Selective Dorsal Rhizotomy For Spastic Diplegic Cerebral Palsy

Background

1. Procedure

Selective dorsal rhizotomy (SDR) is a surgical procedure developed to reduce spasticity and improve mobility in patients with cerebral palsy (CP) and lower limb spasticity. It involves selective division of sensory rootlets in the lumbar spine, maintaining a balance between elimination of spasticity and preservation of function. SDR aims to reduce spasticity, improve function, and improve mobility, reducing dependence and possibly making dependent children independent.

Peacock described SDR performed either through a multilevel L1/2-S1 laminectomy or laminoplasty\(^{27,30}\). Other surgeons have refined the approach to a single level laminectomy at the position of the conus, as determined by intra-operative ultrasound\(^{27,30}\). Between 50 and 70% of the nerve roots are divided. Intra-operative neurophysiology is an integral part of the procedure and allows selective division of nerve rootlets.

2. Current status

SDR is a standard neurosurgical procedure for the treatment of spasticity associated with CP\(^{17}\). NICE issued new guidance for SDR on 15\(^{th}\) December 2010 (http://www.nice.org.uk/guidance/IPG373), stating that evidence of its efficacy is now adequate, and that “healthcare professionals across the NHS can offer the procedure under their hospital’s normal arrangements for consent, clinical governance and audit.” In North America, SDR is recognised and funded by all the major healthcare insurance providers.

3. Patient selection

Selection criteria were defined by Peacock in 1987, and still apply\(^{31}\). The Leeds referral criteria for SDR consideration are:

- a) Child aged 2+ with diagnosis of Spastic Diplegic Cerebral Palsy with
  - a. Dynamic spasticity in lower limbs affecting function & mobility and no dystonia
  - b) Preterm birth or full term with typical signs of spastic diplegia
  - c) MRI shows typical CP changes and no damage to key areas of brain controlling posture & coordination
  - d) GMFCS level: II–III – (but children on either side of these may be considered)
  - e) No evidence of genetic or progressive neurological illness
  - f) Mild to moderate lower limb weakness with ability to maintain antigravity postures.
  - g) No significant scoliosis or hip dislocation (Reimer’s index should be <40%)

Agreement by all involved carers on the goals of treatment for the individual child is crucial. The principal goals of SDR include improved motor function, increased mobility, increased independence and improvement in ease of care\(^{7}\).

Evidence of effectiveness

1. Randomised clinical trials

Three randomised controlled trials have evaluated the effectiveness of SDR for spasticity in children\(^{18,19,39,44}\). Patients with spastic diplegia were randomised between SDR plus Physiotherapy (PT) or PT alone. The individual trials showed improvements in GMFM, spasticity and range of movement were demonstrated in the group undergoing SDR + PT\(^{18,19,39,44}\). When the 3 trials were analysed together in a meta-analysis SDR was confirmed as superior with a 4-point improvement in GMFM score and 1.2 point improvement in Ashworth score at 12-months post-operatively\(^{36}\). None of the studies reported any adverse events. In addition, a direct linear relationship was demonstrated between the extent of dorsal roots cut and magnitude of gain in function\(^{18}\).

2. Prospective non-randomised patient series

Steinbok reviewed outcomes of large published patient series (350 patients) in 2001 and demonstrated a significant reduction in spasticity\(^{36}\). Two prospective studies (45 patients) evaluated the Paediatric Evaluation of Disability Inventory (PEDI) and identified improvements post-SDR in self-care and mobility domains. The Functional Independence Measure for Children was used in two further studies (122 cases); this showed improved motor scores, sphincter function and cognitive scores after SDR. Many prospective case series are reported, and demonstrate:

- Improved range of movement in lower limbs & improvement in formal gait analysis
- Improved qualitative assessment of sitting (70% of cases; 213 patients)
- Improved qualitative assessment of ambulation (50-78% of cases; 346 patients)

When compared with ITB, SDR showed significantly better improvements in Ashworth scale, lower extremity passive range of motion and GMFM scores\(^{13}\).

3. SDR and Orthopaedic Surgery

Orthopaedic interventions for Spastic CP include Derotational Osteotomy, Single Event Multi-Level Surgery (SEMLS), and tendon releases. It is known that it can take 1-2 years for patients to recover fully and gain the full benefit from SEMLS, but this treatment remains popular despite this. However, this does not reverse the spasticity and even if surgery might be beneficial in principle, the tendency for spasticity and its complications to progress over time might hide such benefits. Buckon et al reported a series of 25 children with spastic diplegia, 18 of whom had SDR and 7 of whom had orthopaedic procedures\(^{3}\). Over the 2-year follow-up, children undergoing SDR demonstrated higher improvements on the Gross Motor Function Measure (GMFM). Self-care skills, mobility, social function and independence gains were seen earlier and with greater frequency in the SDR group\(^{5}\).

