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Selective Dorsal Rhizotomy for Spastic Cerebral Palsy

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Cerebral palsy (CP) is a major neurological problem in children; it occurs in 1 of 500 live births and inflicts lifelong disabilities on those with the disorder. Among several factors contributing to the disabilities of patients with CP is spasticity. Spasticity affects nearly 80% of patients with CP, and once fully developed, it never resolves spontaneously. It hinders motor activities in daily living and also causes muscle contractures and orthopaedic deformities in growing children. Selective dorsal rhizotomy (SDR) reduces spasticity in CP; this reduction facilitates patients' motor performance and alleviates orthopaedic deformities. Several surgical techniques for SDR are currently used. The standard technique requires an L1–S1 laminectomy or laminoplasty for visualization of all dorsal nerve roots exiting at their respective foramina. The following is a description of an operation that includes an L-1 laminectomy, ultrasonographic localization of the conus medullaris, and partial deafferentation of L1–S2 roots with electromyographic (EMG) testing under an operating microscope. Advantages of this technique include decreased operative time, reduced postoperative pain, as well as minimal risk of progressive lumbar instability. This procedure is both effective and well tolerated in appropriately selected children and adults.

We have performed this SDR in more than 1500 children and young adults since 1991.

Patient Selection

The primary beneficiaries of SDR are children with spastic diplegia, the most prevalent subtype of CP in which the lower extremities are affected with minimal or no involvement of the upper extremities (**Table 27–1**). Children with spastic quadriplegic CP, in whom all extremities and the trunk are involved, also benefit from SDR. In spastic hemiplegic CP, spasticity is not a predominant cause of motor impairments, and reduction of spasticity does not greatly improve motor functions. Some adults younger than 40 years of age who have relatively mild spastic diplegia and can walk independently are also able to benefit from SDR.

When evaluating a patient, one should first be certain that a patient's motor impairment dates back to infancy and has taken a course of steady improvement rather than progressive deterioration during the preschool years. A careful review of the patient's perinatal history and medical history provided by physicians and therapists is invaluable. A history of premature birth is considered a positive factor in the selection of candidates for SDR. Findings from a neurological examination determine whether spasticity is a major cause or the only cause of muscle hypertonia and significantly hinders motor activities (e.g., sitting, crawling, standing, and walking). In addition, the severity of orthopaedic deformities and their effects on a patient's motor performance should be assessed in detail. A thorough understanding of orthopaedic problems by the neurosurgeons performing SDR is desirable, although orthopaedists and therapists can assist in evaluation. Radiological evaluation includes lumbosacral spine and hip radiographs. The spine radiographs may show the presence of lumbar hyperlordosis, scoliosis, spondylolisthesis, and congenital anomalies. Hip radiographs may reveal hip subluxation and dislocation, deformities that influence the timing of surgical interventions. In children who were delivered at term, head magnetic resonance imaging (MRI) is obtained because diverse abnormalities underlie the diagnosis of spastic diplegia or quadriplegia. In adults with back pain, spine MRI is obtained to rule out herniated disk and other intraspinal disorders. A gait analysis is not routinely obtained, but it helps to con-

Table 27–1 Indications for Selective Dorsal Rhizotomy for Spastic Cerebral Palsy

Children Younger than 18 Years
At least 2 years of age
Diagnosis of spastic diplegia or spastic quadriplegia
Some form of independent mobility (e.g., crawling or walking) with or without an assistive device
History of premature birth; if born at term, child must have typical signs of spastic diplegia
Patients exhibit potential for improvement in functional skills after dorsal rhizotomy
Adults between Ages 19 and 40 Years
Diagnosis of spastic diplegia
History of premature birth
Currently ambulates independently without assistive device
Relatively mild fixed orthopaedic deformities
Patients exhibit potential for functional gains after dorsal rhizotomy
Patients exhibit motivation to perform home exercise program

firm the presence of spasticity before SDR and also to assess changes in motor performance after SDR.

