Combined ventral and dorsal rhizotomies for dystonic and spastic extremities

Report of six cases

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Secondary dystonia is associated most often with CP, but can occur after traumatic brain injuries, strokes, or other disorders that affect the basal ganglia, usually the putamen. Secondary dystonia can be focal, segmental, hemidystonia, or generalized. Dystonic CP is generalized in most cases and affects the face, neck, trunk, and all extremities. Dystonia can be mild or severe, and if severe, it causes discomfort, impedes caregiving, and may interfere with function. In children with CP, dystonia often coexists with spasticity in the extremities.

Children with secondary dystonia are usually treated initially with oral medications such as baclofen, levo/carbidopa, and trihexyphenidyl, and with botulinum toxin injections. If those are insufficient, intrathecal baclofen is often used, particularly for children with severe, generalized dystonia who have some potential for improved function if dystonia is reduced. Because of its effects on ease of care and comfort, intrathecal baclofen is also used for children with severe generalized dystonia who have no reasonable chance for improved function. For children who have a mixture of generalized dystonia and spastic extremities, both can be improved with intrathecal baclofen.

Occasionally, however, children present with severe secondary dystonia in their extremities but hypotonia in their neck and trunk. They may or may not have associated spasticity in their extremities. If those children are severely delayed intellectually, with virtually no likelihood of improved function, a more definitive procedure than intrathecal baclofen may be considered, analogous to the use of lumbar rhizotomies for lower-extremity spasticity. For such individuals, a combined ventral and dorsal rhizotomy appears to be a reasonable option because it addresses their dystonia with ventral rhizotomies and their spasticity with conventional dorsal rhizotomies. In this paper we describe the use and outcome of combined rhizotomies in six children (Table 1).

Case Reports

Case 1

History and Examination. This 13-year-old girl presented to the Spasticity and Movement Disorders Clinic at the Children’s Hospital of Pittsburgh with a history of shunt placement for hydrocephalus, severe mental retardation, blindness, spasticity, and dystonia secondary to an in utero infection with toxoplasmosis. She had undergone a lumbar dorsal rhizotomy when she was 3 years old. She had severe spasticity, with Ashworth scale2 scores of 4/5 in the biceps and 3/5 to 4/5 in the wrist flexors and extensors, but she had no spasticity elsewhere in her body. Her dystonia scores on the BAD scale3 were 2/4 in both upper extremities. The hypertonicity was severely impeding dressing and upper-extremity care, and was causing elbow flexion contractures. A cervical dorsal and ventral rhizotomy was recommended.

Abbreviations used in this paper: BAD = Barry–Albright dystonia; CP = cerebral palsy.
upper extremity and her care was significantly easier. Hypertonicity did not recur during 3 years of follow-up.

**Case 3**

*History.* This 8-year-old girl presented to the clinic because of refractory dystonia. At 1 month of age she had contracted severe pneumococcal meningitis that had caused venous infarctions, global brain damage, and seizures. Following that, she had experienced profound developmental delays and developed generalized spasticity and dystonia that were treated initially by high doses of oral baclofen and multiple orthopedic operations. When she was 5 years old, a baclofen pump and catheter were inserted. Her spasticity improved but the dystonia was unaffected, with BAD scores of 1/4 in the neck and trunk but 3/4 in both upper and both lower extremities. Her intrathecal baclofen dose was gradually increased to 1300 μg/day, without significant improvement.

*Operation and Postoperative Course.* Because of her unresponsiveness to intrathecal baclofen, a combined dorsal and ventral rhizotomy of the cervical and lumbar nerve roots was performed to facilitate care and improve comfort. Osteoplastic laminotomies were performed at C4–7 and L1–S2. Two thirds of the dorsal roots and 50% of the ventral roots were divided bilaterally from C5–8 and L2–S1. Postoperatively, spasticity and dystonia were improved but not eliminated. Three years postoperatively she had no spasticity except in the biceps and adductors (Ashworth Scores 3/5 in both instances); she was dystonic in her extremities less than 50% of the time, and the dystonia did not interfere with caregiving.

