

LONG-TERM FUNCTIONAL OUTCOMES IN PATIENTS WITH CEREBRAL PALSY POST SELECTIVE DORSAL RHIZOTOMY: A SYSTEMATIC REVIEW

Janet P. Slaughter[1] and Corinne A. Sampson[2]

University of Mississippi Medical Center, School of Health Related Professions, Department of Physical Therapy[1], and Children's Rehab Services[2], Jackson, MS 39216

Corresponding Author: Janet Slaughter<jslaughter@umc.edu>

ABSTRACT

Background and Significance: Selective dorsal rhizotomy (SDR) is a surgical procedure that involves the selective cutting of the dorsal lumbosacral spinal nerve rootlets to reduce spasticity. SDR is widely used to treat spasticity in children with cerebral palsy (CP). The purpose of this systematic review was to evaluate the long-term effects of SDR on functional outcomes in children with CP.
Methods: PubMed and Embase databases were searched on November 12, 2018, using terms associated with rhizotomy, cerebral palsy, and activities of daily living. Inclusion criteria consisted of all study designs containing participants diagnosed with CP who underwent SDR at the lumbar or sacral levels on or before the age of 16 and the use of the Pediatric Evaluation of Disability Inventory (PEDI) as a functional outcome measure at least 12 months postoperatively. Risk of bias was assessed with the Joanna Briggs Institute checklist. After performing the electronic search, six articles remained for review.
Results: Whole group scores after SDR in the self-care and mobility domains of the PEDI showed statistically significant improvement ($p<0.05$) through 5 years postoperatively, compared to baseline values. PEDI scores were maintained in the less severe forms of CP through 10 years.
Conclusion: If SDR is performed on well-selected children before severe orthopedic deformities have developed, it has the potential to increase functional independence particularly in the less severe cases through 10 years postoperatively and protect against the functional decline in adolescence typically seen in the more severe cases.

Keywords: Rhizotomy, Cerebral Palsy, Activities of Daily Living

INTRODUCTION

"Cerebral palsy (CP) describes a group of permanent disorders of the development of movement and posture, causing activity limitation that is attributed to non-progressive disturbances that occurred in the developing fetal or infant brain. The motor disorders of cerebral palsy are often accompanied by disturbances of sensation, perception, cognition, communication, and behavior, by epilepsy, and by secondary musculoskeletal problems."[1]. There are several types of tonal abnormalities associated with CP based on whether the damage is pyramidal or extrapyramidal. In this intervention investigation, we are concerned with the tonal presentation from a pyramidal tract injury known as spasticity. Spasticity is defined as a velocity-dependent resistance of a muscle to stretch [2].

Baseline functional levels were established using the Gross Motor Function Classification System (GMFCS), an internationally accepted functional classification system used for individuals with cerebral palsy to classify self-initiated movement based on usual performance at home or in the community. There are five levels in the GMFCS. Abbreviated descriptions of the GMFCS Levels are as follows: Level I-walks without limitations; Level II-walks with limitations, Level III-walks using a hand-held mobility device; Level IV-self-mobility with limitations, may use powered mobility; Level V-transported in a manual wheelchair [3].

Functional status of the participants was assessed using the Pediatric Evaluation of Disability Inventory (PEDI), a comprehensive standardized clinical assessment that assesses functional activities in children with disabilities [4]. The Gross Motor Function Measure (GMFM) was also used. It is a validated criterion-referenced assessment tool designed specifically for children with cerebral palsy to show change over time in gross motor function. It is available in two versions, the GMFM-88, and the GMFM-66. The GMFM-88 is typically used for very young or more severely involved children (GMFCS Level V). It should be used if the child needs assistive devices or orthotics to walk. The GMFM-66 has fewer items, does not allow

the use of orthotics, and is frequently used in research [5]. Gross motor development curves for children with CP have been developed using the GMFM-66 [6]. From these curves, it is known that motor function is stable in GMFCS Levels I and II with no peaks or declines. However, in GMFCS Levels III-V, ninety percent of gross motor development is usually achieved by five years of age and further development peaks at about eight years of age with clinically significant declines seen as they move from adolescence into young adulthood [7].