SDR is not considered for children younger than 2 years because CP cannot be diagnosed with certainty in young children. Dystonia that is concomitant with spasticity is not a contraindication for rhizotomy. Dystonia is exceedingly rare in spastic diplegia but is common in nonambulatory children with whole body involvement. Dystonia becomes clinically evident by age 5 years. SDR in patients with dystonia can improve motor function, and this improvement does not worsen the dystonia. Rigidity is also rare in spastic diplegia, but rigidity cannot be ascertained in the presence of concomitant spasticity. Severe damage to the basal ganglia, as revealed with MRI, is considered a contraindication because of the possibility of concomitant rigidity. A history of multiple orthopaedic operations is generally a contraindication for rhizotomy, mainly because of severe muscle weakness and fixed deformities. Patients with severe fixed joint deformities are excluded from SDR because the deformities limit gains in motor function after rhizotomy. The best example is severe crouch knees due to overstretched or lengthened heel cords after heel cord release procedures. Severe scoliosis is only a relative contraindication for SDR through a single-level laminectomy. Children with increased muscle tone due to severe hydrocephalus, intrauterine and neonatal infections, and head trauma are not candidates for SDR. In general, neuronal migration disorders are contraindications for rhizotomy, but children with typical clinical features of spastic diplegia due to schizencephaly can benefit from rhizotomy.

Preoperative Preparation

Oral midazolam is administered, if deemed necessary. Intubation is performed while the patient is under deep seroflurane anesthesia; sometimes intubation is facilitated by short-lasting muscle relaxants (e.g., atracurium or vecuronium). Anesthesia is induced with seroflurane, and nitrous oxide and is maintained with fentanyl (10 µg/kg), 2% seroflurane, and 70% nitrous oxide. Propofol is avoided because it alters EMG activities. The patient receives a dose of antibiotic before a skin incision. A bladder catheter is inserted.

Operative Procedure

The patient is placed in a prone position on the operating table so that cerebrospinal fluid (CSF) is pooled rostrally and CSF loss from the intracranial compartment is minimized (**Fig. 27–1**). Needle electrodes are placed bilaterally in the adductor longus, vastus lateralis, anterior tibialis, medial hamstring, and medial gastrocnemius muscles in preparation for intraoperative EMG examinations.

Localization of the Conus Medullaris and a Single-Level Laminectomy

Normally, the conus medullaris terminates between the T-12 and L-3 spinal levels. For a laminectomy to be limited to a single level, the conus medullaris must be localized

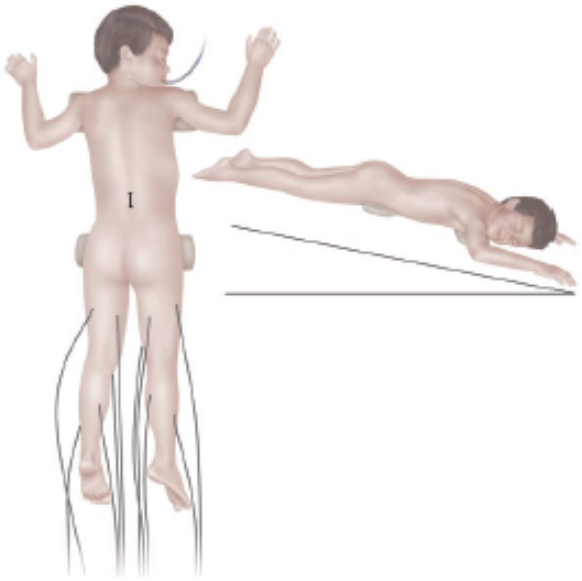

Figure 27–1 After electromyographic electrodes are inserted, the patient is placed prone in the Trendelenburg's position to minimize cerebrospinal fluid loss during the operation.

in several steps with ultrasound before a laminectomy is completed.

In children younger than 10 years, the conus medullaris and cauda equina are localized with axial views of the spinal cord ultrasound through the skin and paraspinal muscles. An ultrasound probe is placed lateral to the spinous process to get axial views of intradural structures at a few levels (**Fig. 27–2A**). On axial views, the conus appears hypoechogenic and circular. The ventral and dorsal roots are attached to the lateral aspects of the conus. Pulsatile movements of the conus are always present. The cauda equina appears as a hyperechogenic mass. It also has pulsatile movements (**Fig. 27–2B**).

In children younger than 10 years, the skin incision is made over the interlaminar space where the conus has been localized with ultrasound. The paraspinal muscles are injected bilaterally with saline solution that contains epinephrine in a concentration of 1:400,000. The interlaminar space is exposed, and the interspinous ligament and ligamentum flavum are removed. The conus is localized with axial and sagittal views of ultrasound. If, with ultrasound, the spinal cord is rostral to the conus or the cauda equina is caudal to the conus, then the skin incision is extended rostrally or caudally. The next interlaminar space is exposed. If the conus is clearly localized with axial and sagittal views of ultrasound, the next caudal interlaminar space is exposed. The cauda equina is localized with ultrasound (**Fig. 27–2B**).

