

Negative Pressure Hydrocephalus—Case Series and Literature Review Illustrating the Natural History and Outcomes of a Rare and Perilous Neurosurgical Entity

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Introduction

Negative pressure hydrocephalus (NegPH) is a rare clinical ventriculomegaly syndrome in which patients require CSF diversion at a less-than-zero pressure. We present seven patients and a literature review, to characterize the natural history, management, and outcomes of this dangerous entity.

Methods

Institutional records were queried to identify patients who had undergone EVD drainage at less-than-zero pressure during the study period, 1995-2015. Chart review captured demographics, diagnoses, treatment modalities, morbidity, mortality, and disposition. Supportive systematic review of published cases and series reporting patient-specific findings in NegPH is further included.

Results

Seven NegPH patients were identified—three SAH, two IVH, one benign tumor, one TBI. One patient was pediatric; five were female. Maximum negative pressure required was -10mmHg. Mortality was 29%; in both patients, care was withdrawn due to severe neurologic disability. Major or minor permanent neurologic morbidity was 100%. Among survivors, 40% required out-of-home disposition. Complications included vasospasm (29%), seizures (43%), infection (43%), shunt revision (43%), and need for secondary neurosurgical intervention (71%). Permanent CSF diversion was required in 57%, with one patient each requiring valveless, antisiphon, Delta 0.5, and Delta 1.0 valves. Literature review identified seven preceding reports, documenting 26 patients.

Conclusions

NegPH is highly variable disease process. Extended clinical course, infection, surgical revision, and IVH predict poor outcome. Early recognition and prompt treatment as the most important factors influencing long-term survival. Patients with underlying CSF leak require surgical repair and titrated EVD wean using flow-directed goals. Pediatric or tumor patients may depend on multi-shunt systems draining ventricular and subdural spaces. SAH portends a grievous prognosis and severe NegPH that cannot be overcome by weaning or valveless shunts—thought attributable to permanent alterations in parenchymal compliance. Vanguard treatments using large-bore catheters or pumpable shunt systems offer approximations of negative pressure drainage, but have not been tested in larger series.

Table 1

Patient	Age	Sex	Diagnosis	Minimum Pressure	Shunt Dependence	Morbidity/Complications	Outcome/Disposition
1	54	F	SAH	-10mmHg	None	Vasospasm; seizures; revisions; secondary intervention;	Death
2	71	M	SAH	-5 mmHg	Anti-siphon	Vasospasm; seizures; infection; revisions	Death
3	48	F	SAH	-10mmHg	None	Seizures; secondary intervention	Home
4	2	F	IVH	-5 mmHg	Delta 1.0	Infection; revisions	Home
5	50	M	Tumor	-5 mmHg	None	Secondary intervention	Home
6	50	F	Tumor	-5 mmHg	Valveless	Secondary intervention	Rehab
7	32	F	IVH	-5 mmHg	Delta 0.5	Secondary intervention	Rehab

Individual patient data in NegPH

Learning Objectives

- 1.) Diagnose NegPH, and recognize clinical scenarios that are likely to precipitate its onset
- 2.) Discuss leading theories regarding the underlying mechanism of NegPH, and how they inform various approaches to clinical management
- 3.) Implement novel treatment plans for major etiologies of NegPH, including flow-directed drainage titration, multi-shunt complexes, large-bore catheters, and pumpable systems

Figure 1



Noncontrast axial CT of the head demonstrating persistent ventriculomegaly in a patient with clinical symptoms of acute hydrocephalus, in spite of external ventricular drainage at -5mmHg