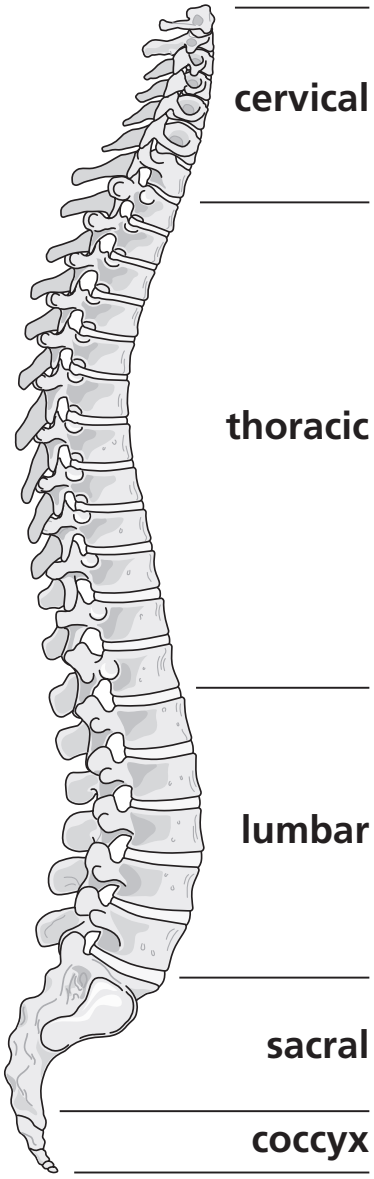
Selective dorsal rhizotomy

This information sheet from Great Ormond Street Hospital (GOSH) explains about the selective dorsal rhizotomy operation for children and young people with cerebral palsy. It details what the operation involves and what to expect when your child comes to GOSH for treatment.



What is selective dorsal rhizotomy?

Selective dorsal rhizotomy (SDR) is an operation used to improve spasticity (muscle stiffness) in cerebral palsy.

Cerebral palsy occurs when a child sustains a brain injury early in life. This most often happens before birth but can happen around the time of birth and even in the first year of life. Although the brain injury is static, that is, it does not get any worse, the difficulties it causes change continuously in the growing child.

One of the commonest causes of cerebral palsy is prematurity. Certain parts of the premature brain are vulnerable to damage, particularly the parts which control leg movement and coordination. In turn, this leads to excessive stiffness or spasticity in the leg muscles and can impair the child's ability to learn to walk. Spasticity also causes pain, and, over time, shortening of muscles and tendons, joint contractures and bone deformities.

Spastic diplegia (which affects the leg muscles more than the arms) is the most commonly occurring type of cerebral palsy. Nerve fibres running from the muscles back to the spinal cord play a major role in maintaining this muscle stiffness. Selective dorsal rhizotomy (SDR), by dividing some of these fibres, is very effective at reducing stiffness and spasticity.

Is SDR suitable for everyone?

Children between three and 12 years of age with typical spastic diplegia may be good candidates for SDR. Children older than twelve years may still be good candidates but are more likely to also need orthopaedic surgery to correct muscle and bone changes.

Children suitable for SDR need to demonstrate adequate muscle strength in the legs and trunk. They are usually able to stand up and support their body, hold their posture against gravity, and make appropriate movements to crawl or walk. These children tend to have delayed physical development, and spasticity interferes with their progress.

Before considering your child for this operation, we will need to make sure that a magnetic resonance imaging (MRI) scan of the brain shows the changes characteristic of spastic diplegia – periventricular leukomalacia. If other areas of the brain involved in balance and coordination are also affected, the child may not be a good candidate for SDR. In addition, we will need to have x-rays taken of your child's hips to confirm that they are stable.

Many children will have already had other procedures for spasticity. It is recommended to wait at least three months from the last Botulinum toxin injection and at least six months from orthopaedic surgery before considering SDR. Regular post-operative physiotherapy is necessary to obtain the best results after SDR so children need to be motivated and show that they are able to cooperate with therapy. We would need to make sure that a definite plan for post-operative physiotherapy is in place before SDR is considered.

Children whose cerebral palsy is related to prematurity tend to have a good response to SDR. This is not the case for all causes of cerebral palsy. Children who have a history of meningitis or hydrocephalus unrelated to prematurity, as well as those with congenital infection or head trauma do not do well with SDR.

Similarly, children with severe muscle rigidity, poor muscle tone or dystonia do not benefit from SDR. It is also not effective for children with severe cerebral palsy involving the whole body. In children with severe scoliosis, SDR is not recommended as it may cause the existing spinal curvature to deteriorate.