In older children (older than 10 years) and adults, ultrasound localization of the conus is impossible. Thus the spinous process of the L-1 vertebra is localized with a lateral radiograph of the lumbosacral spine and marked by ~0.2 ml of indigo carmine. First, the T12-L1 interlaminar space is exposed. The ligamentum flavum is removed to expose the extradural fat tissue. Ultrasound examination of intradural structure is done through the interlaminar space (**Fig. 27–2A**). If the interlaminar space is tight for the ultrasound examination, it is enlarged with a Kerrison punch. Two levels of interlaminar space are examined to localize the conus and cauda equina.

If the conus and cauda equina are localized, a single-level laminectomy is performed with a Midas Rex craniotome with a B5 attachment (Medtronic Powered Surgical Solutions, Fort Worth, Texas) (**Fig. 27–3A**). After the lamina is removed, ultrasound examination of the laminectomy site is obtained again to confirm that the conus and cauda equina are at the laminectomy site. On the ultrasound examination, the conus is distinguished from the cauda equina as follows (**Fig. 27–2B**): a sagittal examination reveals the conus as a hypodense triangle tapering caudally. The ventral and dorsal spinal roots appear hyperdense. When only a caudal end of the conus is in the laminectomy area, sagittal examination does not clearly delineate the conus, and an axial examination is required. A hypoechogenic circular structure on axial view is sought at the center of the dural tube; it is most reliable in localizing the conus. Also, on axial view, one can notice a small cleft between the dorsal and ventral spinal roots on the lateral aspect of the conus. This cleft is an important anatomical landmark because it guides the surgeon in separating the dorsal roots from the ventral roots. Sometimes the patent central canal appears hyperechogenic within the conus.

Even in 2-year-old children, only a single-level laminectomy is needed for SDR. The laminectomy should cover at least 5 mm of the conus so that the dorsal roots are safely separated from the ventral roots at a later stage of operation. If it is needed for the adequate exposure of the conus,

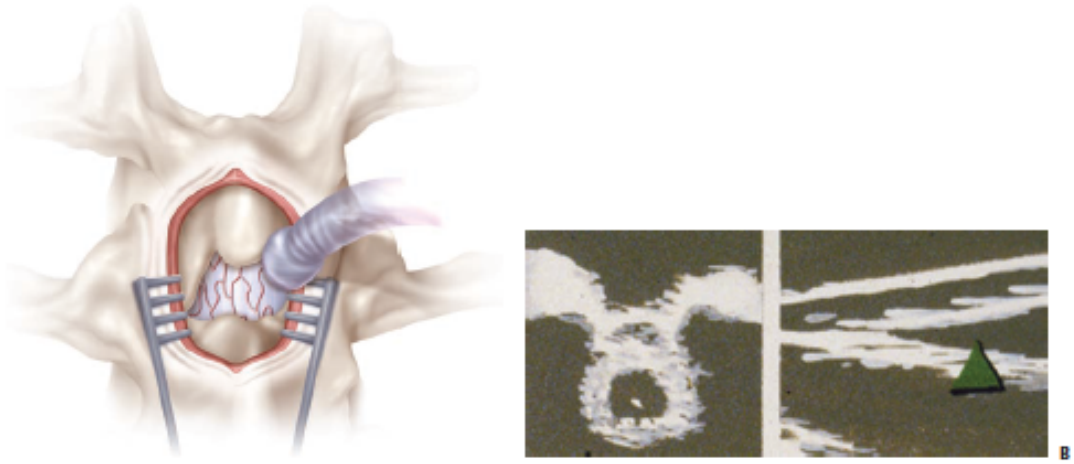

Figure 27–2 (A) Through an L1-2 interlaminar space or a keyhole laminotomy, the conus medullaris is identified with ultrasound. **(B)** The conus appears hypoechogenic (*arrowhead*) in contrast to the hyperechogenic cauda equina. If the conus is not identifiable through the interlaminar space, then an ultrasound examination is repeated through the L1-2 interlaminar space.