A prospective, non-randomised series of 142 patients (71 SDR & 71 ITB) showed that those treated with SDR had a significantly reduced need for orthopaedic surgery compared to similar children undergoing ITB therapy\(^{13}\).
Without SDR, 20-28% patients develop hip dislocation or subluxation. After SDR, hip subluxation tends to stabilise in up to 80% of patients, and up to 40% of patients show improvement in the degree of hip subluxation post-operatively. A review of 158 children undergoing SDR aged 2-3 years, demonstrated a reduced long-term need for orthopaedic procedures. The same group, in their overall study of 178 children and young adults with CP, demonstrated a significant reduction in the need for orthopaedic surgery after SDR undertaken prior to four years of age, with 10-years’ follow-up. The requirements for orthopaedic surgery were associated with the degree of post-SDR mobility – 25% of independent walkers and 44% of assisted walkers required orthopaedic surgery over a nine-year follow up period. This represents a significant reduction from the rate of orthopaedic surgery in Watt’s series. Steinbok reports a reduction in orthopaedic surgery following SDR of at least 35% across all levels of mobility. It is probable that all patients who have had SDR would otherwise have needed at least one orthopaedic procedure before skeletal maturity had SDR not been done. A number of children require more than one orthopaedic correction operation during their lifetime – for example, Ounpuu reports a series of 20 patients undergoing 27 operations.

4. Long-term results
A number of studies have reported the long-term outcomes following SDR – after 5-years, 10 years, and 20 years. Significant and sustained improvements were reported in muscle tone, gait, in GMFM, and in PED scores. An additional study published in 2013 reported 5- and 10-year outcome study demonstrated improvements in GMFM-66 above what normal reference children would have expected. In this study, no children showed deterioration after SDR and 36% (5-year) and 30% (10-year) showed an improvement of more than 20 centiles.

5. Complications and safety
Permanent complications are rare after SDR. Transient dysesthesia is reported in 2.5-40% patients, and lasts up to a few weeks from surgery. Permanent numbness is rare. Transient urinary retention is more frequent, occurring in 1.25-24%, and permanent urinary incontinence is rare. Back pain, (lasting weeks-months), occurs in 4-7% patients. SDR performed through multilevel laminoplasties or laminectomies may increase the incidence of spinal deformity, particularly in children with severe quadriplegic spasticity. These children, however, are often treated with ITB, and a large patient series has shown that limited laminectomies limited to the level of the conus are not associated with significant long-term spinal deformity.

Cost-effectiveness
There are no economic evaluations of SDR alone. There is no current economic evaluation for SDR. Absence of evidence is not of course the same as lack of proof of economic benefit. Indeed there are many interventions in childhood disability which are accepted a standard practice in which there has been no economic evaluation – eg many physiotherapy interventions. There are an increasing number of good quality studies showing long-term clinical benefit of SDR and using appropriate outcome measures. Recent studies use the WHO framework International Classification of functioning, activity and participation (ICF) for outcome assessment – now internationally accepted as Gold Standard. As evaluation of SDR has developed, an increasing number of methodologically sound studies are using the ICF domains and showing positive benefits for SDR.

A number of studies have evaluated cost effectiveness of ITB therapy for severe spasticity. In an attempt to put an economic value onto SDR, one study from Canada has compared the cost-effectiveness of SDR with that of ITB. They concluded that SDR was cheaper to deliver than ITB and thus inferred that SDR was cost-effective.

A matched comparison of SDR and four-monthly botulinum toxin injections in 40 diplegic children showed that improvements in spasticity and gait became insignificant 12-months after commencement of injections; in contrast, those undergoing SDR improved continuously during the 20-month follow up period. Children at GMFCS III-IV accumulate progressive lower limb skeletal, muscular and joint deformities before reaching skeletal maturity. This frequently necessitates multi-level orthopaedic surgery, which often includes soft tissue release surgery in combination with femoral osteotomies and hip reconstruction. Watt et al prospectively studied 74 children with spastic CP and found that 61% had already undergone orthopaedic operations by 8 years-of-age. Excluding gait lab analysis and post-operative rehabilitation, the hospital NHS tariff for femoral osteotomy and internal fixation is up to £9061; that for femoral head relocation and derotation osteotomy is up to £16825. Although these procedures improve the cosmesis of gait, they do not address spasticity, the primary cause of deformity and contracture. LTHHT has performed a cost-benefit-savings analysis based on the current literature and current NHS costs. This demonstrated the potential for significant savings over each patient’s lifetime. The published evidence suggests that undertaking early SDR improves the cost effectiveness of spasticity management. It can also be argued that SDR is analogous to surgery for intractable epilepsy, where early treatment of the pathological process then allows the child’s development to proceed as normally as possible.

Conclusions
1. SDR is safe, and has been approved by NICE for delivery in the UK.
2. Published evidence confirms that SDR provides long-term benefits to the patient.
3. SDR is the only currently available treatment that can permanently reduce or remove spasticity in the legs of suitable children with spastic diplegic CP.
4. Published evidence confirms that SDR can reduce the need for subsequent orthopaedic surgery (and hence both improve the quality of life for the patient and save money for the NHS).
5. SDR appears cost-effective on the basis of the current data.
References


