Selective dorsal rhizotomy (the perspective of the neurosurgeon and physiotherapist)

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Abstract

In this article, the authors review selective dorsal rhizotomy (SDR) as a treatment option for spasticity and its role in the management of bilateral spastic cerebral palsy. The SDR pathway is outlined and referral criteria, patient selection, physiotherapy rehabilitation considerations, and outcomes are discussed.

Keywords Cerebral palsy; multi-disciplinary team; outcomes; patient selection; physiotherapy; quality of life; selective dorsal rhizotomy; spasticity

Introduction

Cerebral palsy (CP) affects approximately 2–3/1000 live births in Europe and is caused by injury or developmental disturbance to the immature brain. A large proportion of cases are caused by events in the perinatal period. The resulting central nervous system damage can lead to multi-system impairment of differing severity and management can be complex. CP characteristically presents with disordered development of movement and posture, but it is important to remember it is not just a movement disorder when choosing treatment options and there is usually, in addition, significant impairment in cognition, behaviour and sensation which can impact on prognosis and functional ability.

Classification and patient selection for different treatments

There have been several classification systems for CP. Generally, however, CP is currently classified into four categories: unilateral spastic, bilateral spastic, dyskinetic and ataxic. Bilateral spastic CP, also known as spastic diplegia, is the most common form and is strongly associated with prematurity. As spasticity is the most important issue in this category of CP, these are the children who are most likely to improve with selective dorsal rhizotomy (SDR). Although spasticity may be present in all four limbs, the lower limbs are affected to a higher degree than the upper limbs, which would usually only demonstrate reduction in fine motor function. Spasticity contributes to muscle imbalance and is implicated in progressive muscle and joint contractures, bony torsions and premature joint degeneration. In children these imbalances are significantly exacerbated by growth. Spasticity and spasms are also recognised as causes of pain and can impact on activities of daily living and cares, with associated effects on emotional development, behaviour and learning.

The severity of the motor disability in spastic CP is classified according to the Gross Motor Function Classification System (GMFCS). Definition of a child's GMFCS level requires detailed physical assessment and carries prognostic implications. Population studies of children with CP have shown that children within GMFCS grades III – V demonstrate significant decline in function over time, with reduced mobility and increasing levels of dependence.

The diagnosis of bilateral spastic CP is clinical and is characterised by the typical physical pattern of movement problems and developmental delay. Magnetic resonance imaging (MRI) demonstrates periventricular leukomalacia (PVL) in over 70% of these children. This appears as periventricular hyperintensity on FLAIR or T2-weighted images, predominantly in the occipital and atrial regions. When the injury is more severe the frontal periventricular white matter is also affected. This white matter injury compromises supratentorial influence to the spinal neuronal pool; abnormal inputs through the vestibular and reticular nuclei and their tracts results in loss of inhibition to the spinal reflex arcs. This results in an increase in tone. In particular, damage to the vestibulospinal tracts increases extensor tone. Due to the topographic arrangement of the periventricular white matter, smaller lesions primarily affect the lower limbs while more extensive lesions also cause upper limb spasticity.

Most children present with a mixed pattern of movement problems with increased muscle tone (hypertonia) and difficulty initiating and sustaining adequate and well-timed voluntary movements. Hypertonia is rarely pure spasticity (velocity dependent muscle resistance) but can include dystonia and inappropriate increased muscle activity associated with other stimuli such as speech or hand function. The mechanisms of these different tonal problems is not clear, and changes in presentation can occur throughout childhood. However differentiating between these problems is an important factor when choosing treatment options as they respond differently.

General management of spasticity

The treatment of spasticity is an important aspect of the overall management of bilateral spastic CP. Removing spasticity alone is not directly related to improved functional achievement. Spasticity treatments must be incorporated within multidisciplinary goals and supported with appropriate therapy and assistive equipment when required.

The national institute for clinical excellence (NICE) recommends that each local paediatric multidisciplinary team should have a pathway for spasticity management, with supporting educational materials to help parents make informed decisions. Usually less invasive management options are considered initially. These include physical therapy, systemic medication,

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postural management, such as specialised equipment and orthotics, as well as localised myoneural blocks such as botulinum toxin injections (BTX).

Medical approaches to management of spasticity

In practice initial improvements with oral baclofen may lessen with growth, and the dose required to reduce spasticity gradually becomes large enough to impair central postural control and alertness. BTX injections may be sufficient to target one or two muscle groups but in more generalised spasticity dose limitations become significant and shorter periods of benefit are reported with repeated use.

