

# Management and Outcome of Post-Infectious Multiloculated Hydrocephalus: A Case Series

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## Abstract

**Background and importance:** Infection following ventriculoperitoneal shunt (VPS) placement is a recognized complication, with variable incidence rates worldwide. Development of post-infectious multiloculated hydrocephalus (MLH) is likely if VPS infection is improperly managed, in turn affecting the prognosis. There is a lack of studies from Saudi Arabia regarding patients' functional outcome in relation to different variables. **Objectives:** To study the causative organisms, related variables and patient outcomes in MLH after VPS infection.

**Methods:** This case series is a retrospective chart review of pediatric patients diagnosed with hydrocephalus from 2011 to 2019. Patients were included if they were aged <18 years, had confirmed cerebrospinal fluid/blood infection with radiological evidence of MLH, and were regularly followed-up. Functional status score was used to evaluate the outcomes.

**Results:** A total of 150 patients underwent VPS insertion during the study period, of which 12 (8%) had postinfection MLH. The mean age at diagnosis and follow-up was 9 and 19 months, respectively. Ten patients developed MLH after their first VPS infection and one each developed MLH following the second and third VPS infections. Cerebrospinal fluid cultures mostly grew only single organisms (6/12), with *Staphylococcus* species being the most common. All patients underwent navigated endoscopic fenestration; nine patients required VPS placement and three required redo endoscopic fenestration surgery. All patients were developmentally delayed, with the majority (75%) having a functional status score of 6–10.


**Conclusion:** Development of MLH after VPS infection is debilitating and requires prompt treatment. Although the overall functional outcome is poor, evolving neuroendoscopic techniques with tailored preoperative planning may play a role in reducing the adverse effect of shunt multiplicity, shunt infections and the higher failure rate among patients with complex hydrocephalus.

**Keywords:** Case series, complex hydrocephalus, multiloculated hydrocephalus, neuroendoscopy, outcome, ventriculoperitoneal shunt infection

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**Submitted:** 31-Jan-2021 **Revised:** 05-Jun-2021 **Accepted:** 02-Aug-2021 **Published:** 31-Aug-2021

Access this article online	
Quick Response Code:	Website: www.sjmms.net
	DOI: 10.4103/sjmms.sjmms_85_21

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**How to cite this article:** Alojan AA, Alotaibi AR, Alalhareth HN, Alwadei AD, Ammar A. Management and outcome of post-infectious multiloculated hydrocephalus: A case series. Saudi J Med Med Sci 2021;9:261-6.

## INTRODUCTION

Multiloculated hydrocephalus (MLH), which is the compartmentalization of ventricles, is a complication that can appear following ventriculoperitoneal shunt (VPS) infection. MLH can manifest as a single or multiple fluid-filled cyst, involving one or more ventricles.<sup>[1]</sup> It is often perceived as secondary to cerebrospinal fluid (CSF) infection or germinal matrix hemorrhage in prematurity. The management paradigm of postinfectious MLH remains a challenge, especially in cases of multiple causative organisms. Psychomotor retardation is an overwhelming adverse effect in postinfectious MLH.

Currently, there is a lack of studies from Saudi Arabia regarding patients' functional outcome in relation to different variables. Accordingly, this case series was carried out to study the causative organisms, related variables and patient outcomes in MLH after VPS infection.

## METHODS

This is a retrospective chart review of pediatric patients who were diagnosed with hydrocephalus between 2011 and 2019 at King Fahd Hospital of the University, a public tertiary care hospital in Dammam, Saudi Arabia. The inclusion criteria of the patients were age <18 years, confirmed CSF/blood infection in correlation with radiological evidence of multiloculation, adequate medical records and regular follow-ups.

The validated functional status score (FSS) was used to evaluate the long-term outcomes. FSS includes six domains: mental status, sensory function, motor function, communication, feeding and respiratory status. Scoring in each domain ranges from 1 to 5, where 1 indicates normal status and 5 indicates severe dysfunction,<sup>[2]</sup> which can reflect the patient outcome in relation to multiple social and functional aspects.

