

Negative Pressure Hydrocephalus: A Rare Diagnosis That Can Be Effectively Treated With Ventriculopleural Shunting Wyatt L. Ramey MD; Willard Stein Kasoff MD; Martin E. Weinand MD

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Introduction

Negative pressure hydrocephalus (NegPH) is a rare entity along the spectrum of hydrocephalic disorders that is very poorly described in the neurosurgery literature and presents a unique challenge in both diagnosis and treatment. A few small case series describe treatment with cerebrospinal fluid (CSF) diversion via ventriculoperitoneal shunting. However, by utilizing the negative intrathoracic pressure gradient necessary for inspiration, we describe the first series of NegPH patients successfully treated with a valveless ventriculopleural shunt.

Methods

All patients with NegPH were retrospectively reviewed between 1999-2015 at a large university teaching hospital. NegPH was diagnosed only if 1) signs and symptoms of hydrocephalus were present, 2) there was radiographic confirmation of ventriculomegaly, and 3) patients were proven with ICP recording via ventriculostomy (EVD) to have negative ICP. Three patients underwent CSF diversion via ventriculopleural shunt insertion and response to treatment was reviewed.

Table 1.				
Patient	Age	Sex	Pathology	Level of Drainage (mmHg)
1	57	М	Cerebellar AVM	-5
2	25	F	Chiari II	-3
3	32	F	Astrocytoma	N/A

Basic demographics, pathology requiring previous intracranial surgery, and level at which EVD placed for CSF drainage prior to shunting.

Results

Three patients were diagnosed with NegPH. Every patient had previously undergone intracranial surgery for various pathologies including evacuation of a cerebellar hematoma. Chiari II decompression, and previous cerebral astrocytoma resection (Table 1). Each patient underwent initial CSF diversion with an extraventricular drain. Following negative ICP recordings, all three patients responded well clinically to CSF diversion with negative pressure drainage via ventriculostomy. The EVD drainage level was set at a level deemed appropriate based on the patients' clinical exam, which was -5 mmHg in Patient 1 and -3 mmHg in Patient 2. This data was unavailable for Patient 3. Once neurologically stable, a valveless ventriculopleural shunt was inserted in all three patients with good clinical and radiographic response to treatment.

Conclusions

The mechanism of NegPH remains unclear. Every reported case thus far, including those in our series, has previously undergone intracranial surgery, which may mean NegPH is a rare complication associated with hemorrhage into the CSF and development of a negative transmantle gradient with ongoing ventriculomegaly. As a result, the pleural space was chosen as the distal shunt terminus in our series because of its negative pressure gradient created for the driving force of inspiration. We feel ventriculopleural shunting offers the best physiologic solution and provides good long-term outcome in the treatment of NegPH.

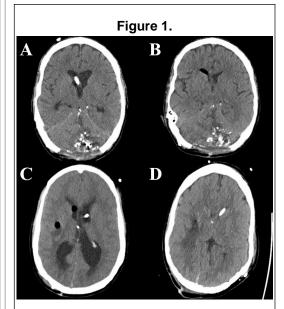
Learning Objectives

By the conclusion of this session, participants should be able to: 1) Describe the importance of negative pressure hydrocephalus and why it is necessary to treat with negative pressure drainage into the pleural cavity, 2) Discuss, in small groups, the pathophysiology of Negative Pressure Hydrocephalus and possible options for its treatment, and 3) Identify an effective treatment for Negative Pressure Hydrocephalus, which we feel is to harness the negative intrathoracic pressure as a distal terminus for a shunting procedure.

References

 Filippidis, Aristotelis S., et al.
"Negative-pressure and low-pressure hydrocephalus: the role of cerebrospinal fluid leaks resulting from surgical approaches to the cranial base: Report of 3 cases." Journal of neurosurgery 115.5 (2011): 1031-1037.

2) Hunn, Benjamin HM, et al. "Successful treatment of negative pressure hydrocephalus using timely titrated external ventricular drainage: a case series." Clinical neurology and neurosurgery 116 (2014): 67-71.



Before and after ventriculopleural shunt CT scans of Patient 1 (A, B) and Patient 2 (C, D).