Catheterization of the cerebral aqueduct for obstructive hydrocephalus in infants

R. M. N. Crosby, M.D., Charles M. Henderson, M.D., and Ronald L. Paul, M.D.
Department of Surgery, Division of Neurological Surgery, University of Maryland School of Medicine, Baltimore, Maryland

Thirty hydrocephalic children with obstruction of the cerebral aqueduct were treated by aqueductal catheterization (interventriculostomy). The complications encountered in the early cases have led to the use of a two-shunt technique. The surgical mortality of 13% compares favorably with that of other methods, and the possibility of a permanent cure of the hydrocephalus eliminating the problems of shunts and shunt dependence makes this approach an appealing one to the authors.

KEY WORDS hydrocephalus · aqueductal stenosis · interventriculostomy · shunt independence

Cannulation of the cerebral aqueduct (of Sylvius) for relief of obstructive hydrocephalus is not a new idea. The first attempts reported by Dandy in 1920 using rubber catheters were not very successful. Although there were isolated subsequent reports of success, most similar trials were discouraging. Recently Greenwood and Hickey and Elvidge have reported more favorable results and have documented the efficacy of this procedure at least in selected cases.

Over the past 10 years, at the University of Maryland, the problem of congenital hydrocephalus has been approached with the idea that certain types of hydrocephalus can be corrected by direct surgical attack, thus giving a permanent cure and obviating indefinite dependence on external shunting devices. During this time, aqueductal cannulations (interventriculostomy) have been performed on 30 infants suffering from congenital aqueductal stenosis. The purpose of this report is to document the results of this direct surgical treatment.

Methods

Clinical Material

The series includes 30 infants and children ranging in age from 7 months to 5 years of age; 23 were less than 1 year old at the time of the aqueductal cannulation. Over half of the patients had at least one other malformation of the nervous system; 12 had meningomyeloceles, and nine had Arnold-Chiari malformations.

Initial Treatment

The protocol for the evaluation and treatment of all types of congenital hydrocephalus, at the University of Maryland, has been reported in detail elsewhere. A brief
Catheterization of the cerebral aqueduct

summary will be outlined here. As soon as the diagnosis of hydrocephalus is suspected, the type of hydrocephalus is identified by appropriate pneumoencephalography and/or air positive contrast ventriculography. After the specific type and site has been defined, a Pudenz ventriculoatrial shunt is inserted. The child is followed until evidence of nonfunctioning of this shunt occurs and signs of increased intracranial pressure appear. At this time, the shunt is revised or another shunt is placed on the opposite side. If necessary, this is followed by the definitive intracranial surgical procedure (interventriculostomy), but a functioning shunt is essential during the immediate postoperative period.

Operative Technique

The operations are performed with the patient in the face-down position. Exposure is accomplished through a midline suboccipital incision. Great care must be taken to cut between preplaced clips when sectioning the dura because large venous sinuses are usually found, perhaps as a result of alternate collateral venous drainage secondary to the shunting procedures. If an Arnold-Chiari malformation is encountered, appropriate exposure is accomplished through a more extensive laminectomy in order to adequately expose the caudal cerebellum. The roof of the fourth ventricle is then retracted upward with a small spatula, and the aqueduct visualized without resection of the midline cerebellar tissue. Stenosis is confirmed by the absence of fluid leaving the lower end of the aqueduct and the impossibility of passing a small round-ended 2.5 mm Silastic tube through it into the third ventricle. After the stenosis has been confirmed, the aqueduct is progressively dilated by passing progressively larger French urethral filiform catheters, starting with a No. 3 and ending with a No. 6. Following dilation, a piece of radiopaque Silastic tube 2.5 cm long with a small rim-like cuff on its lower end made by expanding a thin slice of the tube and placing it over this end is passed through the aqueduct and left in place on a plane parallel to the floor of the fourth ventricle. Proper tube position is heralded by free flow of cerebrospinal fluid (CSF), and droplets of positive contrast medium into the fourth ventricle (Fig. 1). The position of the tube can then be confirmed by a postoperative x-ray film (Fig. 2) and poor position corrected.

