Complications of Ventriculovenous Shunts

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The history of ventriculovenous shunts began with Gärtnner who suggested the concept in 1895, at the National Science Convention at Lübeck. Payr\(^1\) announced the results of his initial attempts at this problem in 1908. The further history of ventriculovenous shunts has been reviewed\(^5\) in the recent past. Ingraham et al.\(^10\) expressed the necessity for a sensitive one-way valve to cope with the problem, but it was not until after the work of Nulsen and Spitz in 1951,\(^15\) and that of Pudenz et al.\(^19\) in 1957 that the procedure met with any degree of success. It has been estimated\(^22\) that over five thousand procedures of this type had been performed by 1960. Little has been published in the English literature about the difficulties which have been, or may be, encountered in the management of the “proximal”-valve (Spitz-Holter) or “distal”-valve (Pudenz-Heyer) ventriculovenous shunts in their various modifications.\(^15,14,19,21,27\)

As has been suggested,\(^1,5\) ventriculovenous shunting is fairly universal in applicability to palliate hydrocephalus, obstructive and communicating, acquired and congenital. Situations that may lead to unfavorable results are those of hydrocephalus associated with increased right atrial and venous pressure from any cause. Thus a higher than desired intraventricular pressure may be necessary to have a sufficient gradient of pressure to initiate flow through the valve. No reports of failure to install a ventriculovenous shunt have been encountered. This possibility could have arisen if one considers a case described by Hooper,\(^9\) when hydrocephalus developed as a manifestation of thrombosis of the superior vena cava.

The mechanically intact device must be placed properly to function successfully. Operators are advised\(^18\) to introduce a 6–8 cm. ventricular segment. This portion may be obstructed\(^6,8,11,18,26\) by fragments of brain or blood clot incident to the trauma of introduction.

The placement of the intravenous portion of the shunt can become involved. Considering the cervical anatomy to be normal, and the isolation of the cervical veins accomplished, placement of the auricular segment may be guided by electrocardiographic\(^14,23\) or roentgenographic\(^19,28\) techniques. The passage of this limb may be complicated by deviations into the subclavian vessels\(^23\) or the tube folding upon itself.\(^13\) The latter circumstance would provide a confusing picture to the operator dependent solely upon electrocardiographic guidance. Roentgenography usually requires the use of a contrast medium, with its attendant dangers, to insure accurate positioning. With placement of the shunt as recommended,\(^1,14,18,19,23,26\) in the right heart, inadvertent entry into the right ventricle may precipitate cardiac arrhythmias\(^14\) leading to ventricular fibrillation. Fortunately this rarely occurs as noted in several series of right-sided cardiac catheterizations.\(^30\) Fixation of the system at various points (dura mater, vein, joints, and valve) may lead to disruption or occlusion of the shunt.

Immediate postoperative difficulties may arise with the rapid conduction of cerebrospinal fluid into the intravascular compartment. In a small hydrocephalic infant this conceivably could precipitate acute heart failure. The rapid changes in the dynamics of the intracranial fluid may result in symptoms of intracranial hypotension.\(^11\) In a child the rapid collapse of the ventricles may be accompanied, as suggested by Dandy,\(^5\) by injury to the bridging veins with the subsequent development of a subdural hematoma. McNab\(^14\) reported 2 deaths with the loss of large amounts of fluid during a shunting procedure. Conversely the valve mechanism may have too high an opening pressure and

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intracranial hypertension may persist.\textsuperscript{11}

Most of the literature regarding complications deals with the more remote postoperative period. Delayed occlusion of the proximal limb may occur\textsuperscript{1,6,8,26} from becoming embedded in the ventricular wall, brain fragments occluding the lumen, plugging by the choroid plexus, delayed hemorrhage and formation of clots.

Placement of the ventricular tube may incite a ventriculitis\textsuperscript{26} or rekindle a dormant meningitis.\textsuperscript{1} Organisms present then may be widely disseminated through the blood by direct introduction into the right atrium.\textsuperscript{1,6,11,15,26}

Infections of the wound in the presence of a foreign body may be stubborn in resolving. The delayed breakdown of thin skin or intertriginous tissues over the implanted tubes, valve and pump occurs occasionally. These lesions may resist healing until the shunt is removed, and the area may act as a focus of infection leading to a devastating bacteraemia.\textsuperscript{6,25}

The valve may become incompetent. Clotting of blood refluxing into the system leads to occlusion. With the shunt occluded, cerebrospinal fluid may escape along the course of the system, forming smoothly lined pseudocysts in communication with the ventricle.\textsuperscript{11}

The Spitz-Holter valve has been shown to act on occasion as a site for bacterial colonization.\textsuperscript{2,4,25} The ensuing bacteraemia is highly resistant to therapy. Cohen and Callaghan\textsuperscript{2,4} have defined a syndrome characterized by pyrexia, splenomegaly, progressive anemia and positive blood cultures associated with Spitz-Holter shunts. Similar studies have not been performed with Pudenz-Heyer valves, but cases of septicaemia developing in their presence are known.\textsuperscript{1}

