Since January 2020 Elsevier has created a COVID-19 resource centre with free information in English and Mandarin on the novel coronavirus COVID-19. The COVID-19 resource centre is hosted on Elsevier Connect, the company's public news and information website.

Shunt Valve Rupture in Ventriculoperitoneal Shunt Failure

Mustafa Güdük[1], Ahmet Akbaş[2], Mürüvvet Ayten Tüzünalp[3], Gürkan Berikol[2], Murat Şakir Ekşi[1]

Key words
- Case report
- Hydrocephalus
- Optical coherence tomography
- Shunt failure
- Ventriculoperitoneal shunt

Abbreviations and Acronyms
CSF: Cerebrospinal fluid
VP: Ventriculoperitoneal
VPS: Ventriculoperitoneal shunt

From the [1]Department of Neurosurgery, Acıbadem Mehmet Ali Aydınlar University, School of Medicine, İstanbul; [2]Neurosurgery Clinic, Taksim Education and Research Hospital, İstanbul; and [3]Ophthalmology Clinic, Acıbadem Healthcare Group, Acıbadem Altunizade Hospital, İstanbul, Turkey

To whom correspondence should be addressed: Murat Şakir Ekşi, M.D. [E-mail: muratsakireksi@gmail.com]

Citation: World Neurosurg. (2021) 145:73-76. https://doi.org/10.1016/j.wneu.2020.08.225

Journal homepage: www.journals.elsevier.com/world-neurosurgery

Available online: www.sciencedirect.com

BACKGROUND: **Shunt complications are common despite advances in surgical techniques and shunting technology. Proximal and/or distal catheter malfunctions are detected in pediatric and adult patients. However, valve dysfunction is rare in such cases.**

CASE DESCRIPTION: **A 24-year-old woman presented with a history of ventriculostomy and ventriculoperitoneal shunt (VPS) secondary to hydrocephalus concomitant with Dandy-Walker syndrome. She has had undulant headache and vision loss episodes in both eyes for 15 days. Her VPS valve was normal when manually checked, and the VPS was observed as intact on x-ray and computed tomography scan. She had high-grade papilledema in both eyes with an optical coherence tomography scan value of 55/99. Lumbar puncture was performed. Cerebrospinal fluid opening pressure was 560 mm H_2O under sedation. VPS exploration surgery was performed. There was a tiny defect over the shunt valve from where clear cerebrospinal fluid was leaking. We revised the old VPS valve with a new valve of 1.5 regular pressure. Her vision improved shortly after the surgery.**

CONCLUSIONS: **This case is a very rare example of shunt valve dysfunction that required further investigation and a new valve replacement even though the preoperative imaging was normal.**

INTRODUCTION

Hydrocephalus is the accumulation of excessive cerebrospinal fluid (CSF) that is due to overproduction, passage obstruction, or lack of absorption problems.[1] The prevalence of hydrocephalus has been reported as 85 persons per 100,000 people in the United States.[1-3] Shunting of CSF is an essential procedure in the treatment of hydrocephalus. The most common route for shunting of CSF is the ventriculoperitoneal (VP) route.[4] Shunt complications are common despite advances in surgical techniques and shunting technology. Shunt failure secondary to proximal (ventricular) or distal (peritoneal) catheter malfunction is common in pediatric and adult patients. So far, however, reports about valve rupture are rare in the English literature. In this case report, we describe a patient who had previously been operated on for hydrocephalus and presented with signs and symptoms of shunt dysfunction despite having radiologic findings of an intact shunt. We also present our diagnostic algorithm for the case.

