Spinal dorsal rhizotomy plus concurrent left and right gastrocnemii releases in a 7-year-old child with haemophilia A and spastic cerebral palsy

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Cerebral palsy (CP) is a permanent disorder affecting the development of movement and posture, caused by non-progressive damage to the developing fetal or infant brain [1]. Spastic CP is one of the most common sub-types affecting more than 50% of these children [1]. Spasticity, often results in joint contractures and bony deformities. These impairments have an impact on a child's function and mobility; activity level and participation in daily life. The current management of spasticity includes: physiotherapy, medical and surgical interventions, such as, baclofen, botulinum toxin, orthopaedic surgery and spinal dorsal rhizotomy (SDR) [2].

SDR is a neurosurgical procedure, which is used to eliminate spasticity in the lower limbs in children with CP. It is an irreversible procedure where 50-70% of the sensory L1-S1 nerve roots are cut permanently at the spinal level [3]. The procedure is followed by a period of intensive physiotherapy. It is assumed that reduction in spasticity will lead to improvements in gross motor function, activity levels, participation and their quality of life.

Several studies have confirmed the long-term effectiveness of SDR in eliminating spasticity [3] and the literature suggests that the improvements gained post-SDR are maintained 10-20 years after the surgery in >50% of patients [4-5].

Children with CP often have other associated impairments of sensation, perception, communication, cognition and behavior. Autistic-spectrum disorders (ASD) are diagnosed by the presence of social and communication difficulties, alongside unusually strong, narrow interests and/or repetitive and stereotyped behaviour.

This paper is a review of a child with severe haemophilia A, CP and ASD who underwent SDR in 2014 and his outcomes following the procedure.

For the purposes of this case study our case is identified as David; which is not his real name. Written consent was obtained from the child's mother to report this case. David was born at 32 weeks gestation in 2007. David received Factor VIII concentrate for a hernia repair at 6 weeks of age. Testing at 3 months of age revealed an inhibitor was present at 4.3 Bu. He had a portacath inserted with Novoseven and then received high dose prophylaxis as immunetolerance. He was inhibitor negative after 3 months and then received normal alternate day prophylaxis. His portacath was removed at age 9 years and he now receives alternate day treatment peripherally. He was diagnosed with CP following delays in his milestones. In 2010 David had an MRI which identified periventricular leukomalacia. He was managed by his local paediatrician and community therapists. In 2015 he was diagnosed with ASD. David at age seven was functionally mobile indoors with the assistance of walking aids and used a wheelchair for outdoor use. He was classified as GMFCS level III using the Gross Motor Function Classification System (GMFCS).

David had received previous injections of botulinum toxin- A to his hip adductors, bilateral hamstrings, gastrocnemii and left psoas as first line spasticity management. These occurred from age 3 -7 years; initially twice in 2010 and then annually. The first 3 procedures were carried out with oral sedation, the final 2 with a general anaesthetic due to difficulty tolerating the procedure. Tranexamic acid was given the night before the procedures and then three times a day for 7 days. A prophylaxis dose of factor was given: on induction; a further dose that evening and then the following day normal prophylaxis was resumed. The effects of botulinum toxin are reversible and enable the reduction in spasticity to be monitored. David received 5 episodes of botulinum-toxin which eventually lost their efficacy. David was deemed a suitable candidate for an SDR and he proceeded to surgery.

The cost of the surgery was covered by the commissioned SDR service. The cost of the factor was covered by haemophilia specialist commissioning.

David had a single level laminectomy performed in 2014 with 59% of sensory nerves being cut between L2 and S2 concurrently with left and right gastrocnemii releases. This technique selectively analyses each individual nerve root with electromyography to separate dorsal and ventral nerve roots through comparison of stimulus responses. Each sensory nerve root is divided into bundles, tested for abnormal electrical response and a percentage between 50-70% cut. Post-surgery David remained in hospital for 3 weeks for rehabilitation.