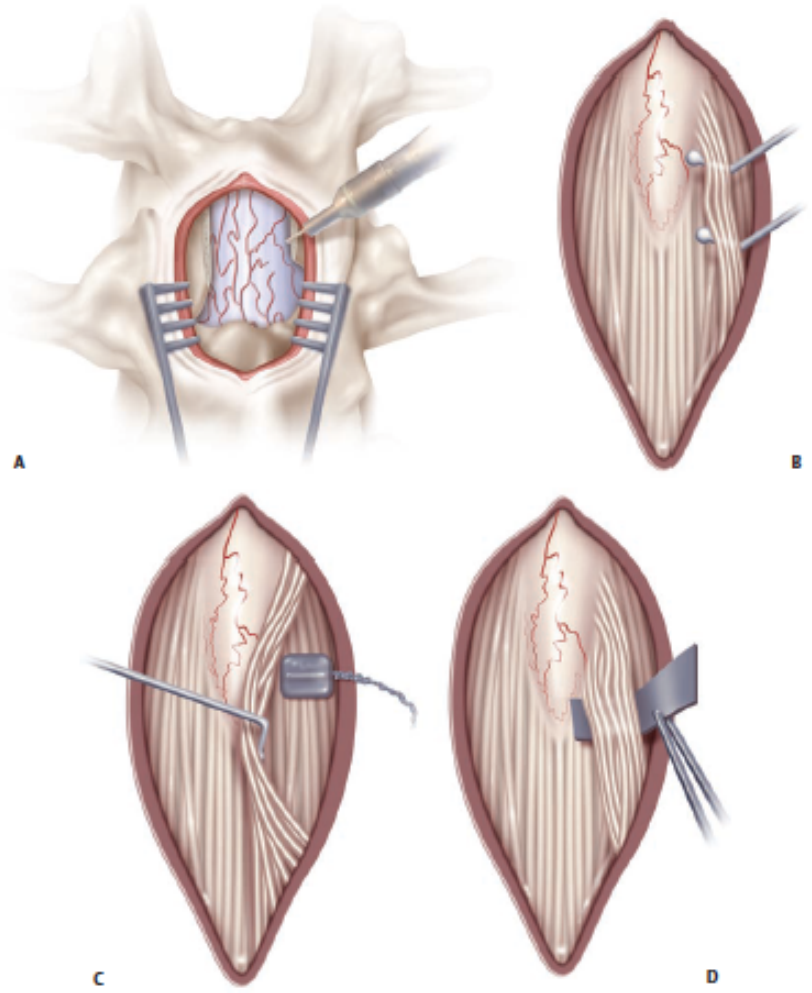

Figure 27–3 **(A)** After the conus is clearly identified, a single laminectomy is done entirely with a Midas Rex craniotome. At least 5 mm of the caudal conus should be exposed. The laminectomy extends laterally close to the facet joint. **(B)** After the dural incision, an operating microscope is brought into the field. The L-1 and L-2 spinal roots are identified at the corresponding intervertebral foramina, and the filum terminale in the midline is found. **(C)** The L-2 dorsal root and the dorsal roots medial to the L-2 root are retracted medially to separate the L2-S2 dorsal roots from the ventral roots. The thin S3-5 spinal roots exiting from the conus are identified. A cotton patty is placed over the ventral roots and lower sacral roots. **(D)** A 5 mm Silastic sheet is placed under the L2-S2 dorsal roots, after which the surgeon again inspects the L-2 dorsal root at the foraminal exit, the lateral surface of the conus between the dorsal and ventral roots, and the lower sacral roots near the filum terminale. The inspection ensures placement of only the L2-S2 dorsal roots on top of the Silastic sheet.

one-third of the lamina that is immediately rostral to the laminectomy is removed. A wide laminectomy is necessary to expose the spinal nerve roots for EMG testing.

Other neurosurgeons perform SDR through L2-S1 or L1-S1 laminectomy or laminoplasty. The multilevel laminectomy makes SDR easy. Nevertheless, the multilevel laminectomy or laminoplasty, when performed in children with spastic diplegia or quadriplegia, can lead to late spine deformities, including spondylolisthesis and increased lumbar lordosis or scoliosis. The multilevel laminectomy may carry a higher risk of spine deformities after SDR in older children and adults than in young children.

