Terminal ventriculostomy for syringomyelia

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The clinical course of 12 patients who underwent terminal ventriculostomy for syringomyelia is presented. Opening the central canal at the tip of the conus medullaris is a relatively benign procedure that improves the symptoms of syringomyelia and syringobulbia. This canal normally terminates at the tip of the conus, but in each of the 12 surgical specimens it continued into the filum terminale for distances up to 8 cm. In most cases the tip of the conus was located more caudally than normal, indicating some degree of tethering in fetal life. This belief is supported by the fact that the newborn, whose conus is tethered to a lipoma at the sacral level, may develop syringomyelia in adult life.

KEY WORDS • terminal ventricle • central canal • tethered cord medullaris • filum terminale • arachnoid diverticula • Pantopaque arachnoiditis

The terminal ventricle is an enlargement of the central canal within the tip of the conus medullaris just below the point of emergence of the last coccygeal nerve root. As early as 1898, this structure had been considered to be analogous to the pathological state of syringomyelia. This ependymal-lined canal seldom extends beyond the conus, whose tip is normally at the lower border of the L-1 vertebra.

Although it was not identified as a case of syringomyelia, the first terminal ventriculostomy on record was described in 1956 by Jones and Love. In their report, six patients with vertebral defects and neurological dysfunction were improved after a tight filum terminale had been divided. Their Case 3 was a 35-year-old woman with a history of numbness of the right arm and thorax for 2 years. In addition to this complaint she had a right-sided limp with hammer toes. There was hypertrichosis in the lumbosacral area, occult spina bifida of L-5 with dysplasia of the centrum, and spondylolisthesis Grade 1 at L5–S1. Myelography showed obstruction of the Pantopaque column opposite the L-3 vertebra, but the cerebrospinal fluid (CSF) was normal. At surgery the loose neural arch of L-5 was removed and a large extradural cyst containing clear fluid was found on the right side. After the dural sac was opened a No. 8 rubber catheter was passed rostrally for a distance of 16½ inches. A cyst containing opalescent fluid involved the lower end of the conus and upper part of the filum. This cyst was opened and a tantalum seton inserted. The filum terminale was taut and 0.5 cm of it was removed between clips, after which the divided ends separated for an additional 1.25 cm.

One year after operation neurological examination showed improvement of the sen-
In addition to the buried rounded nests of ependymal cells in this "syrinx" of the filum (upper right), an apparently communicating canal lined by ependyma is shown in its lower portion. H & E, × 40. Right: This must represent the original central canal. H & E, × 160.

Between the years 1973 and 1975, we have operated on or attended the operations in 12 patients who underwent terminal ventriculostomy. The case histories of the first two of these patients are described.

Case Reports

Case 1

In July, 1973, this 26-year-old woman underwent excision of the terminal ventricle as an alternative procedure during the course of a planned syringotomy of the conus.

History. She had experienced the onset of syringomyelic symptoms at the age of 12 years. A craniovertebral decompression of a hindbrain hernia was performed when the patient was aged 17 years, and for 5 years she noticed some improvement. By the age of 25 she had become seriously disabled and, although continent, was unable to feel the passing of urine or feces. Subsequent to this an unsuccessful attempt at percutaneous punct-
Terminal ventriculostomy for syringomyelia
ture of the cervical syrinx had resulted in paraplegia. She was admitted for evaluation 6 months thereafter.

Examination. The patient was barely able to walk with support. There was a left Horner's syndrome with anhidrosis and the only remaining area of normal cutaneous sensibility was in the right trigeminal distribution. Her previous Babinski responses had disappeared, and she remained unable to feel the passing of urine or feces. These two findings indicated that the syrinx involved the conus. Because in six reported cases of syringomyelia of the conus the patients had experienced excellent relief by a local syringotomy, this procedure was advised.

Operation. At lumbar laminectomy, performed in Cali, Colombia, the conus appeared to be non-fluctuant and attenuated, merging with an enlarged filum terminale at the L3-4 interspace. Since syringotomy was not feasible, 1 cm of the enlarged filum was excised up to the tip of the conus. Grossly, the cut upper end of the specimen resembled a miniature spinal cord. Fluid did not escape, but microscopic sections showed that the specimen contained a dilated central canal and some tiny adherent nerve roots, apparently coccygeal.