Surgical haemostasis was secured with regular Factor VIII concentrate. His trough levels were kept between 75-100% for 10 days post-surgery. Tranexamic acid was given at a dose of 15mg/kg 3 times a day for the duration of David's inpatient stay of 3 weeks. Pain relief was initially managed on day 1 with a PCA of morphine sulphate and then sevredol as required, temazepan, tramadol and gabapentin. On discharge home he remained on tranexamic acid, tramadol and gabapentin.

In accordance with our SDR protocol the Modified Ashworth Scale (MAS) [6], The Cerebral Palsy Quality of Life questionnaire (CPQOL) [7], Gross Motor Function Measure (GMFM) [8-9], 6 Minute walk test (6MWT) [10-11] and Modified Medical Research Council (MRC) Scale for Strength [12] were collected pre-SDR, 6, 12 and 24 months post-SDR by the same physiotherapist.

Before SDR there were dynamic catches present in the hip abductors, hamstrings and calf complex and David's MAS was graded as 3. Post-SDR there were no catches and his score was reported as zero indicating no spasticity. The results from the CPQOL demonstrated that the majority of the domains assessed showed improvement from baseline to 24 months review, with the exception of access to services (see table 1). Overall improvements in GMFM from initial assessment to 1 year post-SDR were observed. These improvements were not sustained at 2 years with the dimensions of standing, walking, running and jumping showing a decline at 2 years. His overall percentile change did however maintain improvement (see table 2).

Comparison with the reference centiles for children who have not undergone an SDR showed that gross motor function had not deteriorated post-SDR [13]. David had shown an improvement of > 20 centiles at 12 months post-SDR compared to pre-SDR (see Figure 1).

For the 6MWT; over the 2 years David's walking speed reduced, but it is important to note that the provision of 2 sticks had only recently been provided to him and as such these results do need to be taken with caution (see table 3). David had a significant weight gain between his 1 and 2 year review. His Body Mass Index (BMI) was 22.18 at baseline and at his 2 year review was 29.2. Height Z score remained close to zero at all the assessments and weight remained > 2 SDs. BMI z score were > 3 SDs post-SDR [14] (see table 4). Compared to pre-SDR, there was an improvement in lower limb muscle strength at hips and knees at 6 and 12 months post-SDR. A decline in muscle strength was noted at 2 years post-SDR (see table 5).

This is the first reported case study of a child with severe haemophilia and cerebral palsy being treated with Spinal Dorsal Rhizotomy for spasticity management. These findings are consistent with the literature in patients where children had good outcomes at 1 year [4]. This paper reports early improvements in motor function at 1 year post intervention, but that David's increased weight gain may have affected his ability to sustain his muscle strength and functional abilities long-term.

Children initially have reduced physical activity post-SDR as they recover from their surgery and regain their muscle strength. Comparison with the reference centiles for children who have not sustained a Spinal Dorsal Rhizotomy showed that David's gross motor function had not deteriorated post-SDR. He showed an improvement of >20 centiles at 1 year post-SDR compared to pre-SDR. Bolster et al [15] evaluated the change in the gross motor function of individual children post-SDR by comparing with reference centiles. A change of > 20 centiles was considered as an increase/decline in motor function. From 1 year on there was a decline in David's outcomes, but overall there was improvement from pre-SDR. This decline in his outcomes may relate to his increased weight.

The literature reports an increased prevalence of obesity in children with disabilities in comparison to children without disabilities [16-17]. Increased weight gain post-SDR has been reported in the absence of identifiable changes in individuals eating habits or in line with expected growth [18]. It has been proposed that changes in spasticity/ decreased muscle tone require reduced calorific intake and as such without changes to diet and physical activity increased weight gain may occur. Guthnecht et al. proposed that body mass increases post-SDR were proportionate to the patients' age and sex. They suggested that decreases in energy expenditure due to reduced spasticity are balanced out by either increased physical activity, reduced calorie intake or both [19]. All patients in this study had a normal BMI at the start, in comparison to our case where obesity was apparent pre-SDR. Future research to identify pre and post-SDR activity levels may be of use in counteracting obesity.

