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Surgery of Cranial Deformity Following Ventricular Shunting: A Multicenter Study

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ABSTRACT

AIM: To review the cases of craniosynostosis secondary to ventricular shunting procedure.

MATERIAL and METHODS: We retrospectively evaluated the medical records of all pediatric patients with hydrocephalus who were treated with ventriculoperitoneal shunt procedure between the years 2017 and 2021 at the Selcuk University, Ankara University, and Bursa Uludag University.

RESULTS: Twenty-one patients were included in the study. The median age at the time of insertion of ventriculoperitoneal shunt for hydrocephalus was 8.1 (range, 1–22) months. Seven patients were shunted because of congenital hydrocephalus. The mean time to development of secondary synostosis was 8.8 (range, 1–36) months. Plagiocephaly was the most common type of secondary synostosis. While shunt revision was performed in 16 patients, cranial vault expansion surgery was performed in 5 patients.

CONCLUSION: Slit ventricle syndrome is a frequent condition at shunted patients, but there is no consensus on identifying patients who require treatment. Using programmable or high-pressure valves, performing cranial vault modeling are possible treatment modalities. Increased awareness of this condition in follow-up may allow early diagnosis and intervention and prevent it from evolving into more serious deformities.

KEYWORDS: Cranial deformity, Ventricular shunting, Craniosynostosis

ABBREVIATIONS: ICP: Intracranial pressure, **SVS:** Slit-ventricle syndrome, **CSF:** Cerebrospinal fluid

INTRODUCTION

Ventricular shunt surgery is frequently performed for the treatment of hydrocephalus associated with high intracranial pressure (ICP) (3,6). The aim is to normalize the ICP and protect the neural tissue from the effects of high pressure. The rationale for hydrocephalus management is to relieve the ICP. However, ICP is also the main force driving the skull growth and expansion, thus counteracting the craniosynostotic process to a certain degree. Therefore, the timing and the type of treatment for hydrocephalus in this context should be properly evaluated in relationship to the surgical expansion of the skull (9).

Ventricular shunt-induced craniosynostosis is a widely recognized cause of secondary craniosynostosis. It was first reported by Strenger, and since then, ventricular shunt surgery has been identified as a common cause of craniosynostosis

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(20), with the reproted incidence ranging between 1% and 10% (7). Some studies have recognized the inherent complexity of repairing the skull abnormality in these patients, with treatment modalities being sought to preserve the skull shape (2,9,13,17). However, the most important goal in craniosynostosis surgery is to decrease the ICP by increasing the intracranial volume (14,15). In surgery for craniosynostosis secondary to ventricular shunt, the goal is not to lower the ICP as these patients are not at risk of increased ICP. Moreover, some authors are not in favor of surgical correction in these patients, arguing that the surgical risks are unacceptable in patients who are not at risk of increased ICP (8). In the contemporary literature, there is a paucity of information on the management of this complication. In this study, we aimed to contribute to the literature by reviewing cases related to craniosynostosis secondary to ventricular shunt.

MATERIAL and METHODS

This study was conducted with the approval of the ethics committee of Uludag University, Faculty of Medicine, decision no 2022-16/31, dated 08/11/2022. We retrospectively evaluated the medical records of all pediatric patients with hydrocephalus who were treated with ventriculoperitoneal shunt procedure between the years 2017 and 2021 at tree academic tertiary care units in Turkey, namely, Selcuk University, Ankara University, and Bursa Uludag University.

The demographic characteristics of the patients, coexistence of spina bifida, type of craniosynostosis, time elapsed between ventriculoperitoneal shunt surgery and detection of secondary synostosis, treatment of secondary synostosis, and follow-up period were recorded.

Patients with hydrocephalus who were treated with endoscopic third ventriculostomy and patients who developed secondary synostosis but who were only followed-up and not treated were excluded from the study.