Separation of Dorsal Roots from Ventral Roots

After bleeding from the epidural veins and bone is controlled, a dural incision is made. Saline irrigation is not used after the dura is opened because it alters EMG responses. An operating microscope is then brought into the field and used during EMG testing and sectioning of dorsal root fascicles. The operating table may be slightly rotated away from the surgeon as the contralateral spinal roots are dissected. The arachnoid is removed, and the conus and filum terminale are identified. At this point, EMG activities are continuously monitored to determine if any movement of the nerve roots evokes EMG activities. Stretching and pressure on the ventral roots but not on the dorsal roots evoke EMG activities and often movement of the patient's lower extremity.

Next, the L-2 spinal roots are identified at the neural foramen, and the L-2 dorsal root is separated from the ventral root (**Fig. 27–3B**). The L-2 ventral and dorsal roots are traced back to the conus until the cleft between the ventral and dorsal roots is identified. Then the L-2 and adjacent dorsal roots are gently retracted medially, and a cotton patty is placed over the ventral roots (**Fig. 27–3C**). The L-1 root is left untouched at this point. Next, the conus and the filum terminale are examined, and the S2-5 sacral roots that exit the conus are identified. The S-2 dorsal root can be bulky, especially in patients with the postfixed lumbosacral plexus, but there is always an abrupt and marked decrease in size of the S-2 root. The individual S3-5 spinal roots appear as thin threads. The dorsal and ventral roots at this level are close together without intervening space between them, so all of the S3-5 spinal roots are left intact. The lower sacral roots can best be identified with a gentle lift at the dorsal roots from the entry zone on the dorsal aspect of the conus. Whenever the surgeon is unsure of the exact identification of the S3-5 spinal roots, then sparing the S-2 dorsal root would be prudent.

Once the L2-S2 dorsal roots are identified, a 5 mm wide blue Silastic sheet (Dow Corning, Midland, Michigan) is placed around all of the dorsal roots and distant from the conus (**Fig. 27–3D**); the Silastic sheet keeps the L2-S2 dorsal roots safely separate from the ventral and lower sacral roots during the rest of operation. Before starting EMG testing, the surgeon reexamines three structures to ensure that no ventral root or lower sacral root is over the Silastic material: the L-2 foraminal exit, the cleft lateral to the conus between the ventral and dorsal roots, and the S3-5 roots.

Identification of Individual Dorsal Roots

A shortcoming of this technique, as compared with alternative techniques, is difficulty in the identification of individual dorsal roots with certainty. Precise identification of the roots, however, is not critical for SDR because all major lower extremity muscles of children with spastic CP receive motor innervation from several segments. As is shown in animal experimental studies, significant somatotopic organization and sprouting may occur in the spinal cord and brain after deafferentation.

The L-2 dorsal root is readily identified at the neural foramen. The L3-S2 dorsal roots below the conus are close together without a natural separation, so unequivocal identification of the individual dorsal root is difficult. Nevertheless, dorsal root fibers of individual segments are roughly identified as follows. First, the dorsal roots are spread on top of the Silastic sheet. The L-3 and L-4 dorsal roots, which are located medial to the L-2 root, are identified; each of the roots consists of two and three naturally separated rootlets. The L-5 and S-1 roots are medial to the L-4 root and largest of all the lumbosacral roots. The L-5 and S-1 dorsal roots consist of three or four rootlets with natural separation. The S-2 root has a single fascicle. Second, an innervation pattern of each root is examined with EMG testing. An individual dorsal root is placed over two hooks of the Peacock rhizotomy probes (Aesculap Instrument Co., Burlingame, California) (**Fig. 27–4A**), and responses to electrical stimulation with a threshold voltage are recorded from the lower extremity muscles. The entire dorsal root is tested at each level immediately before subdividing the dorsal root into rootlets.

