Malfunctioning ventriculoperitoneal shunts

Clinical and pathological features

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The authors report the clinical and pathological findings in 201 procedures to revise malfunctioning ventriculoperitoneal shunts in 114 patients. The revised shunts were predominantly Raimondi three-piece systems. Collection of fluid along the shunt track is emphasized as an early clinical feature of shunt malfunction. In some cases, there are few clinical signs of shunt infection, and the first indication of it may be shunt malfunction. Ventricular catheter obstruction was caused by tissue from the region in addition to choroid plexus. Inflammation was frequently found around both ventricular and peritoneal catheters. Foreign bodies (cotton fibers, hair, and talc) and granulomatous inflammation were often present at both ends. Tissues obstructing the peritoneal catheter included embolic neoplastic cells, choroid plexus, and leptomeninges. There was also more necrotic debris at the peritoneal end. To prevent shunt malfunction, attention should be directed to: 1) optimal placement of ventricular catheters inside the lateral ventricle; 2) prevention of infection; 3) avoidance of contamination by cotton fibers, hair, or talc; and 4) improvements in the biocompatibility of the implanted materials.

KEY WORDS • cerebrospinal fluid shunt • hydrocephalus • spina bifida • ventriculoperitoneal shunt

Mechanical malfunction and infection are the most significant problems associated with shunts for the treatment of hydrocephalus. The Silastic material currently used in shunts is associated with less severe tissue reaction than the polyethylene tubes used earlier.\(^1\),\(^3\),\(^4\),\(^5\),\(^6\) Despite the current use of a variety of mechanical shunting systems, shunt malfunction remains a major problem in pediatric neurosurgical practice.\(^5\),\(^6\),\(^7\),\(^8\),\(^9\),\(^10\),\(^11\),\(^12\) Pathological lesions associated with malfunctioning shunts have been studied only infrequently.\(^2\),\(^3\),\(^4\),\(^5\),\(^6\) We report the clinical features and the pathological nature of obstructing materials in a series of malfunctioning ventriculoperitoneal (VP) shunts.

Materials and Methods

We performed 201 shunt revisions on 119 patients at the Children's Hospital of Pittsburgh over a period of 16 months. In infants, the major indications for shunt revision were: an abnormally enlarging head, full tense fontanels, lethargy and/or irritability, persistent fluid collection along the shunt track, and abnormal neurological signs such as restriction of upward gaze and spasticity. In older children, the indications were persistent headaches, abnormally enlarging head, deteriorating mental function, papilledema, and an increased frequency of seizures. Enlargement of the lateral ventricles was documented preoperatively by computerized tomography (CT) in all of the older children and many of the infants. In a few cases, progressive asymptomatic ventriculomegaly was the indication for shunt revision.

All but five of the shunts we revised consisted of a Codman ventricular catheter connected to a Raimondi peritoneal catheter and slit valve, either through a right-angle connector or through a Selker or Rickham reservoir.* One Hakim Cordis and four

* ACCU-FLO clear ventricular catheter (silicone rubber, 1.3 mm internal diameter), ACCU-FLO distal slit-valve Raimondi peritoneal catheter, right-angle ACCU-FLO connector, and Selker and Rickham reservoirs manufactured by Codman and Shurtleff, Inc., Randolph, Massachusetts.
Holter systems were also revised.† Most ventricular catheters had been inserted initially through parietal or occipital burr holes. Peritoneal catheters had been inserted approximately 25 cm through incisions in the right or left upper abdominal quadrants.

For revision of a shunt, the operative site was prepared by shaving the scalp, and the shaved area and the ipsilateral side of the neck, chest, and abdomen were cleansed with Povidone-iodine surgical scrub for 5 minutes and painted three times with Povidone-iodine solution. The cranial incision was reopened, and, if the shunt was still in continuity, the peritoneal and ventricular ends were disconnected and tested to determine which end was malfunctioning, whereupon that end was removed. If the shunt system had become disconnected, both ends were removed.

The shunts that were removed were submitted for pathological examination. The material found inside and outside the shunt catheters was examined histologically. Sections were routinely stained with hematoxylin and eosin. Special stains, such as Gram, Masson's trichrome, Nissl, and phosphotungstic acid hematoxylin (PTAH) were used in selected cases.

