Dorsal Root Rhizotomy for the Treatment of Spasticity

26

Michael Vassilyadi

Abstract

Dorsal root rhizotomy (*DRR*) surgery is an effective, ablative, long-term treatment in young children with spastic diplegia and has maximal effectiveness when combined with intensive physical and occupational therapies. This neurosurgical operation should be considered when evaluating prospective patients in a multidisciplinary spasticity clinic. DRR provides benefit to carefully selected children with spasticity as a result of cerebral palsy where spasticity is the main factor compromising gait and motor function. The risk of surgical complications is low in experienced centres. Selective dorsal root rhizotomy (*SDDR*), which includes intraoperative electrophysiology and physical muscle monitoring, is strongly recommended as an evidence-based procedure for reducing spasticity and improving gait kinematics. There is also evidence in the literature that SDDR improves gross motor function. Overall, the levels of satisfaction in adults who have undergone SDDR as children have been reported to be generally high, with no negative influence on life satisfaction. In most patients, the benefits remain throughout adolescences and adulthood.

26.1 Introduction

Dorsal root rhizotomy (*DRR*) is an evidence-based neurosurgical procedure that has been proven to offer a reduction in spasticity and an improvement in movement and posture in select patients afflicted with spastic diplegia, and less frequently in those with spastic quadriparesis, in both the short and long term [1–3]. As the proper selection of children is paramount to the success of this irreversible, ablative surgery, it is crucial to have an experienced *multidisciplinary* spasticity team composed of a paediatric neurosurgeon, paediatric neurologist and/or paediatric physiatrist, paediatric orthopaedic surgeon, physiotherapist and occupational therapist, in addition to a nurse to coordinate the clinic and a social worker.

Children who benefit from DRR have a history of prematurity, low birth weight, pre- or

M. Vassilyadi
Division of Neurosurgery, Children's Hospital of Eastern Ontario (CHEO), University of Ottawa, Ottawa, ON, Canada
e-mail: Vassilyadi@cheo.on.ca

C.P. Panteliadis (ed.), *Cerebral Palsy*, https://doi.org/10.1007/978-3-319-67858-0_26

perinatal difficulties (such as prolonged intubation, low APGARS, intraventricular haemorrhage and seizures; see Chap. 6) and delayed motor milestones in the face of relative sparing of speech and intellect and are usually 3–8 years of age with no evidence of an evolving neurological condition. On physical examination there is velocity-dependent increase in tone, increased deep tendon reflexes and clonus with Babinski sign and scissoring gait with exaggerated lordosis; there should be adequate underlying strength, good protective responses, good balance and absence of multiple orthopaedic procedures, fixed severe contractures or dystonic features.

Some centres have widened the age criteria for surgery to include children between 2 and 14 years of age [4]. MacWilliams et al. [5] reported significant functional declines in children with spastic diplegia who underwent *SDDR* (selective dorsal root rhizotomy) after the age of 10 years and that these declines were worse than in children who did not undergo the surgery.

26.2 Patient Selection

In spastic diplegia, spasticity interferes primarily with the function of the lower extremities. Potential candidates for DRR surgery should have minimal evidence of dyskinesia and should be able to walk with or without assistive devices. Head and trunk control should be adequate for sitting upright without support and right in response to lateral challenges. There should be control of quadriceps and hip extensor muscles while rising to stand and returning to sitting, without the reliance on upper extremities for weight bearing. *DRR* is not recommended in children with severe weakness in hip adductor or calf muscles.

Children with spastic quadriparesis (bilateral) can also be candidates for DRR. These children have both upper and lower extremity spasticity that interferes with passive movement, positioning and care. As in children with spastic diplegia, there should be minimal evidence of dyskinesia, no severe truncal hypotonia and a lack of severe fixed contractures at multiple joints requiring orthopaedic surgery. Consideration is given to children who can stand for transfers, but not if lower extremity spasticity aids in performance of the standing transfers.

In a systematic review of the literature, Grunt et al. [6] identified that selection criteria varied considerably with no consensus on the selection process, and most were not based on standardised measurements such as the International Classification of Functioning, Disability and Health. With well-selected patients there would be less heterogeneity with respect to the functional benefits of SDDR.