CASE DESCRIPTION

A 24-year-old woman was admitted to our clinic with undulant headache and vision loss in both eyes. She was alert and cooperative with no additional neurological deficit. Her posterior fossa had been decompressed at the age of 3 years for Dandy-Walker syndrome. She had undergone surgery for hydrocephalus via endoscopic third ventriculostomy and VP shunting at 9 years of age. The distal catheter of the ventriculoperitoneal shunt (VPS) had been revised when she was 14 years old. Since then, she had been very active and alert. Her recent symptoms had begun 15 days ago. Her neurological findings were completely normal except for bilateral papilledema. Her VPS valve was manually checked and observed to be working. There was no sense of fluctuation during palpation of the shunt valve. Plain radiographs were used for tracing the VPS. The VPS was intact on plain radiographs. Cranial computed tomography scan showed no hydrocephalus with the proximal catheter of the VPS within the left ventricle (Figure 1A–C). Optical coherence tomography scan depicted high-grade papilledema with value of 55/99. We performed lumbar puncture under sedation, and her CSF opening pressure was 560 mm H_2O. VPS exploration surgery was scheduled as soon as her test result for 2019 novel coronavirus disease (COVID-19) infection was obtained as negative. Intraoperatively, we observed clear CSF leaking through a tiny defect on the valve of the VPS (Figure 2). We revised the ruptured valve with a new valve of 1.5 regular pressure. Her vision improved shortly after the surgery. Postoperative control computed tomography depicted the ventricular catheter was in place (Figure 1D).

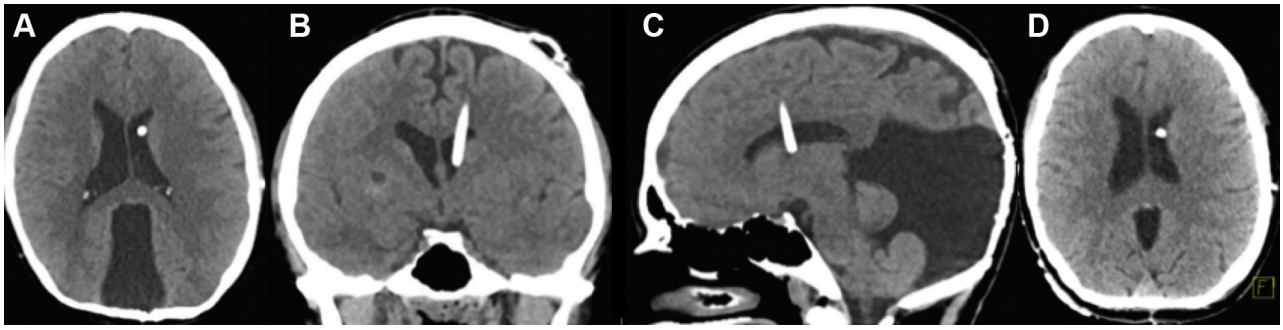

Figure 1. (**A**–**D**) Preoperative (**A**–**C**) and postoperative (**D**) computed tomography images depict the proximal catheter of the ventriculoperitoneal shunt in place.

DISCUSSION

Shunting for hydrocephalus has been in use for >50 years.[1] There are different routes of shunting, including VP, ventriculopleural, and ventriculoatrial. The VPS is the most commonly used shunt in selected patients with hydrocephalus.[1,4,5] The VPS has been modified over the years. However, the current VPS comprises 3 parts: proximal (ventricular) catheter, pressure-sensitive valve with a reservoir, and distal (peritoneal) catheter. A pressure-sensitive valve could be fixed or adjustable.[5] Although VP shunting is well known as one of the oldest procedures in neurosurgery practice, complication rates of VP shunting are 25%–60%.[1]

The most common complications of VP shunting are malfunction and infection, which require further surgical procedures. Shunt failure resulting from proximal or distal catheter malfunctions has been presented in pediatric and adult patients. CSF drainage may be excessive or scant secondary to different types of shunt failures.[6] Among the causes of shunt failure, diagnosis of rare causes, such as shunt valve rupture, may be difficult and overlooked. In such cases, fibrous tissue might have surrounded the dysfunctional shunt part. CSF drainage might still continue through the fibrous tissue even in cases of a broken shunt.[7] Valve failures owing to fracture of the system have rarely been reported in the literature (**Table 1**).[8-11] Woerdeman and Cochrane[10] performed a postmarketing review of U.S. and Canadian databases. They found 58 cases of punctures, cuts, and tears of silicone housing for the SiphonGuard-integrated CODMAN HAKIM Precision (Integra LifeScience, Princeton, New Jersey, USA) valves reported from the United States. Only 1 case of silicone housing separation was found from Canadian database. The problem related to this was that none of the failed valves had been made available for the corresponding manufacturer to make a further analysis; thus, shunt valve ruptures had been attributed to operator-inflicted cuts, handling during implantation or explantation, and trauma to the valve by the patient.[9,10]

