Isolated Unilateral Hydrocephalus Following Ventriculoatrial Shunt*

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A shift of the brain stem may cause sudden death following decompression of one lateral ventricle by a ventriculoatrial shunt if there is a lack of communication between the two lateral ventricles. Isolated loculations of a ventricle following ventriculitis may have a similar effect. This is essentially an acute form of unilateral hydrocephalus.

Post-surgical instances of this type of isolated hydrocephalus are apparently quite unusual. In DeLange's monograph on hydrocephalus, no mention is made of this syndrome. However, in his list of autopsy cases, there were two patients who had an occlusion of the foramen of Monro. One patient had died 8 months after the insertion of the shunt and the second, 17 days after the insertion of the shunt. The clinical course is not given. Emery performed autopsies on 44 patients with shunts all of whom had myelomeningocele and hydrocephalus. Unilateral hydrocephalus was not demonstrated in any of these patients. Numerous articles have been written on complications associated with ventriculostomy shunting of the cerebrospinal fluid, but none refer to post-shunt isolated hydrocephalus.

The fact that we have seen five such patients recently suggests that this condition may be unrecognized rather than rare.

Case Reports

Case 1. This premature infant was transferred to the Children's Hospital on August 11, 1966, 9 hours after birth. The birth weight was 1750 gm, and the head circumference, 30 cm. A 4 X 4 cm intact myelomeningocele in the sacral area was repaired on the day of birth.

When 13 days old, the patient became lethargic. The fontanel was full, and the head circumference had increased to 32.5 cm. Pseudomas aeruginosa and a hemolytic Streptococcus were cultured from the ventricular fluid. Intravenous antibiotics were administered and a Rickham reservoir placed in the left lateral ventricle to instill the antibiotic. Four weeks later a ventriculogram done by injecting air into both lateral ventricles demonstrated massive, symmetrical dilatation of the ventricular system; the septum pellucidum was in the mid-line. At this time, the baby weighed 5 lbs 4 oz, and the head circumference had increased to 38 cm. A ventriculopleural shunt was placed and the reservoir removed on August 30. The head continued to enlarge and a left hemiparesis developed. A second ventriculogram on September 10 demonstrated unilateral ventricular dilatation with a marked left-to-right shift. The obstruction at the foramen of Monro was relieved by fenestration of the septum pellucidum. The patient's hemiparesis improved. A repeat ventriculogram on December 21 demonstrated communication between the ventricles. Marked hydrocephalus was present despite a functioning normal-pressure Holter valve. This was replaced with a medium-pressure valve with further improvement in the hemiparesis; the head circumference became stable.

Comment. Many of the features of isolated or univentricular hydrocephalus following a ventriculoatrial shunt are present in this case. A premature infant developed ventriculitis in the neonatal period, survived after treatment with antibiotics, but subsequently developed hydrocephalus. This was initially symmetrical. Following a shunting procedure, the midline structures were displaced toward the ventricle containing the shunt because of lack of communication be-
FIG. 1. Case 1. Ventriculogram with air injected into each ventricle. The septum pellucidum which was in the midline prior to the ventriculoatrial shunt has shifted toward the shunted ventricle. Obstruction at the foramen of Monro was relieved by fenestration of the septum.

FIG. 2. Case 2. Ventriculogram demonstrating a large porencephalic cyst from which air could not be manipulated into the ventricular system. Air injected into the right lateral ventricle communicated with the small left ventricle. The septum was shifted to the right; the 4th ventricle was not seen.

tween the two ventricles. Although the shunt appeared to be functioning upon palpation, the head continued to enlarge and the patient developed a left hemiparesis due to pressure of the tentorium on the right cerebral peduncle. The failure of communication between the two ventricles could have been recognized at the time of the initial ventriculogram if air had been injected only into the left lateral ventricle.

Case 2. This patient was seen initially at the Children’s Hospital at 13 weeks of age because of increasing head size. Her weight was 800 gm. Pseudomonas aeruginosa was cultured from the ventricular fluid, and the patient was treated with systemic and intraventricular antibiotics. She responded to treatment. When she was 6 months old, a ventriculogram with injection of air into both ventricles demonstrated hydrocephalus and a large porencephalic cyst on the left side (Fig. 2). A right ventriculoatrial shunt was performed at that time. She was readmitted at 20 months of age because of increasing head size and lethargy. The ventricular catheter was repositioned, and a longer atrial tubing was put into place. On the day of operation, the patient developed a low-grade fever which persisted over the next 5 days. During this time, she was alert, ate well, and moved all extremities. She was afebrile on the fifth postoperative day, but was lethargic and quite irritable when aroused. She refused her feedings. She had stertorous respiration; however, the chest x-ray was clear. The patient was found dead in bed on the seventh post-operative day.

