Membranous Occlusion of the Aqueduct of Sylvius

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Membranous occlusion of the aqueduct of Sylvius has been considered a rare cause of hydrocephalus (Fig. 1). The occurrence of 4 cases on this unit within a 6 month period prompted a review of this disorder and further consideration of the pathogenesis, diagnosis and surgical treatment.

Case Reports

Case 1. S.H. (9750-64). This 19-year-old student was well until 2 weeks before admission when she fell on her buttocks while roller skating. Although there had been no head injury she was confused for 2 hours. During the following week she continued her normal activity but felt unsteady and light-headed, and spoke of roaring in her ears. On several occasions she fell suddenly and unexpectedly and after the last fall was admitted with a fractured ankle. Neurological opinion was sought because attempts at ambulation induced a feeling of unsteadiness and fear of falling.

Examination. She had numerous café au lait patches. Early bilateral papilloedema was the only other finding. Skull films were normal. Air encephalography resulted in filling of the 4th ventricle. Ethiodian ventriculography showed the aqueduct to be dilated and bulbous and blocked at the lower end (Fig. 2). Several hours later a few drops of contrast material were found to have passed through into the 4th ventricle.

Operation. With the knowledge that a narrow obstruction existed at the lower end of the aqueduct, exploration of the 4th ventricle was planned before carrying out a Torkildsen procedure.

Through a suboccipital craniectomy the 4th ventricle was inspected. A small No. 8 rubber catheter which initially encountered resistance passed up the aqueduct after moderate pressure was exerted. Upon withdrawal there was a gush of cerebrospinal fluid. Direct inspection of the lower end of the aqueduct revealed that all that remained of the closure was a fringe of thin translucent membrane. Because this region of the aqueduct gripped the catheter upon reinsertion, it was thought that stenosis might recur. Therefore, a 2-inch length of siliconed plastic tubing was placed through the opening into the 3rd ventricle. Aside from a mild postoperative aseptic meningitis the patient’s recovery was uneventful and she remains well.

Case 2. M.M. (14258-64). At birth this 2-year-old boy’s head measured 142 1/2 inches but he walked at 13 months and could put 3 words together at 18 months. One month before admission he fell and struck his forehead. The first of 5 grand mal seizures occurred a week later.

Examination. He appeared well and ran about and talked. The head circumference was 22 1/2 inches. There was fine nystagmus on lateral gaze but no other abnormal neurological signs.

Skull films showed a large vault without separation of the sutures. Lumbar air injection filled only the 4th ventricle (Fig. 3). Ethiodian was placed in the right lateral ventricle together with a little air. There was gross hydrocephalus. The contrast medium filled the dilated aqueduct which was rounded at the obstructed lower end. After the child had been left in the brow up position for 1/2 hour a tiny drop of contrast medium passed into the 4th ventricle (Fig. 4). When the ventriculogram was superimposed on the pneumoencephalogram, the upper end of the 4th ventricle was seen to approximate the lower end of the aqueduct (Fig. 5). A diagnosis of membranous obstruction of the aqueduct was made.

Operation. The 4th ventricle was exposed and in the lower end of the aqueduct a thin translucent membrane was seen which bulged downward with each pulsation of the brain (Fig. 6). A small catheter was easily passed into the aqueduct and when withdrawn a circular opening was visible in the membrane. A larger catheter was then introduced leaving a defect 3 mm. in diameter. A fringe of membrane could be seen around the circumference attached to the wall of the aqueduct.

No catheter was left in the aqueduct. The patient recovered quickly from the operation and has been well since.

Case 3. C.M. (15,310-64). The parents of this 9-year-old girl sought medical advice because of an increasing tendency to turn over her left ankle when walking. She had developed slowly, not walking until she was 18 months old. She had always been clumsy and had recently complained of headache about once a month, occasionally associated with vomiting.

Examination. Her head was enlarged (24 1/2 inches) and her speech was nasal and slurred. Sev-
FIG. 1. Membrane occludes aqueduct of Sylvius at its lower end.

eral café au lait patches were present on the trunk and there was a soft subcutaneous swelling on the left forearm. There was a slight scoliosis with increased lumbar lordosis. The left arm and leg were slightly smaller and weaker than the right and performed coordinated movement less well. She walked unsteadily on a wide base and the left ankle was rotated outwards.