Surgical approaches to management of spasticity

Intrathecal baclofen, through an implantable pump – catheter system, is used to control spasticity and dystonia in children within the GMFCS IV and V levels. Orthopaedic surgery, usually now carried out as a single event as a child approaches skeletal maturity, corrects bone and joint alignment and torsional problems, improves hip stability and reduces fixed contractures. However, the increased tone often remains and can contribute to recurrence of deformity. SDR, usually performed between 3 and 10 years of age, reduces spasticity effectively on a permanent basis.

Selective dorsal rhizotomy – surgical technique

SDR has evolved into its current form over the last 30 years. Following Foerster's initial series published in 1908, and the development of intraoperative neurophysiological monitoring in the 1980's, its modern indications and outcomes were defined by Peacock and Arens in 1987. The traditional procedure involves a four to five-level laminectomy or laminoplasty, with identification of the afferent segmental nerve roots to the lower limbs at their exit foramina. A percentage of the sensory root is divided at each level after dissection of the sensory and the motor roots at each foramen.

The single-level procedure was developed by Park in the early 1990's. A midline fenestration is made at the T12-L1 level and allows identification of the conus using ultrasonography. The conus, and the L2 to S2 nerve roots, are then exposed through a single level laminectomy. There is a clearly identifiable anatomical plane between the ventral (motor) and dorsal (sensory) roots; the motor roots are protected throughout the procedure.

The L2 to S1 sensory nerve roots are identified, divided into rootlets and systematically stimulated using intraoperative electromyography (EMG) to determine their threshold amplitude. Each rootlet is then stimulated at the threshold amplitude at a frequency of 50 Hz. The response generated is graded on a scale of 1–4. It may be confined to the myotome innervated by the stimulated root (grade 1) or it may involve adjacent myotomes (grade 2). More extensive involvement of the whole side, or of the contralateral side and upper limbs, is graded as 3 and 4, respectively. The objective of the procedure is to divide approximately 60–70% of the sensory roots between L2 and S1. Rootlets with grade 3 to 4 responses are preferentially divided. In addition, 50% of the L1 sensory nerve root is divided as it exits its foramen. Intraoperative monitoring of the pudendal nerve together with limited division of the S2 nerve root is now commonly practised to reduce the risk of incontinence.

Acute post-operative complications are rare and can include infection, haemorrhage and cerebrospinal fluid leak. Patients commonly experience transient dysaesthesiae, which usually resolve over a few weeks. Permanent complications, such as incontinence and spinal instability, are now very rare with single-level, monitored SDR.

Patient selection

SDR is a specialist neurosurgical procedure and as such is only carried out in specialist centres where there is a multidisciplinary team with experience of assessing and managing children with cerebral palsy. Immaturity makes understanding a child's individual presentation and the longer term functional implications difficult. Therefore selection for SDR requires multidisciplinary input, involving paediatric physiotherapists with special expertise in movement disorders, neurodisability specialists, paediatric neurosurgeons and orthopaedic surgeons.

Patient selection for SDR remains variable and has not been generally validated (Table 1). Most SDR services use selection criteria that reflect those used by Peacock in the first modern SDR series published in 1987. Similar criteria were also used in the three randomised controlled trials held in North America in 1998.

General selection criteria for SDR

Type of CP:	Spasticity without significant dystonia Classically bilateral spastic diplegia
Severity of CP:	GMFCS II & III (Aims – functional gains in transitions & ambulation with/without mobility aids) GMFCS III & IV (Aims – functional gains in sitting and lying) GMFCS IV (Aims – Comfort and ease of care)
Age	3–14 years
MRI:	No injury to basal ganglia, brainstem or cerebellum Typical periventricular leukomalacia (PVL)
Musculoskeletal:	**GMFCS II & III** – Good trunk control and antigravity strength in legs on clinical examination. **GMFCS II–IV** – Adequate muscle length and joint alignment to allow for rehabilitation and positioning No significant femoral head subluxation on pelvic radiograph (Reimer's index up to or less than 40%) No significant scoliosis
Previous interventions:	Preferably 6 months post BTX injections Preferably 1 year since orthopaedic interventions
Child and family factors:	Motivation to move Ability to cope with the surgery and rehabilitation process (cognitive and emotional)

Table 1

Identifying 'good' candidates for SDR

Typically, good candidates for SDR are aged 3 to 14 years where spasticity is their dominant motor problem without significant ataxia or dystonia. SDR has been shown to be effective in older patients, but with increasing age weakness and musculoskeletal deformities may be more restrictive than spasticity. MRI scan of the brain would show typical PVL, without additional injury to the basal ganglia, thalami, brainstem and cerebellum. The hips would be stable, with Reimer indices below 40%. If this index is higher, hip stabilisation surgery may need to be considered before SDR. It is ideal to defer SDR for at least 6 months after the last botulinum toxin injection, and 1 year after orthopaedic surgery.