EMG Examination and Sectioning of Dorsal Roots

After the innervation of a dorsal root is determined, the root is sharply subdivided into three to five smaller rootlet fascicles of equal size with a Scheer needle (Storz Instruments, St. Louis, MO) (**Fig. 27–4B**). The rootlet fascicles are suspended over two hooks of the rhizotomy probes (**Fig. 27–4C**). Single constant square wave pulses of 0.1 milliseconds duration are applied to the rootlet at a rate of 0.5 Hz. The stimulus intensity is increased stepwise until a reflex response appears from the ipsilateral muscles. After the reflex threshold is determined, a 50 Hz train of tetanic stimulation is applied to the rootlet for 1 second. The reflex response is then graded according to the criteria detailed in **Table 27–2**. Our experience has been that most rootlets produce 1+ to 4+ responses. Thus we base our decision to section a given rootlet on the number of rootlets producing sustained responses at that level and the intensity of the responses. The rootlets that produce a response of 0 are left intact. The rootlets producing 3+ and 4+ responses are cut, and those producing 1+ and 2+ responses are sometimes spared. The dorsal rootlets spared from sectioning are placed behind the Silastic sheet and kept separated from rootlets yet to be tested (**Fig. 27–4D**). If only 1+ and 2+ responses are detected, then rootlets with the most active responses are cut (**Fig. 27–4E**). At least one rootlet is left irrespective of EMG responses to avoid postoperative sensory loss. The procedure is performed in sequence on the remaining L3-S2 dorsal roots (**Fig. 27–4F**).

Using the criteria given in **Table 27–2**, we section 60 to 65% of the rootlets examined. Finally, the L-1 dorsal root is identified at the neural foramen, and half of the dorsal root is cut without EMG testing. In our experience EMG testing of the L-1 root is unreliable. The sectioning of the L-1 dorsal root is necessary to further reduce spasticity in hip flexors, especially in patients with a large L-1 root associated with prefixed lumbosacral plexus. The rhizotomy is repeated on the contralateral side.

The intradural space is irrigated with saline solution. Bipolar cautery is seldom required for control of bleeding from the cut ends of fascicles. The dura is closed in a running fashion with 4–0 monofilament nylon. Clonidine (2 µg/kg up to 7 years of age and 1 µg/kg over 8 years of age) mixed with morphine at 15 µg/kg of body weight is injected intra-