Postoperative Course. At 24 hours there was return of sweating on the left side of the face and within 2 months the patient could feel the passing of urine and feces. A report 21 months after operation indicated that the dissociated sensory loss was less dense and less extensive, the left Horner's syndrome had cleared, and she was walking with the aid of a brace. Although she was able to feel the passage of urine and feces, Babinski responses had not returned.

Case 2

This 49-year-old physician was admitted for terminal ventriculostomy in August, 1973.

History. At the age of 19 years he had discovered his inability to appreciate cold with the right hand. He subsequently developed advanced syringomyelia with bilateral Babinski responses; his symptoms were worse toward the end of the day. He had been impotent for 7 years before admission. Pantopaque myelography, done elsewhere in July, 1973, had outlined a distended cord extending downward from C-2. There were two arachnoid diverticula at the level of T-6 that filled with returning Pantopaque (Fig. 2). The patient elected to have a terminal ventriculostomy, and if improvement did not result, decompression of the hindbrain hernia would be performed.

First Operation. Lumbar laminectomy in August, 1973, disclosed the conus at the level of L-2; the terminal ventricle appeared cystic, and fluid escaped when it was excised together with 1 cm of the filum. Within 24 hours he experienced sweating on the right side of his face for the first time in 9 years. His postoperative improvement was dramatic, the dissociated sensory loss improved remarkably and the Babinski responses cleared. He returned to practice within 6 weeks of the operation.

However, examination 4 months later revealed increasing paraparesis and return of Babinski responses. The spinal fluid protein was elevated. Eight months after operation, although his syringomyelic symptoms were still relieved, his spastic paraparesis was worse. He arrived in a wheel chair. When he

Fig. 2. Case 2. Pantopaque myelogram. The spinal cord, which had been shown to be distended in the head-down position with non-filling of the diverticula, is now collapsed because of the specific gravity of the returning Pantopaque. This factor explains the frequent false negative myelographic findings in syringomyelia.
W. J. Gardner, et al.

Fig. 3. Case 2. With the dura open, one of the formerly opacified diverticula is seen largely overlaying the other. Caudal to the white circular area of thickened arachnoid is an opaque loculated arachnoid cyst that conceals the underlying cord. Three cystic lesions were removed. The patient’s head is to the left.

was first placed on the examining table there was total loss of position sense in the toes, but it had returned on retesting 40 minutes later.

Second Operation. He was admitted to the hospital where myelography was performed to rule out an associated cord tumor. There was no block. The entire syrinx appeared collapsed and, with the patient in the head-up position, the arachnoid diverticula at T-6 again filled with Pantopaque and appeared larger than on the preoperative myelogram. Since Pantopaque myelography is frequently unreliable in syringomyelia, this study was followed by decompression of the hindbrain hernia according to the prearranged plan. This operation was followed by no improvement. The patient had transient paresis of the left arm, and 2 months later he was paraplegic to the level of T-6. The Queckenstedt test disclosed a complete block. There was absence of the CSF pulse waves and the CSF protein content was 796 mg/100 ml. Pantopaque introduced into the lumbar sac stopped at L-2, and when injected into the cistern it stopped at T-6, again with filling of the enlarged diverticula and collapse of the syrinx.

Third Operation. Operation at this level disclosed a collapsed cord, compressed by cystic Pantopaque arachnoiditis (Fig. 3). At examination 13 months after this thoracic cord decompression and 30 months after terminal ventriculostomy, the paraplegia was perhaps slightly improved and there was no recurrence of the neurological symptoms above the T-6 level. The patient reported recurring dislocations of a Charcot joint of the right shoulder which were painless despite the maintained improvement in cutaneous sensory loss. This is consistent with a case of syringomyelia described previously in which deep pain sensation was lost whereas skin sensitivity was retained.