David has ASD and this may have influenced his ability to carry out his exercise regime and fully co-operate with his rehabilitation. David's parents hired a physiotherapist pre and post-SDR ensuring consistency and an established relationship to maximise David's ability to participate in his rehabilitation. At 1 year it was discussed that David should stop walking with his sticks and use a k-walker. David's difficulty adapting to change prohibited him from embracing this therapeutic decision.

To conclude this case study has shown that Spinal Dorsal Rhizotomy has provided a safe and effective means of reducing spasticity in a child with cerebral palsy, severe haemophilia A and ASD, though some improvements were lost due to weight gain and compliance difficulties. Careful selection of individuals appropriate for this intervention is essential to its success. Severe haemophilia is not a contraindication for this procedure if managed by a specialist team.

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Deepti Chugh performed the research
Melanie Bladen designed the research study
Melanie Bladen and Deepti Chugh analysed the data
Melanie Bladen, Deepti Chugh, Eleanor Main and Ri Liesner wrote the paper.

Table 1 to illustrate results of The Cerebral Palsy Quality of Life Questionnaire for Children (CPQoL)

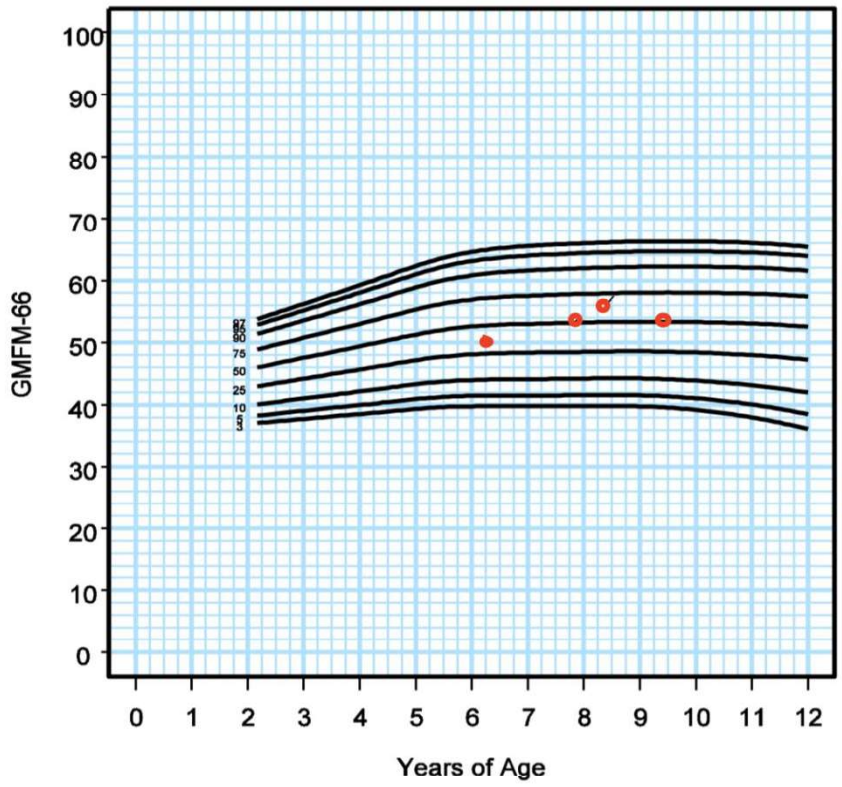
CPQoL	Pre-SDR	6 months post-SDR	12 months post-SDR	24 months post-SDR
Social wellbeing and acceptance	61.5	63.5	62.5	64.6
Feelings about functioning	39.6	59.4	64.6	62.5
Participation and physical health	29.5	47.7	51.1	47.7
Emotional wellbeing and self esteem	54.2	60.4	68.8	58.3
Access to Services	43.8	35.4	37.5	39.6
Pain and impact of disability	56.3	45.3	50	50
Family Health	59.4	43.8	81.3	65.6

The GMFM is a criterion-referenced standardized observational tool used to measure changes in gross motor function in children with cerebral palsy.

