Selective Dorsal Rhizotomy in Spastic Cerebral Palsy

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Introduction

Cerebral palsy (CP) is a neurological disorder encompassing a group of motor conditions that cause physical disability in a developing child. It is caused by a non-progressive damage to the motor control centres of the developing brain and can occur during pregnancy, during labour or after birth up to the age of three.

The incidence of CP is around 3 per 1,000 live births. In a local study, the overall prevalence was 1.3 per 1,000 children. Spastic CP is classified according to the region of the body affected. Spastic diplegia is by far the commonest type of cerebral palsy, accounting for 70% to 80% of all cases. It involves the lower extremities only. In spastic quadriplegia, all four limbs are involved.

Spasticity is defined as a movement disorder characterised by a velocity-dependent increase in tonic stretch reflexes. Children with spastic CP have a neuromuscular mobility impairment stemming from an upper motor neuron lesion in the brain over the areas in the motor cortex or the corticospinal tract, which descends in the spinal cord, in the lateral columns and carries signals for voluntary movement of skeletal muscles. The damage impairs the ability of nerve receptors in the spine to properly receive gamma amino butyric acid, leading to hypertonia in the muscles innervated by these damaged tracts.

In spastic CP, patients also have other signs and symptoms of upper motor neuron syndrome like muscle weakness, paucity of active movement, inability to perform fine movement, brisk tendon jerk reflexes and positive Babinski sign. Consequently, their muscles have reduced dexterity, weakness and fatigability, which will translate into deficient daily activities. Deformities of the bones and joints of the limbs are common after prolonged muscle imbalance and spasticity.

Current treatments for spasticity include oral muscle relaxant medications, physiotherapy, orthotic devices, and repeated intramuscular injections of botulinum toxin. Orthopaedic procedures include tenotomy, tendon lengthening, tendon transfer and osteotomy. Neurosurgical procedures are peripheral neurotomy, selective dorsal rhizotomy and continuous intrathecal baclofen infusion. The spectrum of therapeutic interventions ranges from diffuse to focal, temporary to permanent. A multidisciplinary team approach is needed because these patients often require different treatment modalities at different ages and stages of the disease. In general, stretching exercise, splints and physiotherapy are prescribed for patients at age of 2-3. Botulinum toxin injections, casting and orthosis would be added at age of 3 to 5. A permanent tone reduction procedure like SDR would be performed when these patients show static gross motor dysfunction. SDR is usually performed at the age of 6-7. Soft tissue surgery aiming at fine-tuning joint range and alignment will be done one year after SDR. Bone surgery will be reserved until skeletal maturity is reached.

Selective Dorsal Rhizotomy Clinic

Selective dorsal rhizotomy is a neurosurgical procedure performed in patients suffering from spastic cerebral palsy. The selective de-afferentation of sensory nerve rootlets from L1 to S2 results in a reduction of contracting stimuli to muscles, decreasing spasticity, and thereby improves motor function. A number of case series suggested that selective dorsal rhizotomy reduces spasticity substantially, improves ambulatory function, and involves no unacceptable short-term risk when performed by experienced multidisciplinary teams.

In 1997, the Department of Neurosurgery established the Selective Dorsal Rhizotomy (SDR) Clinic in Tuen Mun Hospital. The team members include physiotherapists from the hospital, special schools and Child Assessment Service of the Department of Health, paediatric neurologists and developmental paediatricians, orthopaedic surgeons, neurosurgeons and urologists. The clinic provides a platform for multidisciplinary approach to patients with spastic CP and their relatives. The clinic provides screening and assessment to select potential surgical candidates for SDR. Setting a treatment plan with realistic therapeutic goal for each individual patient follows. We also provide them with pre-operative training programme and peri-operative rehabilitation protocol. The families have to understand and be committed to the important preoperative physiotherapy training which will last for 2-3 months before a decision on SDR is made. The clinic will follow up patients to monitor their progress after surgery and formulate subsequent treatment plan.

Patient Selection

The selection criteria for Spastic diplegic patients for SDR include:

1. Spastic - Spasticity of the lower limbs interfering normal functions, disturbing fluidity of gait or movement.
2. Strong - Fair to good lower limb muscle strength and control
3. Straight - Fair to good trunk control with no fixed orthopaedic deformity
4. Slim – Not too heavy or obese
5. Smart - Normal to near normal intelligence
6. Social support - A supporting and motivated family

The selection criteria for Spastic quadriplegic patients for SDR include:
1. Significant lower limb spasticity interfering with positioning and care
2. No severe dystonia
3. No fixed contracture at multiple joints

The decision-making on patient selection has to be individualised, as most of the SDR candidates will not meet all criteria. All team members play a part in the decision making process. Unsuitable candidates will be discharged with a rehabilitation plan. They will be reassessed in nine to twelve months time before coming to a final verdict.

SDR is a functional neurosurgical procedure with the purpose of improving lower limb function. In order to monitor and document the clinical progress, we have adopted a series of tools and examinations. Main outcome measures include Modified Ashworth Scale, passive range of joint movement, the Gross Motor Function Measure, the Paediatric Evaluation of Disability Inventory, the Canadian Occupational Performance Measure, urodynamic study, oxygen consumption and three-dimensional gait analysis. We schedule the test immediately before SDR, six months, twelve months and up to six years after SDR.