The venous limb appears to be the principal site of postoperative complications. The cephalad migration of the cardiac segment with growth of the infant has been commented upon.\textsuperscript{1,8,19} Emphasis\textsuperscript{1,14,18,19,23,26,28} has been placed upon positioning the orifice into the atrium to assure function for the longest possible time. Atrial mural thrombi and ulceration are known to occur,\textsuperscript{1,7,11,26} as well as lesions involving the tricuspid valve. These\textsuperscript{6,7} may lead to embolization\textsuperscript{1,7} or serve as a nidus for bacterial colonization leading to a clinical picture resembling bacterial endocarditis.\textsuperscript{2,4,6,25} Techniques had been proposed\textsuperscript{19} to allow for caudal migration of the entire system with growth of the infant. The shunt may shift prematurely, passing into the right ventricle with untoward consequences.

The response of tissues along the course of the plastic system is variable.\textsuperscript{1,6,7,11,26} Thin membranes may invest the entire subcutaneous and intravascular portions of the shunt. Fibrinous sleeves may form or other reactive changes of variable intensity.\textsuperscript{1,7,26} Thrombi may develop along the shunt and propagate, partially or completely occluding the superior vena cava\textsuperscript{7,11} or its tributaries. A local pericarditis was noted by Emery and Hilton\textsuperscript{7} over an area of vegetation possibly resulting from trauma by the catheter. Johnson\textsuperscript{1} reported a transitory pericardial effusion of cerebrospinal fluid relieved by aspiration.

Revision of the system carries additional hazards. The scarified tissues are more difficult to manipulate, especially the fibrosed vessels. Adjustment of the venous segment may require removal and replacement. The Holter vein-bore* when mishandled could result in perforation of a major vessel. A dislodged thrombus could result in massive pulmonary infarction. Transporting the shunt to the opposite side is not without hazard. Depending upon the collateral vessels present, interruption of the second jugular vein may lead to an untoward result.

Pappas\textsuperscript{26} has reported craniosynostosis developing with the approximation of the parietes after a successful ventriculo-atriostomy.

The mortality, incidence of septicemia and venous thrombosis, and the number of initial revisions reported in various series are given in Table 1.

**Case Report**

The patient, a male child, weighed 3 lbs. 15 oz. at birth on Nov. 15, 1960. The circumference of

* Holter Surgical Specialties and Precision Instruments, Catalog #VC-440.
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TABLE 1

Mortality, septicemia, venous thrombosis and initial revisions in various series

<table>
<thead>
<tr>
<th>Series</th>
<th>No. of Cases</th>
<th>Mortality</th>
<th>Septicemia</th>
<th>Venous Thrombosis</th>
<th>Initial Revisions</th>
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<tr>
<td>Anderson</td>
<td>36</td>
<td>12</td>
<td>6</td>
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<td>8</td>
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<td>Carrington</td>
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<td>Cohen &amp; Callaghan</td>
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<td>Elkins &amp; Fonseca</td>
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<td>5</td>
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<tr>
<td>Emery &amp; Hilton</td>
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<td>7</td>
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<tr>
<td>Grotte &amp; Lundberg</td>
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<td>11</td>
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<td>Jensen &amp; Amador</td>
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<td>55</td>
<td>11</td>
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<tr>
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<td>10</td>
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<td>4</td>
<td>2</td>
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</table>

* Necropsy cases only reported.

** Reported as suppurrative emboli in lung, not as septicemia.

† Mortality in 343 cases observed for 1.5+ years.

‡ Mortality in cases with bacteremia, not reported as total.

his head was 27 cm. and of the chest 24 cm. The neonatal period was complicated by a hyperbili-
rubinemia reaching 23.7 mg. per cent (the lowest recorded hemoglobin was 9.4 gm. per cent). He was referred to the Neurological Division on May 21, 1961 because of "an abnormally large head and delayed development." Ventriculograms were interpreted by the neuroradiologist to be consistent with a degree of stenosis of the foramina of exit of the 4th ventricle.

1st Operation. Exploration of the posterior fossa revealed no gross pathology of the foramen of Magendie, and a Torkildsen shunt was installed.

Postoperative course was complicated by meningitis, which responded to penicillin, sulfadiazine and chloramphenicol. The head continued to enlarge.

2nd Operation. On Aug. 16, 1961 (1 month after the exploration) a Pudenz-Heyer shunt was installed and positioned under electrocardiographic guidance.

Postoperatively the child did well, and the size of the head stabilized.

Three months later (Nov. 8, 1961) he was readmitted because of sudden increase in size of head and the development of cystic masses at the site of the occipital scar, and along the tract of the shunt. He had been growing and developing well. Surgical intervention was postponed on this occasion and on a subsequent admission because of intercurrent upper respiratory infections.

