E Editor's Choice

Long-term effects of selective dorsal rhizotomy in children with cerebral palsy: a systematic review

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ABBREVIATION

SDR Selective dorsal rhizotomy

AIM To evaluate the long-term effects of selective dorsal rhizotomy (SDR) 10 years or more after the procedure and complications observed any time after SDR in children with cerebral palsy (CP).

METHOD Embase, PubMed, and the Cochrane Library were searched from their individual dates of inception through 1st June 2018 for full-text original articles in English that described long-term follow-up after SDR in children with CP. The authors independently screened publications to determine whether they met inclusion criteria; thereafter all authors extracted data on patient characteristics, the proportion of the original cohort being followed-up, and the reported outcomes.

RESULTS Of the 199 studies identified, 16 were included in this evaluation: 14 were case series and two studies reported a retrospectively assigned comparison group. Evidence concerning function was limited by study design differences, clinical variability, loss to follow-up, and heterogeneity across trials.

INTERPRETATION At 10 years or more follow-up, available studies generate low-level evidence with considerable bias. No functional improvement of SDR over routine therapy is documented. Furthermore, the long-term effects of SDR with respect to spasticity reduction is unclear, with many studies reporting a high amount of add-on spasticity treatment. More long-term follow-up using robust scientific protocols is required before it can be decided whether the use of SDR as routine therapy for children with CP is to be recommended or not.

Spasticity has traditionally been assumed to be a major cause of functional limitation in individuals with cerebral palsy (CP). Consequently, extensive efforts have been made to alleviate spasticity. Selective dorsal rhizotomy (SDR), the most invasive and irreversible of such procedures, has gained in popularity since the 1990s and is now standard treatment in certain centres. The procedure is typically performed in the preschool years and, consequently, the long-term effects lasting into adulthood are of increasing interest.

Although some reports document the effects of SDR more than 10 years postoperatively, the outcomes measured and their interpretation vary. Some conclude that SDR results in an overall positive effect on activity and function, while others have observed little or no positive impact on function, along with a risk of complications and requirement for major rehabilitation.

Because of the changes in descending pathways, afferent sensory excitation is altered in spasticity.¹ Furthermore, in CP, reciprocal inhibition within spinal circuits is reduced, and it has been conclusively shown that spasticity is associated with augmented excitability of the spinal motor neurons.¹ In CP, the supra spinal input is reduced due to the

underlying central nervous system lesion, and accordingly, spasticity can be hypothesized to be a normal and physiological response to maintaining the ability to generate maximal muscle activation.¹ Supporting this argument is the fact that a child with CP is not born with spasticity but experiences a gradual onset and increase of spasticity during the first years of life, possibly as a physiological response to the increased demands of motor development.^{1,2} When the overall modulation of physiological reflexes is, thus, impaired, normal sensory stimuli cause excessive efferent activation of the motor units and muscle tone becomes spastic. In SDR, these excitatory nerve fibres, emerging from the proprioceptors in the muscle spindles, are cut selectively at the point where they enter the posterior or dorsal root of the spinal cord, thereby attenuating afferent excitation. As a result, SDR provides long-term reduction of spasticity.3,4

Most commonly, the nerve roots to be cut are identified by neurophysiological testing. Only rootlets that respond pathologically to stimuli are cut. Today, no standard procedure, single protocol, or common neurophysiological methodology has been adopted to identify pathological rootlets.

554 DOI: 10.1111/dmcn.14320 © 2019 The Authors Developmental Medicine & Child Neurology published by John Wiley & Sons Ltd on behalf of Mac Keith Press This is an open access article under the terms of the Creative Commons Attribution-NonCommercial-NoDerivs License, which permits use and distribution in any medium, provided the original work is properly cited, the use is non-commercial and no modifications or adaptations are made. This systematic review and evaluation of relevant literature focuses on the assessment of individuals with CP 10 years or more after undergoing SDR, and specifically whether any outcomes related to activity, body function, and structure are included in the assessment. In addition, we reviewed complications observed at any time after SDR. We discuss the potential impact of spasticity in this context, as well as the perceptual or sensory alterations that SDR might cause.