X-rays showed an enlarged cranium and an occult spina bifida at S.1 and S.2. At pneumoencephalography, air entered the 4th ventricle but passed no further. The 4th ventricle was small and displaced caudally (Fig. 7). An air ventriculogram showed the lateral and 3rd ventricles to be grossly dilated. The 3rd ventricle ballooned out below the tentorium and was first mistaken for a dilated 4th ventricle (Fig. 8).

Operation. Utilizing a suboccipital craniectomy, a small rubber catheter was passed through the 4th ventricle. Resistance was encountered at the apex but with firm pressure it passed into the aqueduct. When the cerebellar vermis was elevated, a hole 3 or 4 mm. in diameter was seen in a thin translucent membrane at the lower end of the aqueduct. Fluid now ran freely from the 3rd into the very small 4th ventricle. The hole in the membrane was enlarged with a larger catheter.

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Fig. 3. Case 2. Pneumoencephalogram with filling limited to 4th ventricle and subarachnoid spaces.

Fig. 4. Case 2. Ethiodan ventriculogram after ½ hour in brow up position. A few drops of contrast have escaped into the 4th ventricle. The obstructed aqueduct is dilated and bulbous.
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but while maintaining the exposure, the tip of the narrow retractor pierced the roof of the 4th ventricle just below the aqueduct. There was no bleeding and the craniotomy was closed without a tube in the aqueduct.

She regained consciousness but 24 hours later suddenly became unconscious, then apneic. Artificial respiration was begun and the lateral ventricle, under high pressure, was tapped. The

ventricle meets the contrast in the aqueduct demonstrating the thin occluding septum.

Fig. 5. Case 2. Films of lumbar air encephalogram and ethiodan ventriculogram superimposed (lines drawn on calvaria were used for accuracy). Air in the apex of the 4th ventricle meets the contrast in the aqueduct demonstrating the thin occluding septum.

Fig. 6. Case 2. A. Bulging membrane exposed at upper end of 4th ventricle. B. After perforation, fringe of tissue visible rimming lower end of aqueduct.

Fig. 7. Case 3. Air encephalogram. Aqueduct of Sylvius blocked and 4th ventricle displaced caudally.
craniorrhaphy was reopened immediately and there was no flow of fluid from the 3rd into the 4th ventricle. A catheter was left in the aqueduct as a shunt but she died the following day without having regained consciousness.

Unfortunately the brain was discarded before examination. It was felt that the retractor injury resulted in enough local swelling at the apex of the 4th ventricle to block the lower end of the aqueduct.

Operation. At suboccipital craniotomy the roof of the 4th ventricle was elevated to disclose a thin translucent membrane occluding the lower end of the aqueduct. In an attempt to photograph the region, the membrane was stretched by the retraction and tore allowing cerebrospinal fluid to pour into the 4th ventricle. As the retractor tip had again injured the superior medullary velum, a length of siliconed plastic tube was placed through the aqueduct into the 3rd ventricle. The lower end lay in the upper cervical subarachnoid space. It was felt that this shunt would render harmless the postoperative swelling from the retractor injury that might develop at the outlet of the aqueduct and which had proved so disastrous in Case 3.

The boy recovered rapidly and the 6th nerve palsies disappeared. He played happily with the other children on the ward but his vision remained severely impaired from secondary optic atrophy.

Discussion

A thin septum obstructing the lower end of the aqueduct of Sylvius has been mentioned occasionally as a cause of hydro-
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tcephalus. Russell reviewed the subject in her classic monograph on the pathology of hydrocephalus in 1949 and described in detail 2 cases of her own. Single cases treated surgically have been mentioned by Dandy and Paine and McKissock, and more fully described by Rowbotham, Scheuerman and Groff, Petit-Dutaillis et al., and Leckey and Morley.

