

Selective Dorsal Rhizotomy Clinic Report

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| Date of Assessment: 08.10.2018 | Date of Report: 12.11.2018 |
| Patient's Name: Borys Bartosiak | Date of Birth: 24.06.2013 |
| Age at Assessment: 5 years 3 months | |
| Home Address: ul. Pitsudskiego 38 A, 05-410 Jozefow, Poland | |

It was a pleasure to see Borys and his parents in our multidisciplinary selective dorsal rhizotomy (SDR) clinic at the Portland Hospital on 8th October. Borys' parents were keen to see whether SDR would be beneficial for him.

Medical Background

Borys is currently five years old. During pregnancy, there was some bleeding around the 18th week; this lasted about two to three days, and his mother was hospitalised for one month. Mother was identified as having an infection and given antibiotics. The rest of the pregnancy was unremarkable. He was born at term in good condition by an elective caesarean section due to breech presentation. Apart from torticollis, which appears to have resolved early, there were no issues during the neonatal period.

Borys was diagnosed with cerebral palsy when he was one year old. Although he began to roll at around four to six months, there was a significant delay in his subsequent motor milestones. At one year he started Vojta therapy when he was still not sitting nor crawling. He soon started to commando crawl, and within a short time was able to high kneel and four-point crawl. By 18 months he started cruising with furniture and at two and a half years began to use a Kaye walker. He took his first independent steps (with KAFOs) one year ago.

Currently Borys is able to walk with tripod sticks, up to a distance of around 500m. He can also walk independently, in a protected environment, over thirty to forty steps in his orthoses. He falls frequently and tends to be on tiptoes, not being able to straighten his legs. Climbing stairs is still difficult, although he can manage holding on to one hand. On the floor he prefers to sit in a W position, although his parents try to discourage this. His parents feel that his motor ability is currently stable, and he is not making much progress.

Borys' lower limbs are stiff, and his right is stiffer than the left. He does not complain of leg pain. His upper limbs are affected to a much lesser extent, and his dexterity continues to improve. He is right handed.

Borys is continent. He is on no regular medication and there are no other medical problems. In particular, he is doing well at pre-school and his parents feel that his learning and language are average for his age. There are no concerns with his vision and his hearing is good. He has never been

on medication for tone management.

Interventions

Borys has been having regular Vojta therapy since he was one. He has up to two hours of physiotherapy every day, in addition to daily stretching at home. He has recently also started 'Lokomat' therapy. He is an active boy, and enjoys riding his tricycle.

Borys underwent a course of botulinum toxin injections when he was two years old. This was not felt to have been successful. He went on to have a further course in July 2017 in Germany, when toxin was administered to his adductors and psoas muscles bilaterally. This led to a very significant improvement, which lasted around four months. It was around this time that Borys started to take independent steps. His third course was in January 2018. This also led to some functional improvement, which however only lasted around one month. We noted that following this course lengthening of his adductors and hamstrings has been recommended.

Neuroimaging and Radiography

Borys underwent an MRI scan of the brain and spine, and we had an opportunity to review this in clinic. The brain scan shows only minimal periventricular leucomalacia, involving the occipital poles of the lateral ventricles. The rest of the brain appeared normal. The MRI scan of the spine was unremarkable, with the conus lying at L1.

Regular hip x-rays have shown some progressive migration of the right hip, with a Reimer index of 20%; the left is well-located.

Current Mobility and Posture

Borys uses AFO's for up to 18 hours a day. During some of this time, his orthoses are used as KAFO's, with the addition of a detachable hinged component. He tolerates them well. Parents estimate that Borys can walk up to 500m in his KAFOs with sticks (which stop him from falling forward), and that he can now walk 50-100m unaided – walking is his preferred mode of getting around inside the house. He can go up 3 steps with one hand held, otherwise pulling himself up sideways using the bannister or crawling up. He does not descend stairs independently. He rides a tricycle with foot straps achieving good speed. Parents note that Borys' left hand is worse than his right.

Examination

Muscle Tone:

Brisk reflexes were elicited in both lower limbs, including crossed adductor reflex. Reflexes were less brisk in the upper limbs. Up going plantar reflexes elicited bilaterally, with mild unsustained clonus in the right ankle.

