intracranial pressure was 36.5 percent at the time of diagnosis, which was similar

to the series of Renier et al[4], but different from others reported in the literature[6,9,10,13]. It is important to look for signs of intracranial hypertension especially in older children in order to discover the premature suture closure.

Although some authors have emphasized the coexistence of craniosynostosis with hydrocephalus, there is no mention of hydrocephalus in large series such as that of Schillito and Matson[9,10,18]. We observed only one case of craniosynostosis associated with hydrocephalus. Seizure is rare in patients with craniosynostosis unless increased intracranial pressure is present[6]. The incidence of seizure becomes higher with prolonged duration of increased intracranial pressure. In our series, seizure was found in six patients (4.2%), all of whom also had elevated intracranial pressure.

Papilledema, and optic atrophy which have been reported as rare entities in many series[6,9,10,15], were observed in 44 out of 143 of our patients. After surgical intervention, we observed that papilledema disappeared more in the younger patients than in the older ones. Myriathopoulos[17] proposed that papilledema only occurs when more than one suture is synostozed. We found in our series of 122 patients, that three or more sutures were involved. According to Winston[6], proptosis is associated only with coronal synostosis, which is in contrast to observations, since proptosis is seen in all types of suture closures. Forty-two out of our 63 cases with proptosis had three or more suture closures. Findings at follow-up examination revealed that papilledema disappeared in 15 out of 17 patients (88.2%), and proptosis was no longer evident in 41 out of 52 patients (78.8%). These results are similar to those reported in the literature[6,9,10,13]. Therefore, orbital decompression should be added to removal of sutures in patients with proptosis.

Surgical treatment is undertaken to decrease intracranial hypertension and prevent an abnormal appearance[6,9-11,13]. There are therefore only two surgical indications: increased intracranial hypertension and reconstruction of a cranial deformity. The general strategy regarding surgery is to perform a linear craniectomy. A parasagittal linear craniectomy is one of the preferred surgical techniques used to preserve the sagittal sinus and prevent resynostosis[6,8,9,10,13]. In our study, a parasagittal linear craniectomy, with or without wrapping, was performed in 12 out of 143 patients and resynostosis was observed in one (8.6%). Some authors have advocated several different approaches to prevent resynostosis such as wrapping, application of chemical solutions, and suturectomy[8,9,11,13]. Ingraham et al[8] was the first to advance the opinion that wrapping material was a foreign substance and could be a source of infection in the body. Therefore, wrapping is not necessary if an appropriate craniectomy is performed. In our series, wrapping was performed in 12 patients, and infection was observed in four of them (33.3%).

Suture removal was the preferred procedure in our series. The rate of reoperation was 6.2 percent. In these cases, operative techniques consisted of suture removal (8 patients) and parasagittal linear craniectomy (one patient). It has been reported that the rate of repeat operations ranges from 4 to 40 percent[8-10]. In our series, this ratio was lower than the reports in the literature. It is our opinion that this low ratio is due to wide suture removal performed with a median measuring two cm in width.

In conclusion, we believe that surgical intervention must be performed as early as possible, not only to prevent deformity and improve the appearance of the child, but also to prevent damage to the brain due to increased intracranial pressure. We are also of the opinion that suture removal should be preferred as the surgical technique used to correct this abnormality.

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