Results

Patient Population

Of the 119 patients, 75 were male and 44 female. The hydrocephalus was obstructive in 92, communicating in 21, and of undetermined type in five patients. Associated disorders included aqueductal stenosis, myelomeningocele (with or without aqueductal obstruction), Dandy-Walker malformation, cerebral tumor, intraventricular hemorrhage, encephalocele, and postmeningitic scarring of the subarachnoid space pathways. More than one disorder was present in some patients, such as aqueductal stenosis and intraventricular hemorrhage. The mean age of the patients at the time of revision was 32.6 months, with a range of 1 to 219 months. Seventy-two patients had undergone previous shunt revisions and 62 patients had a history of intraventricular infection.

The presenting signs and symptoms are listed in Table 1. The mean duration of symptoms at the time of revision was 11.5 days. The appearance of fluid around the shunt track deserves special mention, and 43 patients exhibited this finding, usually along the cranial part. In 40 of these patients, shunt malfunction was confirmed at surgery: the peritoneal end was obstructed in 40 instances, the ventricular end in 17, and both ends in nine. In one patient, both ends were patent but malfunction occurred because of disconnection.

Operative Findings

In 186 revisions, the cause of the malfunction was identified. In 163 cases, malfunction was found in shunt systems that had remained connected; the shunt had become disconnected in 18 instances; and the peritoneal catheter had retracted outside the abdomen in five cases. The ventricular end was obstructed in 73, the peritoneal end in 62, and both ends in 47 revisions. In four instances no obstruction was found, but the malfunction was due to disconnection alone. In 15 cases, unequivocal evidence of the cause of malfunction was not found at surgery.

Pathological Findings

Ninety-one ventricular ends and 50 peritoneal ends were submitted for pathological examination. No obstructive lesions were found along the course of the shunt except where there was disconnection, and in those specimens it was not uncommon to find fibrous tissue ensheathing the disconnected ends.

Gray-white to yellow-brown material, sometimes further discolored by hemorrhage, was often found inside the perforated part of the tubing (first 15 mm) at the ventricular end. Material sometimes extruded from the tube through its holes. At the peritoneal end, the most common finding was yellow-white material inside and outside the tubing in the region of the slits. At either end, obstructive material rarely extended inside the lumen of the shunt for more than a few millimeters beyond the region of the holes.

Histopathological findings were categorized under five major headings: 1) tissue normal for the region; 2) pathological tissue; 3) foreign bodies; 4) embolic

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† Hakim Cordis shunt system manufactured by Cordis Corp., P.O. Box 525700, Miami, Florida. Holter shunt system manufactured by Codman and Shurtleff, Inc., Randolph, Massachusetts.
TABLE 2
Histopathology of obstructive lesions