Pathogenesis of Aqueduct Stenosis and Septum Formation. The ventricular system develops from expansions of the rostral central canal of the neural tube. Embryologically it is divided into 3 successive freely communicating chambers. With further development, the middle vesicle remains narrow due to growth of the surrounding nuclear masses and fibre tracts of the mesencephalon, while the vesicles above and below expand to become the 3rd and 4th ventricles. A slight dilatation of the central portion of the aqueduct is reminiscent of its origin and has been called the ventricle of the mid-brain. The lining ependyma may be absent for short distances on one or both sides. Turkewitsch further describes small diverticulae during the development of the aqueduct and demonstrated a number of small wrinkles in its wall. The adult aqueduct is approximately 11 mm. in length and as much as 3 mm. in width. The measurements of Woollam and Millen indicate that in normal subjects the minimum cross-sectional area of the aqueduct occurs at the caudal end and is between 0.4 and 1.5 sq. mm.

There is surprising variation in the size and shape of normal aqueducts. In cross-section the opening may be oval, round, diamond-shaped, T-shaped or slit-like. Frequently there are irregularities in the thickness of the ependyma. Small accessory aqueductules are common as well as a separate large ventral channel or an ependymal cord. The subependymal glial plates present numerous variations in position and cell density.
TABLE 1
Membranous occlusion of the aqueduct of Sylvius

<table>
<thead>
<tr>
<th>Author</th>
<th>Date</th>
<th>Age</th>
<th>Sex</th>
<th>Head Size</th>
<th>Associated Anomalies</th>
<th>Operation</th>
<th>Results</th>
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<td>Orton22</td>
<td>1908</td>
<td>46</td>
<td>M</td>
<td>25 in.</td>
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<td></td>
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<tr>
<td>Versé19</td>
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<td>9</td>
<td></td>
<td>&quot;Enlarged&quot;</td>
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<td>Dandy2</td>
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<td>4 mos.</td>
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<td>8</td>
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<td>Lu et al.11</td>
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<td>Rowbotham15</td>
<td>1938</td>
<td>11</td>
<td>F</td>
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<td>Aneurysm in aqueduct above membrane</td>
<td>Failed to pass catheter</td>
<td>Died</td>
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<td>Dandy3</td>
<td>1945</td>
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<td>1948</td>
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<td>Turnbull &amp; Drake Case 1</td>
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<td>Normal 22½ in.</td>
<td>Café au lait</td>
<td>Perforation of membrane</td>
<td>Well</td>
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<td>Case 2</td>
<td>2</td>
<td>M</td>
<td></td>
<td>Large 22½ in.</td>
<td>None</td>
<td>Perforation of membrane</td>
<td>Well</td>
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<td>Case 3</td>
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<td>Large 24½ in.</td>
<td>Café au lait, spina bifida</td>
<td>Perforation of membrane</td>
<td>Died</td>
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<td>Case 4</td>
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<td></td>
<td>Normal 19½ in.</td>
<td>Café au lait, double aortic arch</td>
<td>Perforation of membrane + tube</td>
<td>Well</td>
</tr>
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The lining ependyma is often incomplete and in these denuded areas glial overgrowths may appear as warty excrescences. This subependymal glial proliferation may be excessive, encroaching on the lumen and even bridging the canal.8

Russell16 felt there was little support for the earlier hypothesis that aqueduct stenosis was usually of inflammatory origin. She described 2 distinct situations. In the first the aqueduct was small, and represented by incomplete reduplications ("forking") which were surrounded by "normal neural tissue". This type was presumed to result
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from an early embryonic defect. The second group was associated with "gliosis"—a progressive bridging and stenosis by glial overgrowth with varying final degrees of stenosis and subdivision. She noted that this disorder could exist in the ventricles but was more significant in the aqueduct because of the calibre. This gliosis appeared to develop in post-natal life and was slowly progressive.

Membranous occlusion was described as a separate entity and felt to be a congenital maldevelopment or, rarely, the result of inflammation.

