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Case report

Long-term benefits from selective dorsal rhizotomy in a young patient with cerebral palsy



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ABSTRACT

Introduction: Spasticity is considered to be one of the most important factors hampering functional abilities among patients with a cerebral palsy (CP).

Aim: The aim of the study was to present results of the selective dorsal rhizotomy (SDR) procedure combined with the physiotherapy process in a 3 year follow-up study, presented from a functional and structural perspective.

Case study: After the diagnosis of CP in the form of spastic diplegia, the 2-years-old patient (GMFCS 4) was directed for a comprehensive rehabilitation. After clinical examination and family consultation, spasticity was found to be important factor limiting patient functional abilities.

Results and discussion: The patient was directed to SDR operation. The patient was evaluated four times: before the SDR, and then 1, 2 and 3 years after the SDR surgery. The spasticity was assessed using the modified Tardieu scale. A functional assessment was done using the gross motor function measure (GMFM) scale, 6-min walk test and functional assessment questionnaire (FAQ-10). A muscle tension remained decreased throughout the 3 years of the follow up period. The achieved reduction in muscle tone was accompanied by a change in a range of motion, an improvement in GMFM total score result, an increased distance in 6-min walk test and in the FAQ-10 questionnaire.

Conclusions: SDR procedure combined with comprehensive rehabilitation programs leads to short- and long-term reduction in the spasticity of a 5-year-old boy with a spastic diplegia. Decreased level of spasticity was accompanied by an increased gross motor functioning and mobility.

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1. Introduction

One of the most common motor disorders and consequently the main cause of functional limitation in cerebral palsy (CP) is spasticity.^{1,2} This physical disability is affecting about 2 of every 1000 live births.^{3,4} CP is a non-progressive condition characterized by a wide spectrum of motor deficits resulting from damages that mainly affect the supratentorial corticospinal tracts and the basal ganglia.^{5,6} Patients often develop debilitating fixed muscle contractures that restrict their normal range of motions around joints, and thus limit their mobility. Contractured muscles have been shown to be shorter and stiffer as a result of both stiffer fibers and stiffening of the extracellular matrix.⁷ Therefore more effective treatment strategies and clinical care are highly desirable for those patients.⁴ Nowadays we are gaining more and more evidence regarding prevention and treatment options for CP. It is already known that cooling prevents the brain in birth asphyxia and due to better management the number and the severity of patients with CP is declining. Therefore, we should seek for more interventions that may help in the management of the disturbances that occur in patients with CP.^{4,8–10} Therefore, we should seek for interventions that may help in the management of the disturbances that occur in patients with CP. Spasticity may be reduced by rehabilitation programs and/or pharmacological interventions.¹¹

Severe involvement of spasticity suggests a multi-professional consultation for a surgical treatment.^{12,13} Selective dorsal rhizotomy (SDR) is an example of such treatment. With its over 100 years of history, SDR has become accepted as a standard neurosurgical procedure for the treatment of spasticity associated with CP.^{14–16} It is used primarily to treat children with a lower-extremity spasticity. In those cases, the primary goal of the surgical intervention is to improve lower-extremity function by reducing spasticity.¹⁷ It is important to note that although the primary insult to upper motor neurons in CP is not progressive, the secondary dysfunction of the musculoskeletal system often progresses. The decline depends, among others factors, on the amount a faulty afferentation of affected muscles. By decreasing the excitatory afferent input from the dorsal roots, the amount of excitation experienced by the lower motor neurons can be reduced, and therefore reducing spasticity.¹⁸

Nevertheless, controversy surrounds the question of whether the loss of spasticity in patients with CP really matters.¹⁹ Potential benefits of therapy chosen for patients with CP should be considered on structure and activity as well as on participation levels.¹⁴ Question still remain whether therapeutic interventions, including SDR, performed during childhood have long-term functional benefits for a patient, his family and patient's functioning in the society.²⁰ To answer those questions the study was undertaken.

2. Aim

The aim of the study was to present results of the SDR procedure combined with the physiotherapy process in a 3-year

follow-up study, presented from a functional and structural perspective.