## RESULTS

A total of 150 patients had ventriculoperitoneal shunt (VPS) inserted during the study period, of which 12 (8%) had postinfection MLH. The mean age at diagnosis was 9 months (range, 1–16 months), the male-to-female ratio was 1:1.4 and the mean follow-up period was 19 months (range, 5–84 months). Six patients were diagnosed primarily with congenital hydrocephalus, three with hydrocephalus secondary to myelomeningocele, two with Grade 4 intraventricular hemorrhage (IVH) of prematurity and one with the Dandy–Walker syndrome. Manifestation of shunt infection or malfunction was noticed in the form of fever,

new-onset seizure, enlargement of head circumference and decrease in oral feeding and activity.

Ten patients developed MLH after their first VPS infection and one each developed MLH following the second and third VPS infections. Most CSF cultures grew single organisms (6/12), with *Staphylococcus* being the most common (5/12) followed by *Klebsiella pneumoniae* (4/12) [Table 1]. All patients required VPS removal and external ventricular drain insertion as a temporizing measure until CSF clearance was achieved with appropriate antibiotic regimens followed by an endoscopic fenestration and VPS insertion (9/12 patients), after three consecutive negative CSF cultures.

The functional status outcome using FSS<sup>[2]</sup> was favorable in this case series, as eight patients (67%) scored 8–10 and one scored 6; three patients scored >10 (i.e., 11, 13 and 15). Finally, one mortality was found to be related to extensive pneumonia. Here, the authors describe three cases of MLH after VPS infection to highlight management with no shunt and navigated neuroendoscopic fenestration as well as a case with multiple causative organisms.

## DESCRIPTION OF SELECTED CASES

### Case 1

Imaging studies of a 3-month-old preterm girl (26 weeks) with an extremely low birth weight of 500 g, due to a progressive increase in head circumference revealed hydrocephalus and she underwent VPS insertion after stabilization. Two years after her first VPS revision, she developed VPS infection (*Pseudomonas stutzeri*). Computed Tomography (CT) scan of brain revealed the development of septate hydrocephalus and cystic encephalomalacia; therefore, she underwent a successful navigated endoscopic fenestration of cysts and third ventriculostomy without the need of VPS insertion [Figure 1]. The patient was followed up for 7 years and is currently responsive and active. Although she has spastic quadriplegia, her FSS was 10 (M: 1, S: 1, C: 1, M: 5, F: 1, R: 1) at the time of reporting this case.

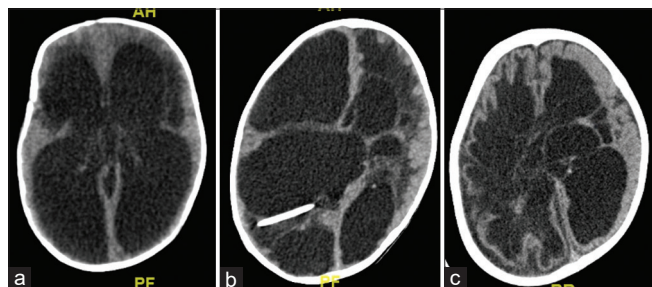
### Case 2

A 10-month-old boy prematurely born at 28 weeks was diagnosed with congenital heart disease and post-hemorrhagic hydrocephalus and underwent VPS insertion after CSF clearance. The patient had a second VPS infection 6 months after the first along with fever, lethargy and decreased oral intake. The CT of the brain revealed a dilated ventricular system with new formation of loculations [Figure 2], and the patient underwent VPS

