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Normal-Pressure Hydrocephalus

Another Treatable "Dementia": Part II

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Learning Objectives: *After reading this article, the participant should be able to:*
1. Identify indications for shunting in treatment of normal-pressure hydrocephalus (NPH).
2. Describe the complications of shunting in patients with NPH.
3. Explain the prognosis after shunting in patients with NPH.

This article is the second of two parts.

Treatment Options

Medical

Acetazolamide (Diamox; Barr Laboratories, Inc.) has been reported to be effective in treating patients with normal-pressure hydrocephalus (NPH) by reducing secretion of cerebrospinal fluid (CSF). Our experience, however, is otherwise: a few of our patients with NPH have tried acetazolamide, but the results have not been encouraging.

Intermittent Lumbar Puncture

Intermittent lumbar puncture occasionally is useful in treatment of NPH. However, this approach requires frequent spinal taps. As a result, patients who initially opt for intermittent lumbar punctures most often request shunting eventually.

Operative

Lumbar peritoneal, ventriculoperitoneal, and ventriculoatrial shunting. Diversion of CSF via shunting is the most direct and efficacious therapeutic modality for treatment of NPH. Unfortunately, potentially serious morbidity is associated with its use. Some morbidity is related to the procedure, but the main risk results from delayed complications such as infection, shunt failure, and subdural hematoma from overdrainage. In the elderly population affected by NPH, in whom the ventricles are enlarged and the brain atrophic, a decrease in ventricular volume from overdrainage may lead to collapse of the cortical surface and hygroma formation as well as stretching of the bridging veins. This may cause those veins to rupture and bleed into the dead space created by the collapsing brain, leading to the formation of a subdural hematoma and its potentially disastrous consequences.

Although studies have shown that use of low-pressure valves is associated with the highest rate of success, they also are associated with the highest rate of complications resulting from overdrainage. The incidence of permanent neurological morbidity and mortality in patients undergoing shunting who have received low-pressure valves is approximately 5%.

It is crucial to discuss the shunt procedure extensively with the patient's family members beforehand, so that they understand the difficulties involved in making an accurate diagnosis and the reality that even with the best preoperative testing, success is not guaranteed. The family also must be made aware of the potential serious long-term morbidities associated with shunting.

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Category: Neural hydrodynamics

