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Original Investigation

Timing of Shunt Insertion in Children with Neural Tube Defects and Hydrocephalus: A Clinical Study

Onur OZGURAL[1], Gokmen KAHILOGULLARI[1], Ihsan DOGAN[1], Umit EROGLU[1], Fatih YAKAR[2], Mustafa Cemil KILINC[1], Emre Yagiz SAYACI[1], Mustafa Agahan UNLU[1]

[1]Department of Neurosurgery, Ankara University School of Medicine, Ankara,Turkey
[2]Department of Neurosurgery, Pamukkale University School of Medicine, Denizli, Turkey

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Corresponding author: Onur OZGURAL ✉ onurozgural@yahoo.com

ABSTRACT

AIM: To define the optimal time of shunt insertion in patients with neural tube defects and hydrocephalus.

MATERIAL and METHODS: In total, 71 patients who underwent operation for neural tube defects and hydrocephalus were retrospectively evaluated between 2012 and 2018. The first group comprised 43 patients who underwent operation at different times (in 10 days after the repair of defect), and the second group comprised 28 patients who underwent operation at the same time. Ruptured and unruptured sacs were immediately considered and operated within 72 hours.

RESULTS: In the first group, 43 patients underwent operation for neural tube defect after birth. Ventriculoperitoneal shunt insertion was performed 10 days after wound healing. Five (11.6%) patients were diagnosed with meningitis on follow-up. Shunt infection or meningitis was not observed on follow-up in the second group, which comprised patients who underwent operation at the same time.

CONCLUSION: The lowest complication rate existed in hydrocephalus management when shunt insertion and myelomeningocele repair procedures were performed at the same time.

KEYWORDS: Hydrocephalus, Neural tube defect, Shunt infection, Ventriculoperitoneal shunt

INTRODUCTION

Neural tube defects (NTD), classified as open or closed, arise in 1/1000 pregnancies (4). Meningeal herniation with myelomeningocele (MMC) or without meningocele (MC) neural tissue is called open NTD in the absence of an overlying skin (6). Hydrocephalus is usually accompanied by MMC (83%–93%) (1), and shunt requirement arises in 84%–89% of these patients (12). The literature generally suggests early NTD repair (21), but shunt timing in NTD patients remains controversial (3,17). On the basis of our experience, this study aimed to suggest a standardization of shunting time when there is coexistence of hydrocephalus and MMC.

MATERIAL and METHODS

In total, 71 MMC patients with hydrocephalus who underwent operation between 2012 and 2018 were retrospectively evaluated. Informed consent was obtained from the families of all individual participants included in the study. The research was reviewed and approved by an institutional review board. The patients were divided into two groups according to their shunt insertion time. The first group comprised patients who had hydrocephalus at birth, but ventriculoperitoneal shunt (VPS) insertion was performed at another time. In the second group of patients, hydrocephalus was radiologically detected and operated at the same time.

Onur OZGURAL : 0000-0003-0592-6139
Gokmen KAHILOGULLARI : 0000-0001-8137-0510
Ihsan DOGAN : 0000-0002-1985-719X
Umit EROGLU : 0000-0001-8623-071X
Fatih YAKAR : 0000-0001-7414-3766
Mustafa Cemil KILINC : 0000-0003-4058-6504
Emre Yagiz SAYACI : 0000-0002-9397-3834
Mustafa Agahan UNLU : 0000-0002-2039-8592

Neurological and general physical examinations were conducted and head circumference was measured every day. All patients were evaluated through fontanel ultrasonography, and cranial computed tomography (CT) was performed in the presence of hydrocephalus. All spinal segmentations were evaluated through magnetic resonance imaging (MRI) in all patients for coexisting pathologies. Patients who had multisystem diseases and weighed below 2.5 kg were excluded.

Modified closure technique of McLone and Naidich (14) was performed to close the MMC sacs, and all operations were performed by the same team. When single-session surgery was preferred, sac repair was performed first, followed by VPS insertion. Operations were performed under general anesthesia, and all unruptured sacs were operated under elective conditions.