RESULTS

Twenty-one patients (14 [66.6%] male and 7 [33.3%] female) were included in the study (Table I). The median age at the time of insertion of ventriculoperitoneal shunt for hydrocephalus was 8.1 (range, 1–22) months. Seven patients were shunted because of congenital hydrocephalus. Six patients were operated for meningomyelocele and hydrocephalus; in 6 patients, the shunt was placed due to hydrocephalus secondary to intraventricular hemorrhage. Two patients were shunted due to infection. The mean time to development of secondary synostosis was 8,8 (range, 1–36) months. Plagiocephaly was the most common type of secondary synostosis (Figure 1). Lambdoid suture was the most affected suture. Two patients had pansynostosis. The average number of shunt revisions in this series was 1.75 (range, 1–7). While shunt revision was performed in 16 patients, cranial vault expansion surgery was performed in 5 patients (Figure 2). In one patient, both cranial vault expansion surgery and shunt valve replacement were performed. In 10 patients, the shunt was replaced with an adjustable shunt, while in 10 patients it was replaced with a higher pressure shunt. The shunt of 1 patient was removed and the patient was followed-up. The mean follow-up duration in our series was 32.1 months. There were no syndromic patients in our study.

DISCUSSION

Craniosynostosis due to ventriculoperitoneal shunt is a serious side effect whose mechanism is not fully understood and its treatment is challenging. Excessive thickening of the cranial vault due to loss of tension across the dura and suture lines is believed to be the underlying mechanism. The reported incidence of craniosynostosis is 1%, but with the increase in the number of ventriculoperitoneal shunt surgeries in recent years, more patients have been reported (16). In the literature, this condition has been associated with slit-ventricle syndrome (SVS). Slit-ventricle syndrome is an entity that affects 1%–5% of shunted patients and only 1%–5% of patients with radiographically small ventricles are symptomatic (10).

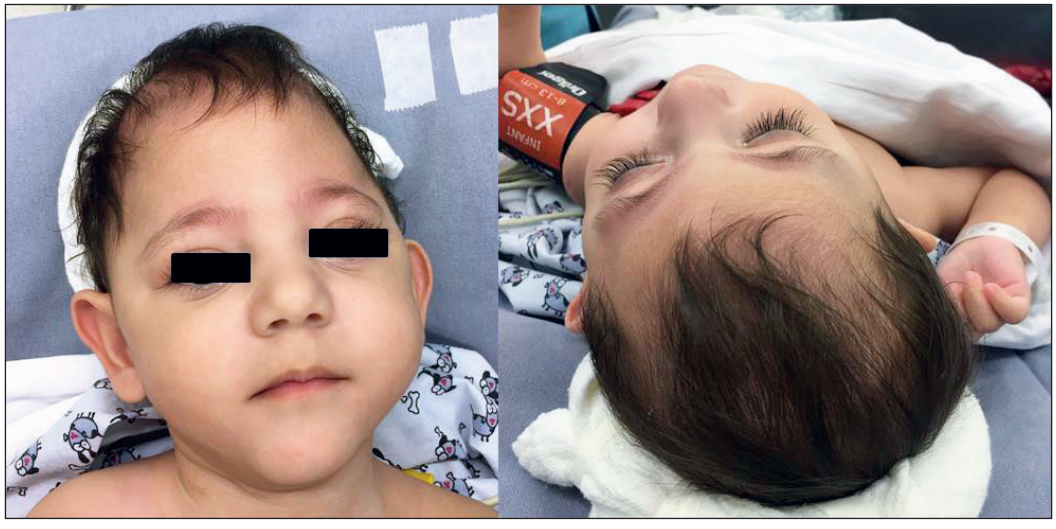

Figure 1: A patient with secondary craniosynostosis due to ventricular shunt who will undergo a surgical intervention.

