

# Chapter 9

## Hydrocephalus



Symptomatic hydrocephalus, whether communicating or non-communicating, in an otherwise healthy and viable patient, is a clear indication for neurosurgical intervention. This classic form of hydrocephalus causes high intracranial pressure, and a deteriorating neurological exam, that can ultimately lead to death if not treated. Typical symptoms include bad headaches, nausea, vomiting, confusion, lethargy, and obtundation. There can also be visual impairment or a sixth nerve palsy. Treatments can include ventriculostomy, if the hydrocephalus is expected to be transient, or a shunt. The generally preferred shunting technique is a right frontal ventriculoperitoneal shunt, as the frontal entry site and the peritoneal distal sites are associated with the fewest complications. A 7 cm right angled ventricular catheter will prevent inadvertent deep or shallow placement of the catheter. The neuronavigation system helps to optimize the placement of the ventricular catheter, and a general surgeon can place the peritoneal catheter laparoscopically (usually through a small incision in the umbilicus) [63]. A programmable shunt valve is also usually desirable. Occipital shunts may be reserved for bald men for whom the frontal shunt valve placement would be cosmetically undesirable.

If a patient with a shunt presents with multiple episodes of what seem to be recurrent shunt infections that are otherwise unexplained, which can include redness over the shunt equipment or wound drainage, the rare diagnosis of silicone allergy should be entertained. Replacement of the shunt with a silicone-free shunt will fix this problem.

In the event the hydrocephalus is due to an obstruction within the ventricular system—in the posterior third ventricle, the aqueduct, or the fourth ventricle—an endoscopic third ventriculostomy (ETV) can be performed instead of a shunt.

In rare cases, a patient can develop many of the same symptoms of “classic” hydrocephalus with normal or even low intraventricular pressures (usually after an intracranial hemorrhage or chronic shunting). These patients also require shunting, but with either low pressure valves or programmable valves set to low pressures. Consideration might also be given here to using a lumbar proximal shunt site (in

cases of communicating hydrocephalus) and/or a pleural distal shunt site. These patients are very challenging to manage and have a significantly poorer prognosis than patients with typical high-pressure hydrocephalus.

## **Normal Pressure Hydrocephalus (NPH)**

This entity is entirely different from “classic” hydrocephalus. In this entity, patients, usually elderly patients in their 60s and 70s, are found to have enlarged ventricles (ventriculomegaly) and associated symptoms that include gait difficulty and may also include memory impairment and/or urinary incontinence. If a lumbar puncture is performed, the pressure is found to be normal.

The problem here is that many elderly patients have enlarged ventricles (due mostly to atrophy) and some gait difficulties or memory loss or urinary incontinence. Nursing homes are likely full of such patients. Yet very few people who have these features will actually benefit from a ventriculoperitoneal shunt. Perhaps a distinction should be made between the term “normal pressure hydrocephalus,” which defines a condition of enlarged ventricles with normal pressure that are causing neurological symptoms and, the much more common “normal pressure ventriculomegaly,” which could define a condition of enlarged ventricles with normal pressure that are not suspected of causing neurological symptoms.

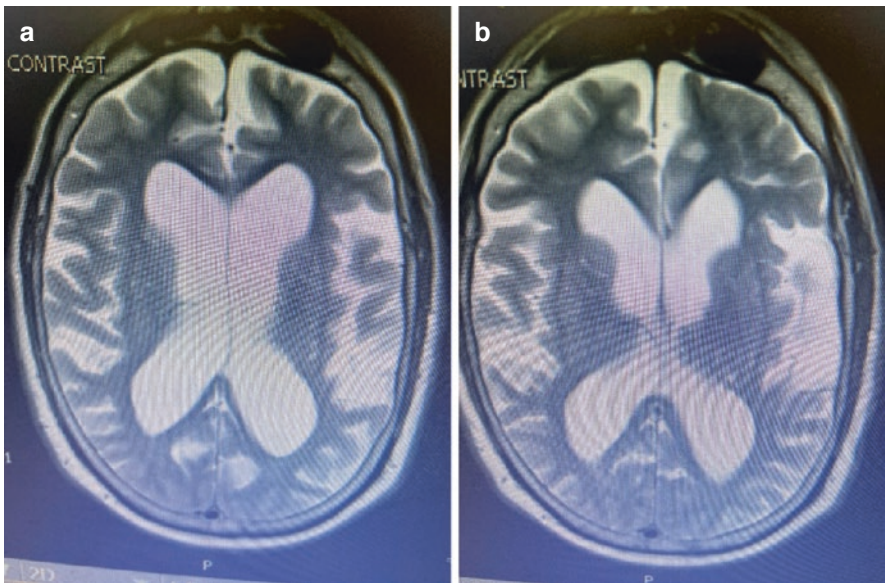
Shunts in these NPH cases are reportedly most beneficial when the gait and memory problems are fairly mild to begin with. Of note, many of these potential surgical candidates are not that bothered by their mild walking issues or their mild memory issues and have little interest in any brain operation. It is also not completely clear why a shunt would help in these cases. The shunt presumably does not drain much fluid as the pressure is normal to begin with, and follow-up imaging often shows no change in ventricular size. Perhaps some patients with NPH who benefit from medium pressure shunts have high/normal intracranial pressures.