An antibiotic was intravenously administered 30 minutes before the shunt surgery, and the same antibiotic was continued for 72 hours in all patients. VPS was the first-choice intervention of our institute for the management of hydrocephalus.

RESULTS

A total of 37 girls (52.1%) and 34 boys (47.9%) were included in the study. The mean age and mean weight were 6.5 ± 2.6 days and 3.1 kg, respectively. Mean follow-up time was 38 ± 10 months for Group 1 and 36 ± 6 months for Group 2. The locations of lesions were cervical in four (5.6%), thoracolumbar in 23 (32.3%), and lumbar in 44 (61.9%). Further, 29 sacs (40.8%) were ruptured, and cerebrospinal fluid (CSF) leak was observed in these patients. All ruptured sacs were located in the thoracolumbar and lumbar regions. There were 14 MC (19.7%) (Figure 1A, B) and 57 MMC (80.3%) cases. Gender, sac location/integrity, shunt infection, and mortality/morbidity in the postoperative period were evaluated (Table I).

The first group comprised 43 patients (60.5%). All shunt insertions were performed 10 days after sac repair. Two sacs (4.6%) were located in the cervical region, 19 (44.1%) in the thoracolumbar region, and 22 (51.3%) in the lumbar region (Figure 2A-E). Moreover, 27 sacs (62.7%) were intact, and 16 (37.3%) sacs were ruptured. Seventeen (39.5%) patients were neurologically intact, ten (23.3%) had paraparesis, and 16 (37.2%) were paraplegic. There was no additional neurological

Table I: Comparison of Two Groups with all Parameters

	Group 1	Group 2
Number of patients	43 (24 F, 19 M)	28 (13 F, 15 M)
Location	2 C 19 ThL 22 L	2 C 4 ThL 22 L
Ruptured/Unruptured sac	16 / 27	13/15
Neurological examination	17 intact 10 paraparesis 16 paraplegia	8 intact 7 paraparesis 13 paraplegia
MC/MMC	8/35	6/22
Shunt infection(Meningitis)	5	-
Mortality	1	-

***F:** Female, **M:** Male, **C:** Cervicale, **Th:** Thoracal, **ThL:** Thoracolumbar, **L:** Lumbar, **MC:** Meningocele, **MMC:** Meningomyelocele.*

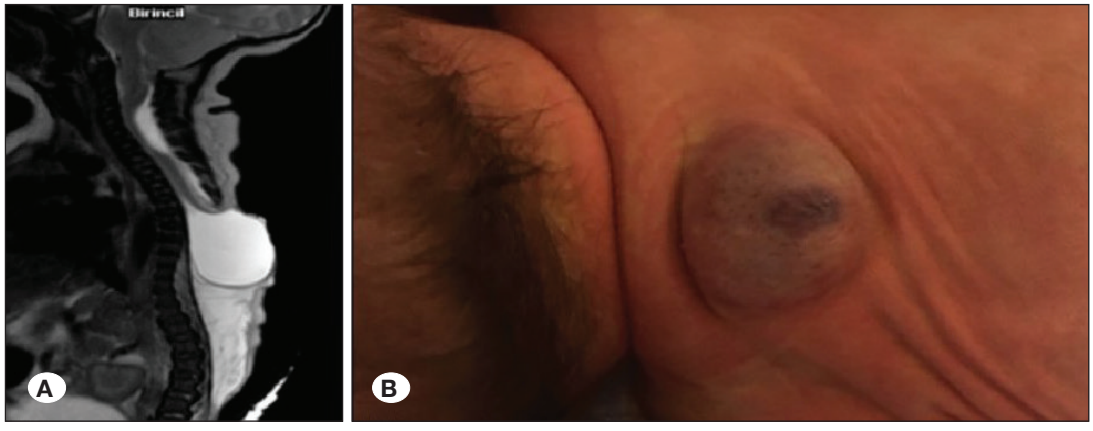

Figure 1: A thoracic meningocele. MRI revealed the meningocele sac **(A)**. The photograph of sac **(B)**.

deficit during the postoperative period. Five (11.6%) patients who had wound healing problem were resutured and antibiotic therapy was continued. Shunt infection was identified in five (11.6%) patients in 2 weeks and all shunts were revised in this group. One patient died due to aggressive meningitis 3 weeks after the first operation.