Key Words: Normal-pressure hydrocephalus, Shunting, Complications of shunting

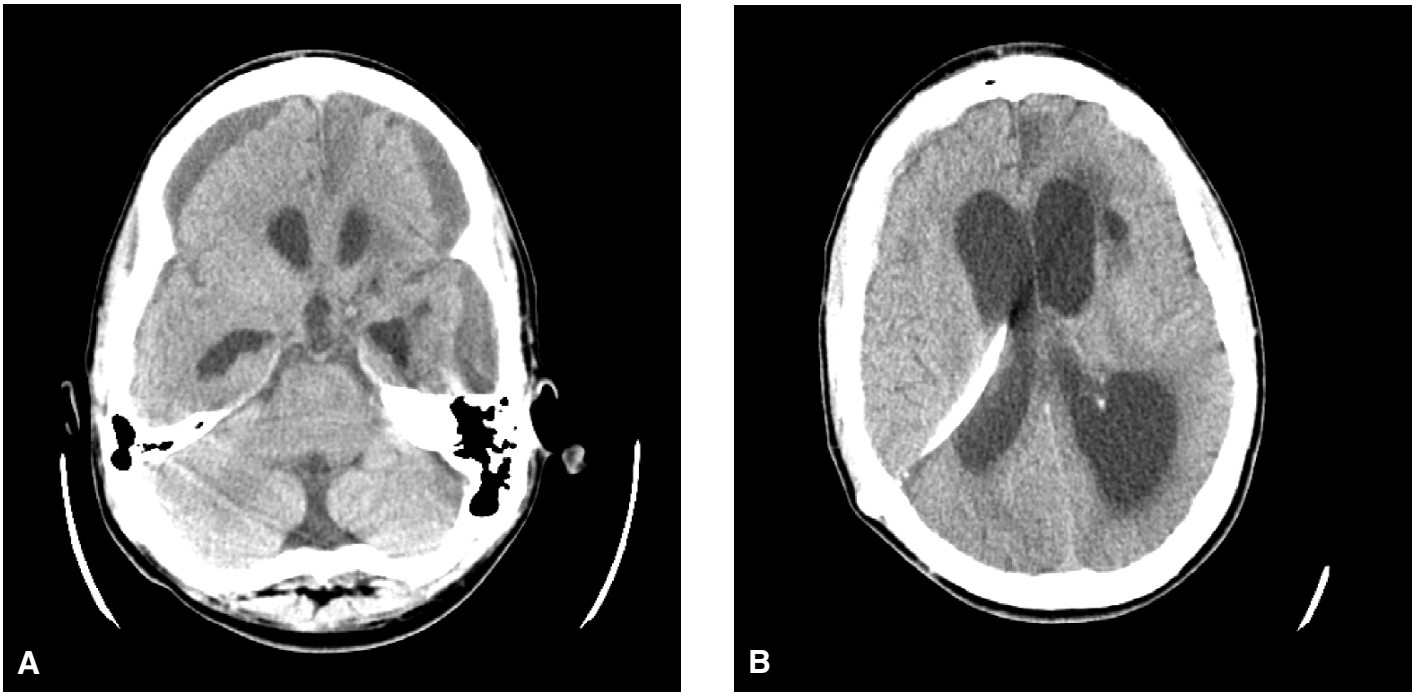

Figure 1. CT scans in an 18-year-old male who had incurred a head injury in a skiing accident developed posttraumatic hydrocephalus. After shunting, he developed subdural collections (*A*). The valve setting on the shunt was increased, and the collections resolved spontaneously 2 weeks later (*B*).

Programmable valves. It is crucial to discuss the risk/benefit ratio. This discussion is very patient-specific and should be individualized. The use of programmable valves has added a new dimension to the treatment of NPH. This technique allows the surgeon to insert a shunt in a noninvasive fashion to modify the amount of CSF drained, so as to obtain the maximum clinical benefit with the least amount of CSF drainage for any particular patient, rather than having to rely on a "one prescription fits all" system.

Traditional valves had a fixed setting. Although valves with different settings were available, after the valve was implanted, the pressure setting could changed only by replacing the valve via open surgery. This created a dilemma in treatment of patients in whom shunting had provided some, but not maximal, improvement. Although these patients might have benefited from a lower-setting valve, in many cases, the risk of another surgery was too great to be justified by the possible improvement. In other patients, however, overdrainage would lead to formation of a subdural hygroma or hematoma, necessitating ligation or removal of the shunt to stop CSF drainage altogether and prevent these collections from becoming larger. Programmable valves allow the drainage settings to be adjusted transcutaneously, without the need for invasive intervention.

Programmable valves have made it possible to take a more aggressive approach to NPH. In a patient who will benefit from drainage of more fluid, the setting is lowered (Fig. 1), whereas in a patient who is overdraining, the pressure setting is increased to decrease the amount of fluid

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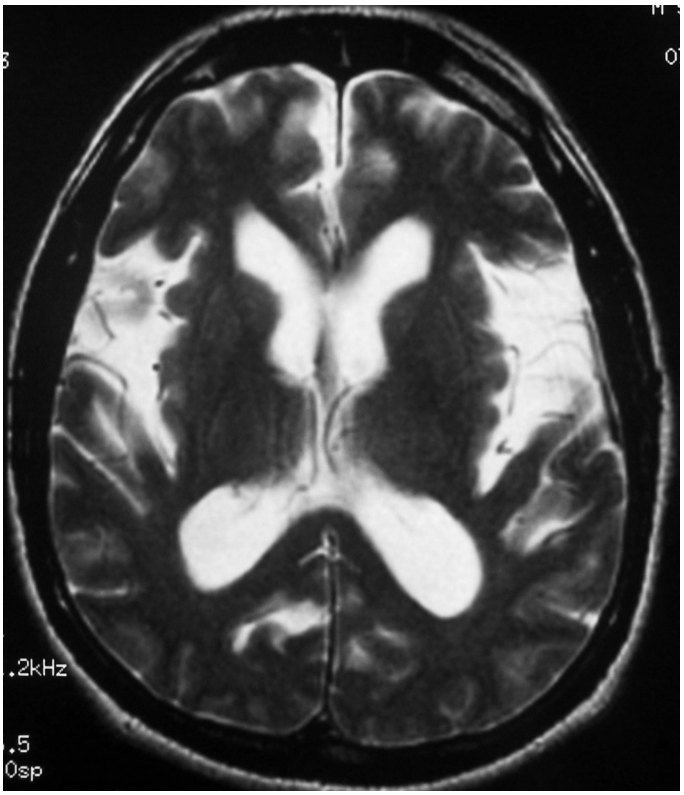

Figure 2. MRI scan in a 50-year-old executive with an intellectually demanding job who experienced dizziness, foggy thinking, and gait unsteadiness. The scan revealed enlarged ventricles with an enlarged sylvian fissure. A shunt was inserted with a programmable valve set at a pressure of 110 mm H_2O. The patient's condition improved but did not return to normal. The valve setting was reduced to 80 mm H_2O, at which level the symptoms resolved completely.

drained (Fig. 2). Pressure adjustments are not performed routinely in every patient, but in many cases, these adjustments can eliminate revision surgery to place a higher- or lower-pressure valve. In addition, now that it is possible to regulate drainage without a second operation, the threshold for shunting has become lower.