The present case was unique, as valve failure had occurred 15 years after the last shunt revision surgery (including the valve itself); this time period is too long to explain any operator-related injury of the system. Besides, the patient had not experienced a recent head trauma. There was no calcification surrounding the shunt valve. The most probable cause in the present case could be degeneration of the shunt valve itself after such a long period since its first implantation. Therefore, necessary handling of the shunt device

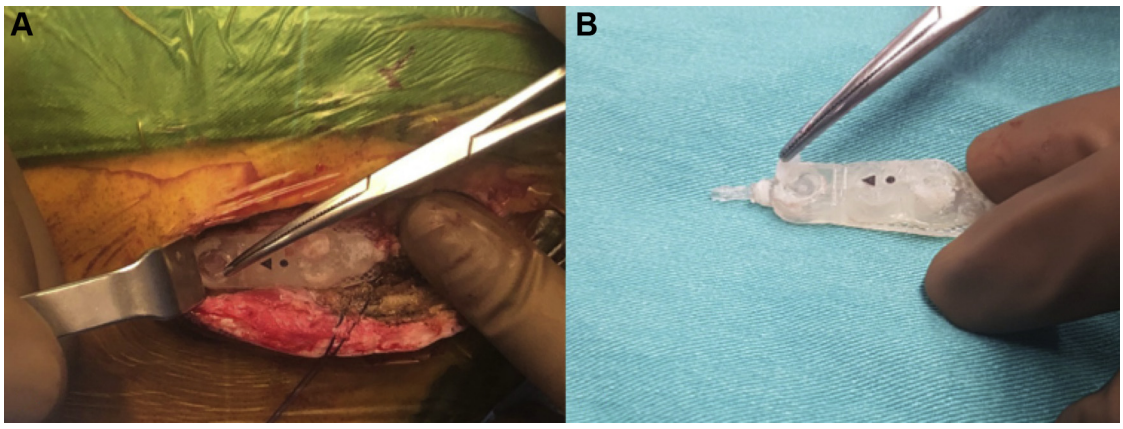

Figure 2. (**A** and **B**) Intraoperative finding of shunt valve rupture.

Table 1. Reported Cases with Shunt Valve Defect in the Literature

Author, Year	Age (years)/Sex	Primary Reason for Shunt Insertion	Number of Shunt Revisions	Time of Dysfunction Since Last Shunt Surgery	Clinical Findings	Imaging Findings	Defect	Defective Shunt Valve Type	Outcome
Hellbusch, 1996[8]	14/M	Aqueductal stenosis	NA (multiple times)	NA	Headache, nausea	Moderately increased ventricular size	Valve fracture	Holter	Good
	11/M	Hydrocephalus secondary to myelomeningocele	NA (multiple times)	NA	Headache, diplopia, papilledema	Mild to moderately increased ventricular size	Plastic of Cordis shunt valve fractured	Cordis	Further revision for ventricular catheter obstruction, then good recovery
	14/M	NA	None	11.5 years	Intermittent headache, decline in school peformance	Increased ventricular size	NA	Holter	Good
Okazaki et al., 2005[9]	7/M	Hydrocephalus secondary to intraventricular hemorrhage	0	7 years	Headache following blunt head trauma	None	Ruptured valve	CODMAN HAKIM	Good
Woerdeman and Cochrane, 2014[10]	12/F	Communicating hydrocephalus	7	6 weeks	NA	Increased ventricular size	Fracture of silicone housing with separation of SiphonGuard from valve	CODMAN HAKIM Precision	NA
	15/F	Posthemorrhagic nonobstructive hydrocephalus	5	9 months	Headache, vomiting	Tangential view of shunt series suggested misalignment of components	Silicone housing for SiphonGuard was fractured	CODMAN HAKIM Precision	NA
Amirjamshidi et al., 2015[11]	31/F	Posterior fossa arachnoid cyst (cystoperitoneal shunt)	0	6 months	Headache, vertigo, blurred vision for 6 months, papilledema, limited visual field, decreased visual acuity	Increased ventricular size and posterior fossa arachnoid cyst	Fractured inlet connector and valve	Fuji (connecting tube of flat bottom flushing device of cystoperitoneal shunt)	Good
Güdük et al., 2020 (present case)	24/F	Hydrocephalus with Dandy-Walker syndrome	2	10 years (15 years since revision of valve)	Headache, vision loss, pailledema	None (normal ventricular size)	Ruptured valve	Medtronic	Good

