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Echols' Procedure for Treating Syringomyelia: Case Report and Historical Review

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Abstract

A 9 year-old girl with syringomyelia and scoliosis was treated with Echols' procedure, a surgical technique that employs a metal stent to maintain drainage of fluid from the syrinx into the subarachnoid space. The patient presented to our institution 34 years later with a history of progressive myelopathy and surgically-treated deformities of the thoracic spine, lumbar spine, and right foot. Computer-assisted myelography indicated that the metal wire remained in place and that the syrinx had collapsed. Neurological examination and neurophysiological testing confirmed the presence of thoracic myelopathy, which may be due to the wire tethering the thoracic spinal cord to the dorsal dura. This is believed to be the sole long-term report of the effects of Echols' procedure. The history of direct treatment of syringomyelia is reviewed and is contrasted with indirect treatment of syringomyelia, which relieves the condition by opening obstructed CSF pathways within the foramen magnum or spine.

Keywords

Syringomyelia; Arnold Chiari Malformation; Type 1; History; Surgery

Introduction

Syringomyelia is a progressive disease that if left untreated often leads to debilitating central myelopathy with permanent sensory and motor deficits. Syringomyelia was identified in the late nineteenth century¹ and its resistance to permanent treatment spurred neurosurgeons in the early and mid-twentieth century to develop innovative therapeutic approaches.^{3,5,11,18,19,23,24,26,27,31,32} One technique that was devised but is no longer practiced was reported by Dr. Dean H. Echols (Fig. 1). His technique to treat syringomyelia involved myelotomy to create a canal for drainage of syrinx fluid into the subarachnoid space and placement of a tantalum wire through the myelotomy as a stent to prevent closure of this fistula.²³ The wire was part of a loop that exited the syrinx through the dorsal column of the spinal cord and was tied to the dura. This report recounts this technique, the clinical and radiographic outcome seen thirty-four years after surgery, and other methods and materials

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that have been reported to promote drainage of fluid from the syring to the subarachnoid space.

Case Report

History and Examination

In 1974, a nine-year-old female presented to an outside institution with progressive thoracolumbar dextroscoliosis. Neurological examination was remarkable for a band of hypalgesia within the ninth through twelfth thoracic dermatomes, which was more pronounced on the left, and for increased deep tendon reflexes in the left lower extremity. Her gait was normal. A myelogram was performed with iophendylate (Pantopaque, Lafayette Pharmacal Company, Lafayette, IN).²⁸ Widening of the spinal cord from the cervical to the upper lumbar segments was reported. The cerebellar tonsils were noted to lie well below the level of the foramen magnum consistent with Chiari I malformation. Pneumoencephalography demonstrated that air entered the basilar cisterns and the 4th ventricle, but not the other ventricles. The diameter of the cervical spinal cord was reduced as the head was elevated; this “collapsing cord sign” indicated that the intramedullary lesion was a syrinx rather than a solid tumor.

Operation

The patient was taken to the operating theater. The eighth through eleventh thoracic lamina were removed and the dura was opened. A needle was inserted through the swollen spinal cord just to the left of midline and clear fluid was removed. The arachnoid membrane was then split and according to the operative dictation an “incision put through the dorsal column into the syrinx cavity.” A stainless steel wire was passed through the myelotomy and into the syrinx, taken caudally about “two spinal segments, brought out of the dorsal column” and tied. The spinal cord “collapsed” in the process. The dura, muscle, fascia, and skin were subsequently closed in separate layers with sutures.

Postoperative Course

Despite the procedure, thoracic scoliosis continued to progress. Internal fixation using a Harrington rod and autologous bone grafting was performed three years later, fusing the thoracic spine in order to prevent further curvature. Since that time the patient has undergone various orthopedic procedures to correct deformities in the right foot secondary to progressive weakness that developed in the right lower extremity beginning five years after the initial surgery. Numbness and tingling subsequently developed in the left lower extremity. Lumbar stenosis and grade II spondylolisthesis eventually developed below her thoracolumbar fusion. She underwent L-3 to S-2 internal stabilization and fusion 27 years after her initial surgery. More recently her gait has become spastic.

The patient was seen at our institution thirty-four years after her initial treatment. No other treatment for syringomyelia or CSF diversion occurred in the interim. She had avoided MRI (magnetic resonance imaging) because the stainless steel loop (Fig. 2) within her spinal cord was confirmed to be ferromagnetic, thereby causing concern that MRI scanning might result in movement or heating of the wire loop within the spinal cord and produce further neurological dysfunction. Her latest radiographic evaluation consisted of a CT-myelogram that was performed with Isovue (Iopamidol, Bracco Diagnostics, Inc., Princeton, NJ) one year ago (Fig. 3). The spinal cord diameter was normal on this study suggesting that the syrinx was absent or collapsed (Fig. 3). Neither the metal loop nor the previous Pantopaque myelogram appeared to have resulted in adhesions or other changes in the subarachnoid space (Fig. 3).^{7,17} Echols’ procedure appears to have resulted in permanent syrinx resolution, but unfortunately did not prevent progressive thoracic myelopathy.