Summary of Cases

The average age of onset of symptoms in these 12 patients was 22 years, and the average interval between onset and terminal ventriculostomy was 17 years (Table 1). Although the question was not asked, four patients volunteered that their symptoms were worse toward the end of the day. The tip of the conus was at or below the level of L-2 in 11 cases; it was opposite the L3-4 interspace in Cases 1 and 7 and the L2-3 interspace in Cases 3 and 6, corresponding to the level in a 25-week fetus. Nine patients were scoliotic and seven had basilar impression. A previous craniovertebral decompression had been performed in seven cases, with improvement in five, no change in one, and worsening in one. One of these (Case 6) had been made
Terminal ventriculostomy for syringomyelia

worse by a cervical syringotomy, but improved 3 years later following a craniovertebral decompression. In six of the seven cases, craniovertebral decompression had disclosed thickened arachnoid-like tissue enclosing the hindbrain hernia. This matted and vascular tissue should not be called "arachnoiditis" since it is not a chronic inflammatory process but is intrinsic in the hindbrain hernia. It has been attributed to eversion of the downwardly dislocated nodulus that causes the vascular fibrous tissue of the developing choroid plexus to remain in the subarachnoid space instead of assuming its intraventricular place on the under surface of the nodulus. Brisk arterial bleeding may be encountered when this fibrous covering is incised, which is not the case in chronic arachnoiditis. Since the posterior choroid plexus is nourished by a branch of the posterior inferior cerebellar artery, it is not surprising that a loop of this latter vessel is frequently extruded with the hindbrain hernia.

Radiographic Studies

Ten of these 12 cases of terminal ventriculostomy had a Pantopaque myelogram performed elsewhere. In one patient the myelogram was negative, although subsequent pneumoencephalography showed the collapsing cord sign of syringomyelia. The two remaining patients (Cases 7 and 8) had received a percutaneous injection of Pantopaque into the syrinx which in each case showed that it was expanded down to the level of T-12. In Case 8, some Pantopaque passed from the syrinx into the fourth ventricle when the patient was placed in the head-down position.

Pantopaque myelography is less reliable than air since its high specific gravity often results in compression of the relatively flaccid syrinx. In Case 2 it resulted in paraplegia from Pantopaque arachnoiditis. In the monograph on syringomyelia by Barnett, et al., it was reported that the initial myelogram in some cases was negative, whereas a second or third one was positive. This suggests increased adhesion of pia to arachnoid because of irritation by microglobules retained from the earlier study. In their table summarizing 100 cases, 94 are shown to have had Pantopaque myelography which demonstrated a wide cord in only 34.

It was pneumoencephalography that first disclosed that the cause of syringomyelia is not in the spinal cord but in the hindbrain. Properly performed, this remains by far the
W. J. Gardner, et al.