Cordis (Hialeah, Florida, USA); CODMAN HAKIM Precision (Integra LifeScience, Princeton, New Jersey, USA); Fuji (Bunkyo City, Tokyo, Japan); Medtronic (Minneapolis, Minnesota, USA); Delta valve (Medtronic, Minneapolis, Minnesota, USA).
M, male; NA, not available; F, female.

during surgery and careful lifelong surveillance of patients would still not be enough to prevent similar cases in the future. More collaboration between surgeons, patients, and manufacturers is necessary to understand the failure mechanism of valve rupture and to take appropriate actions to prevent similar adverse events in the future.

In this report, we describe a patient who had been operated on for hydrocephalus previously and presented with signs and symptoms of shunt dysfunction with intact radiologic shunt patency. The difficulty was in detecting the tiny rupture on the VPS valve using current imaging. Her symptoms and the presence of papilledema in both eyes led us to further analyze the patient with lumbar puncture under sedation, which revealed a very high opening CSF pressure.

CONCLUSIONS

The present case is a very rare example of shunt valve dysfunction requiring further investigation techniques and new valve replacement. Despite normal radiologic findings, further investigation with optical coherence tomography and lumbar puncture should be considered when patients are symptomatic for hydrocephalus. Shunt exploration should be done when such tests are suggestive for shunt dysfunction. Further collaboration between surgeons, patients, and manufacturers is needed to understand the mechanism behind such cases and to prevent future adverse events.

REFERENCES

1. Broggi M, Zattra CM, Schiariti M, et al. Diagnosis of ventriculoperitoneal shunt malfunction: a practical algorithm. *World Neurosurg*. 2020;137:e479-e486.

2. Bondurant CP, Jimenez DF. Epidemiology of cerebrospinal fluid shunting. *Pediatr Neurosurg*. 1995;23:254-258 [discussion: 259].

3. Isaacs AM, Riva-Cambrin J, Yavin D, et al. Age-specific global epidemiology of hydrocephalus: systematic review, metanalysis and global birth surveillance. *PLoS One*. 2018;13:e0204926.

4. Reddy GK, Bollam P, Shi R, Guthikonda B, Nanda A. Management of adult hydrocephalus with ventriculoperitoneal shunts: long-term single-institution experience. *Neurosurgery*. 2011;69:774-780 [discussion: 780-781].

5. Park DB, Hill JG, Thacker PG, et al. The role of limited head computed tomography in the evaluation of pediatric ventriculoperitoneal shunt malfunction. *Pediatr Emerg Care*. 2016;32:585-589.

6. Kestle JR, Garton HJ, Drake JM. Treatment of hydrocephalus with shunts. In: Albright AL, Pollack IF, Adelson PD, eds. *Principles and Practice of Pediatric Neurosurgery*. New York: Thieme; 1999:75-89.

7. Piatt JH Jr. Cerebrospinal fluid shunt failure: late is different from early. *Pediatr Neurosurg*. 1995;23:133-139.

8. Hellbusch LC. Cerebrospinal fluid shunt valve fracture with separation: report of three cases. *Childs Nerv Syst*. 1996;12:142-144.

9. Okazaki T, Oki S, Migita K, Kurisu K. A rare case of shunt malfunction attributable to a broken Codman-Hakim programmable shunt valve after a blow to the head. *Pediatr Neurosurg*. 2005;41:241-243.

10. Woerdeman PA, Cochrane DD. Disruption of silicone valve housing in a Codman Hakim Precision valve with integrated Siphonguard. *J Neurosurg Pediatr*. 2014;13:532-535.

11. Amirjamshidi A, Abbasioun K, Ghassemi B. Fractured inlet connecting tube of the flat bottom flushing device of a posterior fossa cystoperitoneal shunt. *Surg Neurol Int*. 2015;6:36.

Conflict of interest statement: The authors declare that the article content was composed in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

Received 23 June 2020; accepted 31 August 2020

Citation: World Neurosurg. (2021) 145:73-76.
https://doi.org/10.1016/j.wneu.2020.08.225

Journal homepage: ***www.journals.elsevier.com/world-neurosurgery***

Available online: ***www.sciencedirect.com***

1878-8750/$ - see front matter