A common classification used for preoperative ambulatory abilities in children being evaluated for DRR surgery is the five-level NYUMC (New York University Medical Center) system [7] where the best surgical outcome is found in Groups I, II and III [8]. Children that are independent ambulators (*Group* I) have the best chance of improving the appearance and efficiency of their walking. *Group* II consists of children that walk with assistive mobility devices (such as canes, crutches and walkers) and are anticipated to improve the quality of locomotion with less assistance. *Group* III is children that are quadruped crawlers, and the expectation is that they improve at least to the level of using braces or assistive devices. *Groups* IV and V are non-ambulatory patients that are less likely to improve with *DRR* surgery and may be offered alternate therapies such as intrathecal baclofen (see Chap. 25); however, there are recent reports of improvements in these groups with DRR surgery [9, 10]. The Gross Motor Function Classification System (*GMFCS*) is also a five-level evidence-based system for objective classification of motor disability that is used in patients with cerebral palsy; for more see Chap. 22.

Dudley et al. [1] identified a long-term predictive index for ambulation improvement based on four components: preoperative GMFCS assignment, preoperative Gross Motor Function Measure (*GMFM*), distribution of spasticity and the Ashworth scale. The best candidates for surgery had spastic diplegia with GMFM scores >60 and hip adductor tone <3 corresponding to GMFCS groups I, II and III (see Chaps. 17 and 22).

Grunt et al. [11] reported the potential predictive value of brain *MRI* with respect to improvement after SDRR surgery. The improvements in gross motor functioning were best in children with normal imaging and not significantly different in patients with a history of hydrocephalus. The degree of improvement did not correlate with the severity of the periventricular leukomalacia in children with spastic diplegia.

26.3 Surgical Treatment

DRR surgery is usually performed selectively and termed selective dorsal root rhizotomy (*SDRR*). Surgery involves performing a laminotomy, usually between L2 and L5 followed by partial sectioning an average of 15–70% of the dorsal (*sensory*) nerve rootlets from L2 to S2 at the level of the root exit foramina, with most centres cutting more than 40% of the rootlets and limiting the sectioning of the S2 roots [12]. Surgical approaches have also included performing limited laminectomies at either the level of the conus medullaris [13] or more caudal at L5/S1 [14] and various forms of laminoplasty [15]. An absolute requirement for SDDR surgery is intraoperative monitoring with at least an eight-channel muscle response monitor [16]. Intraoperative stimulation of nerve roots and rootlets will differentiate less abnormal from more abnormal responses recorded from the biceps, quadriceps, hamstrings and gastrocnemius muscles on each side. Electrophysiologic abnormalities in dorsal nerve rootlets include:

- Spread to ipsilateral but abnormal myotomes
- Spread to contralateral myotomes
- Sustained, persistent firing throughout the stimulus duration
- Firing after stimulation cessation
- Crescendo/decrescendo responses [17]

During the SDRR surgery, a physiotherapist is present in the operating room to accurately document the lower extremity muscle contractions by palpation. The involved paediatric neurologist or paediatric physiatrist is also present, with the neurophysiology technician, and participates in the decision-making prior to definitive nerve rootlet sectioning. Operating time is usually 6–8 h.

Centres that perform partial, nonselective DRR surgery report similar results to SDRR without the added intraoperative time and cost of all the additional personnel. The scientific validity of neurophysiology monitoring and its role in nerve rootlet sectioning has been questioned [18, 19]. Many of the original criteria have been revised several times and limited to a smaller number to assess if fewer rootlets can be sectioned without changing outcome. Steinbok et al. [20] found that partial spasticity relief may be adequate to achieve a good functional outcome.

Contralateral and suprasegmental spread (*upper extremities*, *neck* and *face*) along with sustained responses with incremental patterns are unique in children with spasticity [21]; all of the other electrophysiology patterns have been identified in children without spasticity, with even contralateral spread being questioned as an absolute criteria for nerve rootlet sectioning [22].