Complications of Shunting

Shunting is not without risks, especially in this elderly and fragile population. Some of these complications are immediate and intraoperative, whereas others are delayed. Immediate complications include mainly intracerebral hemorhage, which occurs at a rate of approximately 3%. Delayed complications include seizures (3%–10% of patients) and infection (approximately 5% of patients). Other delayed complications are related to shunt obstruction, underdrainge, or overdrainage. It is important to keep in mind that these shunts can fail or underdrain; therefore, clinical regression or failure to improve after shunting should warrant an investigation of the shunt function. Shunt failure may occur in up to 20% of patients during long-term follow-up. Overdrainage leads to the most dreaded delayed complication of shunting for NPH: hygroma and delayed subdural hematoma. The incidence of the latter has been reported to vary between 2% and 17%. Additional complications can occur from co-exiting medical conditions.

Outcome

NPH can be frustrating to treat, and the success rate after shunting is extremely variable. Four patterns may be encountered:

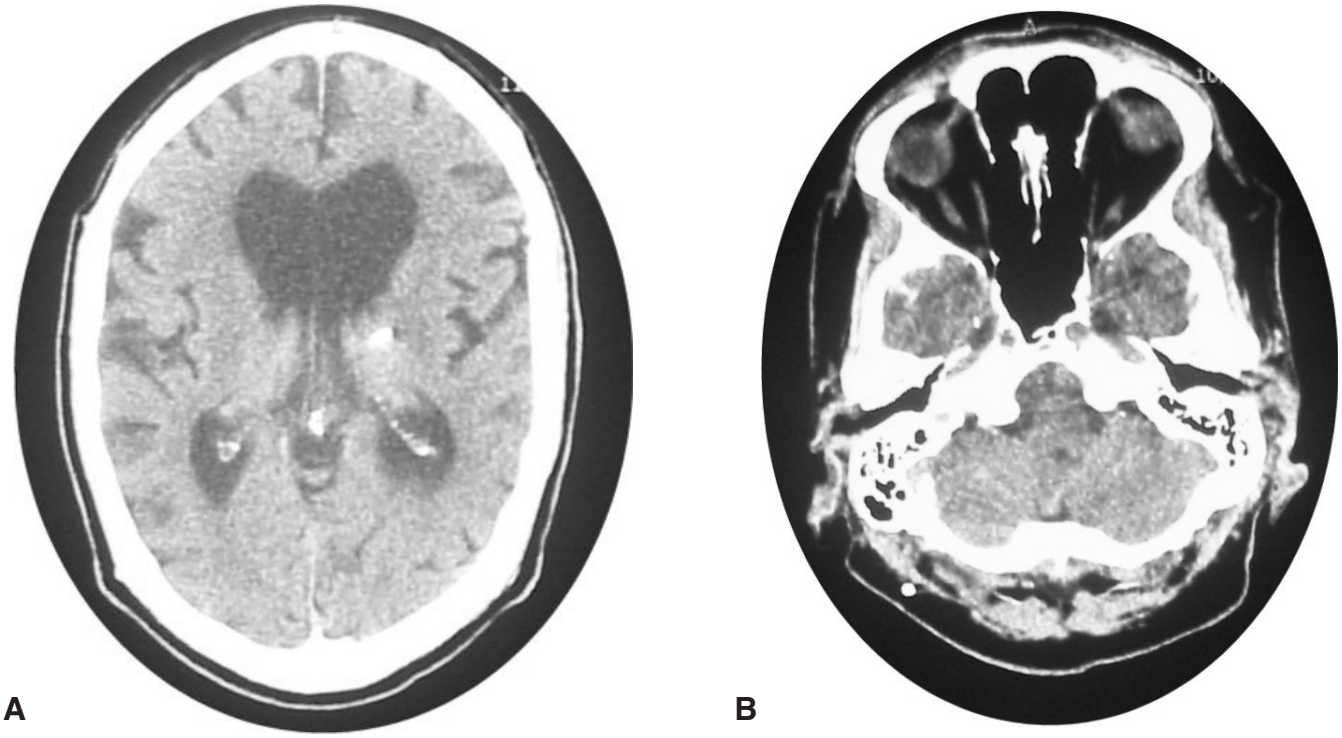

Figure 3. MRI scans in an 80-year-old man had severe gait disturbance, urinary incontinence, and decline in mental status. He was seen by numerous specialists and underwent multiple lumbar punctures without clinical improvement. *A*, Enlarged lateral ventricles. *B*, Small fourth ventricle. A shunt was placed, and the patient's condition improved dramatically. He most likely had aqueductal stenosis, which explains the lack of improvement in response to lumbar puncture, although he did have hydrocephalus.