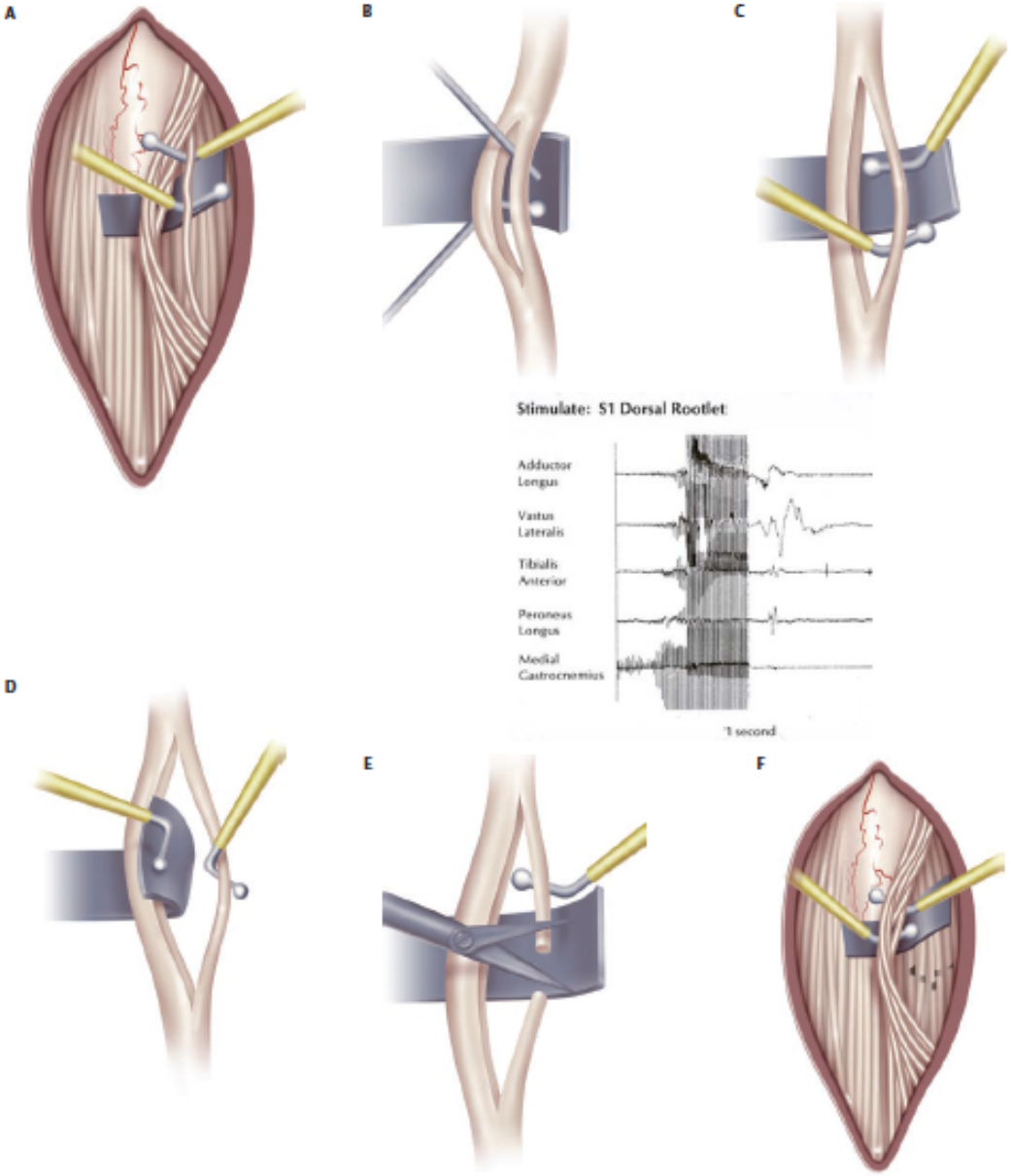

Figure 27–4 **(A)** The L-2 dorsal root is easily identified. In an attempt to identify the L3-S2 dorsal roots, all the dorsal roots are spread over the Silastic sheet and grouped into presumed individual dorsal roots. Then the innervation pattern of each dorsal root is examined with electromyographic (EMG) responses to electrical stimulation with a threshold voltage. **(B)** With a Scheer needle, each dorsal root is subdivided into three to five rootlet fascicles, which are subjected to EMG testing. **(C)** Stimulation of an L-2 rootlet fascicle elicits an unsustained discharge to a train of tetanic stimuli. **(D)** The rootlet is thus spared from sectioning and placed behind the Silastic sheet. **(E)** Stimulation of a rootlet fascicle elicits sustained discharges from multiple muscles. The rootlet is thus sectioned. **(F)** The rootlets spared from sectioning are under the Silastic sheet, and the roots to be tested are on top of the Silastic sheet. Note that EMG testing and sectioning of the dorsal roots are performed caudal to the conus.

Table 27–2 Criteria for Grading Electromyographic Responses in Selective Dorsal Rhizotomy for Spastic Cerebral Palsy

Grade	Electromyographic Response
0	Unsustained or single discharge to a train of stimuli
1+	Sustained discharges from muscles innervated through the segment stimulated in the ipsilateral lower extremity
2+	Sustained discharges from muscles innervated through the segment stimulated and immediately adjacent segments
3+	Sustained discharges from segmentally innervated muscles and muscles innervated through segments distant to the segment stimulated
4+	Sustained discharges from contralateral muscles with or without sustained discharges from the ipsilateral muscles

durally. The Trendelenburg's position is reversed. A strip of Gelfoam is left over the laminectomy defect, and the wound is closed in layers.

Postoperative Management Including Possible Complications

Postoperative Care

Patients stay overnight in the intensive care unit where they receive an intravenous infusion of fentanyl, at a dose of 1 to 3 µg/hour per kg of body weight, and diazepam, as needed. Patients are transferred to the ward the next day, and the fentanyl drip is continued for another 24 to 48 hours. On the third postoperative day, patients are allowed to sit, and physical therapy is started. The patients are discharged to home on the fifth postoperative day and receive outpatient physical therapy from local therapists.

Postoperative Course and Complications

With spastic diplegia the operation invariably reduces spasticity, whereas with spastic quadriplegia there is a small chance of recurrent spasticity. Most patients who were independent walkers preoperatively resume independent walking within 2 weeks after undergoing the SDR technique previously described. Patients who walked with assistance preoperatively take a slower postoperative course. Within 2 months, however, all patients show motor performance exceeding preoperative levels.

The major complications of SDR include paraplegia, sensory loss, bladder and bowel incontinence, CSF leak, and infection. There has been one case of CSF leak requiring operative repair in more than 1500 children and adults who have undergone single-laminectomy SDR at our institution, clear evidence of the safety of the procedures. Also, no patients had late spine deformities that required medical or surgical intervention. Many patients, however, did experience hyperesthesia in the legs for several months.