At autopsy, it was demonstrated that the large porencephalic cyst and the third ventricle did not communicate with the collapsed right lateral ventricle which contained the ventricular catheter (Fig. 3). The left lateral ventricle was displaced into the midline. No
infection was present and no abnormalities were demonstrated outside of the central nervous system.

Comment. This patient had a large left-sided porencephalic cyst which did not communicate with the shunted right lateral ventricle. This lack of communication was not appreciated at the time of the original ventriculogram because air was injected bilaterally.

Case 3. This 1600 gm premature infant was admitted to Children's Hospital when 10 days old because of lethargy and seizures. Klebsiella-Aerobacter meningitis was diagnosed, and the patient was treated by systemic and intrathecal antibiotics. The patient responded slowly to therapy. In the month following admission the head circumference increased 4 cm. Bilateral ventricular punctures were performed, and cloudy amber fluid was obtained, darker on the right. Air injected through the left ventricular needle did not enter the right ventricle even with manipulation of the head. Cultures of the ventricular fluid again grew Klebsiella-Aerobacter organism. A Salmon-Rickham reservoir was placed in the left frontal area, and systemic and intraventricular antibiotics given. The ventriculitis eventually cleared, and the patient was discharged. She was readmitted at 8 months of age because of increasing head size. A ventriculogram demonstrated multiple air-filled chambers. On pneumoencephalography, the fourth ventricle was normal in position and size. The subarachnoid cisterns were partially filled. When she was 11 months old, a ventriculoatrial shunt was performed and the reservoir removed. Although the valve mechanism emptied and filled properly, the shunt had to be revised on two occasions because the fontanel remained tense and the head continued to enlarge. A ventriculogram when she was 13 months old showed large, dilated, loculated ventricles with the ventricular catheter in one of the cysts (Fig. 4). Air was initially injected into the right side. There was a large area in the left frontal region which did not contain air. A second injection was made directly into this area and filled a large cavity which seemed to displace both ventricles to the right. A craniotomy was performed and the isolated porencephalic cyst connected with the ventricular system by making a large fenestration in its wall. Postoperatively, the fontanel became soft and the head circumference stable.

Case 4. This 2700 gm infant was admitted to the Children's Hospital on the day of birth with a myelomeningocele. Pregnancy and delivery had been uneventful. The head circumference measured 33 cm; the chest 31 cm. There was a 3 x 4 lumbosacral myelomeningocele with cerebrospinal fluid leaking through the ruptured sac. A culture of the sac was sterile. The myelomeningocele was repaired on the day of birth.

Five days after the repair, there was leakage of spinal fluid from the wound. The head circumference had increased to 34 cm. Two days later, the patient had a temperature elevation to 102°, and the fontanel was full. A ventricular puncture revealed xanthochromic fluid which contained 60 lymphocytes and 5 polymorphonuclear cells per cu mm. The protein was greater than 300 mg% and the glucose 35 mg%. Paracolon bacillus was cultured from this ventricular fluid. The patient was treated with systemic kanamycin. A ventriculogram showed mild symmetrical enlargement of the ventricular system. On the 16th postoperative day the head circumference had increased to 36 cm. The pa-

Fig. 3. Case 2. Coronal section of brain in Fig. 2 showing that the large left porencephalic cyst and the massively dilated third ventricle are not in communication with the shunted right ventricle which is collapsed.
Patient was lethargic, anorexic, and had frequent tonic seizures associated with opisthotonus. The ventricular puncture was repeated, and thick, yellow, foul-smelling fluid obtained. Paracolon bacillus, sensitive to cephalothin, was cultured. In addition to systemic therapy, 25 mg of cephalothin were introduced into the ventricle utilizing a Rickham reservoir in the right frontal area. The infection was cured after 23 days of this therapy.