**Table 1: Patients demographics, causative organism (s) and functional status score**

Case number	Age at diagnosis (months)	Gender	Diagnosis	CSF culture	Associated sepsis (Blood and/or urine)	FSS	Follow up (months)
1	3	Female	Congenital hydrocephalus	<i>P. stutzeri</i>	<i>S. epidermidis</i> <i>S. hominis</i>	10	84
2	10	Female	Congenital hydrocephalus	<i>S. marcescens</i>	<i>S. capitis</i> <i>S. marcescens</i>	13	65
3	10	Male	Posthemorrhagic hydrocephalus	<i>E. gallinarum</i> <i>S. epidermidis</i> <i>E. Cloacae</i>	<i>S. hominis</i> <i>P. aeruginosa</i>	11	70
4	1	Male	Myelomeningocele with ventriculomegaly	<i>K. pneumoniae</i> (ESBL)	<i>K. pneumoniae</i> <i>E. faecium</i> <i>S. epidermidis</i>	8	20
5	9	Female	Congenital hydrocephalus	<i>K. pneumoniae</i> (ESBL) <i>S. epidermidis</i>	<i>C. albicans</i>	9	22
6	16	Female	Congenital hydrocephalus	-	<i>S. epidermidis</i>	9	20
7	2	Male	Congenital hydrocephalus	<i>S. haemolyticus</i>	<i>E. coli</i>	10	5
8	9	Female	Dandy-Walker malformation with ventriculomegaly	<i>S. capitis</i> <i>S. hominis</i> <i>S. haemolyticus</i>	<i>P. aeruginosa</i> <i>S. capitis</i> <i>K. pneumoniae</i>	8	14
9	11	Male	Congenital hydrocephalus	<i>K. pneumoniae</i>	<i>S. capitis</i> <i>S. hominis</i> <i>S. haemolyticus</i>	15	17
10	7	Male	Myelomeningocele with ventriculomegaly	<i>C. freundii</i> , <i>Aspergillus</i> , <i>A. baumannii</i>	<i>S. haemolyticus</i> <i>S. epidermidis</i>	9	14
11	9	Female	Posthemorrhagic hydrocephalus	<i>K. pneumoniae</i> (ESBL) <i>S. epidermidis</i>	<i>S. epidermidis</i>	6	18
12	6	Female	Congenital hydrocephalus	<i>Aspergillus</i> species	<i>C. parapsilosis</i> Group B streptococcus	9	18

FSS: Functional status score (the lowest score: 6, the highest score: 30). *P. stutzeri* – *Pseudomonas stutzeri*; *S. marcescens* – *Serratia marcescens*; *E. gallinarum* – *Enterococcus gallinarum*; *S. epidermidis* – *Staphylococcus epidermidis*; *S. hominis* – *Staphylococcus hominis*; *S. capitis* – *Staphylococcus capitis*; *K. pneumoniae* – *Klebsiella pneumoniae*; *C. freundii* – *Citrobacter freundii*; *S. haemolyticus* – *Staphylococcus haemolyticus*; *A. baumannii* – *Acinetobacter baumannii*; *E. cloacae* – *Enterobacter cloacae*; *P. aeruginosa* – *Pseudomonas aeruginosa*; *E. faecium* – *Enterococcus faecium*; *C. albicans* – *Candida albicans*; *E. coli* – *Escherichia coli*; *C. parapsilosis* – *Candida parapsilosis*; ESBL – Extended-spectrum  $\beta$ -lactamases



**Figure 1:** Computed tomography brain scan: (a) initial imaging of the dilated ventricular system with transependymal permeation indicating active hydrocephalus, (b) development of multiloculated cysts upon ventriculoperitoneal shunt infection, and (c) final follow-up scan with regression of ventriculomegaly, fenestrated cysts and presence of encephalomalacic changes without the need of a ventriculoperitoneal shunt

removal and external ventricular drain insertion. The CSF culture showed a mixture of *Enterococcus gallinarum*, *Staphylococcus epidermidis* and *Enterobacter cloacae* and had a prolonged course of intravenous and intrathecal antibiotics until his CSF had cleared, and a navigated endoscopic fenestration was performed without the need for VPS insertion.

At 6 years of follow-up, the patient was stable. Follow-up CT showed stable ventricle size with significant leukomalacia.