Variability exists in the way SDRR surgery is performed, and electrophysiology responses obtained may differ substantially with only a slight alteration in technique [12]. Examples include the type of anaesthesia; the dissection of the dorsal root into rootlets; the type and placement of the electrodes, e.g. their distance from the cerebrospinal fluid, from the ventral root, from the root exit foramen and the interelectrode distance; the tension applied; the placement of the cathode/node; the type of stimulator; the definition and determination of threshold; and the tetanic stimulation parameters (how much above the threshold intensity, frequency, duration). Important is the number of muscles used for recording and type of electrodes, the type of recording and interpretation of responses, the correlation with palpable muscular responses and the decision of which rootlets to cut and which to spare [19]. In addition, other factors that may influence outcome include the disease process itself, which is primarily in the brain, corticospinal tracts and disorganised spinal interneuron pools; DRR surgery attempts to reduce the disinhibition by sectioning dorsal nerve rootlets that are quite peripheral from the regions of pathology.

26.4 Effectiveness

DRR provides benefit to carefully selected children with spasticity as a result of cerebral palsy, as spasticity is the main factor compromising gait and motor function in these children. The report by Staudt et al. [16] demonstrates that the surgery is responsible for the improvements seen rather than the maturation of the child or physiotherapy alone. Intensive physiotherapy alone does not improve long-term motor outcome [23]. However, post-operative physiotherapy and occupational therapy are definitely required, usually after a 5-day convalescent period, of which there are three components: (a) muscle stretching to gain mobility and range of motion, (b) muscle strengthening to increase endurance and (c) re-education to impart a better pattern of muscle use. This was also shown by Engsberg et al. [24] who found that the benefits of intensive physiotherapy (gains in strength, gait speed and overall gross motor function) are compounded with the addition of SDRR surgery.

Results from many centres have shown spasticity to be reduced after surgery, with loss of opposition between agonistic and antagonistic muscle groups, allowing for greater range of motion. Deep tendon reflexes decrease significantly or disappear; the Babinski response may also disappear. Tone tends to normalise. Some patients may become hypotonic; muscle re-education and strengthening is particularly important in these cases. Gait velocity is improved with an increased stride length [25]. Assistive devices are reduced and orthotic needs may change. Sitting posture improves, there is no scissoring and movements are more isolated with less energy expenditure. Other improvements include speech, personality, seizure control, upper extremity function and bladder control [26]. Craft et al. [27] indicated that the improvement in cognitive performance may not only be due to improved mood and reduced physical discomfort but also possibly secondary to "*supra-segmental effects*" induced by the DRR surgery. Bloom and Nazar [28] showed improvements in self-care, mobility and social functioning with less caregiver assistance. Mittal et al. [29] identified improvements in activities of daily living using a validated evaluation measure with the functional improvements persisting 3 and 5 years after surgery. In a separate publication, Mittal et al. [30] also showed a sustained upper extremity functional improvement in children in NYUMC Groups I, II and III. Assessments during adolescence and early adulthood have shown lasting benefits [1, 31].

26.5 Complications and Side-Effects

Complications of DRR surgery are related to the medical status of the patients, the surgical exposure and the potential neurological consequences of cauda equina manipulation and sectioning. In 158 children who underwent SDDR, Steinbok and Schrag [32] identified intraoperative, immediately post-operative and postdischarge complications that occurred in 3.8%, 43.6% and 30% of the patients, respectively. The most common intraoperative complication was aspiration pneumonia that occurred in 2 patients (1.3%). Perioperative complications included items such as emesis (59%), constipation (37%), skin rash (10%), dysesthesia (7.6%), headache (2.5%), urinary retention (4.4%), dysuria (1.9%), wound infection (0.6%) and *CSF* leak in one patient (0.6%). *Complications* noted after discharge included back pain (delayed onset in 10.8% and severe in 2.7%), neurogenic bladder/bowel (12.7%; persisted in 5.1%), paraesthesia (6.3%), persistent sensory changes (3.8%), increased seizures (2.5%), increased constipation (1.9%) and root compression in one patient (0.6%).

Golan et al. [33] reviewed the risks of post-operative spinal deformities and found that children with more severe cerebral palsy were more likely to develop scoliosis after surgery. The less affected children that were ambulators were at risk of developing spondylolisthesis. In addition, older age at the time of surgery and female gender were associated with greater post-operative lumbar lordosis (see Chap. 34). Chicoine et al. [34] found that the strongest predictor of improved ability to walk after SDRR was the

preoperative gait score obtained quantitatively by videotaped gait analysis. O'Brien et al. [4] found that children in the 2–5 years age range improved their gait more than children operated between 6 and 14 years of age, with less requirement for future orthopaedic surgery in the younger group (34% versus 70%). In a systematic review, Grunt et al. [2] found that there is lack of evidence that long-term spine abnormalities after SDDR surgery can be attributed to the surgery itself.