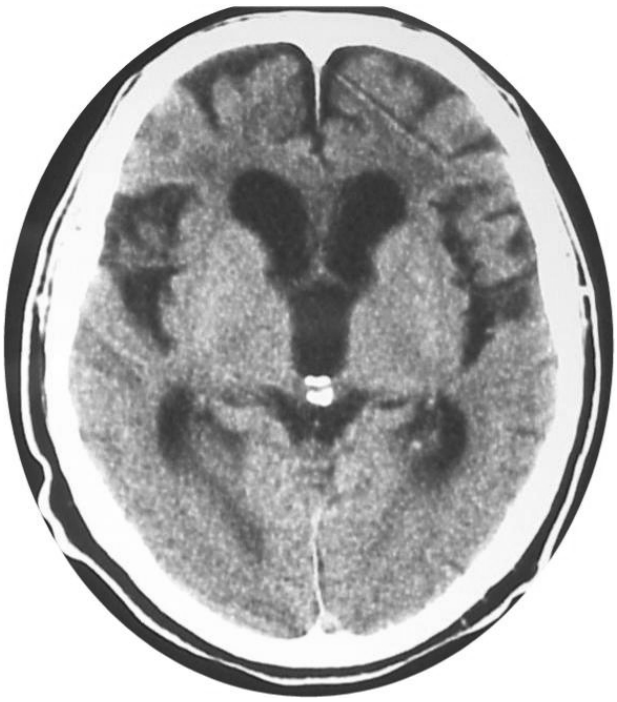

Figure 4. CT scan in a 78-year-old man who exhibited the typical normal-pressure hydrocepalus triad of gait disturbance, urinary incontinence, and dementia. The scan was interpreted as "moderate generalized enlargement of the ventricles. . . mild cortical atrophy. . . the findings are felt to represent atrophy." The patient's condition improved dramatically with CSF drainage.

- Dramatic improvement during the first month followed by stable condition;
- Slow and steady improvement;
- No change or worsening; or
- Transient improvement followed by a return to preoperative baseline.

The size of the ventricle does not necessarily decrease after shunting. Therefore, ventricle size does not have a significant role in the assessment of shunt function. In patients in whom transient improvement is seen, the shunt should be investigated to exclude early shunt failure, which has been reported in this group of patients.

Duration of symptoms is one of the most reliable factors in predicting response to treatment: patients with the shortest duration of symptoms preoperatively have the best results from shunting. However, in some cases, despite extensive investigations, the shunt is determined to be functioning and no other conditions are found, but the patient has regressed to the preoperative baseline after a few years of improvement. These can be challenging cases to manage.

Summary

NPH represents a diagnostic challenge. It is crucial to keep a high index of suspicion when assessing patients who might have the condition. A combination of clinical and radiological findings and drainage interventions must be used to determine which patients may benefit from shunting. There are no absolute diagnostic tests (Figs. 3 and 4); the surgeon must rely on his or her best clinical judgment to make the most appropriate therapeutic decision. The advent of programmable shunt technology has been a major advance in the treatment of NPH and has provided more flexibility in managing patients with the condition.

Readings

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1. Lumbar peritoneal shunting is a therapeutic option for treatment of normal-pressure hydrocephalus (NPH).

   True or False?

2. Subdural hematomas do not occur after shunting.

   True or False?

3. Seizures can occur after shunt placement.

   True or False?

4. Duration of symptoms correlates with prognosis for patients with NPH.

   True or False?

5. Shunt failure can occur in patients with NPH.

   True or False?

6. The risk/benefit ratio has to be individualized for every patient.

   True or False?

7. The outcome of shunting is variable.

   True or False?

8. Ventricular size is a reliable means of assessing shunt function in patients with NPH.

   True or False?

9. Shunting does not entail serious morbidity in NPH.

   True or False?

10. NPH may be very rewarding to treat, with occasional dramatic results.

    True or False?