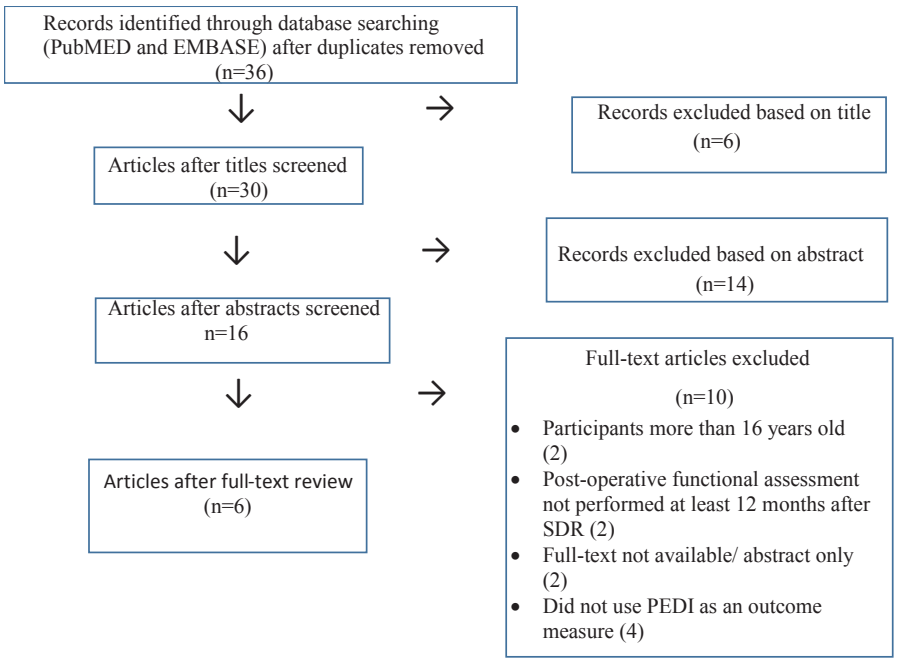
A selective dorsal rhizotomy (SDR) is an individualized and permanent surgical procedure involving the selective cutting of lumbosacral nerve rootlets. It is used as a treatment option to reduce alpha motor neuron excitation. Children with the spastic form of CP commonly undergo selective dorsal rhizotomy procedures in an effort to decrease spasticity that interferes with motor functioning [8]. Intensive physical therapy is required in the postoperative period in an effort to translate spasticity reduction into improved functional skills. Although a selective dorsal rhizotomy procedure is known to reduce spasticity in children with CP, limited studies have focused on its effects regarding long-term functional outcomes.

METHODS

An electronic search was conducted November 12, 2018, using PubMed and Embase, two online databases. In both databases, the following identical search was used ((rhizotomy) AND (cerebral palsy) AND (activities of daily living)). There were no limits placed on the database search. Eligibility criteria consisted of participants that were diagnosed with some form of cerebral palsy. The participants must have received a selective dorsal rhizotomy of lumbar or sacral origin at the age of sixteen years or younger and functional outcome measures must have been performed on all patients at least twelve months following selective dorsal rhizotomy. Among the functional outcome measures performed, all participants must have been assessed using the Pediatric Evaluation of Disability Inventory (PEDI). All study designs were accepted for this systematic review**.** Articles were excluded if they were not available in full text. Thirty-six articles were initially identified from the search. Six articles were removed after the title screen and fourteen articles were excluded based on the abstract screen. After the full texted review, six articles remained for review (Figure 1)

For quality assessment, the literature included in this systematic review was assessed using the Joanna Briggs Institute Checklist for Case Series, a 5-point scale in which a lower score indicates a higher level of evidence. This systematic review was considered a Level 4c.