Confirming SDR is suitable for your child

Once we receive a referral from your paediatrician, orthopaedic surgeon or paediatric neurologist, we will organise an appointment in our movement disorder clinic. Your child will be seen by a consultant in paediatric neurodisability, a specialist paediatric physiotherapist, and a paediatric neurosurgeon with SDR experience and a full assessment will be carried out. Previous investigations, such as hip x-rays and MRI scans, will be reviewed.

At the end of the assessment the team will discuss with you their views regarding your child's suitability for SDR. It is also important for us to understand what you want SDR to achieve and to discuss treatment goals and expectations.

The risks and benefits of the procedure and hospital stay are discussed. The need for a post-operative physiotherapy programme is emphasised. Currently SDR is still not routinely available on the NHS and a request for individual funding may have to be made.

SDR is a definitive procedure and cannot be reversed. We need to be sure that it is the best option for your child at that particular point in their development before recommending it. Sometimes we may advise botulinum toxin injections before deciding that SDR is suitable. This would give us an indication of how your child will respond once some of the spasticity is taken away. We may also recommend a defined period of targeted physiotherapy to increase strength in their body and legs if it is felt that they are still too weak for a final decision to be taken.

Are there any alternatives to SDR?

Early SDR is one option in the management of spastic diplegia in children with cerebral palsy. It is the only procedure to permanently remove the spasticity in the legs, which is the primary cause of disability and long-term deformities. There has, in the past, been some anxiety about the irreversible nature of this procedure. However, its effectiveness at reducing spasticity and improving quality of life has now been clearly demonstrated in several research studies.

Alternatives to SDR in spastic diplegia include long-term physiotherapy alone, use of botulinum toxin injections into the spastic muscles, and multi-level orthopaedic procedures. The latter do not alter the background spasticity and may need to be repeated as the child grows and deformities progress – they may still be required after successful SDR to correct established deformities. Botulinum toxin injections need to be repeated frequently and become less effective over time.

Oral baclofen results in some improvement in spasticity, but doses high enough to do so often cause drowsiness and interfere with the child's ability to learn and concentrate at school. Baclofen given into the spine (intrathecal baclofen therapy) is another possible option. This requires a commitment to regular pump refills and exposes children to the risks of baclofen overdose and withdrawal as well as pump problems. This is generally reserved for patients with severe whole body involvement.

However, not every child with cerebral palsy requires intervention, and many children are able to lead full and happy lives with physiotherapy alone.

Pre-operative assessment

A full pre-operative assessment will be carried out a few weeks before SDR. Unless already done, this will also include formal walking assessment. This assessment serves as a baseline for subsequent follow up reviews after surgery. Your child will be seen by our orthotist who will make plans for their orthotic (splints) needs after SDR. They will have a general medical review to identify any potential difficulties with anaesthesia.

What happens before the operation?

You will be asked to come to Starfish ward at GOSH on the day before the surgery. Your child's surgeon will visit you to explain about the operation in more detail, discuss any worries you might have and ask you to give your permission for the operation by signing a consent form.

It is important that your child does not eat or drink anything for a few hours before the anaesthetic. This is called 'fasting' or 'nil by mouth'. Fasting reduces the risk of stomach contents entering the lungs during and after the procedure.

You will be informed the night before the procedure of the time that your child should be 'nil by mouth' – in other words, have nothing to eat or drink before the anaesthetic. Fasting times are provided in your admissions letter – in broad terms, this is six hours for food (including milk), four hours for breast feeding and two hours for clear fluids before the procedure.

It is equally important to keep giving your child food and drink until those times to ensure they remain well-hydrated and get adequate nutrition. This may involve waking your child in the night to give them a drink which we recommend.

Your child's will be admitted to Puffin Ward on the day of surgery, where you will meet your anaesthetic team, before accompanying your child to the anaesthetic room in the operating theatre.

What does the operation involve?

SDR is carried out while your child is under general anaesthesia and takes around four to five hours. A skin incision is made in the upper lumbar spine. The spinal canal is opened at only one level. An ultrasound probe is used to identify the lower end of the spinal cord. Under the operating microscope, the membrane covering the spinal cord is opened and the lower end of the cord, with the sensory roots entering it, is identified.

Each of the sensory nerve roots is then subdivided into four or five rootlets. Each rootlet is stimulated to identify the ones that contribute most to the spasticity – these rootlets are then divided. The process is repeated for all the other nerve roots, from L1 to S2, on both sides, aiming to divide 50 to 70 per cent of the sensory roots.

At the end of the procedure, the membrane covering the spinal cord is closed again, the back muscles are returned to their original position and the skin is closed with dissolvable stitches.

Are there any risks?

