



Selective Dorsal Rhizotomy (SDR) in SCOTLAND INFORMATION FOR PROFESSIONALS

SDR in Scotland | Information for Professionals | Version 1.0—September 2018

1. Background

A) PROCEDURE

Selective Dorsal Rhizotomy (SDR) is a complex neurosurgical technique developed to reduce spasticity and improve mobility in patients with cerebral palsy and lower extremity spasticity.

It involves the irreversible selective division of dorsal (sensory) rootlets as they emerge from the conus medullaris of the spinal cord. These nerve roots make up the afferent (input) limb of the reflex arc that is exaggerated in spasticity. The procedure takes place under general anaesthesia using intraoperative neurophysiology.

There is close communication between the surgical team and a neurophysiology team during the procedure to map each sensory nerve root to its corresponding motor level and then test the motor response to stimulation. The teams select the most abnormal 50 to 70 percent of nerve roots (those with the most exaggerated responses) at each level for division. All motor nerve roots are preserved.

This procedure decreases spasticity by lessening the sensory input to abnormal reflex arcs whilst preserving voluntary movement and function.

B) SPASTIC DIPLEGIA

Cerebral palsy describes a group of permanent brain disorders originating during fetal development, birth or early childhood. It is associated with abnormalities of movement, balance and posture. The prevalence of cerebral palsy in developed countries is stable at around 2/1000 live births. Approximately 40% of cerebral palsy cases are children who have been born prematurely. Considering the breakdown of cerebral palsy subtypes, around 75% of children will have a predominantly spastic muscle tone of which one third will have a diplegic pattern (lower limb predominant).

Children who are born prematurely and have a very low birth weight (<1500g) are predisposed to periventricular leukomalacia (PVL) and germinal matrix haemorrhage, with post-haemorrhagic ventricular dilatation. As the periventricular white matter is particularly susceptible to injury between 26 and 34 weeks of gestation, the leg fibres are predominantly affected, causing spastic diplegia.

Although the injury to the central nervous system is static, the clinical expression of cerebral palsy evolves as children grow.

Spasticity causes significant discomfort and stiffness, and associated spasms cause pain and interference with function. Growth during childhood, in the presence of spasticity, results in muscle shortening and contractures involving both tendon and soft tissues. In children, spasticity affects muscle growth and torsional abnormalities of long bones, joint instability and premature degeneration. Spasticity may make caring for children with cerebral palsy extremely challenging for their parents and carers.

C) HISTORY

Lumbosacral dorsal rhizotomy for spasticity was first advocated by Foerster in the early 1900's¹.

Partial sectioning of the posterior nerve roots on the basis of intra-operative

electrophysiological stimulation was introduced by Fasano in 1978². The

technique was adopted and popularised by Peacock and Arens³.

D) CURRENT STATUS

SDR has become accepted as a neurosurgical procedure for the treatment of spasticity associated with cerebral palsy in carefully selected children⁴.

It is performed either through a L1 (or L2) to S1 laminectomy or laminoplasty, or more recently (as practiced in Scotland) through a single level laminectomy at the position of the conus, as determined by intra-operative x-rays and confirmed by ultrasound.

Between 50% and 70% of the nerve roots are divided; most surgeons minimise rhizotomy at L4 and S2 to avoid significant quadriceps weakness and bladder dysfunction respectively.

E) PATIENT SELECTION

Agreement by all carers and professionals involved on the goals of treatment for the individual child is crucial. The principal goals may include improved motor function, increased mobility, increased independence and improvement in ease of care. A significant commitment is required due to the amount of postoperative rehabilitation required for up to 2 years following surgery.

SDR would be considered in ambulant children (with or without assistive devices) with spastic diplegia without significant contractures for which orthopaedic surgery was likely. Such surgery would not be undertaken simultaneous to SDR and so its timing would be important to establish if referral was being considered for SDR.

Children usually have a history of preterm birth and exhibit typical PVL changes on brain magnetic resonance (MRI) scan, without significant ataxia, athetosis or dystonia.

Good trunk control, good lower-extremity antigravity strength, no significant scoliosis and the ability to isolate lower extremity movements are also important considerations during the referral/selection process.

A MRI scan is necessary to exclude injury to the basal ganglia and cerebellum.

The optimal age for surgery is believed to be between 3 and 12 years, though it is difficult to assess walking function objectively in children of 3-4 years who are still developing their motor skills. The Scottish SDR service will take referrals between age 4 (for starting the assessment process) and 10 years⁵.

Significant upper limb spasticity with trunk involvement in addition to lower limb involvement is likely to benefit to a greater extent from intrathecal baclofen (ITB) therapy rather than SDR^{6,7}.

If after careful consideration SDR is not recommended for a child, the Multi-disciplinary Team (MDT) will give guidance to the child's referring team on what alternative interventions may be appropriate to improve function and quality of life for child and family.

The selection criteria for the Scottish SDR service have been established based on consideration of the best evidence and can be found on the Neurosurgery <u>MSN website</u>.