Figure 1: PRISMA Diagram



RESULTS

A 2008 study by Chan, et al. [9] included 21 subjects with a mean age at the time of SDR of 8.6 years of age and a range of 6.0 to 11 years. Exclusion criteria included significant mental retardation, significant joint contractures, severe dystonia/dyskinesia, and significant fixed orthopedic deformity. Functional levels at the time of SDR included the following GMFCS levels: Level 1 (n=8), Level II (n=4), Level III (n=7) and Level V (n=2). Twelve patients completed the post-op follow-up at one year. Functional outcome measures completed at follow-up included the PEDI, GMFM, and Observational Gait Analysis. A 3D Gait Analysis was also performed on GMFCS Level I subjects. Significance was set at $p<0.05$ for all outcome measures. Results on the PEDI according to GMFCS levels were as follows: GMFCS I-IV showed significant improvements in Functional Skill (FS) mobility ($p=0.001$) and self-care ($p< 0.0001$) dimension scores, and Caregiver Assistance (CA) mobility ($p= 0.001$) and self-care ($p< 0.0001$) dimension scores; GMFCS I displayed significant improvements in FS mobility ($p=0.032$) and self-care ($p=0.023$) dimension scores, and CA self-care ($p=0.012$) dimension scores; GMFCS II-III exhibited significant improvements in FS mobility ($p=0.032$) and self-care ($p=0.001$), and CA mobility ($p=0.011$) and self-care ($p=0.002$) dimension scores. Results on the GMFM according to GMFCS Levels were as follows: GMFCS I-IV revealed significant improvements in total score ($p=0.001$) in the dimensions of sitting ($p=0.032$), crawling/kneeling ($P<0.0001$), walking/running/jumping ($p=0.027$); GMFCS I showed positive trends but no significant difference in total score ($p=0.059$) in the dimensions of crawling/kneeling ($P=0.058$), standing ($P=0.317$), walking/running ($p=0.199$); GMFCS II-III revealed significant improvements in total score ($p=0.021$) in the dimensions of crawling/kneeling ($p=0.001$). Scores on the Observational Gait Analysis indicated significant improvements ($p<0.0001$) in GMFCS I-IV. The 3D Gait Analysis scores in GMFCS Level I participants revealed significant improvements in knee extension in terminal swing ($p=0.029$) and ankle dorsiflexion in midstance ($p=0.021$).

A study by Dudley et al. in 2013 [10], consisted of 105 subjects whose mean age at the time of SDR was 5.0 years with a range of 3.0 to 10.5 years. Exclusion criteria included children who exhibited low tone, dystonia, or multiple prior orthopedic procedures. Functional levels at the time of SDR according to GMFCS Levels were: Level I (n=11); Level II (n=22); Level III (n=14); Level IV (n=5); 53 subjects were not classified due to the fact that the GMFCS classification was not available at the beginning of the study. Post-op follow-ups were performed at 1 (n=97), 5 (n=62), 10 (n=57), and 15 years (n=14). Functional outcome measures that were completed at follow-up included the PEDI and the GMFM-88 with significance set at $p<0.05$ for both measures. Results from the PEDI revealed that scaled scores in mobility and self-care for GMFCS Levels I-IV showed significant improvements from baseline to 1, 5, and 10 years. The most significant increases occurred in the first 5 years. The authors were unable to stratify by GMFCS Levels at 15 years due to insufficient numbers. Results of the GMFM-88 revealed that GMFCS Levels I-IV had significant increases in total scores in the dimensions of standing as well as walking/running/ jumping at 1, 5, 10, and 15 years. Again, the authors were unable to stratify by GMFCS levels at 15 years due to insufficient numbers. GMFCS Levels I- III demonstrated significant increases ($p< 0.001$) at 1, 5, and 10-year follow-ups compared with preoperative values. GMFCS Level IV did not show significant improvements from preoperative values at any follow-up.

A recent study by Josenby et al. in 2014 [11], included 24 subjects whose mean age at the time of SDR was 4.1 years with a range of 2.5 to 6.4 years. Exclusion criteria included little or no rigidity, dystonia, hypotonia, athetosis, ataxia, and no significant weakness in antigravity muscle groups of the trunk and lower extremities. Functional levels at the time of SDR according to the GMFCS classification were as follows: Level 1 (n=1), Level II (n=7), Level III (n=4), Level IV (n=11), Level V (n=1). Post-op follow-ups were completed at 1 year, 1.5 years, 3 years, 5 years, and 10 years. The PEDI was used to evaluate functional status post-operatively. Significance was set at $p<0.05$. GMFCS Levels I-V exhibited significant increases in FS and CA for self-care and mobility ($p\leq0.007$) dimension scores through five years. GMFCS Levels I-III displayed small, non-significant improvements between 5 and 10 years. GMFCS Levels IV-V showed no significant improvement; however, levels of function at five years were maintained to ten years.