METHOD

The Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) checklist and flow diagram was used.⁵ After assigning the Population Intervention Comparison Outcome (PICO) protocol, PubMed, the Cochrane library, and Embase were searched on two occasions by the authors and a medical librarian (CL) with a combination of keywords and MeSH terms for 'SDR', 'CP', 'follow-up', and 'long-term' to identify articles published from inception to 1st June 2018 (Table SI and Fig. S1, online supporting information). After removing duplicates and reviewing titles and abstracts, all three authors read the remaining full-length articles and agreed on those to be included in the review. Studies that did not assess individuals with CP and where not all individuals had a minimum of 10 years' follow-up were excluded, as were studies not evaluating any aspect of function, pain, fatigue, nor spasticity. Only original, full-length articles in English were included. Additional articles of interest were identified manually. The authors assigned a level of evidence to 16 articles fulfilling these inclusion criteria⁶⁻²¹ based on the guidelines of the Oxford Centre for Evidence-Based

Table I: Selective dereal rhizetemy (SDR); reported complications

What this paper adds

- Ten years after selective dorsal rhizotomy, available studies supply inconclusive evidence on functional outcomes.
- The long-term effect on spasticity is uncertain, studies reported a substantial need for add-on treatment.
- Short- and long-term complications seem frequent but are not reported in a consistent manner.

Medicine.²² Risk of bias was assessed at the study level using a modified version of the Quality in Prognosis Studies tool (Table SII, online supporting information).²³ Data were extracted independently and synthesized by all the authors.

The second search designed to obtain complementary reports of overall complications included a combination of keywords and MeSH for SDR and complications (Table SI and Fig. S2, online supporting information).

Finally, a specific search including ('selective dorsal rhizotomy' OR 'SDR') AND ('scoliosis' OR 'spinal' OR 'bladder dysfunction' OR 'sensory change's OR 'dystonia' OR 'balance') was performed. Only original publications in English and involving 10 or more participants were included (Fig. S2). The procedure for reviewing the 24 included reports^{9,14,20,24-44} was similar to that described above, except that in these cases, neither risk of bias nor level of evidence was assessed (Table I).

RESULTS

Included studies for outcome of SDR at 10 years or more

After removing duplicates, 199 articles were screened and 16 included^{6–21} (Table II and Fig. S1); none involved a randomized control trial nor prospective evaluation of a control group. Fourteen were case series and two involved

Table I: Selective dorsal ri	nizotomy (SDR):	reported complications
Type of complication	Incidence, %	Reporting study
Short term (<1y after SD	R)	
Pulmonary	~9	Abbott; ²⁴ Steinbok and Schrag; ³⁵ Nordmark et al. ³⁷
Gastrointestinal	20-80	Steinbok and Schrag; ³⁵ Abbott ²⁴
Urinary tract	7–12	Steinbok and Schrag; ³⁵ Abbott; ²⁴ Nordmark et al. ³⁷
Problems with the surgical wound	1	Abbott; ²⁴ Peter and Arens; ⁴⁴ Steinbok and Schrag ³⁵
Sensory alterations	~50	Abbott; ²⁴ Steinbok and Schrag; ³⁵ Peter and Arens; ⁴⁴ Tedroff et al. ²⁰
Leakage of cerebrospinal fluid	11	Nordmark et al. ³⁷
Late (≥1y after SDR)		
Hyperlordosis	16–40	Golan et al.; ²⁶ Crawford et al.; ²⁵ Bolster et al.; ⁹ Johnson et al.; ²⁹ Grunt et al.; ⁴⁸ Spiegel et al.; ³² Steinbok et al.; ³³ Langerak et al.; ³⁰ Turi and Kalen ³⁸
Scoliosis	11–57	Spiegel et al.; ³² Steinbok et al.; ³³ Grunt et al.; ⁴⁸ Bolster et al.; ⁹ Nordmark et al.; ³⁷ Ravindra et al.; ³⁴ Turi and Kalen ³⁸
Spondylolisthesis, Spondylolysis	1–20	Grunt et al.; ⁴⁸ Johnson et al.; ²⁹ Li et al.; ³¹ Peter and Arens; ⁴⁴ Peter et al.; ⁴⁹ Spiegel et al.; ³² Nordmark et al.; ³⁷ Bolster et al.; ⁹ Peter et al.; ⁴⁰ Langerak et al. ⁴¹
Spinal stenosis	27	Gooch and Walker ²⁷
Impaired bladder control	1–29	Montgomery; ⁴² Steinbok and Schrag; ³⁵ Abbott; ²⁴ Langerak et al.; ¹⁴ Park et al. ⁴³
Increased dystonia	20–60	van de Pol et al. ³⁶
Muscle spasms	14–57	Montgomery; ⁴² Nordmark et al. ³⁷
Leg and back pain	29-71	Park et al., ⁴³ Peter and Arens; ⁴⁴ Langerak et al. ³⁰
Sensory changes – paresthesia	4–86	Montgomery; ⁴² Nordmark et al.; ³⁷ Park et al.; ⁴³ Peter and Arens; ⁴⁴ Grootveld et al. ²⁸
Obesity	15	Westborn et al. ⁶⁸

