Reexpansion of previously collapsed ventricles: the slit ventricle syndrome

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This study reports six cases of hydrocephalic children with the “slit ventricle syndrome” who evidenced reexpansion of the ventricular system following insertion of high-resistance valves and anti-siphon devices. The authors contend that slit ventricles and subsequent ventricular coaptation can be prevented by elimination or early replacement of low-resistance valves, and maintenance of normal- or nearly normal-sized ventricles by shunt revision with valve upgrade and/or an anti-siphon device, as judged by the appearance of the ventricles on computerized tomography.

KEY WORDS • slit ventricle syndrome • hydrocephalus • anti-siphon device • high-resistance valve • shunt

Since the development in 1952 of the valve-regulated shunt by Nulsen and Spitz, hydrocephalus has been successfully treated by various procedures. One of the well recognized complications of shunting has been that of overdrainage, resulting in slit or subnormal-sized ventricles. These events have created a relatively new phenomenon, the “slit ventricle syndrome.” This syndrome occurs in shunt-dependent patients and is characterized by the following triad of events: 1) intermittent or chronic headaches secondary to episodic ventricular catheter obstruction; 2) small ventricles documented by computerized tomography (CT) scan or ventriculogram; and 3) slowed refill of the palpable valve mechanism.

Presently, various therapeutic measures are employed in attempts to solve the problem of overdrained ventricles. We would like to recommend another therapeutic modality that should be added to the armamentarium of the neurological surgeon: the use of the anti-siphon device and high-resistance valve.

Summary of Cases

Patient Population

The six patients in our series (four males and two females) were aged 3 to 7 years. The hydrocephalus was of varied etiologies (Table 1). All of the patients had previously undergone multiple revisions of the shunt’s proximal end because of ventricular catheter obstruction. All patients had both CT and ventriculographic evidence of extremely small ventricles. In addition, they all presented with headaches, vomiting, malaise, and slowed refill of the palpable valve. This group of patients underwent placement of both high-resistance valves (pressure range 140 to 165 mm H₂O) and anti-siphon devices, with subsequent resolution of their symptomatology and reexpansion of the ventricular system. The following case is representative of the group.

Illustrative Case Report

This 5-year-old girl was initially treated as a neonate for hydrocephalus secondary to aqueductal stenosis. A ventriculoperitoneal shunt with a low-pressure (40 mm H₂O) Holter valve was placed (Fig. 1 left). At the age of 1 year, she presented with proximal ventricular obstruction, evidenced by extreme lethargy and vomiting (Fig. 1 right). The valve was upgraded to a medium-pressure Holter valve with a pressure of 95 mm H₂O, and she became asymptomatic. After an additional 3 months, she again suffered a ventricular block proximally, and her shunt was upgraded to a high-pressure (120 mm H₂O) Holter valve. One month later she was again brought for
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TABLE 1

Summary of six cases of “slit ventricle syndrome” with valve upgrade and placement of anti-siphon device

<table>
<thead>
<tr>
<th>Age (yrs)</th>
<th>Etiology of Hydrocephalus</th>
<th>Original Valve Type</th>
<th>Postop Headache</th>
<th>Postop Computerized Tomography Appearance</th>
</tr>
</thead>
<tbody>
<tr>
<td>5</td>
<td>aqueductal stenosis</td>
<td>low-pressure Holter (40 mm H₂O)</td>
<td>none</td>
<td>increased ventricular size</td>
</tr>
<tr>
<td>3</td>
<td>Arnold Chiari II</td>
<td>low-pressure Holter</td>
<td>none</td>
<td>increased ventricular size</td>
</tr>
<tr>
<td>4</td>
<td>4th ventriculocoele</td>
<td>low-pressure Holter</td>
<td>none</td>
<td>increased ventricular size</td>
</tr>
<tr>
<td>7</td>
<td>neonatal intraventricular hemorrhage</td>
<td>low-pressure Holter</td>
<td>none</td>
<td>increased ventricular size</td>
</tr>
<tr>
<td>3</td>
<td>aqueductal stenosis</td>
<td>low-pressure Holter</td>
<td>none</td>
<td>increased ventricular size</td>
</tr>
<tr>
<td>6</td>
<td>neonatal intraventricular hemorrhage</td>
<td>low-pressure Holter</td>
<td>none</td>
<td>increased ventricular size</td>
</tr>
</tbody>
</table>

FIG. 1. Left: Original air-Conray ventriculogram of neonatal aqueductal stenosis with hydrocephalus. Right: Air ventriculogram showing markedly small ventricles causing proximal ventricular catheter obstruction.