The second group comprised 28 (39.5%) patients. MMC repair and shunt insertion were performed at the same time. Two sacs (7.1%) were located in the cervical, four in the thoracolumbar (14.2%), and 22 (78.7%) in the lumbar regions. Eight (28.5%) patients were neurologically intact, seven (25%) had paraparesis, and 13 (46.5%) were paraplegic. Fifteen sacs (53.5%) were unruptured, and 13 (46.5%) were ruptured. No CSF leakage and shunt infection were observed in this group. Two patients (7.1%) had a mild wound healing problem that did not require surgical intervention. There was no mortality in this group during follow-up.

■ DISCUSSION

Although MMC is one of the most complex congenital malformations, the chances for prolonged life may be provided with an aggressive treatment approach (1,13). Hydrocephalus is a disease that has two forms, which are congenital and acquired, and could accompany MMC at a rate of approximately 80% in children (19,20). There is no consensus on shunting time; therefore, some authors recommend simultaneous surgery (5,11,15,21), whereas some authors adopt two-stage surgery (2,7,12,16). In our retrospective study, we compared the shunt insertion time in patients with repaired sacs.

In the literature, MMC frequency is reported to be higher in females than in males (18), and our study had similar results (female, 52.1%; male 47.9%). MMC is usually located in the lumbar region at a rate of approximately 60%–70%, which is similar to the observations in our study (61.9%).

Machado et al. (11) emphasized that single-session surgery might provide good wound healing of the sac and low infection rates because shunting reduces CSF pressure on the wound; a second surgery is then not necessary. It seems to be logical that early shunting procedure protects the brain from ventricular dilatation; some authors agree with this theory, whereas some disagree (8). In addition, there is no consensus on the surgical sequence. In our study, we found that performing the procedure at the same time was better; however, we performed sac repair first and then inserted the shunt, unlike Machado et al. (11), who aimed to decrease the pressure on the sac before repair. However, we did not want to take the risk of sac damage during shunt surgery (11); therefore, we repaired the sacs first and then inserted VPS.

Following MMC repair, CSF leakage is a major cause of morbidity (9). Margaron et al. (12) found that compared with