comparison to a group with CP assigned retrospectively.^{6,17} One study had controls of comparable age, but the SDR and comparison group were significantly different with respect to Gross Motor Function Classification System (GMFCS) level. Moreover, the comparison group included individuals with all subtypes of CP, even subtypes purposely excluded from SDR intervention, such as dyskinetic and unilateral CP.⁶ In the other investigation with a control group, Munger et al. made retrospective comparisons (indicated by a propensity model to be highly accurate) to individuals whose clinical presentation was appropriate for SDR, but who, for a variety of reasons, did not undergo surgery.¹⁷ In one study, the use of validated motor development charts for long-term assessment of gross motor function after SDR allowed comparison with the manner in which a child with CP is expected to develop.⁹ Two studies involved surveying patients: in one case online⁶ and in the other predominantly by telephone.⁷

As assessed according to the Oxford Centre for Evidence-Based Medicine, the level of evidence for the studies included was low, with two at level 3 and 14 at level 4 (Table SIII, online supporting information).

The overall risk of bias was medium to high in all cases (Table SIII). All sample sizes were small (n=11-95), being less than 50 in 13 of the 16 studies. All were conducted in clinical settings. Loss during follow-up, in part due to retrospective study designs and retrospective data, resulted in attrition bias in a vast majority of the reports: only four studies followed-up more than 80% of the original cohort^{11,14,20,21} and four lost more than 50% of the initial participants (Table II). Several of the instruments employed were valid and reliable, but none of the investigations was conducted with randomization or blinding. Different approaches to assessment resulted in heterogeneous data.

The outcomes assessed

Because of the heterogeneous nature of the outcomes, a descriptive analysis was performed. The outcomes most relevant and most frequently reported are listed below.

Spasticity

Seven out of the 16 studies evaluated the effect on spasticity specifically, and all reported sustained long-term reduction or even normal muscle tone.^{8,10,11,14,17,20,21}

Additional antispasticity treatment

Five articles,^{7,9,10,17,18} including two of those that documented long-term reduction in spasticity,^{10,17} noted a need for additional treatment such as botulinum neurotoxin A (BoNT-A) injections. Bolster et al. reported that 45% of the children had received BoNT-A treatment, mostly in the gastrocnemius muscle,⁹ while Dudley et al. reported 12.5%.¹⁰ Hurvitz et al. reported that 53% had BoNT-A treatment: 38% were on oral treatment and 15% had an intrathecal baclofen pump placed after SDR.⁷ Munger et al. documented an average of 7.5 and 19 BoNT-A injections in the SDR and comparison groups respectively.¹⁷ Finally, Park et al. reported that 22% of their participants still required oral antispasticity medication and 3% were treated with intrathecal baclofen at long-term follow-up.¹⁸

Gross Motor Function Measure assessments

Six studies used the Gross Motor Function Measure (GMFM), all of which showed an early improvement in GMFM score.^{8–11,20,21} However, only two^{9,11} compared this improvement to the gain anticipated as a result of the natural development of the child.^{45,46}

In one investigation originally involving 29 children with CP, among the 20 assessed 10 years after SDR, six had improved scores based on their GMFM motor curve (centile ranking), with those classified in GMFCS levels I to II^{47} (*n*=6) having developed largely as expected and those (5/14) in GMFCS level III improving more than expected.⁹ A 10-year follow-up of 29 children in southern Sweden concluded that 'the mean maximum development of GMFM-66 in children undergoing SDR seems to be better than, or at least as good as, the one found in the motor curves'.¹¹