<table>
<thead>
<tr>
<th>Pathological Findings</th>
<th>Ventricular End</th>
<th>Peritoneal End</th>
</tr>
</thead>
<tbody>
<tr>
<td>tissues normal for the region</td>
<td>35 0</td>
<td>0 0</td>
</tr>
<tr>
<td>choroid plexus</td>
<td>36 0</td>
<td>0 0</td>
</tr>
<tr>
<td>glial tissue</td>
<td>21 0</td>
<td>0 0</td>
</tr>
<tr>
<td>leptomeninges</td>
<td>15 0</td>
<td>0 0</td>
</tr>
<tr>
<td>ependyma</td>
<td>6 0</td>
<td>0 0</td>
</tr>
<tr>
<td>brain</td>
<td>49 33</td>
<td>0 17</td>
</tr>
<tr>
<td>connective tissue</td>
<td>0 17</td>
<td>17 3</td>
</tr>
<tr>
<td>mesothelial cells</td>
<td>17 22</td>
<td>5 24</td>
</tr>
<tr>
<td>pathological tissues</td>
<td>10 3</td>
<td>17 7</td>
</tr>
<tr>
<td>fibrin</td>
<td>13 17</td>
<td>45 28</td>
</tr>
<tr>
<td>necrotic debris</td>
<td>6 7</td>
<td>44 23</td>
</tr>
<tr>
<td>hemorrhage</td>
<td>7 0</td>
<td>6 7</td>
</tr>
<tr>
<td>inflammation</td>
<td>6 4</td>
<td>4 1</td>
</tr>
<tr>
<td>acute</td>
<td>10 7</td>
<td>5 4</td>
</tr>
<tr>
<td>chronic</td>
<td>5 4</td>
<td>6 7</td>
</tr>
<tr>
<td>granulomatous</td>
<td>3 1</td>
<td>1 1</td>
</tr>
<tr>
<td>calcification</td>
<td>5 12</td>
<td>0 1</td>
</tr>
<tr>
<td>neoplastic cells</td>
<td>0 1</td>
<td>0 2</td>
</tr>
<tr>
<td>foreign bodies</td>
<td>0 1</td>
<td>0 1</td>
</tr>
<tr>
<td>cotton fibers</td>
<td>8 15</td>
<td>0 1</td>
</tr>
<tr>
<td>hair</td>
<td>0 1</td>
<td>0 1</td>
</tr>
<tr>
<td>talc granules</td>
<td>0 1</td>
<td>0 1</td>
</tr>
<tr>
<td>bacteria</td>
<td>0 1</td>
<td>0 1</td>
</tr>
<tr>
<td>unidentified material</td>
<td>0 1</td>
<td>0 1</td>
</tr>
<tr>
<td>no luminal tissue</td>
<td>0 1</td>
<td>0 1</td>
</tr>
</tbody>
</table>

FIG. 1. Material from the ventricular end of a shunt, with hair (arrow) and cotton fiber (arrowheads) embedded in the tissue. Polarized light. H & E, × 80.

FIG. 2. Material from the peritoneal end of a shunt with talc granule exhibiting a typical Maltese cross appearance (small arrow), cotton fiber (large arrow), and an unidentified foreign body (arrowhead). Note the foreign-body giant-cell reaction. Polarized light. H & E, × 80.

FIG. 3. Computerized tomography scans of a patient with tuberous sclerosis and an intraventricular giant-cell astrocytoma (arrowheads). Left: Unenhanced scan showing the shunt catheter passing directly through the tumor mass (arrowheads). Right: Contrast-enhanced scan at a slightly different level showing the enhancing tumor mass (arrowhead) and the top of the ventricular catheter (arrow). This patient had tumor cells in the ventricular catheter on two occasions and embolized tumor cells in the peritoneal end on one occasion.
in the patient with the giant-cell astrocytoma (Fig. 4). This was not associated with clinical evidence of metastasis, presumably because this tumor is not as invasive as medulloblastoma.

Discussion

The presenting signs and symptoms of shunt malfunction in our patients were similar to those recorded in other reports. However, since the Raimondi three-piece shunt system was used in almost all of our patients, it is possible that some of the clinical and pathological findings of the study were peculiar to this system. In particular, the appearance of fluid along the shunt track was considered reliable evidence of shunt malfunction, but this inference may not be applicable to other systems.

There is evidence that low-grade infection is an important cause of shunt malfunction. It may occur without clinical evidence of sepsis. Acute inflammatory cells were found in the shunts of 28 of our patients, but in only two was infection proven by culture of pathogenic organisms. On the other hand, seven patients with evidence of infection established by culture showed only chronic inflammation or none at all.

Malfunction of shunts may also be considered separately as occurring at the ventricular end, valve, and peritoneal end. Ventricular catheters contain side holes in the first 15 mm from their tip, and the patency of these holes is essential for proper function. Astrocytes and fibroconnective tissue are especially capable of proliferation and may fill the holes and the inside of the catheter. Choroid plexus and ependymal cells may also have a proliferative capability under some pathological conditions. Choroid plexus has generally been considered to be the most frequent cause of ventricular catheter obstructions. Hakim found plexus to be the obstructive material in 80% of "15 or more" catheters examined, but he also found other tissues including brain, hyalinized connective tissue, hemosiderin, and a mixture of protein, fibrin, and leukocytes. Collins, et al., reported scanning electron microscopic characteristics of the surface tissue on 16 obstructed ventricular catheters. Choroid plexus was found frequently in catheter tips placed in behind the foramen of Monro, but ependyma predominated in catheters placed in front of it. Blood clot was present in both locations. Their observations suggested that the nature of the obstructive agent was related to the position of the catheter; but the number of observations was too small for statistical analysis.