**Case 4**

*History and Examination.* This 7.5-year-old girl developed severe generalized spasticity and dystonia secondary to anoxic event caused by a supraglottic papilloma when she was 5 years old. She was becoming progressively tighter in her extremities and more difficult to dress. Her full leg extension made going through doorways difficult.

She had a mixture of spasticity and dystonia in her upper extremities, with Ashworth scores of 3/5 in the pectorals, 4 in the triceps, and 3 in the wrist flexors. The BAD scores were 4/4 in both upper and both lower extremities.

*Operation and Postoperative Course.* At operation, 80% of the dorsal roots and 66% of the ventral roots at C5–T1 and at L2–S1 were divided. One month postoperatively, her Ashworth scores were 1/5 in all muscle groups in the upper and lower extremities. Her BAD scores were 1/4 in the upper and lower extremities. She was then lost to follow-up.

**Case 5**

*History and Examination.* This 12-year-old boy presented to the Spasticity and Movement Disorder Clinic with refractory spasticity and dystonia in the extremities secondary to CP. At the age of 4 years a baclofen pump and intrathecal catheter had been inserted, but his dystonia worsened and was refractory to intrathecal baclofen at a dose of 1490 μg/day. His Ashworth scores were 4 in the biceps, adductors, and quadriceps. He had significant contractures of the hamstrings and plantar flexors. His BAD scores were 1/4 in the upper extremities and 3/4 in the lower extremities.
Operation and Postoperative Course. At operation the patient underwent an L1–S1 osteoplastic laminotomy in two sections to preserve the intrathecal catheter. He had a combined sensorimotor rhizotomy of L2–S1, dividing 60% of the dorsal roots and 50% of the ventral roots at each level. Postoperatively he had no evident spasticity or dystonia in the lower extremities; tone in the upper extremities was unaffected. Six months postoperatively his care was significantly easier and he was more comfortable. He had no dystonia or spasticity in the lower extremities.

Case 6

History and Examination. This 9-year-old boy with Pelizaeus–Merzbacher syndrome had dystonia in his lower extremities and chorea of his head, trunk, and upper extremities. He was treated initially with intrathecal baclofen with the catheter tip at C-5, and his maximum daily intrathecal baclofen dose was 830 μg/ml. He had insufficient reduction in lower-extremity hypertonicity, with Ashworth scores of 5/5 in the adductors and quadriceps, 3/5 in the plantar flexors, and BAD scores of 3/4 in the lower extremities.

Operation and Postoperative Course. At operation the patient underwent a T12–S1 osteoplastic laminotomy and an L1–S1 combined dorsal and ventral rhizotomy was performed, dividing 50 to 75% of the dorsal and 50 to 100% of the ventral roots. Postoperatively, he had no residual spasticity or dystonia in his legs. He developed a cerebrospinal fluid infection with *Enterococcus* sp. and the pump and intrathecal catheter were removed. At his 6-month follow-up visit, his lower-extremity spasticity and dystonia was resolved. Since removal of his pump he has experienced the return of mild upper-extremity chorea.

Discussion

These six cases demonstrate the potential of combined ventral and dorsal rhizotomies to improve severe dystonia affecting the upper or lower extremities in children with concomitant spasticity. Ventral rhizotomies would be expected to help not only secondary dystonia but hereditary degenerative dystonia as well. There are obviously few cases in which this operation is indicated, but for those few it can be helpful to the patient and family and gratifying to the physician. Most children with secondary dystonia can be effectively treated with intrathecal baclofen; however, this treatment does not alleviate dystonia satisfactorily in approximately 10% of cases. In other cases, intrathecal baclofen is used and is effective, but must be discontinued because an infection develops and parents are not willing to allow another pump to be implanted.