3rd Operation. Revision of the shunt was performed on Jan. 4, 1962. The cardiac limb of the shunt had been cut through by the ligature at the connector (a cuff of attached plastic could be seen). The distal limb was found in the neck, at the base of the smoothly lined pseudocyst, which communicated with the ventricle through the connector. This limb of the shunt was found obstructed in situ. Withdrawing it from the anterior facial vein, a new shunt (opening at 10 cm. water pressure) was introduced. Manipulation tore the fibrotic wall of the vein, and the internal jugular vein was cannulated directly. Electrocardioscopy revealed no changes in pattern as more than the anticipated length of shunt was inserted. Introducing positive contrast medium (2 cc. of 60 per cent Renografin) into the shunt, roentgen-ray films showed it folded upon itself in the superior vena cava. The limb was withdrawn and replaced with ease. Further radiological studies revealed its orifice in the right ventricle. It was retracted until a biphasic p-wave was noted on the cardiogram. No arrhythmia was noted. Securing the shunt in the jugular vein, the central end was attached to a Heyer pump (the cephalic limb being modified electively). A redundancy was permitted to exist in the subcutaneous cervical segment.

Postoperatively therapy with tetracycline was instituted. On the 3rd postoperative day the patient became irritable and a spiking fever (102.8°F.) developed. Roentgenogram of the chest revealed no pneumonia. The cardiac limb of the shunt was not identified. A hemogram revealed the count of white blood cells to be 8,600/mm.3 with a marked lymphocytosis. The fever subsided on the 6th postoperative day, but the child remained irritable. Tetracycline was discontinued.
Films of the chest on Jan. 15, 1962 revealed an increased cardiac silhouette suggestive of a pericardial effusion. The hemogram now showed a count of white blood cells of 21,900/mm$^3$ with a polymorpholeukocytosis. Electrocardiography showed a sinus rhythm.

A pericardiocentesis was performed and also a cisternal puncture. The occipital fluid (presumably in communication with the lateral ventricle) and pericardial fluid were grossly, microscopically and chemically dissimilar. Transitory improvement followed the pericardial tap, but it had to be repeated the next night. The cardiac silhouette then appeared stable by roentgen ray. A persistent low-grade fever developed. Serial cultures of blood and pericardial fluid were sterile.

On Feb. 11, 1962 the patient suffered an episode of cyanosis and dyspnea. The cardiac silhouette had increased in size. Pericardiocentesis was performed with no gross decrement in the pericardial shadow being noted on a subsequent film. He deteriorated and died on Feb. 18, 1962.

The shunt functioned well throughout the course of the illness. The pseudocysts collapsed; the fontanelle was soft; the suture lines were closed.

Autopsy (Figs. 1 and 2) demonstrated the patent shunt had passed through the tricuspid valve and insinuated itself between the chordae tendinae of the marginal cusp. The right ventricular wall had been perforated, and 2 cm. of the limb had passed into the pericardial sac. The length of the shunt in the superior vena cava was coated with fibrinous material and had been incorporated partially into the wall of the vessel. The brain showed a patent, enlarged ventricular system and evidence of the shunting procedures. The suboccipital pseudocyst was in communication with the cisterna magna. Atelectasis of 90 per cent of both lungs was noted.

Discussion

Ventriculo-venous shunting offers the most physiologic approach available to palliate hydrocephalus. In view of the evidence
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The dissected specimen demonstrates the shunt locked between the chorda tendinae of the marginal cusp and penetrating the ventricular wall. Note the coated caval segment of the shunt.

Presented by Laurence,12 the thickness of existent cortex in infants offers no indication of prognosis in any individual case arrested. Taylor et al.29 commented upon the remarkable expansion of cerebral cortex in arrested hydrocephalus. Attempting to establish the etiology of the condition, and attacking the problem at that level is obvious. Increasing numbers, however, will be treated by the procedure in any one of the modifications.

Contraindications to the performance of the operation are less clear. Patients with severe brain damage and profoundly moribund individuals are not good candidates. Introduction of a shunt in the presence of infectious intracranial disease may lead to its generalized dissemination. The cardiopulmonary status of the subject must be evaluated carefully, especially in the old and very young. The presence of myocardial failure or cor pulmonale would indicate using a very low-pressure valve or selecting a different form of shunt. In the infant, hydrocephalus may be associated with other anomalies including those of the cardiopulmonary system. The case described by Hooper9 obviates the current mode of ventriculovenous shunt.

Pudenz and associates13,19 and Nulsen and Spitz15,25 described placement of the cardiac limb under radiographic control, the latter utilizing an image intensifier. Ease of positioning would be enhanced were the end fitted with a radio-opaque marker. Tracking the orifice with growth in children would be facilitated, and it would have helped provide the answer to the problem posed in the case presented.

The patency of the Spitz-Holter and Pudenz-Heyer systems can be verified by the behavior of their pumping action. Sites of obstruction can be deduced by analyzing the nature of the malfunction. At times a sluggish shunt will perform better after being flushed. The advisability of percutaneous needling of a system is questionable because of possible contamination of the shunt.
When the need for revision arises the case should be reevaluated. It may be advisable to resort to a different route. This is true especially when difficulty is encountered in recanulatting the same system of veins.

Meticulous asepsis and scrupulous technique help assure the best results in each case.

Summary

The possible complications arising from ventriculo-venous shunts were discussed.

An unusual case of ventricular perforation by the cardiac limb of the shunt was presented.

Suggestions in evaluating candidates and improving results were made.

References