3. Case study

The participant was recruited into this study after obtaining age-appropriate assent from both a child and parents. History of his disorder has been analyzed first. The patient was born at week 39 of gestation. His gestational age was evaluated according to Dubowitz classification²¹ for 33 weeks. Apgar score in 1st, 3rd and 5th minute was evaluated respectively for 8, 7, and 8 points. Motor milestones like rolling, creeping and crawl were achieved approximately at age of 12 months. Independent sitting, standing next to furniture were achieved at approximately 2 years of age. Computed tomography scanning did not reveal presence of damage to the basal ganglia. After the diagnosis of CP in the form of spastic diplegia, the 2-years-old patient was directed for a comprehensive rehabilitation. The patient's at-home rehabilitation program had been carried out systematically three times a week for 1 h per day. The patient participated also in a 3-week long rehabilitation in hospital stays, twice a year for 4 h therapy per day. The rehabilitation programs, at home and hospital, were based on individually tailored therapeutic goals and conducted using neurophysiological methods. Generally, the home-based therapy was focused on gait parameters improvement. On rehabilitation stays at hospital, despite individual therapy for 1 h, the patient also undergo additional exercises and training for 3 h a day. The following therapy procedures were used: progressive strength training (30 min), aquatic exercises (45 min), balance training using virtual reality game-based therapy (30 min), group therapy focused on mobility using competition approach (45 min) and stretching (30 min).

At the age of 4 years, functional status of the patient was classified at level 4 according to gross motor function (GMF) classification system (GMFCS). The patient was able to support his full weight on feet using a walker. After the SDR procedure a spasticity in lower limbs, measured by modified Tardieu scale, was reduced in plantar flexors, knee flexors, hip flexors and hip adductors from 2 (clear catch at a precise angle, followed by release) to 0 (no increase in muscle tone).²² After clinical examination and family consultation, spasticity was found to be important factor limiting patient functional abilities. Therapeutic team introduced the SDR procedure and its treatment goals were explained to the patient and his family. Proposed therapy matched current needs and expectations of the patient, family and the therapeutic team as well (increase mobility and independence in everyday life). The patient was directed to SDR operation according to Park technique,²³ combined with gastrocnemius muscle and gracilis tendon lengthening procedure. The operation took place at the 5 years of patient's age. After the operation, patient returned to his rehabilitation programs as described above.

4. Results

The patient was evaluated four times: before the SDR, and then 1, 2 and 3 years after the SDR surgery. The spasticity was

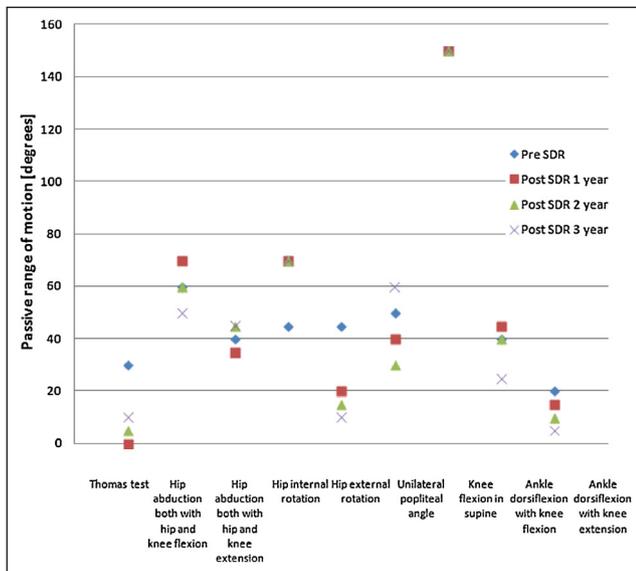


Fig. 1 – Changes in PROM.