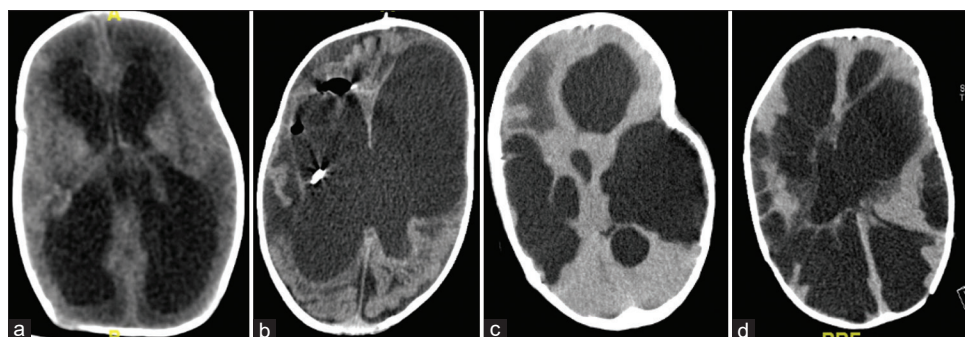
He was developmentally subnormal, and wheelchair bound with lower-limb spasticity; the FSS score was 11 (M: 2, S: 2, C: 2, M: 3, F: 1, R: 1).

### Case 3

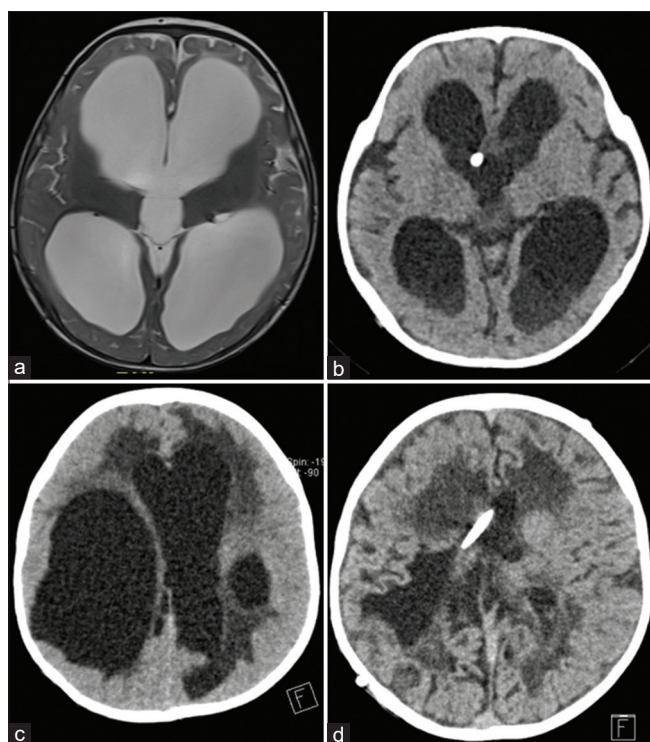
A newborn full-term baby girl was delivered by cesarean section due to hydrocephalus. She developed fever (39°C) and VPS infection 9 months after its insertion. CSF culture showed *K. pneumoniae* extended-spectrum  $\beta$ -lactamase and the ventricular catheter tip showed *S. epidermidis*. *Candida albicans* was detected from urine culture. The VPS was removed and external ventricular drain revision was done every 2 weeks upon unresponsive medical treatment. During hospitalization, she developed MLH [Figure 3]; navigated neuroendoscopic fenestration and VPS insertion were performed [Figure 4]. She was followed up for 2 years, and presented with interactive facial expressions but poor head control; her FSS score was 9 (M: 1, S: 1, C: 1, M: 4, F: 1, R: 1).