Range of Movement

| | Range-Right (R1/R2) | Range-Left (R1/R2) |
|------------------------------|---------------------|--------------------|
| Hip | | |
| Flexion | Full | Full |
| Extension | - 10 | 0 |
| Adduction | Full | Full |
| Abduction (hips flexed) | 15 / 45 | 20 / 55 |
| Abduction (hips extended) | 5 / 10 | 10 / 25 |
| Internal Rotation | 80 | 70 |
| External Rotation | 45 | 45 |
| Knee | | |
| Flexion | Not tested | Not tested |
| Extension | 0 | 0 |
| Popliteal Angle | 75 / 60 | 70 / 45 |
| Ankle | | |
| Dorsiflexion (knee flexed) | 20 / 30 | 15 / 30 |
| Dorsiflexion (knee extended) | - 5 / 10 | - 5 to - 10 / 15 |
| Plantarflexion | Full | Full |

Duncan Ely test was positive on both sides with a value of 40 degrees. There were mild fixed flexion contractures at the hips, right more than left, but none at the knees. Full passive dorsiflexion could be achieved at the ankles.

Strength and Selectivity:

Formal assessment of strength via manual muscle testing was not possible in clinic. On observation, the right lower limb is weaker than the left, with proximal hip muscles and knee extensors muscles showing moderate weakness.

Patterns of Movement:

Sitting: In sitting he was mildly kyphotic but there was no scoliosis.

Standing: Borys tends to hold onto a supporting surface when standing still and may lean against the supporting surface with his abdomen or weight bear on one hand to free the other hand to play. He stands with both hips and knees in moderate flexion (right knee more flexed than left), with a midfoot break and mild equinus bilaterally. Both hips are in mild internal rotation, the right more than the left.

Gait: Borys had a classic spastic diplegic gait, with hip and knee flexion, internal rotation at the hips, and equinus at the ankles when out of orthoses, his hands tending to splay during walking. He had

marked truncal lean to the right. He demonstrated a reduced stride length on his left compared to the right possibly due to reduced stability during stance phase on the right leg.

When walking, wearing his KAFOS and tripod sticks, heel strike is achieved on initial contact on both sides. During midstance, there is marked knee flexion on either side. By initiating forward truncal lean with weight taken through the tripod sticks in front, he is able to bring his centre of gravity over his base of support during terminal stance to help to straighten his hip and knee. He then demonstrates trunk overextension so as to clear his foot and enable hip and knee flexion for swing phase on the opposite side.

When walking, wearing his ankle foot orthoses alone and tripod sticks, foot flat is achieved at initial contact on both sides and his hips and knees are in marked flexion during midstance, right more than left. Forward truncal lean and weight bearing through the tripod sticks is required to assist in hip and knee movement into extension at terminal stance. There is marked internal rotation of the right hip throughout stance phase. He relies on momentum to propel him forward and trunk overextension to clear either foot and enable swing phase.

Opinion and Recommendations

Borys has spasticity throughout his lower limbs, involving all the muscle groups. He has made good progress over the last couple of years, and is now able to walk independently over short distances indoors. Outdoors he is able to walk using tripod sticks. He tends to walk in equinus, with stiff knees and hip flexion. His deep tendon reflexes are brisk and sustained clonus was identified in his left ankle.

We felt that Borys would benefit from SDR. The procedure reduces lower limb spasticity permanently. His spasticity is clearly an obstacle to further motor progress at the moment and we would expect that after SDR he would be able to increase his independence and stamina. We would also expect that his balance will improve, and his falls when walking independently will reduce. We cautioned that he will be weaker after SDR, and it would be beneficial for him to focus on straightening therapy for a few months. If the family wishes to proceed, we would then schedule surgery for early next year.

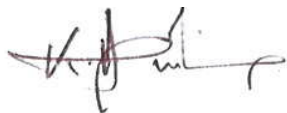
We also discussed potential risks of surgery, which would include infection, bleeding, and leakage of spinal fluid from the wound. These are all very rare. In the single-level procedure, the risk of destabilizing the spine is very low; in addition we find that children recover quickly and are able to resume their physiotherapy at around three days after surgery. We monitor all the lower limb muscle groups, as well as the sphincters, during surgery, and the risk of incontinence is very low.

In our service, the child is on the ward for six days. Our package then includes a further two weeks of out-patient intensive physiotherapy, at two sessions a day.

Borys' parents will be thinking about this and will get back to us once they have made a decision.

With many thanks and best regards,

Yours sincerely,



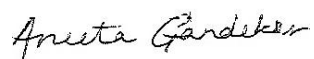
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