Even for those patients who are reportedly better with a shunt, the improvements are often very subtle and can be documented only with detailed testing. Furthermore, many of these patients, at long-term follow-up after shunting, will show minimal improvement, no improvement, or worsening of their clinical conditions. One analysis that reviewed 44 publications showed only 29% of shunted patients experienced prolonged or significant improvement [64]. This same study showed a 38% complication rate, a 22% rate of re-operation, and a 6% rate of major complication or death. A more recent study showed that of patients shunted for NPH, only 43% had a clinically significant improvement in health-related quality of life at 1 year follow-up [65]. Another study showed little difference at long-term follow-up between such patients who were shunted compared with those who were not shunted [66].

There is also some evidence that patients suspected of having NPH might benefit from oral acetazolamide (a medicine that decreases CSF production), just like patients with idiopathic intracranial hypertension. One study showed many patients

with presumed NPH improved with oral acetazolamide, and that there was often persistent benefit at 1 year follow-up [67]. Another small study also showed many patients with presumed NPH improved with acetazolamide, and that patients who improved showed a significant decrease in the volume of periventricular white matter hyperintensity volume [68]. Further study of acetazolamide in this context would make sense.

All that said, criteria for even considering a shunt surgery for possible NPH should be stringent and should include (1) the patient has clearly enlarged ventricles in the presence of limited generalized brain atrophy; (2) the patient has few confounding medical co-morbidities; (3) the patient has insidious onset of mild to moderate gait difficulties and possibly memory problems or urinary incontinence that have no other obvious explanations as to their cause; and (4) the patient shows dramatic improvement of their symptoms after a high volume lumbar puncture (see Fig. 9.1). At the very least, more study is needed here to better select the patients who will significantly benefit from shunt operations.



**Fig. 9.1** This is a 70-year-old man with the main complaint of some mild intermittent memory issues for about a year. He also had periodic imbalance for several years. He had spinal stenosis and episodes of pain radiating down his legs that had improved with epidural steroid injections. He also had very rare mild episodes of urinary incontinence. Brain MRI (a, b) showed enlarged ventricles and moderate diffuse brain atrophy that was unchanged compared with an MRI from 6 years earlier. Medical history was also noteworthy for hypertension being treated with multiple medicines and trigeminal neuralgia controlled on a low dose of carbamazepine (200 mg BID). After consultation, the patient's carbamazepine was switched to gabapentin. The patient quickly noted a significant improvement in his alertness and his memory. He did not wish to pursue a diagnostic spinal tap

## **Idiopathic Intracranial Hypertension**

Also known as “pseudotumor cerebri,” this condition involves high pressures in the brain without any discernable mass. It frequently affects young, overweight women and can cause headaches, papilledema, and visual loss. A common cause is thought to be a narrowed transverse sinus, and sinus stenting is another consideration in refractory cases. Usually, this condition can be managed with acetazolamide and weight loss. For patients with deteriorating vision or intractable headaches despite medicines, a shunt can be offered. A right frontal VP shunt can be placed with neuronavigational guidance, unless the ventricles are very small, in which case a lumbo-peritoneal shunt can be offered. Surgery is rarely needed in this condition.