Complications after SDR are rare but you need to be aware of them. Complications include infection, leak of cerebrospinal fluid from the wound, development of a fluid collection below the skin, severe leg weakness and incontinence. As all the nerve roots are carefully checked by stimulating them during the operation and monitoring their response, severe weakness and incontinence are very rare complications. In addition, there are risks

associated with general anaesthesia, but these, as well as the risk of long-term spinal deformity, are very rare.

What happens after the operation?

After surgery, you will be called to the recovery room to be with your child as they wake up. Once awake and stable, your child will be returned to Sky ward. Close monitoring throughout the first 24 hours ensures good pain control, usually using patient controlled analgesia (PCA) given into a vein (intravenously or IV). Your child will be encouraged to lie on their back, but will be helped to turn from side to side every four to six hours. While your child remains in bed, they will have a bladder catheter in place draining off urine into a collecting bag. It is normal for children to complain of mild headache at this stage. Some children will experience uncomfortable leg spasms in the first few days after surgery, but this can be managed with muscle relaxants.

Over the next day, the PCA is reduced slowly. Gentle physiotherapy in the bed is started on day three and the bladder catheter is removed on day three. It is usual for children to have some numbness in their legs in the first week. Your child's legs will be less stiff than before surgery, but may also be significantly weaker at this stage.

Your child will be encouraged to start sitting out of bed on day three. Physiotherapy is then gradually increased, paying particular attention to maintaining good trunk balance and range of movement in the lower legs. Muscle strengthening exercises are begun. On day five or six, we would expect that your child's discomfort will be easily controlled by simple pain relief given by mouth. At this stage, your child is discharged to our patient hotel, located just outside the main hospital entrance.

A two-week programme of physiotherapy then begins, with two daily one-hour sessions in our gym. The aim of this programme is to continue to develop strength in the legs, trunk and pelvis, increase range of movement in the legs, develop better leg movement, and to develop and improve walking. All this takes time, and will be continued after hospital discharge through your local physiotherapy service.

Most children will require physiotherapy sessions at least three times a week for the first six months. Parents will have the opportunity to learn how to participate in the rehabilitation process and will be given ideas for starting their home programme which can then be adjusted by their home physiotherapist. The physiotherapists at GOSH will be in contact with your child's local physiotherapist and there will be opportunity to discuss any concerns as they arise.

Follow up

After discharge, we will see your child again at three, six and 12 months. Assessments, similar to the baseline review, will be carried out so that their progress can be closely monitored. We will also plan to see you on an annual basis after that. Formal walking assessment will be repeated at two years. You will be seen directly by our

paediatric orthopaedic surgeon if any concerns with alignment arise during follow up. Progress and the team's recommendations will be discussed regularly with your local physiotherapists.

What is the outlook for children who have had SDR?

SDR is not a cure for cerebral palsy. Reduction in spasticity is immediately apparent after the procedure, but SDR unmasks any weakness and difficulties with co-ordinating movements common in cerebral palsy. It takes time for the strength in the legs to return. Through the physiotherapy programme, your child will learn to use their body in a new way. Leg movement will become easier and the level of control, dexterity, range and speed will increase over time, although it may take up to two years for the full benefit of the procedure to become clear. Many children develop hypersensitivity in the soles of their feet after surgery – this is temporary and will improve. There may also be short term bladder disturbance, which may cause a change in toilet habits and could be frustrating for both you and your child.

There is now enough evidence to demonstrate that SDR is associated with substantial long-term benefits. These are not only related to reduction in spasticity, but also relate to improved movement and walking as well as improved quality of life for both the children and their families. One research study has shown that the benefits of SDR one year after surgery were still there twenty years afterwards. A recent large study has shown that children undergoing SDR at a young age continue to improve walking in childhood, without the expected decline in adolescence. Although SDR may not rule out the need for orthopaedic surgery later, a reduction in the need for these procedures after early SDR has been shown.

Further information and support

If you have any questions about the SDR operation, please contact the SDR Pathway Coordinator on 020 7405 9200 extension 1532 who will pass your queries on to the correct member of the team.

For further information about cerebral palsy, contact SCOPE – the main organisation in the UK supporting anyone affected by cerebral palsy and other disabilities. Call their helpline, SCOPE Response on 0808 800 3333 or visit their website at www.scope.org.uk

The latest guidance from NICE on the management of spasticity in children and young people, from July 2012, may be found at pathways.nice.org.uk/pathways/spasticity-in-children-and-young-people/spasticity-in-children-and-young-people-overview.

Adapted from an original leaflet produced by North Bristol NHS Trust by the Selective Dorsal Rhizotomy Service in collaboration with the Child and Family Information Group at Great Ormond Street Hospital

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