One stratification of 44 patients on the basis of their GMFCS level⁸ concluded that the function of those in GMFCS levels II and III improved initially and later declined somewhat; while the gross motor function of those in GMFCS levels IV and V had declined to less than the baseline value at the 10-year follow-up.⁸ Of the 102 patients initially included by Dudley et al., 57 were still being followed up 10 years postoperatively and 50 of these had GMFM scores. GMFCS levels I to III were associated with significant improvement at the 10-year follow-up in comparison to the corresponding preoperative scores, whereas GMFCS level IV was not associated with any change at all.¹⁰ Finally, two assessments of the same cohort found that GMFM scores were significantly lower 10 and 17 years after SDR than the peak value at the 3-year follow-up.^{20,21}

Gait analysis

A broad range of gait data were reported in four studies, with wide variation in the specific measures of outcome and their presentation. Walking speed improved in one case,¹³ while another article described improved steplength and velocity at the 3-year follow-up, with a subsequent decline to baseline after 10 years.¹⁹ One description of gait lacked any comparison to baseline.¹⁶ Munger et al. found that the Gait Deviation Index for both the SDR group and those who were not operated on improved, more so in the latter case.¹⁷

Gait or ambulatory ability

Park et al. reported improved ambulation in 42% of participants' gait, similar to the preoperative situation in 42%, and worsened ambulation in 14%.¹⁸ Tedroff et al. reported improvement in short-term ambulatory status,

Study	LOE	Fraction of study population evaluated	Population by GMFCS level	Control group	Age at the time of SDR (range), y:mo	Follow- up (y)	Outcome of interest	Measures
Ailon et al. ⁸	≥	44/142	ll (8), lll (16), lV (17), V (3)	No	Mean 4:6 (2:11– 7:8)	>10, mean 14·6	Spasticity, GMF, ROM, strength	MAS in hip adductors, ROM hip abduction, quadriceps strength, GMEM
Bolster et al. ⁹	≡	20/36	−II (6), III (14)	Comparison to motor	Median 6:4 (2:10 12:1)	10	GMF	GMFM-66 - centiles
Daunter et al. ⁶	≥	38/unknown	I (2), II (7), III (11), IV (14), V (4)	50 age matched patients with all CP subtypes	Mean 5:5, all <10y	22	Pain, fatigue, function	Internet survey: PROMIS, FSS, self- reported change in function and walking ability
Dudley et al. ¹⁰	≥	57/105	Only 52 of 105 classified	No	Mean 5:0 (3:0- 10:6)	10	Spasticity, GMF, ADL	MAS, GMFM-88, PEDI
Hurvitz et al. ⁷	≥	88/271	I (7%), II (18%), III (23%), IV (36%), V (16%)	No	Mean 6:0 (SD 3:11)	19±3	Function, pain	Telephone or clinic interview, SWLS, NPRS
Josenby et al. ¹¹	≥	29/35	I (1), II (8), III (7), IV (12), V (1)	No	Median 4:4 (2:6– 6:7)	10	Spasticity, ROM, GMF	MAS, ROM, GMFM-66
Josenby et al. ¹²	≥	24/35	I (1), II (7), III (4), IV (11). V (1)	No	Median 4:1 (2:5– 6:4)	10	ADL	PEDI
Langerak et al. ¹³	≥	13/unknown	Walkers	None with CP, 12 age- matched typically developing controls	Mean 7:4 (2:0- 14:0)	20	Gait, ROM	2D gait analysis, ROM
Langerak et al. ¹⁴	≥	14/14	I (7), II (3), III (3), IV (1)	No	Mean 7:4 (2:0- 14:0)	20	GMF, ICF	GMFCS, ICF
Langerak et al. ¹⁵	≥	31/47	All ambulant (I–III)	No	Median 5:2 (2:0- 27:0)	17–26	Activity and participation	Interview: FMS, Life Habit
Langerak et al. ¹⁶	≥	31/47	All ambulant (I–III)	None with CP, 43 age- matched typically developing controls	Median 5:2 (2:0– 27:0)	17–26	Gait	3D gait analysis, GDI
Munger et al. ¹⁷	≡	24+11/96 ^a	Walkers	11 matched controls fulfilling criteria for SDR	Median 4:9 (SD 2:11)	10–17	Gait, spasticity, QoL, pain, participation	3D gait analysis, GDI, MAS
Park et al. ¹⁸	≥	95/431	I (21%), II (28%), III (31%), IV (14%), V (5%) ^b	No	Mean 6.0 (2:0– 17:11)	20-28	Ambulation, pain, satisfaction with life	SWLS, GMFCS, NPRS
Subramanian et al. ¹⁹	≥	11/14	Walkers	None with CP, 12 age- matched typically developing controls	Mean 7:10 (2:6– 13:2)	10	Gait	2D gait analysis
Tedroff et al. ²⁰	≥	19/19	I (3), II (4), III (2), IV (10) ^b	No	Mean 4:7 (SD 1:8)	10	GMF, spasticity, gait, ROM	GMFM-88, Wilson gait scale, MAS, ROM
Tedroff et al. ²¹	≥	18/19	I (3), II (5), III (3), IV (6), V (1) ^b	No	Mean 4:7 (SD 1:8)	15–19, median 17	GMF, spasticity, gait, ROM, OoL,	GMFM-88, Wilson gait scale, MAS, ROM, SF-36, Brief Pain Inventory