An earlier study conducted in 2002 by Mittal et al. [12], consisted of 41 subjects with a mean age at the time of SDR of 4.8 years with a range of 3.0 years to 7.5 years. Exclusion criteria encompassed children who exhibited low tone, dystonia, multiple prior orthopedic procedures, and double hemiplegia. Functional levels at the time of SDR were not classified according to GMFCS levels. The following groups were formed: Group I, walks no AD (n=16); Group II, walks with AD (n=18); Group III, quadruped crawler (n=6), Group IV, belly crawler (n=1), and Group V, no locomotion (n=0). Post-op follow-up was conducted at 1 year and 3 years with all 41 patients and at 5 years with 30 patients. The PEDI was used as the functional outcome measure. Significance was established at $p<0.05$ with the following results: Groups I-IV: significant improvements in raw, normative, and scaled scores ($p<0.001$) in the FS dimension of the self-care and mobility domains from baseline to 1, 3, and 5 years. Between 3 and 5 years, results were not significant ($p=0.223$). In general, Groups I-II exhibited greater benefit in both self-care and mobility scores when compared to Group III.

Thirty-five subjects participated in a study by Nordmark et al. in 2008. [13] The mean age at the time the SDR was performed was 4.5 years with a range of 2.5 to 6.6 years. Exclusion criteria included the presence of dystonia, ataxia, fixed contractures, and earlier orthopedic operations other than an adductor tenotomy. Functional levels at the time of SDR according to GMFCS classification were as follows: Level 1 (n=1); Level II (n=8); Level III (n=10); Level IV (n=15); Level V (n=1). Postop follow-ups were completed at 1, 1.5, 3, and 5 years. Functional outcome measures completed were the PEDI, GMFM-88, and GMFM-66 with significance set at $p<0.05$ for all measures. Results of data obtained from the PEDI demonstrated significant changes ($p < 0.001$) in FS and CA scaled scores for the self-care and mobility domains from baseline to 1, 1.5, 3, and 5 years in the whole group and in all GMFCS Levels except GMFCS Level III. GMFCS Level III showed no statistically significant improvements in FS and CA in the self-care domain. The GMFM-66 revealed significant changes from baseline to 1, 1.5, 3, and 5 years in the whole group, GMFCS Levels I-V. GMFCS Levels I-II did not show significant changes during follow-ups. GMFCS Level III was significant after 3 and 5 years. GMFCS Levels IV-V showed significant changes at 1.5, 3, and 5 years. The GMFM-88 showed significant changes ($p<0.001$) in total and goal total scores for GMFCS Levels I-V at 1, 3, and 5 years. Scores at 1.5 years were not significant.

The final article in this review was conducted by Van Schie et al. in 2005 [14]. There were nine subjects with a mean age at the time of SDR of 5.4 years with a range of 3.6 to 6.8 years. Subjects were excluded from the study if they had contractures that limited hip, knee, or ankle function. GMFCS functioning levels prior to SDR were as follows: Level II (n=1) and Level III (n=8). Postop follow-up was conducted at one year using the PEDI, GMFM-88, and Edinburgh Visual Gait Score (EGS) with significance set at $p<0.05$ for all. The PEDI showed significant improvement ($p=0.012$) for GMFCS Levels II-III in mean scaled scores in the FS and CA dimensions. The GMFM-88 was assessed bi-monthly and revealed significant improvement ($p=0.008$) in GMFCS Levels II-III in mean scores in dimensions of crawling, standing, and walking/running/jumping. The Edinburgh Visual Gait Score revealed significant improvements in mean totals ($p=0.021$) in GMFCS Levels II-III. Improvements were seen in initial contact and heel-lift position of the ankle.

DISCUSSION

In general, the majority of participants' scores after SDR in the self-care and mobility domains of the PEDI showed statistical improvement through five years postoperatively, compared to baseline values.

PEDI scores were maintained in all GMFCS levels through ten years. Changes in GMFCS levels from baseline were sparse, as most subjects maintained their preoperative functional classifications. Functional improvements were most notably observed within the first five years in Levels I, II, and III and no trends toward a functional decline in adolescence were noted. Four of the six studies also used the GMFM-66 and/or GMFM-88 to evaluate the change in gross motor function over time. Increases were noted in GMFM total scores from baseline through ten years for GMFCS Levels I-III and maintained through adolescence for GMFCS Levels IV-V.