In our current practice we recommend SDR across GMFCS II – IV with differing aims based on their GMFCS level. The largest group comprises children within the GMFCS II and III levels. Features associated with good functional improvement in GMFCS II are minimal musculoskeletal deformity, good selectivity and dissociation of movement at the foot and ankle, antigravity muscle strength throughout the trunk and lower limbs, adequate walking balance to manage without walking aids, minimal visual perceptual problems, as well as physical determination with a disposition to be active.

For GMFCS III with good musculoskeletal alignment, the difference usually lies in their significantly lower core strength and muscle selectivity; they tend to retain their antigravity strength. They therefore require walking aids for balance and to compensate for muscle weakness at the pelvis, even after SDR. This group may make greatest gains in sitting and floor mobility. For most GMFCS IV children, SDR aims to improve comfort, posture and ease of care.

Definition and clarification of objectives with the family, and, where appropriate, with the child, within the context of surgical risk, is crucial. Ensuring parents, carers and therapists understand potential prognosis following surgery ensures the child is offered appropriate post-surgical support. It is our experience that where too much is expected of the surgery this leads to emotional and psychological difficulties for the child and their family. Clear realistic goals support the rehabilitation process, but allow for unexpected improvements should these occur.

Outcomes

Effects of SDR

SDR is very effective at eliminating spasticity. In our experience, in ambulant children, SDR leads to improved mobility, increased stamina, better balance and fewer falls. Children who walk with assistance become more independent. Sitting and standing posture improves. In GMFCS IV children, floor mobility and improve after SDR. Pain and discomfort attributed to spasticity are removed across all GMFCS groups and parents have reported improvements in comfort and ease of movement with better sleep and generally raised mood. Patients and their carers also report an increased ability to manage activities of daily living and improvements in hand function.

The evidence from randomised control trials suggests that functional gains following SDR are likely to be moderate but significant. The Gross Motor Function Measure (GMFM) is a clinical tool designed to evaluate change in gross motor function in children with CP. A meta-analysis of three randomised controlled trials, comparing SDR combined with physiotherapy to physiotherapy alone, found that GMFM increases by approximately 4% following the combined treatment, which is almost double what physiotherapy alone would achieve. In a separate randomised controlled trial comparing SDR to botulinum toxin injections, the effects of SDR were more enduring than the injections and SDR patients had fewer orthopaedic interventions.

Long term outcomes

Evidence on the long-term effects of SDR is beginning to emerge. A recent study with a 2-year follow-up found that children aged between 4 and 7 years old with pre-operative GMFM scores between 65% and 85% benefited most from SDR. One group has reported on children with spastic diplegia at 5 and 10 years after surgery. There was immediate reduction in lower limb muscle tone and no recurrence of spasticity with improvement in passive range of movement and GMFM. Notably, the Paediatric Evaluation of Disability Inventory (PEDI) scores of children classified as GMFCS I–III showed improvement in the functional skills, mobility and caregiver domains. Children classified as GMFCS IV–V showed small changes.

Another study that followed patients to 15 years post-surgery found similar improvements in lower limb muscle tone, GMFM and ADL. The authors found that improvements in PEDI scores persisted to 15 years. There was also a significant reduction in the requirement for further orthopaedic intervention compared to other studies of children of similar ages who did not undergo SDR. The Cape Town experience found that 20 years after SDR there was a 58% improvement from baseline in activities of daily living, with no worsening of GMFCS grade. Moreover the improved walking speed and knee range of movement had been maintained to 20 years.

Who will be less likely to benefit?

Less satisfactory long-term outcomes have been reported and provide a substantial contribution to the evidence base for patient selection. Children with spastic quadriplegia had poorer outcomes compared to those with diplegia. Children over 10 years of age had better long-term outcomes with multi-level orthopaedic surgery than with SDR. This underlines the particular challenges related to patient selection in the older age group.