Table I: Summary Clinical Characteristics and Treatment Details of Patients

Patient #	Sex	Age at ventricular shunting (months)	Etiology of Hydrocephalus	Time interval to the development of secondary synostosis (months)	Type of craniosynostosis	Number of shunt revisions	Treatment	Follow-up (months)
1	F	6	Meningomyelocele	6	Plagiocephaly	2	Shunt revision	20
2	F	2	Congenital	3	Plagiocephaly	2	Shunt revision	9
3	M	3	Congenital	3	Plagiocephaly	1	Shunt revision	3
4	M	9	Congenital	7	Plagiocephaly	2	Shunt revision	26
5	M	18	Congenital	3	Brachycephaly		Calvarial vault expansion	120
6	M	22	Meningomyelocele	22	Plagiocephaly	7	Shunt revision	80
7	M	12	Meningitis	27	Plagiocephaly	2	Shunt revision	34
8	M	22	Congenital	9	Plagiocephaly	3	Shunt revision	16
9	F	15	Intraventricular hemorrhage	36	Scaphocephaly	1	Shunt revision	72
10	M	13	Intraventricular hemorrhage	9	Plagiocephaly	1	Shunt revision	78
11	M	6	Meningitis	6	Plagiocephaly	1	Shunt revision	6
12	F	18	Intraventricular hemorrhage	16	Plagiocephaly	3	Shunt revision	36
13	M	4	Intraventricular hemorrhage	6	Scaphocephaly	1	Shunt revision	20
14	M	5	Congenital	4	Coronal + sagittal synostosis	1	Calvarial vault expansion	36
15	M	3	Meningomyelocele	6	Scaphocephaly	1	Calvarial vault expansion	28
16	F	2	Intraventricular hemorrhage	4	Coronal synostosis	2	Shunt revision	36
17	M	4	Meningomyelocele	4	Scaphocephaly	1	Calvarial vault expansion	28
18	F	3	Intraventricular hemorrhage	8	Coronal + sagittal synostosis	1	Calvarial vault expansion + shunt revision	12
19	M	1	Congenital	1	Pansynostosis	1	Shunt revision	12
20	F	2	Meningomyelocele	1	Pansynostosis	1	Shunt revision	1
21	M	1	Meningomyelocele	4	Plagiocephaly	1	Shunt revision	3

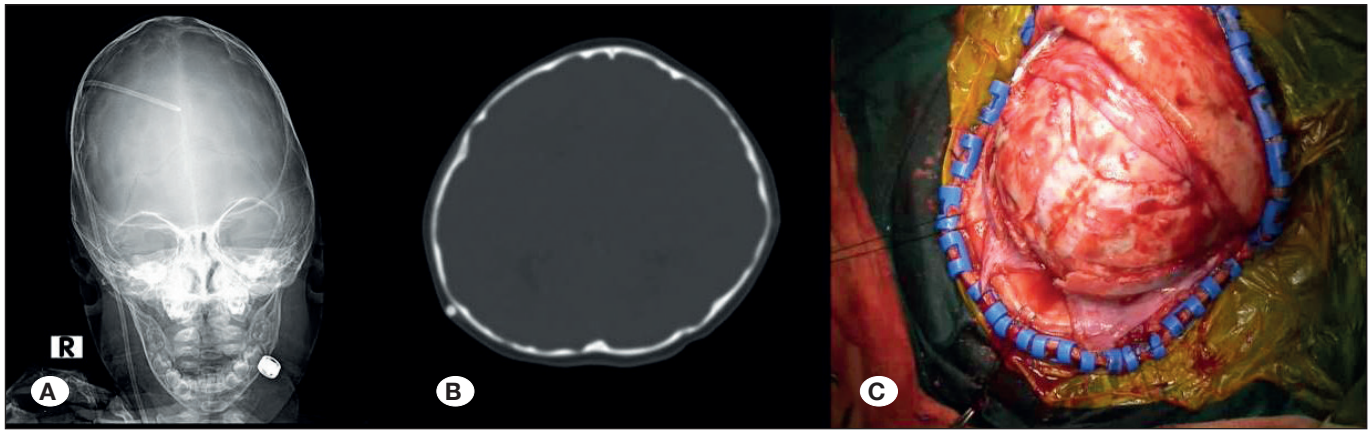

Figure 2: A) Copper-beaten appearance on plain radiography; **B)** scalloping of the inner calvarium on axial computed tomography scan; and **C)** intraoperative photograph showing osteotomies during cranial vault expansion.

The revision rate is especially high for shunts inserted before the age of 6 months, which increases the rate of secondary synostosis (1). In our series, the mean age at insertion of ventriculoperitoneal shunt was 8.1 months. Shunts were placed at an earlier age in 7 patients with congenital hydrocephalus.