In their critical review of the literature, Drachman and Richardson1 questioned the sharp distinction between forking and gliosis in view of the wide variation in the abundance of glia surrounding the aqueduct. They noted that many cases in the literature and one of their own had histological features of both. Russell noted the high incidence of associated congenital abnormalities and this appeared to be unrelated to the degree of gliosis present.

We believe that membrane formation might result from a small bridging glial occlusion at the caudal end of the aqueduct which gradually becomes an attenuated transparent sheet from prolonged pressure and dilatation of the canal above.

Descriptions of the membrane are similar.17,15,16,19 It has occurred at the lower end of the aqueduct in all cases. Thin, translucent, it is composed of loose-textured fibrillary neuroglia. Scattered clumps of ependymal cells have been noted on it and in the brain adjacent to its attachment. A pin-hole opening was described in one case (Russell16) and in another (Orton12) small ependymal canaliculi provided routes by which cerebrospinal fluid could trickle past the obstruction. Such minute openings were demonstrated radiographically in 2 of our cases (Cases 1 and 2).

The ages of the reported cases warrant consideration. Only one case was reported under 1 year of age while the remainder varied between 2 and 46 years (average 16 years). This supports the hypothesis of post-natal maldevelopment, although a tiny opening or canaliculus may provide an incomplete by-pass to delay symptom-producing or fatal hydrocephalus. It is interesting that in a large series of aqueductal stenoses in which infants and patients with tumors were excluded, the approximate average age was 16.13

That this septum has an origin in common with other forms of aqueductal stenosis is supported still further by the similar incidence of associated congenital abnormalities. The presence of café au lait patches in 3 of our 4 cases is noteworthy.

Diagnosis. The signs and symptoms of aqueductal stenosis have been described in detail.15 The diagnosis should be suspected in children and young adults who develop increased intracranial pressure without definite localizing signs.

The clinical picture in cases with membranes did not differ essentially from that seen in aqueductal stenosis in general. There was no apparent sex difference. As mentioned the average age was 16 years. Ten of the 14 cases in which age was stated were between 5 and 23 years. In all but 3 the head was enlarged. Where recorded, an associated congenital abnormality was present in 5 of 11 patients.

The presence of a membrane occluding the aqueduct can be established tentatively with positive contrast ventriculography. The aqueduct has a typical appearance. It is long, dilated and has a bulbous expansion at the lower obstructed end. In both cases so studied, delayed films revealed that a drop or two of contrast had trickled past the membrane into the 4th ventricle. Lumbar encephalography may fill the 4th ventricle and then the narrow septum can be demonstrated by superimposition of the ventriculograms. However, a spinal air injection may be dangerous in the presence of papilloedema and is unnecessary. The appearance of the aqueduct should be sufficient to justify exploration of the apex of the 4th ventricle.

The ventriculographic appearance of aqueductal stenosis has been described by Paine and McKissock under 4 headings: partial occlusion, funnelled aqueduct, bulbous type and atresia of the aqueduct.13 Their radiographs illustrating the bulbous type are identical to those in the present series. Although an operation on a membrane occlusion was mentioned it was not included in the paper. It is probable that the patients
represented by Figs. 7 and 8 in that paper did have membranous occlusion and might have been treated successfully by perforation of the membrane instead of the Torkildsen procedure.

Surgical Treatment. Simple perforation of the membrane appears to be the ideal surgical management. Our Case 3 was the only death in the 7 cases which have been treated in this fashion. The prolonged firm retraction of the roof of the 4th ventricle, which resulted in injury to the outlet of the aqueduct and fatal postoperative hydrocephalus, was unnecessary. Only a glimpse of the membrane is needed to warrant its perforation with a catheter.

The decision as to whether a tube should be left in the aqueduct deserves comment for it was done in 2 of our cases. Where the opening is large, simple perforation appears to be adequate. However, when the opening is small and grips a small catheter, or where some contusion of the apex of the roof of the 4th ventricle has occurred from retraction, it might be wise to leave a tube in place. Modern plastic tubing evokes little or no reaction and seems to be tolerated indefinitely. The tube in Case 1 was not anchored, the ends lying free in the 3rd and 4th ventricles. In Case 4 a suture was used to secure the lower end to the arachnoid in the upper cervical canal.