Prolonged shunt function appears to follow placement of ventricular catheters in the frontal horn, anterior to the choroid plexus. Catheters placed through occipital or parietal burr holes will migrate with head growth from an initial frontal horn position into the body of the lateral ventricle, where they may be obstructed by choroid plexus. The tips of frontally placed catheters may migrate into the frontal brain substance as the ventricles diminish in size with resolution of hydrocephalus. Vries advocated the use of ventriculoscopy to place ventricular catheters in an optimal position inside the frontal horn through a parasagittal burr hole at the level of the coronal suture. With this technique, he reported prolonged shunt function in 85 patients treated over 18 months.

In the patients reported here, not only choroid plexus and ependyma, but also leptomeninges, glial tissue, connective tissue, and brain tissue (ependymal astrocytes, neurons, and white matter) were found. Leptomeninges may have been pushed into the shunt tubing as it was inserted or derived from the inadvertent placement of the catheter in the subarachnoid space. Brain tissue was probably introduced into the catheter during insertion or removal, or resulted from subependymal placement of the catheter.

The frequency of granulomatous inflammation with foreign-body giant cells is not surprising in view of the variety and number of foreign bodies (Table 2). The fact that they were embedded in the surrounding tissues and surrounded by multinucleated giant cells indicates that they were not artifacts picked up during the processing of slides. Cotton fibers were frequently seen and presumably originated from cottonoids or cotton sponges used during the original insertion of the shunt. Human hair may be from the patient or from operating room personnel, most likely the former. Talc granules, which have a typical Maltese cross appearance, were also found frequently and probably came from either the gloves of operating room personnel or from the talc used to powder the infants.

There is little information about pathological lesions obstructing the peritoneal end of the catheter or the factors that govern continued function of the peritoneal end. Acute and chronic inflammatory changes were as frequently present at the peritoneal end of the shunt as at the ventricular end. It is not surprising that chronic inflammation continues in a fluid-filled system as a barrier to spread of infection.

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dend as at the ventricular end in our patients. Our comments about low-grade infection and shunt malfunction apply to the peritoneal end also, where granulomatous inflammation and foreign bodies were frequently found.

Hoffman, et al., reported the possibility of embolization of medulloblastoma cells through VP shunts. Peritoneal metastasis without local recurrence or other systemic metastasis suggested that seeding of the neoplasm to the peritoneal cavity occurred via the shunt. We have provided confirmatory evidence for this hypothesis by finding tumor cells within the shunt catheter. In our patients, embolization of normal tissues, such as choroid plexus and leptomeninges, via the shunt catheter probably contributed to peritoneal end malfunction. Embolic material may also become necrotic inside the catheter, which may explain why necrotic debris is more frequent at the peritoneal end than at the ventricular end.

Catheter obstruction due only to a thin film of tissue on the surface, with apparently “no tissue inside the lumen,” occurred more commonly at the peritoneal end than at the ventricular end. However, many shunts showed evidence of invasion of the lumen by adjacent tissues. Such cellular invasion may be stimulated by proteinaceous fluid, embolized from the ventricular end, or may be due to local tissue reactions to the plastic catheter.

Conclusions and Recommendations

Previous studies have emphasized the importance of optimal placement of the shunt catheter and the avoidance of infection in order to reduce the incidence of shunt malfunction. Low-grade chronic infection may also be an important factor in shunt malfunction. This fact emphasizes the need for scrupulous attention to aseptic neurosurgical technique and comprehensive microbiological studies. In addition, we frequently found foreign-body contamination. To prevent such contamination, we suggest the following: 1) avoid the use of talc on the anterior surface of the trunk in babies in the 48 hours preceding a shunt operation; 2) wet the patients’ hair thoroughly before shaving; 3) use cottonoids and sponges minimally during the operation. Improving the biocompatibility of implanted materials may reduce proliferative reactions of tissues. Optimal positioning of ventricular catheter tips may not only improve ventricular end function, but may also reduce obstruction of the peritoneal end by reducing embolization of tissue through the catheter.

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References


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