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Andrew Guillotte

Abdul-Rahman Alkiswani

Kathryn Keeler
Children's Mercy Hospital

Michael D. Partington
Children's Mercy Hospital

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Symptomatic spinal arachnoid cyst with spastic diplegia secondary to cerebral palsy: illustrative case

Andrew Guillotte, MD,[1] Abdul-Rahman Alkiswani, BS,[1] Kathryn A. Keeler, MD,[2] and Michael D. Partington, MD, MS[1,3]

[1]Department of Neurological Surgery, University of Kansas Medical Center, Kansas City, Kansas; [2]Department of Orthopedics, Children's Mercy Hospital, Kansas City, Missouri; and [3]Division of Pediatric Neurosurgery, Children's Mercy Hospital, Kansas City, Missouri

BACKGROUND Selective dorsal rhizotomy (SDR) can improve the spastic gait of carefully selected patients with cerebral palsy. Spinal arachnoid cysts are a rare pathology that can also cause spastic gait secondary to spinal cord compression.

OBSERVATIONS The authors present an interesting case of a child with cerebral palsy and spastic diplegia. He was evaluated by a multidisciplinary team and determined to be a good candidate for SDR. Preoperative evaluation included magnetic resonance imaging (MRI) of the spine, which identified an arachnoid cyst causing spinal cord compression. The cyst was surgically fenestrated, which provided some gait improvement. After recovering from cyst fenestration surgery, the patient underwent SDR providing further gait improvement.

LESSONS SDR can be beneficial for some patients with spastic diplegia. Most guidelines do not include spinal MRI in the preoperative evaluation for SDR. However, spinal MRI can be beneficial for surgical planning by localizing the level of the conus. It may also identify additional spinal pathology that is contributing to the patient's spasticity. In rare cases, such as this one, patients may benefit from staged surgery to address structural causes of spastic gait prior to proceeding with SDR.

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KEYWORDS spinal arachnoid cyst; selective dorsal rhizotomy; spastic diplegia; spasticity; cerebral palsy

Cerebral palsy is a common condition with incidence of 2–3 per 1000 live births.[1] The clinical manifestations of cerebral palsy are highly variable, with spastic diplegia being the most common clinical phenotype.[2,3] Spastic diplegia can result in varying severity of gait impairment due to lower extremity weakness, increased tone, spasticity, and orthopedic deformity. Cognition is often spared in the spastic diplegia form of cerebral palsy. Spastic diplegia is most commonly a result of insult to the developing brain between 20 and 34 weeks of gestation. This typically results in periventricular leukomalacia, which can be seen on magnetic resonance imaging (MRI). Treatment of spastic diplegia requires a multidisciplinary approach involving combinations of physical therapy, oral medications, botulinum toxin injections, intrathecal baclofen therapy, and surgical intervention.[2–5] Orthopedic surgery is often required to correct lower extremity and/or spinal deformity. Selective dorsal rhizotomy (SDR) can improve the gait and quality of life in carefully selected patients.[5–10]

There are several proposed criteria for the selection of patients for SDR with spastic diplegia.[5,11] Some of common criteria include patient age, ability to tolerate intense physical therapy, lower extremity strength greater than 3 out of 5, supportive home environment, moderate balance and limb control, absence of severe ataxia or dystonia, and absence of severe orthopedic deformity. Most authors recommend brain MRI prior to performing SDR to evaluate for basal ganglia injury, which would be a contraindication to proceeding with SDR, as well as to assess for hydrocephalus and to check shunt function, when present.

Spinal MRI prior to SDR is not routinely recommended in the literature. However, we present a case that demonstrates that it may have utility in the preoperative evaluation prior to SDR. Our patient was a 4-year-old male with spastic diplegia who was determined to be a good candidate for SDR. MRI of his spine was performed as part of the preoperative evaluation to localize the level of the conus and to evaluate for spinal cord pathology that could be contributing to

ABBREVIATIONS CSF = cerebrospinal fluid; GMFCS = Gross Motor Function Classification System; MRI = magnetic resonance imaging; SDR = selective dorsal rhizotomy.

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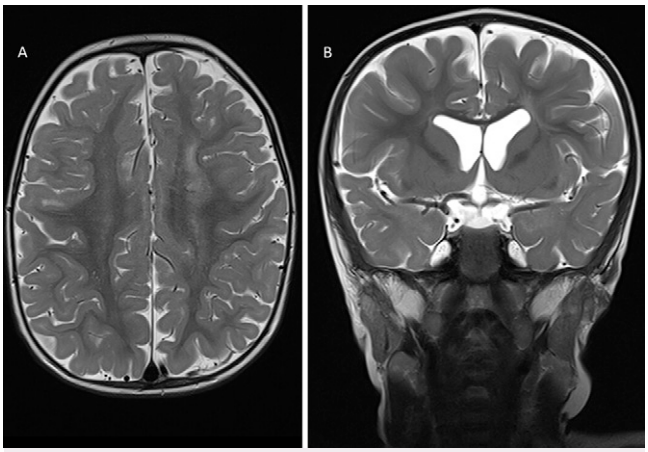

FIG. 1. The patient's magnetic resonance imaging (MRI) of the brain, demonstrating periventricular leukomalacia. **A:** Axial T2-weighted image. **B:** Coronal T2-weighted image.

his gait dysfunction. A large, compressive, dorsal intradural arachnoid cyst was identified spanning the lower cervical and upper thoracic region. The arachnoid cyst was fenestrated prior to proceeding with SDR.