Pre-surgical assessment

Information sharing and careful assessment are crucial

Prior to surgery children should undergo a full assessment with information shared between the hospital and local physiotherapy and occupational therapy teams and school/nursery staff. The assessment battery includes environmental and personal factors, age appropriate activity and participation measures, measurement of biomechanics, muscle tone, strength, selectivity, coordination and functional balance. Motor planning and visual perception are also considered, with more formal assessment recommended if issues are highlighted. Depending on GMFCS level and Functional Mobility Scale (FMS), speed and endurance measures are used for transitions and walking. Questionnaires regarding perceived quality of life and support required for daily living are undertaken.

Children with cerebral palsy may need additional psychological evaluation to prepare for surgery and rehabilitation. For the more able child, coping with a period of reduced function following surgery is challenging, especially for transfers and mobility; this must be anticipated and managed sensitively. A period of more intensive pre-surgical therapy has been shown to improve early surgical gains, and can be helpful to prepare the child and their family for what will be expected after surgery. The child's relationship with their therapy team is crucial in the process. Motivation to move may have more influence on outcome than cognitive ability, especially in the more severely affected child.

Orthopaedic issues raised during selection assessment are discussed with the child's local orthopaedic surgeon or specialist centre, and a plan for management is outlined. It is typical to review the child's current orthotics, equipment and home and school situation to pre-empt any issues and recommend additional equipment.

Post-operative care is critical to achieving the best outcomes

Research evidence supports intensive physiotherapy rehabilitation for up to 2 years post-SDR and this must be confirmed with the local therapy team prior to surgery. Additional therapy funding is usually provided on request from the local commissioning body, although this is not guaranteed. Children will require a period of gradual reintegration back into school/nursery and this should be planned for with the special needs coordinator.

Early post-operative recovery and rehabilitation

Following surgery children are nursed flat for 24 hours; gentle bed mobility is then allowed. Pain is managed with nurse or patient controlled analgesia and is not usually significant after the first 3 days. Anxiety about potential pain however often persists. Knee gaiters are prescribed for daily use to reduce early leg spasms. Physiotherapy begins on day 3 with active movements, sitting and tilt table standing. Most children attend therapy sessions in the gymnasium by day 4. The orthopaedic element is managed in a similar way to other single level spine procedures; spine movement should be active rather than passive with extremes avoided in the first 6 weeks. Hamstring stretches can cause neural tension at the surgical site and should be based on pre-operative length during soft tissue healing (usually 3 to 4 weeks).

Spasticity is reduced immediately after surgery and children often report that their legs feel looser and more comfortable. Dysaesthesia, usually described as itching, hypersensitivity to touch, or patchy numbness, is common in the first few weeks. Sometime this can be distressing and gabapentin is offered to all children, but is not always well tolerated. Only a small percentage of children experiencing symptoms at discharge will have ongoing problems at 6 months post-surgery.

Most surgical centres offer 2 to 3 weeks of intensive rehabilitation after SDR. Immediately following SDR reduced sensory feedback means that children struggle to initiate and sustain movement control. This can be distressing especially if the child was previously ambulant. This lack of control is often mistaken for structural muscle weakness; however it is the sensory input of the movement that is missing. In particular, they lack muscle tone control with poor awareness of reciprocal activation and inhibition. They may rely on their vision to a greater extent and this can be a significant limiting factor for children with marked visual perceptual problems.

Optimal muscle length and joint alignment are important in supporting function where children have to re-learn muscle control. To compensate for lack of control they often use joint end of range or relatively tight muscles to provide stability – this can be a problem where structures are short or over-lengthened. Well considered orthotic provision is essential; this not only provides distal stability and appropriate knee and hip alignment but also facilitates activity. Knee extension should be prioritised over ankle dorsiflexion. A residually tight calf complex can be accommodated in orthotics as necessary to facilitate standing and walking.

The decision to consider early surgical muscle lengthening is highly individual and is usually recognised during pre-SDR assessment. If alignment surgery is not considered essential for rehabilitation it is preferable to defer surgery. Standard strategies for maintaining muscle length, such as serial casting and orthotics, should be part of the post-SDR regime. It is usual to see improvement in muscle length within the first 6 months if the muscle is used functionally; however this should be closely evaluated at each post-operative assessment. If there is no improvement, or if the structural tightness is limiting expected progress, then earlier surgery may be required.