In a *recent* systematic review of systematic reviews, SDDR was strongly recommended for reducing spasticity (moderate quality of evidence in the literature) and improving gait kinematics, e.g. low quality of evidence. There is evidence for SDDR to improve gross motor function, but weakly recommended for improving function and participation, such very low quality of evidence [35]. Overall, the levels of satisfaction in adults who have undergone SDDR as children have been reported to be generally high, with no negative influence on life satisfaction [31]. In Chap. 25 "Intrathecal baclofen therapy for the control of spasticity" will discuss the treatment of spasticity using intrathecal baclofen and will compare the two treatment options of DRR surgery and implantation of a programmable baclofen pump in children with cerebral palsy.

References

1. Dudley RW, Parolin M, Gagnon B, et al. Long-term functional benefits of selective dorsal rhizotomy for spastic cerebral palsy. J Neurosurg Pediatr. 2013;12:142–50.
2. Grunt S, Becher JG, Vermeulen RJ. Long-term outcome and adverse effects of selective dorsal rhizotomy in children with cerebral palsy: a systematic review. Dev Med Child Neurol. 2011;53:490–8.
3. McLaughlin J, Bjornson K, Temkin N, et al. Selective dorsal rhizotomy: meta-analysis of three randomized controlled trials. Dev Med Child Neurol. 2002;44:17–25.
4. O'Brien DF, Park TS, Puglisi JA, et al. Effect of selective dorsal rhizotomy on need for orthopedic surgery for spastic quadriplegic cerebral palsy: long-term outcome analysis in relation to age. J Neurosurg Pediatr. 2004;101:59–63.
5. MacWilliams BA, Johnson BA, Shuckra AL, D'Astous JL. Functional decline in children undergoing selective dorsal rhizotomy after age 10. Dev Med Child Neurol. 2011;53:717–23.
6. Grunt S, Fieggen AG, Vermeulen RJ, et al. Selection criteria for selective dorsal rhizotomy in children with spastic cerebral palsy: a systematic review of the literature. Dev Med Child Neurol. 2014;56:302–12.
7. Abbott R, Johann-Murphy M, Shiminski-Maher T, et al. Selective dorsal rhizotomy: outcome and complications in treating spastic cerebral palsy. Neurosurgery. 1993;33:851–7.
8. Ailon T, Beauchamp R, Miller S, et al. Long-term outcome after selective dorsal rhizotomy in children with spastic cerebral palsy. Childs Nerv Syst. 2015;31:415–23.
9. Ingale H, Ughratdar I, Muquit S, et al. Selective dorsal rhizotomy as an alternative to intrathecal baclofen pump replacement in GMFCS grades 4 and 5 children. Childs Nerv Syst. 2016;32:321–5.
10. Kan P, Gooch J, Amini A, et al. Surgical treatment of spasticity in children: comparison of selective dorsal rhizotomy and intrathecal baclofen pump implantation. Childs Nerv Syst. 2008;24:239–43.
11. Grunt S, Becher JG, van Schie P, et al. Preoperative MRI findings and functional outcome after selective dorsal rhizotomy in children with bilateral spacticity. Childs Nerv Syst. 2010;26:191–8.
12. Steinbok P, Kestle JR. Variation between centres in electrophysiologic techniques used in lumbosacral selective dorsal rhizotomy for spastic cerebral palsy. Pediatr Neurosurg. 1996;25:233–9.
13. Park TS, Gaffney SE, Kaufman BA, Molleston MC. Selective lumbosacral rhizotomy immediately caudal to conus medullaris for cerebral palsy. Neurosurgery. 1993;33:129–34.
14. Lazareff JA, Mata-Acosta AM, Garcia-Mendez MA. Limited selective posterior rhizotomy for the treatment of spasticity secondary to infantile cerebral palsy: a preliminary report. Neurosurgery. 1990;27:535–8.
15. Funk JF, Haberl H. Monosegmental laminoplasty for selective dorsal rhizotomy-operative technique and influence on the development of scoliosis in ambulatory children with cerebral palsy. Childs Nerv Syst. 2016;32:819–25.
16. Staudt LA, Nuwer MR, Peacock WJ. Intraoperative monitoring during selective posterior rhizotomy: technique and patient outcome. Electroencephalogr Clin Neurophysiol. 1995;97:296–309.
17. Logigian EL, Shefner JM, Goumnerova L, et al. The critical importance of stimulus intensity in intraoperative monitoring for partial dorsal rhizotomy. Muscle Nerve. 1996;19:415–22.
18. Cohen AR, Webster HC. How selective is selective posterior rhizotomy? Surg Neurol. 1991;35:267–72.
19. Vassilyadi M, Ventureyra ECG. Treatment options for spasticity in children. Crit Rev Neurosurg. 1998;8:193–200.
20. Steinbok P, Gustavsson B, Kestle JR, et al. Relationship of intraoperative electrophysiological criteria to outcome after selective functional posterior rhizotomy. J Neurosurg. 1995;83:18–26.