When 8 weeks old, the infant again had tonic seizures which were associated with laryngeal stridor. The head circumference was now 39 cm, the fontanel tense, and the eyes showed the “setting sun” sign. A ventriculooatrial shunt was inserted and seemed to be functioning properly on palpation. The head, however, continued to enlarge. The valve was replaced, and the patient improved. One month later, the patient was readmitted because of respiratory stridor and progressive enlargement of the head. Skull roentgenograms showed that the ventricular catheters were displaced laterally as compared to an earlier study (Fig. 5). A ventriculogram was performed by injecting air into the left lateral ventricle; this ventricle was greatly enlarged and extended beneath the falx. There was no communication with the right lateral ventricle which contained the shunt catheter. The ventricular catheter was repositioned into the left lateral ventricle. This system was functioning satisfactorily 10 months later.

Comment. Isolated hydrocephalus was considered when the patient’s head continued to enlarge despite a functioning shunt, there was also lateral displacement of the ventricular catheter.

Case 5. This obviously hydrocephalic 7-month-old girl was first admitted to the Children’s Hospital because of progressive enlargement of the head. She was born after 7 months’ gestation with a birth weight of 2050 gm. When 1 month old, she had developed meningitis which was quite resistant to treatment with antibiotics over a 2-month period. The head began to enlarge at a faster
rate than expected and a ventriculoatrial shunt was performed when she was 4 months old. The shunt functioned poorly and was revised when the infant was 5 months old and again at 6 months.

The patient was quite alert and would follow objects with her eyes. The head circumference was 52 cm and there was venous distension over the scalp. The patient could not support her head or sit even with support. The Holter valve refilled very slowly on palpation. At surgery there seemed to be poor drainage of fluid from the ventricular catheter. This was replaced and a Rickham reservoir included in the system. The post-operative x-rays showed that the ventricular catheter was placed quite far laterally and posteriorly; the fontanel remained full and bulging, and the patient began to vomit. A transcoronal ventriculogram then demonstrated an extremely large and distorted cavity which was felt to represent the left lateral ventricle which had herniated across the midline under the falx (Fig. 6). Air was injected only on the left side. The left ventricular fluid protein was 54 mg%. A dilute solution of water soluble contrast material was injected through the reservoir, and demonstrated that the right lateral ventricle was displaced laterally and posteriorly.

At a second operation the markedly displaced septum pellucidum was then fenestrated through a left frontal craniotomy. One week after the surgery, the fontanel was soft and the head circumference had decreased to 50 cm. A repeat ventriculogram showed that the septum pellucidum had returned nearly to the midline as had the tip of the ventricular catheter.

Discussion

Symptoms. The syndrome of unilateral hydrocephalus was described by Cushing as follows: "Thus, a patient some months after an acute attack of so-called ‘brain fever,’ presented symptoms that were supposed to be those of tumor-profound headache of varying intensity, located on the left side;
choked disc, more pronounced in the left eye; attacks of vomiting, aphasia and weakness of the body musculature on the right side." This patient died some weeks later of bulbar paralysis due to pressure. At the autopsy, dilatation of the left ventricle alone was found. The left foramen of Monro was occluded and there were granulations of the ependyma.

The syndrome is usually incomplete in infants, and the symptoms may be quite nonspecific prior to a sudden death. Table 1 lists the symptoms of isolated hydrocephalus in this group; most reflect increased intracranial pressure. Only one patient showed signs of unilateral involvement. Laryngeal stridor may indicate inadequate decompression of a portion of the ventricular system. The onset of symptoms came 1 to 4 weeks after manipulation of the shunt insertion or revision.

**Mechanism.** All of the patients in this series had a severe infection of the central nervous system which responded to vigorous and prolonged antibiotic therapy. In at least four of the five, the ventriculitis was proved. As the ventriculitis healed, the granulation tissue interfered with the flow or absorption of cerebrospinal fluid, with resultant hydrocephalus. All of the patients showed some degree of disability or retardation as a result of the ventriculitis and hydrocephalus. The hydrocephalus apparently had been symmetrical prior to the shunt, but heavy purulent exudate in the ventricle resulted in loculated cavities on healing.