Illustrative Case

The patient is a 4-year-old male with history of prematurity (29 weeks) and spastic diplegic cerebral palsy. He was referred to the multidisciplinary spasticity clinic for evaluation for surgical management of increased tone that significantly affected his gait. His gait abnormalities included toe-walking with excessive plantar flexion, increased pelvic rotation to compensate for reduced hip extension, and excessive hip and knee flexion during swing phase. He had previously been treated with oral baclofen, botulinum toxin injections, and physical therapy with some benefit. In addition to his spastic gait, the patient had been endorsing progressively worsening balance difficulty and tingling in his bilateral feet for 7 months prior to his neurosurgical evaluation. Intrathecal baclofen and SDR were among the surgical options considered for him. He was thought to be a good candidate for SDR. MRI of the brain and spine was performed during preoperative planning and evaluation. The brain MRI showed periventricular leukomalacia, which is typical for spastic diplegia–type cerebral palsy (Fig. 1). However, the spinal MRI showed a large thoracic arachnoid cyst causing compression of the spinal cord (Fig. 2). It was decided that the arachnoid cyst should be surgically treated prior to proceeding with SDR.

T2 and T3 laminectomies were performed for fenestration of the arachnoid cyst with the use of motor and sensory monitoring. The cyst was identified upon opening the dorsal dura. The cyst was widely fenestrated and much of the wall was excised. The spinal cord was observed to begin to return to a normal position within the center of the spinal canal. T2 and T3 laminoplasties were performed by securing them to the adjacent cranial and caudal vertebrae. Figure 3 shows the postoperative MRI of the thoracic spine demonstrating the decompressed cyst and normal cerebrospinal fluid (CSF) flow voids within the subarachnoid space. Despite good decompression and return of CSF circulation, the spinal cord has not completely returned to a normal shape due to the chronic compression caused by the arachnoid cyst. Formal gait analysis was performed pre– and post–arachnoid cyst fenestration. The patient

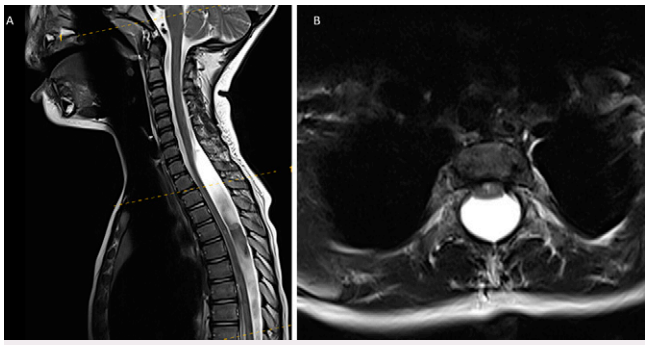

FIG. 2. Preoperative MRI of the spine, demonstrating dorsal thoracic arachnoid cyst with spinal cord compression. **A:** Sagittal T2-weighted image. **B:** Axial T2-weighted image.

achieved some improvement, which was greatest in the left lower extremity, postoperatively. After an initial improvement in his gait after arachnoid cyst fenestration, his gait improvement plateaued. He continued to have significant gait difficulties consistent with spastic diplegic cerebral palsy. He was fitted for ankle foot orthotics to assist with his gait. After he made a full recovery from the arachnoid cyst procedure, he was scheduled for SDR.

Eight months after the arachnoid cyst fenestration, the patient underwent SDR with intraoperative electromyography monitoring. The laminae of T12 and L1 were removed. The dura was opened, and the conus was visualized. The right L1 nerve root was stimulated to confirm sensory and motor function. The sensory root was dissected from the motor root and stimulated to confirm that no motor fibers were included. The sensory root was subdivided into rootlets and stimulated to determine which fibers demonstrated pathologic response. The pathologic fibers were sectioned. The process was repeated for each cauda equina root bilaterally. Thirty-eight percent of the sensory rootlets were sectioned on the right side and 40% on the left. Laminoplasty was then performed to repair the T12 and L1 vertebrae prior to closure. He again demonstrated significant improvement in his gait following the SDR. He showed a stepwise improvement in his gait following the arachnoid cyst fenestration followed by a plateau period in his improvement. He had further stepwise improvement after the SDR. Formal gait analysis was repeated 1 year after SDR demonstrating further improvement in his bilateral lower extremity function. At his final gait analysis, he was making flat-foot contact and had overall improved gait mechanics. His gait was still somewhat impaired by plantar flexion and hip abduction weakness that had