CONCLUSIONS

When performed on well-selected children diagnosed with spastic cerebral palsy before the development of severe orthopedic deformities, SDR has the potential to not only increase functional independence in GMFCS Levels I-III through ten years post-operatively but also protect against the functional decline typically seen in GMFCS Levels III-V during adolescence. However, the majority of the studies lacked adequate participation in long-term follow-ups partly due to the fact that not enough children had yet reached the postoperative years. This fact may limit the accuracy of data regarding changes in functional outcomes.

ACKNOWLEDGMENTS

The authors would like to acknowledge Tylyn Brumfield, Collin Anderson and Kollin Cannon for their assistance in gathering the information for this review.

DISCLOSURES

Janet P. Slaughter has no financial disclosures or conflicts of interest to declare. Corinne A. Sampson has no financial disclosures or conflicts of interest to declare.

REFERENCES

[1] P. Rosenbaum, et al. "A report: the definition and classification of cerebral palsy," *Dev Med Child Neurol Suppl,* vol 109, pp. 8-14, 2007.

[2] T.D. Sanger, et al. "Classification and definition of disorders causing hypertonia in childhood," *Pediatrics,* vol. 11, pp. e89-e97, January 2003.

[3] R.J. Palisano, et al. "Content validity of the expanded and revised Gross motor function classification system," *Dev Med Child Neurol,* vol 50, pp. 744-50, March 2008.

[4] S. M. Haley, et al. *Pediatric Evaluation of Disability Inventory.* San Antonio, TX: The Psychological Corporation, 1992.

[5] CanChild Centre for Childhood Disability Research. Retrieved from: https://canchild.ca/en/resources/44-gross-motor-function-measure-gmfm .

[6] P. L. Rosenbaum, S. D. Walter, S. E. Hanna, R. J. Palisano, D. J. Russell, and P. Raina. "Prognosis for gross motor function in cerebral palsy: creation of motor development curves. *JAMA,* vol. 288, pp. 844-852, September 2011.

[7] S. E. Hanna, P. L. Rosenbaum, D. J. Bartlett, R. J. Palisano, S. D. Walter, L. Avery and D. J. Russell, "Stability and decline in gross motor function among children and youth with cerebral palsy aged 2 to 21 years," *Dev Med Child Neurol,* vol. 51, pp. 424-28, June 2006.

[8] P. Steinbok, "Selective dorsal rhizotomy for spastic cerebral palsy: a review," *Childs Nerv Syst,* vol. 23, issue 9, pp. 981-90, September 2007.

[9] S. H. S Chan, K. Y. Yam, B. P. H. Yiu-Lau, C. Y. C. Poon, N. N. C. Chan, H. M. Cheung, M. M. F. Wu, and W. K. Chak. "Selective dorsal rhizotomy in Hong Kong: multidimensional outcome measures," *Pediatr Neurol,* vol. 39, no. 1, pp. 22-32, 2008.

[10] R.W. R. Dudley, M. Parolin, B. Gagnon, R. Saluja, R. Yap, K. Montpetit, J. Ruck, C. Poulin, M-A. Cantin, T. E. Benaroch, and J. Farmer. "Long-term functional benefits of selective dorsal rhizotomy for spastic cerebral palsy," *J Neurosurg Pediatrics,* vol. 12, pp. 142-50, August 2013.

[11] A. L. Josenby, P. Wagner, G-B Jarnlo, L. Westbom, and E. Nordmark. "Functional performance in self-care and mobility after selective dorsal rhizotomy: a 10-year practice-based follow-up study," *Dev Med Child Neurol,* vol 57 no. 3, pp. 286-93, October 2014.

[12] S.Mittal, J. Farmer, B. Al-Atassi, K. Montpetit, N. Gervais, C. Poulin, T. E. Benaroch, and M. Cantin. "Functional performance following selective posterior rhizotomy: long-term results determined using a validated evaluative measure," *J Neurosurg,* vol. 97, pp. 510-18, September 2002.

[13] E. Nordmark, A. L. Josenby, J. Lagergren, G. Anderson, L. Stromblad and L. Westbom. "Long-term outcomes five years after selective dorsal rhizotomy," *Pediatrics,* vol. 8:54, December 2008.

[14] P. E. M. van Schie, R. J. Vermeulen, W. J. R. van Ouwerkerk, G. Kwakkel and J. G. Becher. "Selective dorsal rhizotomy in cerebral palsy to improve functional abilities, evaluation of criteria for selection," *Childs Nerv Syst,* vol. 21, pp. 451-57, February 2005.