### TABLE 1

**Summary of data in 12 patients undergoing terminal ventriculostomy**

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Sex, Age†</th>
<th>Operations‡</th>
<th>Preop. Neurological Status</th>
<th>Follow-Up Findings</th>
<th>Postop. Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>F, 12</td>
<td>CVD at 17, TV at 26, see case report</td>
<td>21 mos; see case report</td>
<td>good</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>M, 19</td>
<td>TV at 49</td>
<td>see case report</td>
<td>30 mos; see case report</td>
<td>good</td>
</tr>
<tr>
<td>3</td>
<td>M, 43</td>
<td>TV at 46</td>
<td>weakness, atrophy, fasc shoulder girdles; DSL bilat C2-10 &amp; lt trigem; absent gag &amp; abdominal reflexes</td>
<td>25 mos; weakness, atrophy, fasc relieved; DSL greatly improved; return of gag &amp; abdominal reflexes</td>
<td>good</td>
</tr>
<tr>
<td>4</td>
<td>M, 14</td>
<td>TV at 26, CVD at 15, TV at 49</td>
<td>bulbar sx &amp; ataxia increasing since birth; DSL both trigem, shawl distrib, &amp; rt leg; hands tightly clenched</td>
<td>29 mos (phoned report): bulbar sx &amp; ataxia improved; appreciates heat rt hand; clenched hands relax at rest; drives car; working as bank clerk</td>
<td>good</td>
</tr>
<tr>
<td>5</td>
<td>M, 15</td>
<td>TV at 44</td>
<td>bulbar sx; DSL below elbows, also lt T6-L5, &amp; rt leg; fasc legs; spastic ataxic gait</td>
<td>10 mos: writing &amp; gait improved; 26 mos: DSL relieved in legs; improved in arms; no fasc; ataxia worse</td>
<td>good?</td>
</tr>
<tr>
<td>6</td>
<td>F, 23</td>
<td>syringotomy at 32, CVD at 35, TV at 33</td>
<td>DSL bilat C2-T5: fasc pain, weakness, shoulder girdles; weak grasp; sweats only in axillae; severe urinary frequency</td>
<td>12 mos: DSL top level at C-6; urinary frequency &amp; manual dexterity improved; 26 mos (phoned report): no fasc; astereognosis persists</td>
<td>good</td>
</tr>
<tr>
<td>7</td>
<td>M, 24</td>
<td>TV at 43</td>
<td>DSL shawl distrib; weak hands, spastic legs; astereognosis; shoulder fasc; burning inter- scapular pain</td>
<td>improvement in DSL, strength of hands &amp; astereognosis; no fasc; pain relieved; see text</td>
<td>good</td>
</tr>
<tr>
<td>8</td>
<td>F, 17</td>
<td>TV at 49</td>
<td>DSL bilat C2-T5 &amp; rt trigem; paralyzed rt arm; progressing paresis of lt arm; pain lt deltoid</td>
<td>26 mos (phoned report): no further progression; pain less; working daily</td>
<td>fair</td>
</tr>
<tr>
<td>9</td>
<td>F, 16</td>
<td>TV at 17</td>
<td>DSL rt T4-L3; absent abdominals; hypoactive rt patellar</td>
<td>13 mos: upper &amp; lower level of DSL has receded; return lt abdominal reflex; 25 mos (phoned report): feeling fine</td>
<td>good</td>
</tr>
<tr>
<td>10</td>
<td>F, 22</td>
<td>CVD at 23, TV at 27, recurring symptoms for 4 mos; DSL lt T8-L1; right spastic hemiparesis</td>
<td>48 hrs: improved DSL and hemiparesis; 14 mos: improvement maintained</td>
<td>good</td>
<td></td>
</tr>
<tr>
<td>11</td>
<td>F, 29</td>
<td>CVD at 32, TV at 35</td>
<td>paralyzed rt arm; all cutaneous sensibility absent bilat C2-T10, impaired below; severe burning pain both palms; anhidrosis; astereognosis; fasc shoulders</td>
<td>18 mos (phoned in Greece): depressed; believes symptoms progressing again; see text</td>
<td>probable recurrence</td>
</tr>
<tr>
<td>12</td>
<td>M, 28</td>
<td>CVD at 42, cerv. disc at 43, TV at 50</td>
<td>quadriparesis worse on rt; DSL C2-9 bilat, also both trigem; fasc both arms; complete anhidrosis; bilat Charcot shoulder joints</td>
<td>16 mos: postoperative complication; see text</td>
<td>poor</td>
</tr>
</tbody>
</table>

*DSL = dissociated sensory loss; fasc = fasciculations; sx = symptoms.
†Age at onset of symptoms.
‡CVD = craniovertebral decompression; TV: terminal ventriculostomy. The age at which the patient underwent the operation is given in years.
most reliable radiographic procedure; the following technique is advised: It should not be employed within 10 days of a preceding lumbar puncture in order to exclude the possibility of continuing leakage that may result in collapse of the syrinx. With the patient seated, the head is flexed to widen the interspace between the foramen magnum and C-1. The chair is then tilted backward until the cervical spine is vertical. This is important since otherwise the preliminary air injection, instead of outlining the cord, may disclose only its posterior surface. The patient’s head is maintained in this flexed position, and the film cassette is placed on the shoulder in contact with the head (Fig. 5); in this way its proximity to the cervical spine eliminates the need for tomography. A scout film is made with the central ray directed through the foramen magnum. If this film is satisfactory, only then is the patient anesthetized, preferably with intravenous Pentothal (thiopental), after which the lumbar puncture is performed. The depth of anesthesia should be adequate to insure that movement of the patient will not interrupt the continuity of the procedure. Air (30 ml) is then rapidly injected and a film exposed immediately, but the procedure continues while this film is being developed. Such continuity is important because the picture changes rapidly. Fluid is replaced with air in 10-ml equivalents and a film exposed after each 30 ml. The first film should disclose a hindbrain hernia and a swollen cord. When later films disclose a collapsed cord, the exchange is discontinued. The elapsed time of anesthesia need not exceed 10 minutes and an endotracheal tube is not needed. Of greater importance, no foreign material is left in the already narrowed subarachnoid space.