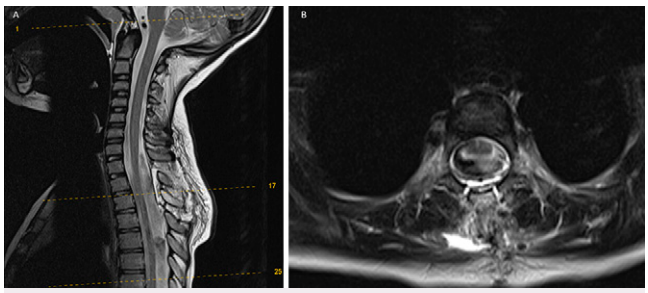

FIG. 3. Postoperative MRI of the spine, demonstrating fenestrated arachnoid cyst with decompressed spinal cord. **A:** Sagittal T2-weighted image. **B:** Axial T2-weighted image.

TABLE 1. The patient's tone measured by the modified Ashworth scale prior to intervention, after thoracic arachnoid cyst fenestration, and after SDR

Muscle Group	Preop	After Cyst Fenestration	After SDR
Lt hip abduction	2	1+	0
Rt hip abduction	2	1+	0
Lt hip flexion	0	0	0
Rt hip flexion	1	0	0
Lt knee flexion	1	0	1
Rt knee flexion	1+	0	1
Lt knee extension	1+	2	1+
Rt knee extension	1+	2	1
Lt ankle dorsiflexion	2	2	0
Rt ankle dorsiflexion	3	2	0
Lt ankle plantarflexion	0	0	1
Rt ankle plantarflexion	0	0	1

Modified Ashworth scale definition: 0 = no increase in muscle tone; 1 = slight increase, end range; 1+ = slight increase, half range; 2 = more marked increase; 3 = considerable increase; 4 = affected part rigid.

been recognized on previous evaluations. Table 1 summarizes the modified Ashworth score for each major muscle group in the lower extremities prior to surgical intervention, after arachnoid cyst fenestration, and after SDR. All three evaluations were performed by the same physical therapist. The modified Ashworth scores showed improvement after each intervention. His family reported that they had noticed subjective improvement in his gait and endurance. Video 1 shows a lateral view of his gait before and after SDR obtained during formal gait evaluation. The video before SDR was taken after thoracic arachnoid cyst fenestration.

VIDEO 1. Clip showing the lateral view of the patient's gait before and after selective dorsal rhizotomy (SDR). The video before SDR was taken after thoracic arachnoid cyst fenestration. Click here to view.

The patient was seen in the neurosurgery clinic at 1 year following the SDR. He will continue to follow up with the rehabilitation medicine clinic and spasticity clinic for long-term monitoring. He will follow up in the neurosurgery clinic on an as-needed basis.

Patient Informed Consent

The necessary patient informed consent was obtained in this study.

Discussion

Observations

Selective dorsal rhizotomy is a well-established surgical treatment for the spastic diplegia subtype of cerebral palsy. This procedure involves the selective transection of dorsal rootlets in the lower thoracic and lumbar segments of the spinal cord. By selectively cutting the sensory nerve fibers that transmit abnormal signals from the muscle spindles to the spinal cord, SDR reduces the level of spasticity in the lower extremities through interruption of the afferent limb of the reflex arc. The reduction in spasticity is expected to improve mobility, gait pattern, and reduce the risk of joint contractures and deformities.[12]

At the time of its inception by Förster[13] in 1908, the surgical approach involved multilevel laminoplasty with complete transection of dorsal nerve roots. This often left patients with marked muscle weakness, sensation and proprioception loss, and other surgical complications such as spinal deformity and infections. It was undoubtedly, however, effective in reducing the spasticity that characterizes the disease process. With its re-conception starting in the 1960s, SDR transitioned to division of only a fraction of the dorsal rootlets, such that proprioception and sensation could be maintained through a sufficient subset of afferent inputs.