The speed of regaining muscle control is usually related to GMFCS level, with children in GMFCS II regaining some walking after 3 weeks, usually with aids for balance and to improve stamina. Children within GMFCS III and IV take longer to regain previous activity. Additional postural support at discharge is usually required. Young children with CP are highly variable and unpredictable. Associated learning & behavioural issues may make rehabilitation more challenging. Fatigue can be a considerable problem, and children require plenty of rest between therapy sessions. It is important that parents and carers recognise that relearning movement can be a slow process.

Once children become more active it is common to see re-emergence of previous dystonia or associated movements. Most commonly these are seen in the feet, quadriceps and arms. Parents and children need to be reassured that this is not a return of spasticity. If these persist, and interfere with rehabilitation, they should be managed in an appropriate way most commonly with orthotics or BTX.

Post discharge rehabilitation

Guidance for early rehabilitation is based on the child's abilities at discharge. Individual aims and areas that require close monitoring are highlighted. There are some physical limitations in the first six weeks related to surgical healing, so contact sports, trampolining and horse riding should be avoided. The local therapy team will need to continue to assess risk during this period and advise on mobility aids and equipment. This is particularly important for ambulant children as it is not unusual for children to have sudden loss of antigravity control especially if they are distracted or trying to do another task while standing.

This is usually related to the sensory strategies they are using and should not be seen as an ominous predictor of future function. Experience shows sensory feedback modification and therapy techniques such as proprioceptive neuromuscular facilitation (PNF) can improve this.

The first 3 to 6 months are usually a period of most rapid change particularly for children within the GMFCS II and III levels, as they recover from the surgery and regain functional skills. It is essential to provide progressive rehabilitation that includes movement control, graded resistance strengthening, fitness and endurance. The optimal dose of therapy remains debated but key elements are repetitive guided practise with appropriate motivational goals to ensure motor learning and musculoskeletal change. Timely orthotic and equipment changes will maximise functional gains and assistive modalities such as electrical stimulation have been used successfully to support therapy aims.

Evaluation of outcomes and longer term monitoring

Children have formal assessment at 6, 12 and 24 months post-surgery with repeated outcome measures and goals setting for ongoing rehabilitation. As children regain skills more formal therapy is substituted by fitness activities and physical leisure pursuits. This aids motivation and supports long-term gains. Specialist therapy can then be targeted at specific issues or used to support additional management strategies.

Longer term evaluation and monitoring remains essential for children with cerebral palsy until skeletal maturity. Removal of spasticity can impact on, but not fully negate, other associated factors which can still cause problems as children develop and grow. Biomechanical problems may be less dramatic in this post-SDR group but the effects of gravity coupled with relative weakness and joint instability still cause deformity and misalignment issues. These require timely therapeutic management and appropriate orthopaedic surgery to maintain mechanical advantage. Therefore it is not uncommon for children to require foot stabilisation procedures and muscle lengthening or tendon plication during their growing years.

Summary

SDR is an effective and permanent treatment for spasticity in young children with bilateral CP. Historically mobility and functional improvements were the prime aims of SDR in more functionally able children. However, for carefully selected children, SDR can bring sustained improvements in quality of life for GMFCS levels II–IV. SDR should not be seen as a treatment without risk and should be considered with clear aims on a case by case basis, as part of a wider management pathway. Rehabilitation is considered essential to support functional gains. Children undergoing SDR will retain many of the problems related to their brain injury and may require ongoing interventions for these even after spasticity is removed. There remain many unanswered questions about the mechanisms of altered muscle tone and movement control in children with CP, and longer term evaluation will ensure that the most appropriate children are selected for this intervention. ◆

FURTHER READING

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Practice points

- Current evidence shows that single level selective dorsal rhizotomy is a safe, effective and permanent treatment for generalised lower limb spasticity in carefully selected children with bilateral spastic cerebral palsy
- Patient selection and treatment should be only be carried out by a multidisciplinary team with training and experience in treating children with cerebral palsy and movement disorders. This usually includes physiotherapists, paediatrician and surgeons, all with training and expertise
- Parents/carers should be informed that surgery is irreversible and clear goals based on a child's level on severity should be discussed and agreed prior to surgery
- Prolonged post-SDR physiotherapy is essential to achieve functional goals and long term follow up through childhood is necessary
- Reduced spasticity can limit orthopaedic problems however other factors in cerebral palsy can mean that alignment surgery may still be required to optimise functional gains