### DISCUSSION

The development of MLH has been described following different etiological diseases, such as neonatal



**Figure 2:** Computed tomography brain scan: (a) initial imaging after birth with ventriculomegaly and transependymal permeation, (b) imaging after first ventriculoperitoneal shunt infection with distorted ventricles, (c) imaging after the second ventriculoperitoneal shunt infection with development of multiple loculations and tense brain parenchyma, and (d) final imaging follow-up post neuroendoscopic fenestrations with stable appearance of ventricles and significant leukomalacia



**Figure 3:** (a) Magnetic resonance imaging brain T2-weighted image axial view scan at diagnosis showing triventricular hydrocephalus. Computed tomography brain scan showing: (b) infected debris at dependent occipital horns with external ventricular drain insertion, (c) development of multiloculation compartments and (d) post endoscopic fenestration and single ventriculoperitoneal shunt insertion

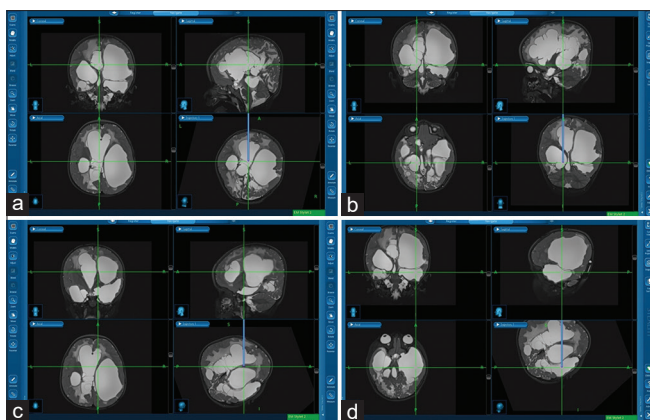
meningitis (bacterial or fungal), IVH of prematurity, traumatic head injuries, post-shunt infection and secondary to other inflammatory processes.<sup>[3]</sup> The underlying pathophysiological processes are not yet clearly understood. However, the organized formation of exudates and debris secondary to ventriculitis with the formation of subependymal gliosis that disrupt the normal ependymal lining plays a role in the development of compartmental intraventricular septations, and thus is the source for the development of such septations in the ventricular system.<sup>[4]</sup>

Four histopathological stages of MLH have been described (A. Ammar, 2017, p. 88):<sup>[5]</sup> Stage 1, formation of fibrinous, intraventricular membranes; Stage 2, increased infiltration of the membranes by inflammatory cells (lymphocytes accompanied by plasma cells and foamy histiocytes); Stage 3, gliosis and early (perivascular) fibrosis of membranes; and Stage 4, diffuse collagenous fibrosis of membranes with eventual transformation of membranes into dense, fibrotic septa with softening of the brain, loss of integrity, ventricular dilatation and low intracranial pressure.

In 1980, Kalsbeck *et al.* classified MLH as multiple septations, isolated lateral ventricle, entrapped temporal horn and isolated fourth ventricle.<sup>[6]</sup> However, “complex hydrocephalus” is another term that has been interchangeably used in the literature to describe a complicated hydrocephalus as a combination of outlet obstruction and defective absorption of the CSF.<sup>[5,7]</sup> It can be further subcategorized into temporary or permanent, based on whether the defect in absorption and/or permeability of the CSF through the subarachnoid space is transient or constant.<sup>[7,8]</sup>

Several risk factors have been attributed to the development of MLH and reported in the literature; history of shunt revision/infection, low birth weight (<2000 g), prematurity, age at the time of the initial shunt procedure, history of IVH or myelomeningocele and coinfection were significant risk factors.<sup>[9-11]</sup> In our case series, prior shunt infection was present in all cases and the average time of shunt infection until the development of MLH ranged from 2 weeks to 2 years.