While there is no consensus on selection criteria for SDR, the guidelines initially proposed by Peacock and Arens[14] have long outlined the "ideal" candidate for the procedure. These include suggestions around birth events, functional level emphasizing Gross Motor Function Classification System (GMFCS) level and spasticity via the Ashworth scale, gait analysis, head imaging, prior interventions, and child and family factors.[15] Favorable outcomes have been repeatedly shown in patients who undergo both SDR and physiotherapy, as evidenced by three seminal randomized controlled trials in the field.[16–18] While long-term data show generally positive results in a wide range of patient subsets, with reduction in muscle spasticity and tone, as well as improvement in muscle range of motion, Gross Motor Function Measure, and functional status,[19] it appears that young, diplegic children with GMFCS grade II or III function are most likely to benefit from SDR.[20]

Spinal arachnoid cysts are a rare spinal pathology that most commonly occur in the thoracic region and can be extradural, intradural, intramedullary, or a combination.[21] The trapped pockets of CSF result in localized abnormal CSF circulation. In some cases, the arachnoid cysts can cause compression of the spinal cord. They most commonly arise dorsal to the spinal cord but can also be located ventrally. There are several proposed causes, including congenital, infectious, posttraumatic, postsurgical, and idiopathic etiologies. Spinal arachnoid cysts are often classified as primary or secondary. Primary arachnoid cysts are of idiopathic etiology. Secondary arachnoid cysts are the result of infectious, hemorrhagic, or other inflammatory etiologies.[22] Asymptomatic spinal arachnoid cysts may be observed. Common clinical symptoms of spinal arachnoid cysts include back pain, radiculopathy, lower extremity weakness, bladder dysfunction, and myelopathy.[23] Surgical treatment is recommended for symptomatic arachnoid cysts. Extradural arachnoid cysts may be resected. Intradural arachnoid cysts are typically fenestrated or marsupialized to decompress the spinal cord and restore CSF flow.[23–25] Primary arachnoid cysts typically respond well to fenestration or marsupialization while secondary arachnoid cysts are challenging and have high rates of recurrence.[26,27] Cyst shunting may be considered for recurrent or surgically inaccessible cysts.

Patients with intradural spinal lesions, such as the arachnoid cyst in the case of our patient, can present with confounding examination findings. In a review by Bond et al.,[24] approximately two-thirds of the 31 patients who had undergone operations for spinal arachnoid cysts were found to have symptoms of myelopathy or radiculopathy, with pain, lower extremity weakness, gait instability, spasticity, and sensory loss chief among their complaints. Additionally, 10% of patients were incidentally diagnosed after an MRI for further evaluation of another central nervous system or spine abnormality. Although Park and Johnston[28] suggest radiological evaluation with radiographs of the lumbosacral spine and hips to assess for pathologies such as hyperlordosis, scoliosis, spondylolisthesis,

hip subluxation, dislocation, or deformities, they do not comment on exclusion of cord pathologies with MRI.

However, supplementing with spinal MRI may hold great potential in delineating possible multifactorial contributions to weakness and gait deficits secondary to spasticity. Early identification of confounding spinal pathologies can aid in the optimal choice of surgical method (excision, marsupialization, fenestration, partial excision, or shunting) as well as surgical timing in the context of planned SDR. Additionally, it may help in preoperative planning of the SDR itself as the definitive method of localizing the conus. By identifying and appropriately treating said spinal pathologies, improvements in motor function and spasticity can be gained that exceed those of SDR alone, as evidenced by improvements in American Spinal Injury Association Impairment Scale[29] and in the Neurological Scoring System[22] that have been observed following surgical intervention for spinal arachnoid cysts.

Lessons

Spastic diplegic cerebral palsy is a common condition that is sometimes treated with SDR. The preoperative evaluation for SDR commonly includes brain MRI to confirm the typical findings of periventricular leukomalacia and to ensure there are no additional brain lesions that would be a contraindication for SDR. We have presented a case in which a preoperative spinal MRI revealed a compressive thoracic spinal arachnoid cyst that was contributing to the patient's lower extremity spasticity and gait difficulty. He had spasticity and gait improvement following cyst fenestration and further improvement following SDR. This case indicates that spinal MRI prior to SDR may be beneficial. It allows for preoperative planning of the laminectomy or laminoplasty level through clear identification of the conus. Preoperative MRI may also identify spinal cord pathology, such as compressive lesions, neoplasms, tethered spinal cord, or other spinal cord lesions that may be contributing to, or mimicking, the spasticity of cerebral palsy. The primary limitation of this case study is that it is a single case study. We are not able to provide any analysis of the frequency that preoperative MRI before SDR would alter the surgical plan but acknowledge that it is likely rare.

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Disclosures

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions

Conception and design: Partington. Acquisition of data: Guillotte, Keeler, Partington. Analysis and interpretation of data: Alkiswani, Keeler. Drafting of the article: Guillotte, Alkiswani, Partington. Critically revising the article: all authors. Reviewed submitted version of the manuscript: Guillotte, Alkiswani, Partington. Approved the final version of the manuscript on behalf of all authors: Guillotte. Administrative/technical/material support: Guillotte.

Supplemental Information

Videos

Video 1. https://vimeo.com/875593563.

Correspondence

Andrew Guillotte: University of Kansas Medical Center, Kansas City, KS. aguillotte@kumc.edu.