21. Steinbok P, Keyes R, Langill L, Cochrane DD. The validity of electrophysiology criteria used in selective functional posterior rhizotomy for treatment of spastic cerebral palsy. J Neurosurg. 1994;81:354–61.
22. Warf BC, Nelson KR. The electromyographic responses to dorsal rootlet stimulation during partial dorsal rhizotomy are inconsistent. Pediatr Neurosurg. 1996;25:13–9.
23. Steinbok P, McLeod K. Comparison of motor outcomes after selective dorsal rhizotomy with and without preoperative intensified physiotherapy in children with spastic diplegic cerebral palsy. Pediatr Neurosurg. 2002;36:142–7.
24. Engsberg JR, Ross SA, Collins DR, Park TS. Effect of selective dorsal rhizotomy in the treatment of children with cerebral palsy. J Neurosurg Pediatr. 2006;105:8–15.
25. Peacock WJ, Staudt LA (1991) Functional outcomes following selective posterior rhizotomy in children with cerebral palsy. J Neurosurg 74:380–5.
26. Gigante P, McDowell MM, Bruce SS, et al. Reduction in upper-extremity tone after lumbar selective dorsal rhizotomy in children with spastic cerebral palsy. J Neurosurg Pediatr. 2013;12:588–94.
27. Craft S, Park TS, White DA, Schatz J, et al. Changes in cognitive performance in children with spastic diplegic cerebral palsy following selective dorsal rhizotomy. Pediatr Neurosurg. 1995;23:68–75.
28. Bloom KK, Nazar GB. Functional assessment following selective posterior rhizotomy in spastic cerebral palsy. Childs Nerv Syst. 1994;10:84–6.
29. Mittal S, Farmer JP, Al-Atassi B, et al. Functional performance following selective posterior rhizotomy: long-term results determined using a validated evaluation measure. J Neurosurg. 2002;97:510–8.
30. Mittal S, Farmer JP, Al-Atassi B, et al. Impact of selective posterior rhizotomy on fine motor skills: long-term results using a validated evaluative measure. Pediatr Neurosurg. 2002;36:133–41.
31. Hurvitz EA, Marciniak CM, Daunter AK, et al. Functional outcomes of childhood dorsal rhizotomy in adults and adolescents with cerebral palsy. J Neurosurg Pediatr. 2013;11:380–8.
32. Steinbok P, Schrag C. Complications after selective posterior rhizotomy for spasticity in children with cerebral palsy. Pediatr Neurosurg. 1998;28:300–13.
33. Golan JD, Hall JA, O'Gorman G, et al. Spinal deformities following selective dorsal rhizotomy. J Neurosurg Pediatr. 2007;106:441–9.
34. Chicoine MR, Park TS, Vogler GP, Kaufman BA. Predictors of ability to walk after selective dorsal rhizotomy in children with cerebral palsy. Neurosurgery. 1996;38:711–4.
35. Novak I, McIntyre S, Morgan C, et al. A systematic review of interventions for children with cerebral palsy: state of the evidence. Dev Med Child Neurol. 2013;55:886–910.