A) THE NATIONAL INSTITUTE FOR HEALTH AND CLINICAL EXCELLENCE (NICE)

NICE has provided guidance for SDR as a treatment option in the management of spasticity in children. SDR has undergone extensive review by NICE as part of a standalone evaluation, which was published in December 2010: <u>https://www.nice.org.uk/</u> <u>guidance/ipg373</u>

In this guidance, NICE acknowledged the increased evidence on efficacy and safety of the procedure, published since its last guidance in 2006. The guidance states that evidence on its efficacy is now adequate, and stated that 'healthcare professionals across the NHS can offer the procedure under their hospital's normal arrangements for consent, clinical governance and audit.

A wider review of the treatment of spasticity in children was published in July 2012 which was subsequently updated in November 2016: <u>https://www.nice.org.uk/guidance/cg145</u>

The NICE guidance strongly advises that SDR should be undertaken by a multidisciplinary team with specialist training and expertise in the care of spasticity in patients with cerebral palsy, and with access to the full range of treatment options and that long term outcomes should be audited in a standardised manner.

In view of this evidence and a period of time gathering evidence, the NHS England Specialised Services Clinical Reference Group for Paediatric Neurosciences produced an evidence based clinical commissioning policy in July 2018:

https://www.england.nhs.uk/publication/clinical-commissioning-policy-selective-dorsalrhizotomy-sdr-for-the-treatment-of-spasticity-in-cerebral-palsy-children-aged-3-9-years

B) EVIDENCE IN THE LITERATURE

A systematic review of interventions for children with cerebral palsy published by Novak in 2013 included selective dorsal rhizotomy as a "green" intervention⁸.

A meta-analysis of three randomised controlled trials which evaluated the effectiveness of SDR for spasticity in children published in the late 1990's pooled the baseline and 9 to 12 month outcome data. Results showed only a small functional benefit⁹.

However, to determine whether applying stricter criteria for patient selection led to improved outcomes, ambulant children with cerebral palsy were selected for SDR using very strict clinical criteria in Oswestry. It was concluded that application of strict selection criteria when considering children for SDR leads to encouraging results as demonstrated by gait analysis and other measures¹⁰.

Long Term Outcomes:

More recent publications have been reporting on long term outcomes post SDR surgery.

Daunter et al in 2017 reported on a cohort of patients an average of 22 years following SDR surgery, finding adults with CP who had SDR in childhood had less motor decline and fewer daily assistance needs suggesting the functional impact lasts long after surgery¹¹.

At 10 year follow up after SDR Ailon et al in 2015 noted that the functional benefits were best seen in patients in GMFCS II and III but were not sustained in GMFCS IV and V¹².

Ranges of motion for the hips and knees were within normal limits at ten years, and were maintained at a normal midrange point, confirming long-term gait improvements. Locomotor function was also shown to be improved 20 years post-operatively in a cohort of 13 children who underwent SDR for spastic diplegia in 1985. These children also demonstrated sustained improvements in their functional status, as measured on the International Classification of Functioning, Disability and Health model, at 20 years^{13,14}.

McFall reported in 2015 on the adolescent growth spurt in young people who had SDR all had maintained or improved their GMFCS level and their gait profile score during their adolescent growth spurt and this paper also highlighted the importance of careful patient selection and the importance of MDT approach to the post-op management of these children including orthoses, physiotherapy and orthopaedics¹⁵.

Other Effects:

Suprasegmental effects have also been reported after SDR.

Gigante found a reduction in upper limb tone with improved motor control and function in 92% of SDR cases¹⁶.

There have also been reported changes in improved block stacking, improved upper limb tone and better manipulation patterns¹⁷.

Orthopaedic considerations:

SDR has been noted to have a positive effect or no effect on hip joint subluxation.

SDR however does not lead to avoiding the need for further orthopaedic interventions, although children who become independent ambulators following SDR have a significantly reduced need for orthopaedic procedures^{18,19}.

Buckton et al reported differences within 25 children with spastic diplegia, of whom 18 underwent SDR and 7 underwent appropriate orthopaedic procedures, as decided by informed parents. Children undergoing SDR demonstrated higher improvements on the Gross Motor Function Measure (GMFM), as well as significantly better gains in self-care skills, and a decrease in care-giver assistance throughout the two year follow up period. The orthopaedic surgery group showed significant improvements in some aspects of function. There may be some children for whom orthopaedic surgery is a better option²⁰.

C) COMPLICATIONS AND SAFETY

SDR is an irreversible surgical operation and carries some risks. Although complications are rare they include:-

- Infection
- Leak of cerebrospinal fluid from the wound or development of a fluid collection below the skin
- Neurological injury: Injury to a motor nerve root or nerves innervating the sphincters could occur during surgery and would potentially result in severe leg weakness or incontinence. All nerves are checked carefully before division, however so that these are very rare complications.
- Spinal deformity: Thoracolumbar Scoliosis, thoracic kyphosis and lumbar lordosis in the long-term follow up of SDR cases has been reported in medical literature. This

is generally associated with the multi-level lumbosacral laminoplasty technique. This technique is not used in our centres^{21,22}.