Operative Technique

The lumbar laminectomy should be wide and long enough to make allowance for a low position of the conus and to aid in the identification of the filum terminale. This structure was plainly visible through the dura in two cases because it was adherent to the posterior arachnoid. If enlargement of the opening in the bone proves necessary after the dura has been incised, any entrance of blood will make identification of the structures more time consuming. The filum is elevated on a nerve hook at the lower end of the exposure and traced up to the tip of the conus (Fig. 4). The filum is grasped with a hemostat at the lower end, clipped, cut, and retracted upward. This will usually disclose a vein on the undersurface of the terminal ventricle. To control bleeding, a piece of Gelfoam is placed beneath it, the filum is cut with scissors, a moist cotton patty is promptly applied, accompanied by gentle pressure for a few minutes with a suction device. The dura should be carefully closed and the patient kept relatively flat for a week to avoid leakage of CSF. In six of the 12 patients, fluid was not seen to escape from the divided terminal ventricle, presumably because of the head-down position of the patient during the operation. In one such case, the surgeon (HSB) elevated the head of the table and flow began. This change of position is recommended if spontaneous flow does not occur. In Case 12, the conus was swollen when viewed through the intact arachnoid, but collapsed promptly after evacuation of the CSF.

In Case 7, the conus was distended and remained so after section of the terminal ventricle. This syrinx was then incised causing escape of the previously injected Pantopaque together with syrinx fluid. A Silastic tube was introduced to drain the cavity. This conus was at the L3-4 level, and there were severe arachnoid adhesions involving the low-lying conus and cauda equina. A piece of the thickened arachnoid was excised for microscopic study, but unfortunately did not
reach the laboratory. This specimen was taken to search for heterotopic nests of glia which have been demonstrated at autopsy in the subarachnoid space in cases of syringomyelia and also in infants with myelocle. Such glial heterotopia, together with the pia-arachnoid diverticula described in Case 2, have been interpreted as evidence of impaired dissection of this space by the subarachnoid fluid pulse wave, caused by overdistention of the neural tube in embryonal life.

Operative Results

Patients were followed for up to 30 months, and all but two have shown improvement (Table 1). In cases where anhidrosis and muscle fasciculations were described, these signs cleared immediately. In Case 1, return of facial sweating was followed later by clearing of the ocular portion of the Horner's syndrome. All patients showed improvement in other signs and symptoms, although in Case 8 the improvement, as determined by a telephone interview, consisted of lack of further progression of arm weakness and lessening of shoulder pain. In Case 5, despite maintained improvement in other signs of syringomyelia, the overall physical capability was deteriorating because of increasing ataxia in all extremities. In Case 7, the patient was using his hands better and both proprioception and analgesia improved within 2 weeks of his operation. This improvement was maintained when examined at 26 months and the constant, interscapular burning pain, present before operation, had not recurred. His performance, however, was still impaired by frozen shoulder joints and an only partly effective operation for replacement of a painful left hip joint.

In Case 11, the excised portion of the filum was not carried up to the tip of the conus. This patient showed striking improvement for 48 hours followed by relapse. Fourteen days after this operation, the terminal ventricle was excised, together with the remaining 1 cm of the filum. There was prompt improvement which was maintained for at least six months, but 18 months after operation a follow-up telephone call to her home in Greece revealed that she was depressed and believed that her symptoms were again progressing. This may represent the only recurrence. However, the longest follow-up period is only 30 months. In the first 11 cases, no conus symptoms followed the operation. Recurrence of muscle fasciculations or anhidrosis was not described in any case. Eight of the patients lived far from Cleveland which in some instances posed a problem for adequate postoperative evaluation.