Why not use Torkildsen's procedure? Perforation of the thin membrane is so straightforward that it is difficult not to recommend this direct approach in all such cases. The single death in this series resulted from retractor injury to the superior medullary velum with subsequent swelling and acute hydrocephalus. Although this injury is unnecessary, when there is suspicion that a postoperative outlet occlusion might occur, a tube can be left in place.

This direct approach described by Dandy and Fraser and Dott fell into disrepute because attempts were made to cannulate all stenotic aqueducts, the length of which was unknown. Even so, Greenwood and Hickey showed that rubber tube shunts through the aqueduct can be practical, safe and enduring. Recently Elvidge reported on 10 patients with aqueduct stenosis in whom he probed the canal. In order to maintain patency he performed interventricularostomy with rubber and more recently plastic tubing. There was an operative mortality of 20%. He did not encounter a membrane but on 1 or 2 occasions he saw a tiny amount of grey tissue pointing out of the lower end of the aqueduct.

The ventriculo-cisternal shunt described by Torkildsen has become the standard procedure in cases of aqueductal obstruction. It is safe, relatively simple and highly successful. Yet it is to be noted that in one large series of aqueductal stenosis this procedure was successful in only 16 of 25 cases.

Perhaps the difficulties are associated with the length of the tube and the fact that it must be carried out of the ventricle circuitously and reinserted into the subarachnoid space. If the ventricular portion of the tube is short, subsequent contraction of the ventricles may occlude it. At times it is difficult to fix the lower end in the narrow cervical subarachnoid space. Any withdrawal will make the shunt ineffective if the fluid is diverted to the subdural or extradural space.

Following simple perforation none of these problems exist but it must be admitted that exploration of the 4th ventricle carries additional hazard. If a tube is used to maintain the opening, it should be short and connect the 3rd directly to the 4th ventricle.

Summary and Conclusions

1. We have reported 4 cases of membranous occlusion of the aqueduct of Sylvius treated surgically. Three patients are well while one died from an operative mishap.

2. We believe that this thin septum results from the same developmental defect that produces the more severe degrees of non-tumorous aqueductal stenosis. When glial overgrowth is restricted to the lower end of the aqueduct, it gradually becomes an attenuated transparent sheet from prolonged pressure and dilatation of the canal above.

3. The presence of a membrane occlusion can be tentatively established from the typical appearance of the aqueduct in positive contrast radiography. Appreciation of the ventriculographic findings may result in the more frequent recognition of this cause of aqueductal occlusion.

4. Simple perforation of the membrane appears to be the most satisfactory treat-
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References

6. Elvidge, A. Personal communication.

Addendum

Re Case 1. S.H. 9750-64. This patient remained well for 10 months, then after a "flu-like" illness developed recurrence of nausea, vomiting, episodic vertigo and ataxia. After a month she became bedridden with intractable vomiting and was readmitted to the neurosurgical service. There was early recurrence of papilloedema. Skull x-rays showed that the tube had not shifted position but lumbar air injection succeeded in filling only the lower part of the cisterna magna.

With the thought that the tube was obstructed, re-exploration of the posterior fossa was carried out. On opening the dura the cause of the trouble was immediately apparent. The foramen of Magendie was contracted and obliterated, except for a pinpoint opening in the centre, by a film of thickened arachnoid which also matted the cerebellar tonsils together. When this membrane was incised cerebrospinal fluid gushed out reducing the pressure. Inspection of the 4th ventricle now revealed the lower end of the tube from which spinal fluid was dripping continuously. With this evidence of its patency the tube was found to be freely movable in the aqueduct. It was removed and replaced through the foramen of Magendie so that the upper end lay in the 4th ventricle and the lower in the upper cervical subarachnoid space (4th ventricle cisternal shunt). The patient recovered completely. It was concluded that postoperative arachnoiditis had obliterated the outlets of the 4th ventricle. This probably would not have occurred if the tube had been longer so that it passed from the 3rd ventricle, through the aqueduct and 4th ventricle to end in the upper cervical subarachnoid space as in Case 4.