Table 2 to illustrate Gross Motor Function Measure 88 (GMFM) and GMFM-66 and GMFM-66 percentile results

	Lying and rolling	Sitting	Crawling and kneeling	Standing	Walking, running and jumping	GMFM 88-Total Score	GMFM 66 Score	GMFM-66 centiles
Pre-SDR	100%	95%	95.24%	17.95%	13.89%	64.42%	50.85	35th
Post-SDR	100%	96.67%	95.24%	43.59%	16.67%	70.43%	54.15	55th
12 months post-SDR	100%	100%	100%	56.41%	16.67%	74.62%	57.68	60th
24 months post-SDR	100%	100%	100%	38.4%	13.89%	70.47%	53.38	50th

Figure 1 Graph to illustrate David's plotted normal trajectory without SDR



Gross Motor Function Measure (GMFM-66) Percentiles by Age
Gross Motor Function Classification System (GMFCS) Level III
(Red icons indicate David's GMFCS score percentile pre-SDR, 6, 12 and 24 months post-SDR)

GMFM-66 percentiles measures relative ability compared with other children of the same age and GMFCS level [13]

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Table 3 to illustrate Functional Mobility

	Timed Up and Go (seconds)	2-minute walk test (meters)	6-minute walk test (meters)
Pre- SDR	14.49secs Bilateral pod sticks and AFO'S *	60m Bilateral pod sticks and AFO's *	190m Bilateral pod sticks and AFO's *
6 months post-SDR	20.39 secs Bilateral pod sticks and AFO'S *	60m Bilateral elbow crutches and AFO's *	Clinically a 2minute walk test was completed to avoid fatigue
12 months post-SDR	13.38 secs Bilateral elbow crutches and "Aircast walking splints"	75m Bilateral elbow crutches and "Aircast walking splints"	215m Bilateral elbow crutches and "Aircast walking splints"
24 months post-SDR	22.86 secs Bilateral sticks; in GRAFOs Δ	30m Bilateral sticks; in GRAFOs Δ	88m Bilateral sticks; in GRAFOs Δ

*AFO's= Ankle Foot Orthoses ΔGRAFOs =Ground reaction ankle force orthoses

Table 4 to illustrate Body Mass Index (BMI) in relation to height and weight

	Height cm			Weight kg			BMI		
	Height	SDS	Centile	Weight	SDS	Centile	BMI	SDS	Centile
Pre-SDR	124	-0.02	49	34.1	2.17	98	22.18	2.81	99.8
6 months post-SDR	129	0.28	61	41	2.62	99.6	24.64	3.16	99.9
1 year post-SDR	131	0.09	53	43.4	2.5	99.4	25.29	3.11	99.9
2 years post-SDR	136.1	0.08	53	54.6	2.78	99.7	29.48	3.4	>99.9

SDS = Standard scores

Manual muscle testing was performed by the same physiotherapist utilising the classification shown in table below. Measurements are taken at the mid-range of the active range and categorised on an 8 point scale from; no movement; flicker; gravity eliminated; gravity counterbalanced; minimal resistance; against gravity and moderate resistance; against gravity and moderate to maximal resistance and normal strength.

Table 5 to illustrate Modified Medical Research Council (MRC) Scale for strength pre and post Spinal Dorsal Rhizotomy (SDR)

	Pre-SDR		6months post-SDR		12months post-SDR		24months post-SDR	
	R	L	R	L	R	L	R	L
Hip flexors	4	4	4	4	4	4+	4-	4-
Hip extensors	4	4	4+	4	4+	4	4	4
Hip Abductors	2	3	3	3	3	3	2	2
Knee Flexors	3	4	4	4-	4	4-	4-	4
Knee Extensors	3	3	4	4	4+	4+	3	4-