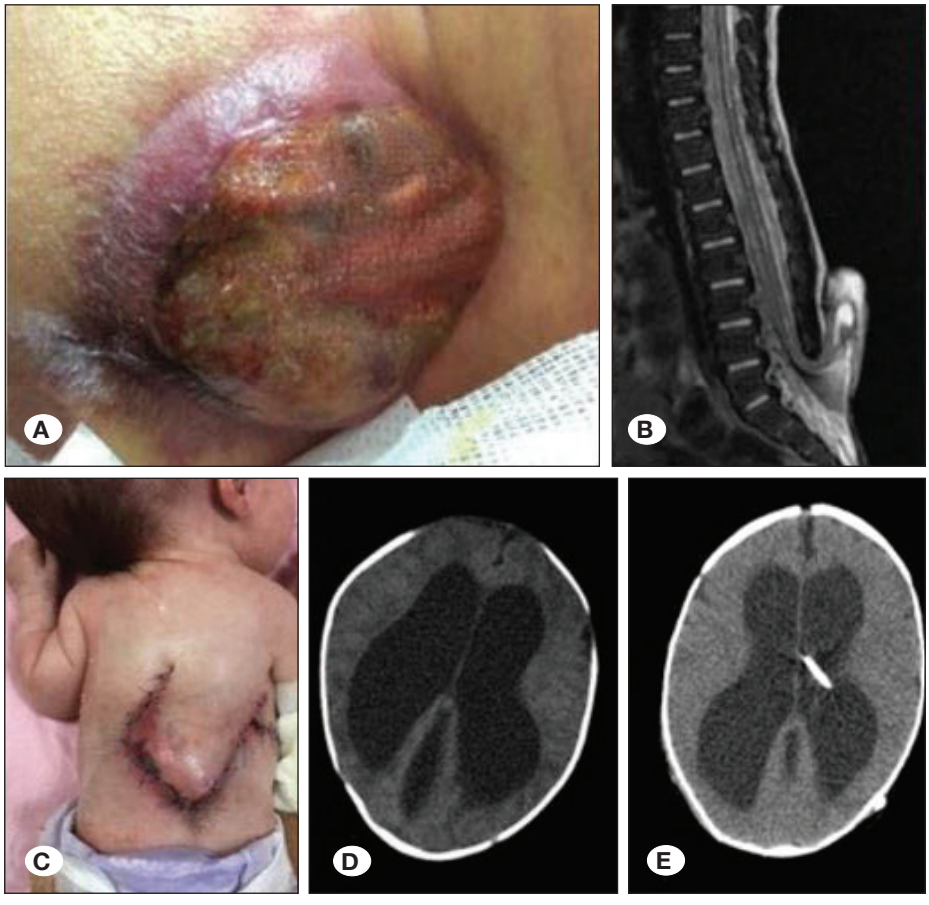

Figure 2: The photograph of the lumbar myelomeningocele sac **(A)**. MRI revealed myelomeningocele **(B)**. The postoperative photograph of sac wound **(C)**. CT revealed hydrocephalus **(D)**. CT after shunt operation **(E)**.

patients who underwent shunting 5 days after repair, patients who underwent shunting prior to sac repair simultaneously and 4 days after repair showed higher shunt infection rates; therefore, they suggested shunt surgery to be performed 5–10 days after MMC repair. Lee et al. (10) suggested that early simultaneous repair of MMC and VPS in patients with MMC and hydrocephalus not only decreases further brain damage but also eliminates the risks of wound break down, CSF leak, and secondary CSF infection. Conversely, Oktem et al. (16) evaluated 94 patients and compared the surgical results of the single-session and two-stage surgery groups. Their findings demonstrated higher wound infection, CSF leakage, and meningitis rates in the single-session surgery group than in the two-stage surgery group. Thus, they proposed that VPS insertion should be performed in a different session, especially for ruptured sacs, because it must be proven that there is no infection (16). This study is completely different from ours because our findings showed that single-session surgery prevented meningitis or any shunt infection. In single-session surgery, the pressure in the ventricular system after sac repair was lowered; therefore, CSF leakage rate decreased, and another surgery for newborns was not required. Gamache (7) suggested that the simultaneous MMC repair and VPS insertion may be performed in the first 36 hours of life, but after 48 hours, the infection rates increase; thus, external ventricular drainage (EVD) with MMC repair should be preferred. As a different approach, we operated patients whose sacs were ruptured in 72 hours under intravenous antibiotic. We did not perform EVD because it might cause the contamination of CSF.

The first group in our study comprised 17 (39.6%) patients with ruptured sacs. Antibiotic therapy was immediately initiated after birth, and the patients were operated within 72 hours. Five (11.6%) patients developed meningitis due to the ruptured sacs. Although antibiotic therapy was initiated right after birth, it did not prevent the development of infection in the first group. The second group comprised 14 (50%) patients with ruptured sacs. They were administered the same antibiotics at the same time as the first group. However, we did not observe meningitis in both groups. We believed that despite no real evidence, success in single-session surgery is related to a very low colonization. Oncel et al. (17) studied 30 MMC patients and found that performing surgery within 5 days provided lesser hospitalization and antibiotic therapy. Thus, the procedure described in this study correlates with that in our study, especially in case of patients with ruptured sacs as an early intervention.

CONCLUSION

Literature review had no consensus on VPS timing for MMC patients. The results of our study supported that simultaneous MMC repair and VPS insertion in the hydrocephalic newborns, especially those with unruptured sacs and those who are appropriate for general anesthesia, decreases meningitis rates. However, a consensus will be possible on the basis of studies involving larger case series.

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