Follow-up Evaluation

Follow-up evaluation of the patients was performed in serial examinations of the function or nonfunction of the external shunting device.

Nonfunction was defined as the inability to compress an expanded Pudenz-Heyer clearing device, indicating occlusion of the cardiac end, or persistent nonexpansion of the clearing device indicating occlusion of the ventricular end. Patients judged to have successfully arrested hydrocephalus, based on clinical evaluation and head growth curves, and in whom the external shunting device was no longer functioning, were considered to have had a successful result from the aqueductal cannulation and to be shunt-independent.
Results

Mortality

Two deaths occurred in the immediate postoperative period; in both, CSF fistulas led to meningitis. The fistulas occurred probably as a result of persistent increased intracranial pressure due to conversion of an internal hydrocephalus to a communicating type. This problem will be discussed in more detail below. Two late deaths occurred at 1 month and 3½ years as a direct result of failure to control the hydrocephalus. Autopsy in these cases revealed obstruction of the midline fourth ventricle foramina due to adhesions to the cerebellum, presumably secondary to the resection of the vermis at the time of surgery. Resection of cerebellar tissue is now avoided for this reason. The overall surgical mortality rate was, thus, 13%. In no case did death occur as a consequence of surgical brain-stem injury.

Five subsequent deaths occurred that were unrelated to surgery or failure to control hydrocephalus. Three died between 1 to 3 years postoperatively from sepsis. All three had meningomyeloceles. Autopsy in these cases revealed no evidence of significant hydrocephalus; the interventriculostomy tubes were in their appropriate place, and were presumably functioning. Two children died suddenly at 3 and 4 months postoperatively. In one, autopsy again revealed no significant hydrocephalus, and the interventriculostomy tube was in its proper place. In the other, autopsy was not performed; however, there were no symptoms of acute increased intracranial pressure prior to its death. The overall mortality rate in this series was 30%.

Morbidity

Surgical Complications. Most surgical complications were due to improper interventriculostomy tube placement, postoperative migration of the tube, or a postoperative CSF fistula. There was no instance of permanent neurological impairment caused by cannulation of the cerebral aqueduct. In a few cases, transient impairment of conjugate eye movements, tachycardia, tachypnea, moderate hyperthermia or instability of systemic blood pressure were seen; these phenomena usually abated within the first 48 hours and presented no significant management problems.

In one case, improper tube placement occurred. The tube was apparently angled too far dorsally, and appeared to have entered the subarachnoid space in the region of the quadrigeminal cistern, presumably passing through the quadrigeminal plate (Fig. 2 right). Interestingly, no impairment...
Catheterization of the cerebral aqueduct

of ocular function was detected. The tube was subsequently correctly repositioned.

In two of the earlier cases in the series, migration of the interventriculostomy tube occurred, in one into the 3rd ventricle, in the other into the lateral ventricle. Although the first patient was asymptomatic, another tube was positioned correctly; the second patient had aseptic meningitis, requiring transcortical removal of the tube. It is interesting that both of these tubes migrated rostrally instead of caudally, as one might expect. Since this experience, the cuff already described has been placed on the caudal end of the tube to prevent rostral migration. There has been no subsequent instance of tube migration.

Cerebrospinal Fluid Fistulas. Cerebrospinal fluid fistulas in the immediate postoperative period occurred in four of the first 10 cases. Two died of meningitis as described, and two were successfully managed by placement of external shunting devices. As a result of this experience, posterior fossa exploration has been done only if there is a functioning external shunt to prevent symptomatic conversion from an obstructive hydrocephalus to a communicating type.

Failure to Control Hydrocephalus

Of the first 10 patients subjected to aqueductal cannulation in the absence of functioning external shunts, two died from meningitis as described, and two were successfully managed by placement of external shunting devices. As a result of this experience, posterior fossa exploration has been done only if there is a functioning external shunt to prevent symptomatic conversion from an obstructive hydrocephalus to a communicating type.


discussion

It has been postulated that in instances of congenital obstructive lesions, the subarachnoid space fails to develop in utero.\textsuperscript{1,8,10,11} Therefore, correction of an internal obstructive process would merely result in conversion of an internal hydrocephalus to a communicating variety.