Secondary synostosis generally affects the patients who are shunted before the age of 6 months (17). The reported timing of operation has also varied in the literature. Abnormal head shape can be recognized and diagnosed at follow-up from 1 to 7 months after shunting. In the literature, there is no consensus on the optimal time for the second surgery. In our series, the time to development of secondary synostosis in patients was 8.8 months. Compared to the literature, the delayed detection of secondary synostosis may be attributable to the non-inclusion of asymptomatic patients in our study. Sun et al. reported that occipital cranial deformities may be encountered especially in posteriorly inserted shunts (19). We could not detect a relationship between shunt insertion site and secondary synostosis in our series. This may be due to the relatively small number of patients.

In the literature, headache, nausea, vomiting, and altered mental status are frequent indications for shunt revisions (10). In our series, surgery was decided for the patients because of detection of skull deformity during outpatient follow-up. There was no neurological deficit in our series.

There is no clear consensus on the optimal time-point for the installation of shunts. In the series reported by Golinka et al., 10 out of 13 patients were shunted from parietal of occipital (10). This may also be one of the causes of secondary synostosis, as caregivers of the patients typically do not want to lay the patient on the shunt valve.

The sagittal suture is the most affected suture in SVS (21). In our series, plagiocephaly was the most common skull deformity, which may be attributable to the fact that caregivers of the patients tend to tilt the patient head to the other side of the shunt valve after the insertion of the ventriculoperitoneal shunt.

Spring-assisted cranioplasty, posterior cranial vault distraction, shunt revisions, and cranial vault expansion are the possible treatment modalities (4,5,12,18). Yan et al. mentioned that tension on the sutures and dura are the major mechanism of skull growth and spring assisted distraction osteogenesis can keep the force on the dura under the craniotomy area for minimizing the premature fusion (22). Weinzwig et al. reviewed 12 SVS cases that were treated by cranial vault expansion (21). Seven of the 12 patients underwent shunt revisions following cranial vault expansion surgery. Doorenbosch et al. reported 8 patients with secondary craniosynostosis, all of whom were treated by cranial vault remodeling together with insertion of programmable shunt valve (7). Habibi et al. treated 16 shunted patients with intractable headache during 10 years period and performed a combination of a hinge multiple-strut decompressive craniectomy and internal cranial flap thinning by drill (11). In our series, 5 patients were treated with cranial vault expansion surgery, but only one patient had the shunt valve replaced with an adjustable one.

Golinko et al. found that choice of an adjustable or non-adjustable shunt had no remarkable effect on the development of secondary synostosis (10). Generally, neurosurgeons prefer non-adjustable shunts in the initial surgery. In our country, the social security system covers the cost of fixed-pressure shunts for first shunt surgery in the pediatric age group.

Cranial vault expansion surgery is the preferred method for the treatment of secondary synostosis due to shunt (10). In our series, shunt revision was applied more frequently. Only 5 patients required cranial vault expansion. If the sutures of the patient with secondary synostosis were open, the valve of the shunt was revised, while cranial vault expansion was performed in patients with premature fusion of the sutures. More frequent postoperative follow-up of patients in the outpatient clinic may help avoid procedures such as cranial vault expansion.

The most notable limitation of this study is its retrospective nature and the small sample size. The heterogeneity with respect to shunt preference, timing of reoperation, reason for shunting, and decision differences between the 3

surgeons are the other main limitations. There may also be an element of selection bias as we excluded patients who were followed-up in the outpatient clinic but were not treated. Cosmetic evaluation of patients is also lacking in the study. Moreover, there were some missing data such as serial head circumference and ICP measurements.

CONCLUSION

It is challenging to determine the appropriate shunt pressure in pediatric patients, especially in infants. Despite optimal shunt function and cerebrospinal fluid (CSF) flow dynamics, cranial suture pathology may occur secondary to abnormality of the brain and, therefore, the underlying dura. SVS is a frequent occurrence, but there is no consensus on identifying patients who require treatment. Using programmable or high-pressure valves may be an alternative, but increased awareness of this condition in follow-up may allow early diagnosis and intervention and prevent it from evolving into more serious deformities.

AUTHORSHIP CONTRIBUTION

Study conception and design: BG, MOT,
Data collection: BG, MZ, TMG
Analysis and interpretation of results: GK, HK
Draft manuscript preparation: BG, MOT
Critical revision of the article: HK
All authors (BG, MOT, MZ, TMG, GK, HK) reviewed the results and approved the final version of the manuscript.

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