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Shunt Valve Rupture in Ventriculoperitoneal Shunt Failure

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

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 ² 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 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 H₂O 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.
DISCUSSION

Shunting for hydrocephalus has been in use for >50 years. There are different routes of shunting, including VP, ventriculopleural, and ventriculoatrial. The VPS is the most commonly used shunt in selected patients with hydrocephalus. 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. Although VP shunting is well known as one of the oldest procedures in neurosurgery practice, complication rates of VP shunting are 25%–60%.

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. 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. Valve failures owing to fracture of the system have rarely been reported in the literature. Woerdeman and Cochrane 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 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.

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...
| 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[^5] | 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 |
| 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, papilledema | None (normal ventricular size) | Ruptured valve | Medtronic | Good |

[^5]: 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); 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.

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