Recently, Milhorat and his coworkers\textsuperscript{14} have demonstrated that the subarachnoid space does, in fact, develop normally in the presence of obstructive congenital lesions; 52 such cases examined histologically had a subarachnoid space. Twelve were perfectly normal and 40 showed various degrees of patchy fibrosis, the severity of which could be correlated with the duration of the hydrocephalic process, failure of adequate shunt control, or a preceding history of infection or subarachnoid hemorrhage. Milhorat, \textit{et al.},\textsuperscript{15} have also done radioisotope studies which demonstrate that in the presence of obstructive hydrocephalus the subarachnoid space does not function. It does, however, begin to function after successful external shunting procedures. They conclude, therefore, that the reason the subarachnoid space does not function in the presence of obstructive hydrocephalus is simply due to mechanical compression of the space by the expanded cortical mantle. Relief of the mechanical compression causes the space to open.

It is apparent that in certain cases surgical correction of obstructive hydrocephalus can immediately relieve the hydrocephalus. In our first 10 cases in which aqueductal cannulations were performed in the absence of functioning external shunts, seven required external shunting procedures subsequent to the aqueductal cannulations because of continued head enlargement. Other reports regarding operative correction of obstructive hydrocephalus reveal similar experiences. Matson\textsuperscript{13} reported eight cases
of direct surgical attack on obstructions to outflow from the fourth ventricle; 50% of these required subsequent external shunts. Turnbull and Drake reported four cases of direct surgical correction of congenital membranous occlusion of the cerebral aqueduct; one of these required postoperative shunting to control progressive head enlargement. Jones reported 10 cases of surgical extirpation of posterior fossa cysts, six of which required postoperative shunting. Whitten, et al., reported four cases of the Dandy-Walker syndrome corrected surgically, and two of these required postoperative shunts. Of these collected cases, exactly one-half manifested continued head enlargement and needed external shunting procedures.

Surgical correction of internal congenital obstructive lesions can effect immediate control of hydrocephalus in roughly 30% to 50% of the cases; the need for external shunting devices and all their complications can be eliminated in this group of patients. The low mortality rates appear to justify these operations. Furthermore, since the severity of the fibrosis of the subarachnoid space correlates with the duration of the hydrocephalus, early operation might increase the chance for success.

The question remains as to whether the subarachnoid space, which initially functions poorly following restoration of normal flow of fluid from the ventricular system, can ultimately function normally, particularly in the presence of a functioning external shunt. Does shunt dependency remain permanent? Milhorat, et al., have demonstrated that in the presence of a functioning external shunt the normal CSF pathway is not completely short-circuited. The potential for re-establishment of normal CSF absorption is therefore present.

Our clinical experience does suggest that restoration of normal subarachnoid space function can occur in some cases. We have seen, for example, several patients in whom an external shunt stopped functioning during the postoperative period and who then developed acute signs of increased pressure; their subsequent spontaneous remission was not associated with restoration of shunt function. These patients had, presumably, become shunt-independent.

By subjecting these patients to aqueductal catheterization (interventriculostomy) we have obtained a higher percentage of patients who are shunt-independent (63%) than could have been expected by chance or any other means. Ostensibly this means their hydrocephalus is cured. The remaining 37% who still have functioning external shunts could conceivably become shunt-independent in the future. With the passage of time, the percentage of shunt-independent patients will therefore rise from its already acceptable level. Ideally, one would like to be able to predict the patients whose subarachnoid space would function normally immediately following definitive surgical correction of the obstructing lesion. This, however, is not possible at current levels of knowledge.

References
Catheterization of the cerebral aqueduct


16. Rivila RA: Cited by Greenwood and Hickey, ref 7


This paper was presented in part at the meeting of the Society for Research into Hydrocephalus and Spina Bifida, London, June 19–21, 1969.

*Address reprint requests to: R.M.N. Crosby, M.D., 1010 St. Paul Street, Baltimore, Maryland 21202.*