-z4 selective dorsal mizotomy (SUR). It controls, out of 96 invited. Pollow-up with gait analysis at 10y; 13/24 SUK group, 8/11 control group, total not separated. "Data shown are Gross Motor Function; Motor Function System (GMFCS) levels at follow-up. LOE, Level of Evidence (according to Oxford Center for Evidence-based Medicine 2011); GMF, Gross Motor Function; ROM, range of motion; MAS, Modified Ashworth Scale; GMFM, Gross Motor Function Measure; CP, cerebral palsy; PROMIS, Patient-reported Outcome Measurement Information System; FSS, Fatigue Severity Scale; ADL, activities of daily living; PEDI, Pediatric Evaluation of Disability Inventory; SD, standard deviation; SWLS, Satisfaction With Life Scale; NPRS, Numeric Pain Rating Scale; ICF, International Classification of Functioning, Disability and Health; FMS, Functional Classification of Function of Paciation of Disability Inventory; SD, standard deviation; SWLS, Satisfaction With Life Scale; NPRS, Numeric Pain Rating Scale; ICF, International Classification of Functioning, Disability and Health; FMS, Functional Mobility Scale; GDI, Gait Deviation Index; OoL, quality of life.

followed by a decline to baseline levels at 10 and 17 years' follow-up. 20,21

Self-care and mobility

The two evaluations of self-care using the Pediatric Evaluation of Disability Inventory, an instrument for which normative scores are available for children aged up to 7 years 6 months, both revealed increased scores throughout the period of follow-up.^{10,12}

Fatigue

The level of fatigue reported by patients who had undergone SDR was similar to that of a comparison group in one investigation.⁶

Pain

Although pain was not consistently evaluated or reported, two articles stated that the SDR group reported pain similar to the comparison groups.^{6,17} Pain comparable to norm values, as assessed by SF 36v2 (Short Form Health Survey 36 version 2), were reported in one study²¹ and pain 'last week' was reported by 44% of the participants in another study.⁷

Range of joint motion

Five studies reported on range of motion, four of which observed early improvement followed by later reduction.^{8,13,20,21} In one case, the range of motion at the 10-year follow-up was similar to the preoperative values.¹¹

Orthopaedic surgery after SDR

Although taken into consideration in most of the studies, this was not reported in a consistent manner.^{7–11,13,14,16–21} Altogether, 28% to 94% of the patients underwent orthopaedic surgery subsequent to SDR, often soft tissue surgery to correct contractures. There was no report of the potential presence of residual musculoskeletal deformities, that were present at long-term follow-up.

Complications or adverse events

Long-term complications not related to loss of function were addressed in three of the studies addressing functional outcome.^{7,9,14} The second search retrieved supplementary reports (Table I and Fig. S2). Only short-term and/or specific complications were described in some of the reports, making determination of the incidence difficult. The complications described in a total of 24 articles are summarized below.

Perioperative pulmonary complications

Judging from three comprehensive studies, the risk of acute pulmonary complications, such as bronchospasm and pneumonia, is approximately 9%.^{24,35,37}

Postoperative spinal complications

In four case series involving 553 patients, one deep wound infection, six dural leaks, and two wound hematoma were observed.^{24,35,37,44}

Acute neurogenic bladder dysfunction was reported in 7% to 12% of participants^{24,35,37} and permanent bladder dysfunction in 29% of children with CP who underwent SDR.^{14,24,35,42} The longest follow-up available, a questionnaire administered 24 years after rhizotomy, documents an 11% incidence of urinary incontinence.⁴³