Her recent examination was remarkable for weakness of the distal right lower extremity where plantar flexion strength was 4/5, ankle dorsiflexion strength was 2/5, and extensor hallucis longus muscle strength was 0/5. Other extremities were of normal strength. On sensory examination, she had anesthesia distal to the right midcalf level. Hypalgesia to pinprick was noted in right C5 through right L3 dermatomes and left C2 through C6 dermatomes. Deep tendon reflexes were hypoactive in the left upper extremity, normoactive in the right upper extremity, and hyperactive in the left lower extremity. Clonus was present in the left ankle. Right patellar reflex testing resulted in adduction of the hips and extension of the left knee. Right ankle reflex was absent. Babinski sign was present on the left and absent on the right.

Clinically, she had suspended and dissociated sensory loss and hypoactive left upper extremity reflexes compatible with central cervical myelopathy. Lower extremity spasticity and hyper-reflexia could arise from cervical or thoracic myelopathy. Distal right lower extremity weakness and hyporeflexia is compatible with myelopathy affecting the thoracic spinal cord.

To clarify the sites of neurological involvement, somatosensory evoked potentials (SSEP), nerve conduction studies (NCS) and transcranial magnetic stimulation for motor evoked potentials (MEP) were performed. There was no recordable SSEP from the left posterior tibial nerve. The right posterior tibial nerve showed a normal peripheral peak, but no reproducible cortical peak. SSEP performed with stimulation of the right and left median nerves was normal. These findings suggested that dysfunction of the right dorsal column of the thoracic spinal cord caused her proprioceptive sensory deficit in the right lower extremity. The MEP potentials showed normal central motor conduction times for the right and left lower extremities, recording at the tibialis anterior, suggesting that the corticospinal pathways to both lower extremities were intact though mild degrees of axonal damage could not be accurately assessed. The right peroneal motor NCS showed signs of peripheral nerve axonal damage with no recordable CMAP (compound muscle action potential) at the distal extensor digitorum brevis muscle (EDB) and a small CMAP at the more proximal tibialis anterior muscle. The abnormality was probably the result of multiple orthopedic procedures to the right lower extremity causing direct damage to the peroneal nerve.

Her history of slowly progressive myelopathy suggested that spinal cord tethering at the site of the previous surgery might be present. Upper extremity strength was normal indicating preservation of lower motor neurons near the cervical portion of the previous syrinx, so it is unlikely that more laterally located long tracts would have been injured by the syrinx at the cervical level. If tethering were present, removal of the wire could potentially arrest the progression of thoracic myelopathy. Because Echols' procedure was performed before puberty, one could argue that the wire loop had prevented the attached spinal cord segment from reaching its normal adult position. The counter argument is that her subsequent thoracolumbar fusion procedure eliminated movement of the spinal cord relative to the spinal column at the level of the wire loop and therefore prevented repetitive traction on the spinal cord and the effects of tethering. The patient was informed of the potential benefits and risks of removal of the wire loop and has declined to pursue this treatment.

Discussion

Myelography was the method for radiographic diagnosis of syringomyelia until the widespread availability of magnetic resonance imaging rendered the radiographic diagnosis of syringomyelia completely non-invasive. Magnetic resonance imaging also provided a convenient tool to monitor changes in diameter and length of the syrinx after surgery, which is one method of evaluating the effectiveness of surgery.^{4,5,19,21,29,32,33,36} Myelotomy for

syrinx drainage was first reported by Abbe and Coley in 1892, and later practiced by others including Elsberg, Pousepp, Peiper, and Frazier. A myelotomy that entered the syrinx created a drainage channel between the syrinx and the subarachnoid space that would drain the syrinx.^{1,10,12,22} It was widely believed in the 1930's and 40's that if a method were developed to permanently maintain patency of the fluid pathway between the syrinx and subarachnoid space that the primary obstacle to successful syringomyelia treatment would be overcome.²³ In a number of studies from that era, surgeons would open the spinal cord, usually a bit lateral to the midline, enter the syrinx, and perform some intervention to maintain drainage of the syrinx.^{1,11,12,23} Objects of various types were placed in the myelotomy with the intent to keep the incision open permanently.¹¹ Some of the materials used were: silk thread, dura mater, muscle, silver clips, Teflon, and gutta percha secured by a silver clip.²³ An alternative approach was to widen the myelotomy by removing a window of dorsal column.^{11,23,24} Most of these treatments were reported to provide short-term improvement, but thereafter neurologic progression resumed unabated. Often a second surgery was performed and inspection of the previous myelotomy revealed that the spinal cord incision had closed.^{12,23} Success at maintaining patency of the syringostomy was elusive.²³

Echols and Kirgis first reported their procedure in 1949 as treatment for a patient with recurrent syringobulbia and syringomyelia after previous cyst aspiration.²³ The authors reported:

The previously visualized portion of the brain and cord were reexamined and the remaining cervical segments of the spinal cord were exposed. It was obvious that the cyst had re-expanded...An incision was made in the cyst at the level of the 5th cervical segment. A catheter was passed upward in the cyst from this point well beyond the previous opening. It was apparent that the cyst...extended from some point inferior to the midthoracic level, superiorly to the junction of the pons and mesencephalon...A drain, made by twisting several tantalum sutures together, was inserted into the cyst and brought out through another opening, which was made about 2 cm. inferior to the previously made opening. The ends of the drain were sutured to the deep surface of the dura and the dura was closed tightly.²³

Their patient had "slow but steady improvement" after this procedure. The main complaints that persisted were "stiffness and weakness of the posterior cervical muscles."²³ The patient was released to home.²³

At the time that Dean Echols and Homer Kirgis introduced their procedure, Echols was a practicing physician at Tulane University School of Medicine and Kirgis was his resident.¹⁸ Their procedure for syringomyelia treatment became known as Echols' procedure and was reported to provide relief for those suffering from the effects of the fluid collection within the spinal cord. In their original paper, Echols and Kirgis state, "It has been advocated that a foreign body be inserted into the cyst in such a way that the permanency of the communication between the cavity of the cyst and the subarachnoid space could be assured."²³ He used this procedure and stated that "...the intracystic fluid pressure had become equalized with that of the subarachnoid space undoubtedly as a result of the maintenance of the free transmission of fluid between the two spaces along the tantalum drain."²³ Tantalum wire was selected by them because the metal did not react with body tissue, was resistant to corrosion, strong, and very ductile.⁶

Our patient's procedure took place in 1974 and was not performed by Dr. Echols. In 1974, spinal cord tethering was considered to be a consequence of developmental disease of the nervous system and not surgical procedures. The effects of restriction of normal spinal cord motion by a developmental condition was reported in 1910 by Fuchs who explained that

increased tension in the spinal cord caused incontinence in myelomeningocele patients.^{13,25} Garceau used the terms “filum terminale syndrome” and “cord-traction syndrome” to describe the syndrome.^{2,14,25} Hoffman coined the term “tethered spinal cord” in 1976.^{2,20,25} Thus, when the wire was inserted into our patient’s syrinx and tied to the dura in 1974, it was not appreciated that a surgical procedure that would result in restriction of the movement of the spinal cord might result in delayed spinal cord injury. In fact, Kirgis and Echols explained that they expected that cord motion against the wire was beneficial because it would keep the fistula open.²³

For a chronic disease such as syringomyelia, the follow-up period of two years reported by Echols seems relatively short to claim permanent success. We could not find any other reports of the results of this procedure in the medical literature. The lack of general adoption of Echols’ procedure by the neurosurgical community suggests that if this procedure were employed by others, the results of this treatment were inferior to other treatments in terms of efficacy and safety. The reporting of the results of unsuccessful treatments has recently been encouraged,^{8,9,34} but negative studies have been difficult for an investigator to report and for an editor to publish. We report that in this case the method resulted in permanent collapse of the syrinx as assessed by CT-myelography thirty-four years after treatment. The stainless steel wire did not evoke an inflammatory reaction in the surrounding subarachnoid space which compares favorably to silastic catheters which may evoke fibrosis that results in occlusion of the syringe-subarachnoid shunts within the spinal cord or subarachnoid space.³⁰ However, the requirement to penetrate the spinal cord twice with Echols’ technique rather than once with myelotomy and shunting makes Echols’ procedure potentially more likely than shunting to cause injury to the dorsal columns of the spinal cord.

Improved imaging technology, understanding of syringomyelia pathogenesis, and surgical equipment has resulted in the preference of many neurosurgeons to treat syringomyelia by relieving obstructions in the CSF pathways that cause syringomyelia.^{15,16,37} Because of this, direct drainage of the syrinx has become less popular as a primary treatment for syringomyelia.^{12,23} Treating syringomyelia using surgical procedures that open obstructed CSF pathways and that do not require entrance into the spinal cord parenchyma is a practical way to avoid surgical trauma to the spinal cord.

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Figure 1.
Dean H. Echols, MD. Courtesy of The Ochsner Journal³⁵

Figure 2.
(Left) Sagittal view of the stainless steel wire (arrows) with the Harrington rod (arrowheads) running posteriorly to it. (Right) Coronal view of the spinal cord where the steel wire at T10-11 is seen (arrow). Arrowheads show the Harrington Rod.

Figure 3.

Upper and Lower Left: CT cuts at the T10-T11 level. A single Harrington rod (arrowheads) rests on a dorsal fusion mass. Metal wire (arrow) extends between the spinal cord and dura.

Upper and Lower Center: CT Post-Myelogram, Axial Images. The metal wire (arrow) is again seen through the subarachnoid space and into the cord. The Harrington rod is represented by arrowheads. *Right:* Sagittal view of CT myelogram showing 2 mm of tonsillar ectopia and normal diameter of the cervical spinal cord.