evaluation. A second coronal ventricular catheter was placed in the opposite ventricle, and the valve was changed to a high-pressure (140 mm H₂O) Hakim valve. Nine months later the patient developed proximal blockage, and a third posteriorly placed catheter was added (Fig. 2). She became asymptomatic. Subsequently, she complained of chronically intermittent headaches, and CT scans showed persistently slit ventricles (Fig. 3 left). Her valve, although functioning, refilled very slowly. The anti-siphon device was added 2 years after valve upgrade to a high-resistance valve. Subsequent CT scans have shown ventricular reexpansion of the occipital horns. She has remained asymptomatic over the following 2 years (Fig. 3 right).

Results

We have achieved ventricular reexpansion of slit ventricles by implementing the anti-siphon device and the high-resistance valve. This method has allowed ventricular reexpansion after failure with either valve upgrading or placement of an anti-siphon device alone. There was definite evidence of increased ventricular size. Interestingly, all of these patients had

FIG. 2. Air-Conray ventriculogram showing three ventricular catheters in place.
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FIG. 3. Plain computerized tomography scans. Left: The patient has persistent slit ventricles. Right: After placement of the anti-siphon device, the uncontrasted scan shows biparieto-occipital reexpansion of the ventricular system.

low-pressure valve systems at the time of their original shunting. Retrospectively, it has been our observation that patients who develop slit ventricles, whether symptomatic or not, had either low-pressure valves or valves that through incompetence registered low pressures. The patients in this group have been followed for 3 to 6 years and the results suggest a successful treatment.

Discussion

Inadequate ventricular catheter patency secondary to small ventricles was noted in the early 1960’s, as reported by Becker and Nulsen. They recommended upgrading valves when confronted with small ventricular systems. In our large population of shunted patients in Cleveland, we have often succeeded in resolving symptomatology by simply placing a higher-resistance valve; however, the clinical im-

FIG. 4. Computerized tomography scans showing slit ventricles (left), temporary bilateral occipital horn dilatation immediately following subtemporal craniectomy (center), and recurrent slit ventricles 3 years after subtemporal craniectomy (right).

provement was usually not accompanied by ventricular enlargement.

In 1974, Epstein and his co-workers suggested that subtemporal craniectomy would alleviate recurrent shunt obstruction secondary to small ventricles, and concomitantly would allow ipsilateral ventricular dilation for catheter replacement. Subsequently, other authors have supported subtemporal decompression in the treatment of slit ventricles. We elected to try this approach in one patient with aqueductal stenosis who, from the neonatal period to adolescence, had experienced multiple episodes of ventricular catheter blockage. This patient had essentially nonvisible ven-

FIG. 5. Subtemporal craniectomy for decompression of proximal shunt obstruction.
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Ventricles (Fig. 4 left). Since infancy, he had been managed by our conventional approach, that is, replacement of ventricular catheters and valve upgrading. At 17 years of age, when he again presented with proximal obstruction, he underwent subtemporal decompression (Fig. 5). There was clinical improvement and temporary bilateral occipital horn dilatation (Fig. 4 center). However, he subsequently presented with several episodes of proximal blockage and was last treated by valve upgrading. Two years later, his CT scan shows extremely slit-like ventricles (Fig. 4 right).

We agree that this approach vented the acutely increased intracranial pressure (ICP), and that the surgical fontanel provided an excellent gauge for checking ICP; however, only transient ventricular dilatation resulted, and the patient was still susceptible to proximal shunt dysfunction. Thus, subtemporal decompression did not solve this patient's problem of recurrent ventricular catheter obstruction. We concur that subtemporal craniectomy can be a valuable maneuver when one is confronted with rapid decompensation secondary to shunt obstruction and concomitant acutely increased ICP.

We have accomplished ventricular reexpansion of slit ventricles by implementing the anti-siphon device and the high-resistance valve. The anti-siphon device is placed in series distal to the valve. This device was first described in 1973 by Portnoy, et al., and was designed to reduce the negative intraventricular pressure noted when the patient is in a sitting or standing position.

We believe that one should employ prophylactic measures to prevent slit ventricles by avoiding the use of low-pressure valves and by follow-up review of all shunted patients with CT scans at the age of 1 year and serially as necessary. Whenever slit ventricles are encountered, even in asymptomatic patients, one should upgrade the valve and add an anti-siphon device. We are not suggesting that our proposed method of management is a simple cure to this complicated problem; however, for us, it has proven to be the most effective approach in combating the iatrogenic complication of slit ventricles.

References


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