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