Gastrointestinal dysfunction

Constipation, the most common gastrointestinal problem reported, occurred in approximately 26% of patients.^{24,35}

Sensory changes

Acute postoperative sensory changes, such as hypersensitivity, numbness, and paresthesia, were reported in approximately 50% of the patients, with most of these resolving within 1 to 2 years after SDR.^{20,28,43,44} However, some of these changes were permanent.^{28,37,42–44}

Spinal deformity

Spinal deformity is the most commonly reported long-term complication, with increased risks of lumbar lordosis,^{9,25,26,29,30,32,33,38,48} scoliosis,^{9,32,33,37–39,48} spinal stenosis,^{27,30} and spondylolisthesis.^{9,29,31,32,37,40,41,44,48,49}

Loss of motor function

The main complication related to gross motor function is unmasking of weakness.^{34,44,50}

Unmasking of dystonia

A recent study in the Netherlands found that despite careful selection, nine of 24 non-walking patients exhibited some signs of dystonia after SDR. Not surprisingly, when dystonia was present, the caregivers tended to be less satisfied with the procedure.³⁶

Pain

Long-term back and leg pain have been reported, occasionally caused by recurrent muscle spasms after SDR.^{7,37,42} However, it is often unclear whether such pain is related directly to the operation itself.^{7,30,43,44}

DISCUSSION

The current review of available literature on the long-term effects of SDR on children with CP includes 16 studies with a low level of evidence and a moderate-to-high risk of bias. Their study designs and heterogenic assessment tools limit the possibility of analysing data and, consequently, a descriptive and narrative analysis and discussion are presented here.

Impaired motor control, muscle weakness, cocontraction, dystonia, spasticity, and abnormal posture are all motor features that coexist in CP. In the decision to perform SDR, spasticity and strength are considered to be the most important of these. Indeed, spasticity is regarded as the major indication for SDR in at least 94% of relevant publications.⁵¹

In children, body weight and muscle strength typically increase with age. In CP, weakness is pervasive⁵² and the increase in muscle strength may not keep pace with weight gain. This has been demonstrated in children with CP as a reduced strength normalized to weight ratio,⁵³ and is believed to be one explanation for diminishing ambulatory ability with age.⁵⁴ The weakness associated with CP is sometimes clinically hypothesized to be compensated for, to a certain extent, by the hypertonicity of spasticity.¹ For instance, children with spastic CP may use the spasticity in their legs to transfer themselves from a wheelchair to a bed or chair.

Infants with prenatal or perinatal CP do not have spasticity at birth; however, spasticity first appears towards the end of the child's first year. A cross-sectional and longitudinal investigation on all children in Sweden with CP (covering 57 953 measurements on 4162 individuals), using the Modified Ashworth Scale, found that the level of spasticity in the calf of most of these children increased until they reached 5 years of age.² Thereafter, the spasticity declined and at 15 years of age only 22% recorded Ashworth scores of 2 or more, in contrast to 38% at 5 years. Similarly, another population-based Swedish study (n=3028), found that BoNT-A treatment was most frequent among 4- to 6year-old children, with such treatment becoming less common with increasing age.⁵⁵

Such findings have several implications with respect to SDR. Evaluation for and performance of this operation often occur at the age when spasticity, the most consistent indicator for SDR, is most pronounced.^{15,56} Thus, the reported short-term improvements after SDR correspond temporally both to a period of anticipated natural development of motor skills, as well as an expected reduction in spasticity. The impaired gross motor function reported in connection with follow-ups for 10 years or more^{8,18–21} can be explained, at least in part, by less strength relative to body weight. For example, Park et al. found that 30% of the almost 300 individuals they followed for 2 to 28 years after SDR reported weakening of their muscles as they matured into adulthood. As adults, their spasticity may help compensate for this weakness.⁴³

Another difficulty is the frequent presence of both spasticity and dystonia in CP.^{1,36,57} Dystonia often develops at a later age than spasticity and its severity is related inversely to function.^{1,57} As described above, spasticity is believed to be caused by a reduction in spinal inhibition by altered descending tracts and can thus be treated by altering spinal activation with SDR. However, in contrast, although the exact pathophysiology of dyskinesia or dystonia is unknown, current hypotheses involve altered activity of basal ganglia circuits, which as an effect, erroneously modulate the higher level of cortical activity.¹ Consequently, operating at the spinal level with SDR does not alleviate dystonia and, indeed, the unmasking of dystonia as the result of such an operation can be a serious problem.³⁶

A systematic review has shown that the interventions that reduce spasticity most effectively are not those that

give the greatest improvement in activities or gross motor function.⁵⁸ Moreover, as observed in children with CP who ambulate with or without an assistive device, strength is closely related to function and explains considerably more of the variance in performance than does spasticity.⁵⁹ Accordingly, motivating reduction of spasticity in terms of an anticipated improvement in function can be questioned.