Ventral rhizotomies appear to be effective for dystonia in the extremities, and can be done either in the cervical region, the lumbar region, or both. However, ventral rhizotomies do not alter dystonia in the trunk, and if truncal dystonia is severe, the procedure is probably not indicated. For those children, if intrathecal baclofen is not an option, bilateral deep brain stimulation is probably the only viable alternative.

For the children in this series, dystonia was their predominant movement disorder, but they also had spasticity. Their dystonia was treated with ventral rhizotomies and their spasticity with dorsal rhizotomies. If no spasticity had been present, ventral rhizotomies alone would have been performed, because there is no evidence that dorsal afferent impulses have anything to do with the pathophysiological mechanisms of dystonia. In fact, the patient in Case 2 in this series underwent an initial dorsal rhizotomy only, and there was no change in her dystonia postoperatively.

The evaluation of hypertonicity in children who have both dystonia and spasticity requires the differentiation of the two disorders. Spasticity is a velocity-dependent increased resistance to passive muscle stretch, typically of agonists but not antagonists. It is graded by passively moving a joint through its range of motion at different velocities, and cannot be graded when a muscle is being contracted by dystonia. In contrast, dystonia is characterized by sustained muscle contractions, generally of agonist and antagonist muscles simultaneously, resulting in twisting and abnormal postures; it is graded by observing abnormal movements and their interference with function, not by palpating resistance to passive muscle stretch.

In our series, parents and caregivers consistently reported less patient discomfort and improved ease of care postoperatively. Despite the high proportion of dorsal rootlets that were divided, no child developed decubitus ulcers postoperatively. As Case 2 in this series demonstrates, the percentage of ventral rootlets cut is important when the goal is to alleviate dystonia. The more severe the dystonia, the higher the percentage of rootlets that need to be divided.

Earlier cases include those reported in 1970 by Kottke, in which dystonia and athetosis improved in six children with CP after bilateral dorsal rhizotomies of C1–3. In 1973, Heimburger et al. reported improved dystonia, athetosis, and spasticity in 15 children after dorsal rhizotomies of C1–3. In 1977 and 1978, Fraioli et al. reported sectioning of the dorsal nerves of C1–5 to treat dystonia and athetosis in 16 patients with CP, and observed moderate improvement in eight, slight improvement in five, worsening in two, and death in one. Truncal dystonia improved in a few patients.

Other practitioners have used ventral rhizotomies to treat dystonia. Taira and Hori treated 30 adults who had torticollis with intradural ventral rhizotomy of C1–2, extradural denervation of the C3–6 posterior rami, and contralateral sectioning of branches of the spinal accessory nerve to the sternocleidomastoid muscles. They observed results that were comparable to the traditional Bertrand procedure, but with less sensory deficit than after that procedure. Chen et al. performed selective ventral rhizotomies in three patients, with “satisfactory” results. In our cases, rhizotomies were done nonselectively, without intraoperative stimulation. We saw no clear indication for performing selective motor rhizotomies.

One of us (A.L.A.) performed a combined dorsal and ventral rhizotomy in a 13-year-old boy with severe hemidystonia in Africa, dividing two thirds of the dorsal roots and 85% of the ventral roots from C5–8. The child had excellent reduction in his dystonia at his 2-year follow-up visit, with no adverse side effects and a small amount of retained volitional movement. Because intrathecal baclofen is rarely available in third-world countries, rhizotomies may be a more applicable option there—either ventral rhizotomies for dystonia or combined rhizotomies for dystonia and spasticity.
Conclusions

The long-term effects of cervical and lumbar rhizotomies in this population are unknown. The patients are being asked to make follow-up visits to determine if they experience known potential complications of cervical and lumbar rhizotomies such as swan neck deformity, scoliosis, or lordosis. We have not observed decubitus ulcers or deep venous thrombosis postoperatively, probably because although the patients’ legs have less movement than preoperatively, they are not flaccid.

References


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