The Procedure

We performed the first SDR procedure in December 1996. We modified the procedure and used multiple level laminoplasty (L2-S1) for dura exposure after Dr. Peacock’s surgical demonstration during the neurosurgical commissioned training in 1997 (figure 1). In October 2006, we used a less invasive single level laminectomy at the level of conus medullaris for SDR (figure 2). After laminectomy, the surgeon will open the dura and arachnoid at the midline. The dorsal roots will be separated from the anterior motor roots by using patties. The dorsal root will be further subdivided into 4-5 rootlets and their clinical response to electrical stimulation tested. Abnormal rootlets will be cut. The procedure will be performed on L1 to S1 dorsal roots on both sides.

The term “selective” is used because we rely on the Intra-operative Trigger Electromyography (EMG) response to find the rootlets causing abnormal muscle spasms before cutting them. We insert paired needle electrodes into the deltoids, hip adductors, vastus medialis, hamstrings, gastrocnemius and the external anal sphincter for EMG monitoring during SDR. A pair of active electrodes is used to stimulate individual rootlet during surgery.

The EMG criteria for selecting an abnormal nerve rootlet include:
1. Low stimulation threshold
2. A tetanic or polyphasic to a tetanic stimulation of a 50Hz chain lasting for one second
3. A spread of EMG response to the contralateral side (figure 3)

Besides EMG, we also use the on-table clinical response during dorsal rootlet stimulation as selection criteria. It has been observed that according to the segmental innervation of the lower limb, the higher the muscle tone, the more extensive the rhizotomy would be. In general, 40-60% of the rootlets would be cut during SDR. In the first two postoperative days, the patient is nursed on lateral position and turned every 2 hours. A Foley’s catheter is removed on day 3. The patient is mobilised on day 4 and both patient and parents will participate in intensive physiotherapy programme later on as outpatients in the following two months.

Figure 1. A. Skin incision for multiple level laminoplasty. B. Dorsal rootlets at the S1 level were exposed after multiple level laminoplasty. The paired electrodes were used to apply electrical stimulation during surgery.

Figure 2. A. Skin incision for single level approach targeting at the conus medullaris. B. The conus medullaris was exposed and SDR was performed in this 2 cm dura opening under operating microscope.
Clinical Outcomes

We have performed around eighty SDR procedures since 1996. 80% of the surgical candidates belong to the spastic diplegic group and 10% belong to the spastic quadriplegic group. 75% of them had surgery done before the age of nine. There was no major surgical complication. Wound pain and postoperative low-grade fever were fairly common, usually lasting a few days and improved after acetaminophen. There was no wound infection, cerebral spinal fluid leak or pseudomeningocele after surgery. A small proportion of them developed dysuria after surgery but no urinary tract infection was found. Around 20% of the SDR patients complained of some degree of lower limb numbness that usually resolved after a few days to weeks.

There was no long-term complication such as spinal deformity. The progression of hip joint pathology was in accordance with their initial status according to the Gross Motor Function Classification System (GMFCS) and was not related to surgery itself. There was no deterioration of sphincter function seen. We observed that the reduction of lower limb muscle tone was long lasting. The patients have no recurrence of lower limb spasticity after surgery for a mean follow up of 6 years. We found that these patients experienced substantial reduction in spasticity after SDR, as documented by a marked reduction in Modified Ashworth Score of the lower limbs when the baseline Modified Ashworth Score was compared with findings 12 months after SDR. There was significant improvement in combined hip abduction range with the knee in extended position (R2) and in selective control scoring. The patients exhibited significant improvement in Gross Motor Function Measure total score and in dimensional scores in crawling, kneeling, walking, running, and jumping after selective dorsal rhizotomy plus physiotherapy. Improvements in walking were also reflected by significant improvements in Observational Gait Scores. Changes and improvement in instrumental gait analysis and oxygen consumption were also observed**. Patients usually developed transient lower limb weakness after SDR. The weakness might last for 4-8 weeks and the GMFM also dropped during this period of time. In all patients, the weakness recovered after physiotherapy and training. Most of them showed improvement in GMFM after two to three months of intensive physical therapy.

Our orthopaedic colleagues will look into the musculoskeletal issues of spastic CP patients. For post SDR patients, the orthopaedic surgeons would decide on the appropriate intervention such as iliopsoas release, heel cord release, derotational osteotomy, foot/ankle stabilisation procedures, and muscle transfer procedures at around a year after SDR. We believe musculoskeletal surgery and SDR are complementary procedures with additional benefits over ambulatory function and will enhance the quality of life of our patients. It has been shown that SDR performed in patients at a young age – 2 to 4 years old - can prevent subsequent soft tissues and joint deformity and reduces the need of future orthopaedic intervention.

Around 50% of our surgical candidates have abnormal urodynamic study such as detrusor instability and hyper-reflexic bladder before SDR. None reported any deteriorating urinary symptoms and signs after SDR. Fifty percent of patients with detrusor instability, as confirmed by a pre-SDR urodynamic study, demonstrated improvements in their urinary symptoms and signs after SDR. This finding has been confirmed in some patients with repeated urodynamic studies.

Conclusion

We conclude that selective dorsal rhizotomy, together with intensive postoperative physiotherapy improve the lower limb muscle tone, range of motion across joints and leads to subsequent improvements of lower limb ambulatory function. SDR is a safe procedure and operation-related complications are uncommon. A multidisciplinary team capable of providing the whole spectrum of spasticity treatments is mandatory for quality management and care of patients suffering from spastic cerebral palsy.

References