Treatment of Infantile and Childhood Hydrocephalus

The development of ventricular shunt tubing with one-way valves has opened the way to successful treatment of hydrocephalus. The vajve can be set at a desired pressure, allowing the CSF to escape directly into the bloodstream or peritoneal cavity whenever the pressure level is exceeded. The authors have obtained gratifying success, often a complete or nearly complete restoration of mental function and gait, by the placement of a ventriculoatrial or ventriculoperitoneal shunt. As a group, these patients all had the first of the clinical triad described above (only half of our patients were incontinent), and their lateral ventricular span at the leyel of the anterior horns was in excess of 50 mm (a true dimension calculated from CT scans), equivalent to approximately 18 mm on the usual X-ray film with relatively little cerebral atrophy. Deviations from this syndrome such as the occurrence of dementia without gait disorder or the presence of apraxias, aphasias, and ther focal cerebral signs should lead one to a diagnosis other than hydrocephalus. The most consistent improvement has been attained in those hydrocephalus patients in whom a cause could be established (subarachnoid haemorrhage, chronic meningitis, or third ventricular tumour). Uncertainties of diagnosis increase with advancing age owing to the frequent association of senile dementia and vascular lesions. However, age alone does not exclude the existence of hydrocephalus as a cause of gait disorder, and long duration of symptoms does not preclude a salutary outcome from shunting.

The potential failures of shunting must be anticipated in patients who do not conform to the typical syndrome, or whose disease has advanced to the stage of longstanding incontinence or dementia. As noted further on, a good clinical response is attended by some reduction in ventricular size. In some instances, a lack of improvement is explained by inadequate decompression and this situation justifies a revision of the shunt with a valve that drains at lower pressures. Overdrainage, in contrast, causes headaches that may be chronic or orthostatic and may be associated with small subdural collections of fluid. These are generally innocuous and do not require drainage unless they enlarge, cause focal neurological symptoms, or rarely, seizures.

Although relatively simple as a surgical procedure, there are pliations of shunting, the main ones being a postoperative subdural hygroma or hematoma (if the ventricular pressure is reduced or rapidly, the bridging dural veins may stretch and rupture); infection of the valve and catheter, sometimes with septicemia; occlusion of the tip of the catheter in the ventricle; and, particularly uitants and children, the "slit ventricle syndrome" (see below). The incidence of complications can be considerably reduced by placing the catheter in the anterior horn of the right ventricle, where there is no choroid plexus, and by using the latest type of Hakim valve, which permits extracranial electronic control of valve pressure. Meticulous aseptic technique and the preoperative postoperative administration of oxycillin and gentamicin havl duced the incidence of shunt infections. Once the CSF is shul the ventricles diminish in size within 3 or 4 days even when! hydrocephalus has been present for a year or more. This indil that the so-called hydrocephalic compression of the cerebrue largely reversible. Indeed, in Black's series, the ventricles failed return *to* normal in only 1 of his 11 shunted patients, and in this patient there was no clinical improvement. Clinical improvement occurs within a few weeks, the gait disturbance being slower to reverse than the mental disorder. Symptoms of cerebral atrophl due to Alzheimer'i disease and related conditions are not alteree by ventriculoatrial shunting. Of course, if the hydrocephalus is caused by an operable tumour, surgical removal is the procedure of choice.

Here one encounters more difficulties than in the treatment off the adult disorder. The ventricular catheter may wander or become obstructed and require revision. Peritoneal pseudocysts may form (most shunts in children are ventriculoperitoneal). Another unexpected complication of shunting has been coaptation of the ventricles, the so-called slit-ventricle syndrome. This occurs more frequently in young children, though we have observed it in adults as well. Some patients may develop a low-pressure syndrome with severe generalized headaches, often with nausea and vomiting, whenever sitting or standing. Some children become ataxic, irritable, or obtunded or may vomit repeatedly. The CSF pressure is extremely low and the volume of CSF much reduced. In babies, the cranium may fail to grow even though the brain is of normal size. In most shunted patients, the intracranial pressure in the upright position is

diminished to 30 mm H_2O, but in this condition it may reach 100 to 150 mm. In order to correct the condition, one would imagine that replacing the shunt valve with another one that opens under a higher pressure would suffice. Indeed, it may be preventive. But once the condition is already established, the most effective measure has been the placement of an antisiphon device, which preserves valve flow when the patient stands.

Whether or not to shunt all hydrocephalic infants soon after birth is a controversial issue. In several large series of cases that have been treated in this way, the number of surviving with normal mental function has been small. The series of Dennis and his associates is representative. They examined 78 shunted hydrocephalic children and found that 56 (72 %) had full-scale IQs between 70 and 100; in 22 patients, it was between 100 and 115; in 3 orients, lower than 70, and in 3 others, it was above 115. Mental junctions improved unevenly and performance scores lagged behind verbal ones at all levels. The use of the carbonic anhydrase inhibitor acetazolamide to inhibit CSF formation has not been successful in our hands, but a number of reports attest to its value. These authors believe that by giving 250 to 500 mg of acetazolamide orally each day, shunting can be avoided in both normal pressure and infantile hydrocephalus.