The isolated lateral ventricle or a loculated collection of cerebrospinal fluid acts as an enlarging mass. This mass causes a pressure differential between the two hemispheres and leads to a shift of the midline structures that is encouraged by the continuous draining of cerebrospinal fluid through a functioning ventriculooarial shunt. The clinical picture varies according to the rapidity with which the shift develops. In Dott's case of unilateral hydrocephalus, which pre-

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**Fig. 6. Case 5. Upper photographs:** Air injected into the left ventricle which has herniated beneath the falx. Note the lateral and posterior displacement of the catheter. **Lower photograph:** The septum pellucidum was fenestrated on May 13, 1968. The septum and the tip of the ventricular catheter now approach midline.
### TABLE 1
Clinical course in five cases of isolated unilateral hydrocephalus

<table>
<thead>
<tr>
<th>Clinical Features</th>
<th>Case 1</th>
<th>Case 2</th>
<th>Case 3</th>
<th>Case 4</th>
<th>Case 5</th>
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<td>Symptoms of isolated hydrocephalus*</td>
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<td>HC increased; stridor, lethargy</td>
<td>HC increased; retarded; lethargy, full fontanel</td>
<td>HC increased; stridor</td>
<td>HC increased; full fontanel, vomiting</td>
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</table>

* HC = maximum head circumference.  
† LV = left ventricle; RV = right ventricle; IC = isolated cavity.  
‡ SP = septum pellucidum.

sumably was the result of an undiagnosed ventriculitis, the ventricular surface was covered by a thick layer of granulation tissue in which the choroid plexus was buried. It is reasonable to assume that this would result in a decrease in the rate of formation of cerebrospinal fluid.

**Differential Diagnosis.** A subdural hematoma in a patient with a ventriculoatrial shunt may cause symptoms similar to those seen in our cases. Displacement of the ventricular system during air encephalography has been described in patients with congenital arachnoid cyst. The proper diagnosis can be established by tapping the suspected area and injecting air for radiographic study, if necessary.

The differential diagnosis in a patient with a ventriculoatrial shunt with signs of increased intracranial pressure whose head is continuing to enlarge is usually considered under the heading of a "malfunctioning shunt." This was certainly true in the present series. There were seven shunt revisions in this group of patients before the proper diagnosis was made. In most of these patients, the valves appeared to be functioning normally on palpation. It was assumed that the symptoms were due to too high an opening pressure in the valve or to only intermittent function of the system.

Patency of the ventricular catheter can be shown by aspirating cerebrospinal fluid from the Salmon-Rickham reservoir, and competency of the valve and atrial catheter determined by contrast radiography. The radiographic demonstration of shunt patency can be accomplished by tapping the reservoir with a syringe containing 1.0 cc of 60% meglumine iothalamate (Conray); the valve is then "pumped," and under fluoroscopy the system is seen to fill toward the heart with each pumping movement. Upon complete filling, small clouds of contrast material can be seen in the atrium. If the contrast material is injected it will enter the ventricle, not the valve. If the system is found to be patent in a symptomatic patient, a ventriculogram, accomplished by injecting air only into the left lateral ventricle, should be considered. With
a history of neonatal ventriculitis, a ventriculogram is essential prior to shunt revision. Reevaluation by contrast studies should be considered in any patient who has required multiple shunt revisions in a short period of time.

A spontaneous change in the position of the ventricular catheter was very helpful in suggesting the presence of an isolated cavity within the ventricular system. Utilization of this clue, of course, presupposes that skull x-rays demonstrating the position of the catheter were obtained at the time the shunt was placed.

Prevention. Identification of a potential isolated hydrocephalus can be accomplished by injecting air only into the left lateral ventricle at the time of the initial air study. If an obstruction at the foramen of Monro or an isolated cavity is demonstrated, bilateral ventricular catheters may then be inserted and connected to a single valve using a “T” connector.

Summary

Five cases have been presented in which a portion of the ventricular system was isolated from the ventricular catheter of a functioning shunt. This isolated cavity continued to enlarge, eventually causing collapse of the shunted ventricle and a shift of midline structures. The typical patient, frequently born prematurely, develops meningitis, ventriculitis, and subsequently, hydrocephalus. This hydrocephalus is initially symmetrical. Despite the insertion of a ventriculocisternostomy shunt which seems to function on palpation, the head continues to grow, and several revisions are done. The patient may develop a hemiparesis or have non-specific signs of increased intracranial pressure including laryngeal stridor. The diagnosis of isolated hydrocephalus is suggested by roentgenograms of the skull which show lateral displacement of the ventricular catheter. This is confirmed by ventriculography with air being injected only into the left ventricle initially. Fenestration of the wall of the isolated cavity provides drainage and results in a prompt return of the septum pellucidum to the midline. This condition should be treated promptly since minimal general or focal symptoms precede sudden death.

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References