It is often suggested that reduction of spasticity with SDR lowers the need for future orthopaedic surgery and prevents contractures. To some extent Munger et al. support this finding by showing that the 24 participants who had SDR underwent 10.8 orthopaedic surgeries on average during the follow-up compared to 13.5 orthopaedic surgeries in the 11 retrospectively assigned control participants.¹ However, in most of the articles analysed here, the need for orthopaedic surgery and contracture release after SDR remained high. Overall, the findings presently available do not indicate either an increase or decrease in the need for further orthopaedic surgery.^{7–11,13,14,16,19–21}

With respect to the duration of the reduction in spasticity after SDR, only seven of the 16 studies analysed took a long-term perspective.^{8,10,11,14,17,20,21} A sustained reduction of spasticity in most of the muscles evaluated was most often reported. Interesting, however, was the somewhat contradictory finding in five of the articles that 22% to 59% of patients required additional treatment with oral medication, BoNT-A, or intrathecal baclofen pumps.^{7,9,10,17,18} A possibility is that either their spasticity had returned or had not been fully resolved by the SDR. Supporting this is one paper stating that BoNT-A treatment were 'mostly gastrocnemius' and another describing the use of intrathecal baclofen pumps.^{7,9} However, from available data in the remaining three papers, it is not possible to establish if muscles treated were innervated by rootlets not sectioned during the SDR, such as muscles in the upper extremity. It is also possible that some individuals might have been treated for dystonia.³⁶

As indicated, the advantages and disadvantages of spasticity and the need to alleviate spasticity may vary with age.^{1,2} Consequently, it appears advisable to reduce spasticity, when necessary, employing reversible treatment options, the nature and degree of which can be adjusted as the child grows.

Assessment of the outcome of SDR by comparison with an appropriate control group is particularly important when the patients are children, whose functional motor skills improve as they age. Charts depicting the motor development anticipated in children with CP at different GMFCS levels have been constructed.⁴⁶ For example, children in GMFCS level II improve an average of 25.1 points on the GMFM-66 scale between the ages of 2 and 12 years.⁴⁶ However, the difficulties in creating and followingup an appropriate control group for 10 years or more are immense and, thus, the vast majority of studies that do so are case series. Only two of the studies included had a somewhat higher level of evidence when evaluating the long-term effects of SDR on gross motor function and in neither case was this effect clearly beneficial.^{9,17} One study concluded that the results obtained indicate that different methods of treatment provided similar outcomes into early adulthood. $^{\rm 17}$

Clinicians and researchers who are responsible for the care of children with CP have developed a wide variety of tools for assessment and classification of both severity and function. However, in the 1980s and early 1990s, when the use of SDR became more widespread, most of the instruments used today, including the GMFCS, the Manual Ability Classification System, the Functional Mobility Scale, and even the GMFM were not yet available nor commonly utilized. For instance, the GMFCS was first described in 1997, while the GMFM was published in 1989.^{47,60}

In addition, in earlier times medical records were often somewhat unspecific and/or subjective, including vague descriptions (such as 'moderately affected' or 'severe type') of function and severity,⁴⁴ precluding retrospective assignment of GMFCS levels. Nevertheless, many of the relevant studies published to date have assigned baseline GMFCS levels retrospectively when evaluating the outcome of SDR performed in the early 1980s and throughout the 1990s.^{6,8,10,12,14–16,18} Such an approach can introduce a substantial risk of detection bias, especially when changes in GMFCS level are used as a measure of outcome.