- Other papers have reported mild scoliosis, hyperlordosis and spondylolisthesis although no clinically significant deficits were associated with these findings²³.
- There are also risks associated with general anaesthesia.

D). PATIENT INFORMATION AND CONSENT

Parents and families should have ample opportunity to explore and consider all appropriate tone management strategies in discussion with their local MDT. The aim of SDR is to reduce spasticity.

For those who are candidates for SDR surgery and wish to consider this option, discussion also takes place in neurosurgical clinic.

The MSN for Neurosurgery website contains information on SDR for patients and families that can be accessed and discussed with parents. <u>http://www.msn-neuro.scot.nhs.uk/</u> services/selective-dorsal-rhizotomy/

The expected benefits and outcome from the procedure, as well as issues related to post-operative pain and discomfort, rehabilitation and the need for regular intensive physiotherapy will also require discussion.

The standard consent form used for paediatric neurosurgical procedures is appropriate for use in SDR surgery.

3. SCOTTISH CLINICAL CRITERIA FOR REFERRAL FOR SELECTIVE DORSAL RHIZOTOMY

The clinical criteria for referral were developed in partnership with Paediatric Neurologists, Neurosurgeons, Orthopaedic Surgeons, Orthotists, Physiotherapists and the Allied Health Professional National Lead for Children and Young People at Scottish Government.

The criteria were developed to be used in discussion with families. The goal is to provide equitable access for SDR procedures that is based upon sound clinical decision making. Although developed for SDR it is envisaged that the criteria could be used for the management of tone using a variety of modalities. The pre-op and post-operative physiotherapy management should be discussed at the outset.

The MSN website hosts all relevant information required with regard to the referral criteria and the pathway:

https://www.msn-neuro.com/services/selective-dorsal-rhizotomy/docs/SDR_Pathway_ V1_0.pdf

Here is a brief summary of referral criteria:

- 1. Age: children should have a confidently established GMFCS level (usually between 5-10 years old).
- 2. The child should have capacity to engage and be compliant with physiotherapy and wearing of orthoses

- **3.** Family should understand and agree to a commitment in active participation in the rehabilitation programme following surgery.
- 4. Absence of significant concerns about weight management and obesity.
- 5. Ambulant Spastic diplegia Cerebral Palsy with clearly demonstrable spasticity (moderate to severe)
- 6. GMFCS II-III
- 7. No predominant motor pattern of dystonia / involuntary movements.
- 8. Movement control –isolate hip flexion and if possible knee extension and ankle dorsiflexion
- 9. Balance at least moderate.
- 10. Be reasonably strong particularly in knee extensors and hip flexors.
- **11.** Absence of significant joint contracture or abnormal torsional profile for which orthopaedic surgery was likely.
- 12. Absence of significant upper limb dysfunction

4. PATIENT PATHWAY IN SCOTLAND

The pathway is available on the MSN website:

https://www.msn-neuro.com/services/selective-dorsal-rhizotomy/docs/SDR_Pathway_ V1_0.pdf

All decision points assume full involvement and agreement with the child and their family or carers.

Only referrals made through specialist MDT, that meet the Scottish Clinical Criteria, <u>and</u> who have identified appropriate resource for community physiotherapy, will be accepted for

consideration.

A brief summary of the SDR patient pathway:

- 1. Local team discuss with families who express an interest in SDR and decide if clinical criteria are met or not
- 2. If criteria are met, local team ensure physiotherapy resource is available and organise specialist reviews, MR imaging and gait analysis
- 3. Following gait analysis and imaging, results are discussed at regional level with local team and decision made to refer to National SDR service or not
- 4. If agreed may be suitable for SDR surgery, referral is made to SDR coordinator
- 5. Child is discussed at National SDR MDT meeting which meets 3-monthly with relevant local/regional team members
- 6. If not accepted for surgery, recommendations will be given for ongoing care/ tone management with local team

- 7. If accepted for SDR surgery/ decision to proceed is made at National meeting, appointment is made to meet the neurosurgeon as an outpatient (Glasgow or Edinburgh Children's hospital- family preference)
- 8. Once date set for surgery, child has a pre-operative physiotherapy assessment and orthotic review
- 9. Admission for surgery
- 10. 3 weeks inpatient stay for intensive post-op rehabilitation
- **11.** Handover to local Team for ongoing rehabilitation (resource for additional intervention will have already been made) with recommendations
- 12. Follow up assessments: a. 3 months neurosurgeon review
 - b. 6 months physiotherapy assessment
- c. 1 year, 2 years. 5 and 10 years reassessment to review outcomes
- 5. REFERENCES
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Who can I contact for further information/advice?

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Further information is also available from:

Managed Service Network for Neurosurgery SDR webpage:

https://www.msn-neuro.com/services/selective-dorsal-rhizotomy/