Despite the detection of single organism was marginally higher than multiple organisms in this case series, the unusual fulminant organisms that are encountered is a serious problem on the overall outcome, and it must be



**Figure 4:** Sets of intraoperative electromagnetic navigation photographs (a-d) of multicompartmental hydrocephalus using single left-sided frontal approach neuroendoscopy through Kocher's point (a), which shows fenestration of all compartments, reaching to the temporal horn of the contralateral side (d)

addressed properly and aggressively. Coagulase-negative *Staphylococcus* and *Staphylococcus aureus* were the most commonly detected organisms in the CSF samples of patients with MLH presenting with shunt infections, accounting for approximately two-thirds of all infections.<sup>[12]</sup> Another case series determined that Gram-negative bacteria were the most commonly detected organisms accounting for the majority of shunt infections: six cases had *Escherichia coli*, two cases had *Klebsiella* species, one case had *Protens* species and one case had *S. aureus*.<sup>[6]</sup> In our series, the most commonly detected organisms were *K. pneumoniae* (four patients), followed by *S. epidermidis* (three patients), *Staphylococcus haemolyticus* and *Aspergillus* species (two patients). Notably, one of our cases had *Serratia marcescens*, a rare opportunistic organism,<sup>[13]</sup> in which the patient experienced a very severe course of infection and passed away.

Traditionally, the treatment of MLH is carried out using multiple shunt systems, aiming to drain each cyst separately into a confined peritoneal space. This carries a potential risk of failure, which often requires multiple shunt revisions, which can affect the morbidity and mortality rates.<sup>[6]</sup> Alternatively, navigated neuroendoscopic fenestration is an effective approach. This is done by converting the isolated multiloculations into a single compartment through the fenestration of the septations to insert a single shunt instead of multiple shunt insertions. One study compared two groups of children who had complex hydrocephalus; the first group was treated using the endoscopic approach, whereas the other group was treated with a complex shunt system.<sup>[14]</sup> Patients in the endoscopic arm had better postoperative outcomes and fewer complications compared with those who underwent the complex shunt placement procedure. Moreover, the endoscopic approach

was found to have fewer shunt revision rate (0.25/year) compared with (3.04/year) the complex shunt group.<sup>[14]</sup> In addition, improved overall clinical outcomes were achieved in the majority of children who underwent navigated endoscopic fenestration.<sup>[15]</sup> In our case series, all children underwent navigated endoscopic fenestration, nine patients required VPS placement, two did not require VPS (Cases 1 and 3 in Table 1), one required two shunt systems and three patients required redo endoscopic fenestration surgery.

The functional outcome in the literature is still unfavorable. The majority of our patients had mild developmental delay based on an average FSS of 9 [Table 1]. The initial diagnosis and the concurrent systemic illnesses accompanied by a very fulminant causative organism are correlated adversely with outcome. This is clearly seen in one of our patients (Case 2 in Table 1), who had recurrent shunt infections (*S. marcescens*) complicated by the development of multiloculation and died from extensive pneumonia after an aggressive course of management. In a study examining 13 infants with MLH who underwent endoscopic fenestration of multiple ventricular cysts, it was reported that seven patients died, three survived with severe developmental retardation and three were noted to have a developmental delay in the last follow-up visit.<sup>[6]</sup> The initial diagnosis, unusual causative organism and overall health condition all influence the overall patient outcome.<sup>[16]</sup>

## CONCLUSION

Development of MLH post-VPS infection is debilitating and requires prompt treatment. Although the overall functional outcome is poor, the evolving neuroendoscopic techniques with tailored preoperative planning may play a role in reducing the adverse effect of shunt multiplicity, shunt infections and the higher failure rate among patients with complex hydrocephalus.

## Ethical considerations and declaration of patient consent

The institutional review board of Imam Abdulrahman Bin Faisal University, Dammam, Saudi Arabia, provided ethical approval for this study (Ref. no.: IRB-2020-01-230) in July 2020. The authors also certify that they have obtained all appropriate patient consent forms. In the form, the parents/guardian of the patients have given consent for the images and other clinical information to be reported in the journal. The parents/guardian understand that names and initials will not be published, and due efforts will be made to conceal the identity of the patients, but anonymity cannot be guaranteed.

**Peer review**

This article was peer-reviewed by two independent and anonymous reviewers.

**Financial support and sponsorship**

Nil.

**Conflicts of interest**

There are no conflicts of interest.

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