In a commendable attempt to overcome this problem, one of the 16 articles described a machine learning algorithm based on the Gillette Functional Assessment Questionnaire and gait data collected in connection with a medical visit.¹⁷ The GMFCS levels arrived at by this algorithm agreed in 80% of the cases with those assigned by trained clinicians. These authors concluded that, 'of those study participants who had both baseline and follow-up GMFCS values, five out of nine improved their GMFCS level and one out of nine worsened, while all of those without SDR remained at their baseline GMFCS level'.¹⁷

One aspect of SDR is the potential negative impact of irreversibly cutting afferent fibres on future development. Sensory input is generally considered to be essential for refining and consolidating neuronal connections during development of the nervous system,⁶¹ with sensory feedback contributing to the strength and timing of muscle activity.⁶² Selective presynaptic control of the Ia afferents has been proposed to contribute to the acquisition of new visual and motor skills and alterations in interneurons between sensory afferents and motor efferent neurons play a central role in modulating reflexes during motor learning.⁶³ Accordingly, SDR may influence motor learning negatively, contributing to a lack of long-term improvement in gross motor function or mobility.

Another aspect of the cutting of sensory afferents is the 'tremendous variation' and very poor reproducibility of the responses obtained when stimulating one and the same rootlet in preparation for SDR reported by several investigators.^{64–66} One study reported that in only 16% of the patients did the relevant rootlets display pathological reflex responses and, as a result, when performing the SDR, 84% of the children underwent a non-selective procedure.⁶⁷ At

the 1-year follow-up these children displayed outcomes similar to those who had undergone more selective surgery, raising concerns over the 'selectivity' of this approach and questioning the belief that selective rhizotomy is more beneficial than the random procedure.⁶⁷

Although complications and adverse events secondary to SDR occur, the prevalence of acute complications has not always been reported. In this review, only a minor portion of the 16 long-term studies reported on complications and side effects. When a search was conducted aiming to identify these issues several identified papers specifically focused on complications not always addressing the baseline situation. Additionally, some of the reported complications, such as constipation, are frequently encountered in all children while, for example, spondylolisthesis or spondylolysis are rare. This can, as an effect, over- or underestimate the true rate of complications.

The major late complication observed in this review, reported in 11% to 57% of all individuals, is spinal deformity, although most of the case series involved X-ray follow-up at a time-point clearly before skeletal maturity.^{9,25–27,29–33,38,39,48,49} The highest risk of skeletal deformity occurs during adolescent growth. Therefore, any reports of a risk of developing deformity should include a radiological evaluation at skeletal maturity. Presently, such information is not available.

The overall risk for complications is higher in children with more neurological involvement, which may contribute to the focus on performing SDR in children who demonstrate better gross motor and cognitive function-ing.^{9,24,33,35,36,48}

SDRs are costly for families, insurance companies, and the healthcare system and, furthermore, this procedure involves a major investment of time by both the child and his or her parents. Moreover, lifelong strength training after the procedure is often recommended for maintenance of motor function.

CONCLUSION

In summary, although the evidence available is limited by clinical variability and heterogeneity across trials, this review demonstrates that conclusive data on the long-term effects of SDR performed on children with CP are still lacking. Low-quality evidence suggests that the long-term effect on spasticity remains uncertain, with many investigators reporting a considerable need for subsequent additional treatment of spasticity. The possibility that SDR reduces or eliminates contractures and/or orthopaedic surgery is challenged by the large number of such patients who later develop contractures that require surgical treatment. The currently available studies involving follow-up for 10 years or more do not indicate that the improvement in function obtained is any better than with routine therapy and orthopaedic management. Further prospective, long-term evaluation based on global registries of children with CP who have received SDR, or other treatment, is highly recommended.

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SUPPORTING INFORMATION

The following additional material may be found online: **Table SI:** Searches in PubMed and Embase

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Table SII: 'Risk of bias' modified version of the Quality in Prognosis Studies tool

Table SIII: Level of evidence and risk of bias for the studies involving long-term follow-up after selective dorsal rhizotomy

Figure S1: Search strategy for long term effects of selective dorsal rhizotomy.

Figure S2: Search strategy for identifying publications that report complications associated with selective dorsal rhizotomy.

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ECEditor's Choice

Editor's Choice

Thousands of individuals have undergone selective dorsal rhizotomy (SDR) in the last 30 years to reduce their spasticity. My Editor's Choice for the May 2020 issue is this systematic review of long-term outcomes of the procedure in cerebral palsy. The authors suggest that the effectiveness of SDR has not been assessed soundly, including in the long term,¹ and they conclude to uncertainty of its effects. The review of published studies is generating discussion from a number of perspectives represented in this issue, including neuroscience,² clinical experience,³ and lived experience.⁴

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