The only postoperative complication occurred in Case 12 and it contributed to a poor result. This was a man of 50 whose syringomyelic symptoms had begun at 28 years of age. He had syphilis, diabetes, and severe generalized cervical spondylosis. A craniovertebral decompression at the age of 42 years was followed by little change, and the next year he had a Pantopaque myelogram followed by an anterior discectomy and interbody fusion at C3-4. Although the greatest narrowing of the spinal canal was at a lower level, his findings implicated the C3-4 level. The CSF protein was 170 mg/100 ml. Terminal ventriculostomy at the age of 50 years disclosed the tip of the conus at L-2. Through the unopened arachnoid the conus appeared swollen but collapsed after evacuation of CSF. The filum terminale was divided at the lower end of the exposure, and a piece 3.7 cm in length was excised up to the tip of the conus. Fluid did not escape spontaneously nor on digital compression of the conus. Within 24 hours the preoperative fasciculations in the arms had ceased: the grip in the left hand had become somewhat stronger and sweating had returned above the level of T-6. However, at 48 hours there was urinary retention and flaccid paresis of the legs and muscle fasciculations had developed in the calves. The wound was reopened and the conus was found to be distended. Incision released a quantity of bloody fluid and some clot. After this the conus symptoms improved, although the right leg remained weaker than it had been before. Since a syrinx is frequently bridged by naked, thin-walled blood vessels, this hematomyelia may have resulted from the digital compression of the collapsed syrinx. Sixteen months after operation there was no return of fasciculations in any area and sexual potency was intact. The patient continued to sweat above the level of T-6 and the area of dissociated sensory loss was somewhat smaller than before operation. The CSF protein was down to 72 mg/ml. However, although he was still able to drive his car, he functioned less well because of progressing weakness in the left hand and right leg, possibly related to his cervical spondylosis. The improvement in
Terminal ventriculostomy for syringomyelia

sweating and dissociated sensory loss and the lowered CSF protein suggest that the syrinx was draining into the lumbar sac.

Pathological Findings

The surgical specimens were compared (MS) with the terminal ventricle and filum in autopsy specimens from non-syringomyelic patients. The surgical specimens consisted of slender, conical structures varying in length from 8 mm to 8 cm. After fixation, the diameter of the rostral end was 2 to 3 mm, and the caudal end 1 mm. Multiple cross sections showed a central canal, patent throughout and increasing in size rostrally in all but two; these latter had an apparently closed canal at the rostral end, but with a patent and probably dilated canal in the more caudal sections. Both patients improved and this apparent closure at the cut end was attributed to the crushing effect of the scissors.

In a control series, the spinal cord and filum terminale were obtained at autopsy from 11 adult patients with no known neurological disease. The conus terminated at or above the level of the L1–2 disc in nine. The specimens, examined microscopically, included the tip of the conus and several centimeters of the filum. Cross sections at the junction of the conus with the filum terminale showed a closed central canal in eight specimens, patency in two, and apparent dilatation in one. In no specimen was there a patent central canal below this level. Thus, in our small series of syringomyelia patients there was a difference between the level of the conus and also of the patency of the central canal in the surgically excised specimens as compared with those obtained at routine autopsy.

Comment

Terminal ventriculostomy deserves a further trial as a substitute for craniovertebral decompression in the treatment of syringomyelia. The operative, radiological, and pathological findings in this series constitute further evidence that all syringomyelia is symptomatic hydromyelia, that "non-communicating" syringomyelia is a non-existent entity, and that intradural arachnoid diverticula, filling also from above, constitute the subarachnoid counterpart of the syrinx. The erect posture may aggravate the symptoms of syringomyelia, perhaps by increasing the downward thrust of the ventricular pressure wave. Pneumoencephalography is superior to Pantopaque myelography in demonstrating syringomyelia.

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References


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