assessed using the modified Tardieu scale.²⁴ Passive range of motion (PROM) was measured by an experienced physician to define the degree of muscle shortening.^{24,25} PROM measurement included: the Thomas test, hip internal and external rotation, hip abduction both with hip and knee flexed and extended, knee flexion in supine position, unilateral popliteal angle, and ankle dorsal flexion both with knee flexed and extended. A functional assessment was done using the GMFM measure (GMFM) scale which is a standardized instrument designed and validated to measure changes in GMF in 5 domains (lying and rolling; sitting; crawling and kneeling; standing and walking; running and jumping) over time. The total result of GMFM scale presents spectrum of functional abilities of patient.²⁶ A gait function was evaluated using the 6-min walk test (6MWT) which measures the distance a person can walk at an unhurried, self-determined pace in 6 min.^{27,28} Changes in functional mobility were also assessed from a family perspective using functional assessment questionnaire (FAQ-10). The FAQ is a 10-level, parent-report walking scale covering a range of walking abilities from non ambulatory to ambulatory in all community settings and terrains.²⁹

After the SDR procedure a spasticity in lower limbs was reduced from 2 i.e. clear catch to 0 – no increase in tone, using the modified Tardieu scale. A muscle tension remained decreased to 0 throughout the 3 years of the follow up period. The achieved reduction in muscle tone was accompanied by a change in a range of motion (Fig. 1), an improvement in GMFM total score result, an increased distance in 6MWT and in the

FAQ-10. Functional patient's abilities changed also in the GMFCS classification (Table 1). Detailed results are presented in Fig. 1 and Table 1.

5. Discussion

A balance between elimination of spasticity and preservation of motor functions were observed in short and long-term studies published so far.^{14,30-33} In the present study, the elimination of spasticity allowed the patient to achieve new functional skills (Table 1). Before the operation, patient was classified at level IV according to the GMFCS. Children classified at this level may continue to walk for short distances using a walker or rely on wheeled mobility at home and school and in the community. After a year from the SDR procedure functional mobility skills of our patient were assessed on GMFCS level III. This means that he was able to walk indoors on a level surface with an assistive mobility device. Children assessed on the level III according to the GMFCS may climb stairs holding onto a railing, may propel a wheelchair manually or are transported when traveling for long distances or outdoors on uneven terrain.³⁴ Results of this study show also changes in PROM which are the most common postoperative changes in the structure domain of the International Classification of Functioning (ICF) by World Health Organization (WHO).^{14,17,35,36} In this study, changes in the range of motion, turned out not to be fixed (Fig. 1). This may result, among others contextual factors, from the compensatory mechanisms common in CP and from individual adjustment to altered biomechanics of patient move. Such result may indicate that the spasticity is not the only factor limiting the PROM in the presented case.

Another aim of this follow-up study was also to examine effects of structural alterations after the SDR in relationship with potential gains of functional abilities. Despite the lack of a positive trend in the PROM changes, functional benefits were observed, both in the GMFM, the 6MWT and in the FAQ-10. Presented results may suggest that the optimal range of motion is not the most important variable influencing the functional status of the CP patient.

The members of the National Institute for Clinical Excellence in United Kingdom (NICE) published guideline based on literature review where the use of the SDR surgery in patients with CP was recommended.³⁷⁻³⁹ Changes in gait function mostly refer to increase of the stride length^{34,40,41} gait speed,^{36,40} and the improvement of the biomechanical gait parameters.^{40,41} In our study, 3 years after the operation when the patient was 8, walking distance increased more than 6 times (from 20 m to 132 m) in the 6MWT. In our opinion, the cause of the progress in mobility and GMF in our patient may be at least twofold. To a certain extent, the improvement results from both the SDR surgery and the holistic rehabilitation programs. However, all measurements and procedures were performed; however, in the patient's age range from 4 to 8 years. Thus, such functional improvements may be also due to natural motor development. As it was published in 2002, natural development in patients with CP is characterized by the largest increase in GMFM-66 total score in the mentioned period of life.⁴² On the other hand, after observed reduction of

Table 1 – Functional changes in time after SDR.

	GMFM	FAQ-10	6MWT, m	GMFCS
Pre SDR	32.6	2	20 (walker)	IV
Post SDR 1 year	41.5	3	40 (walker)	III
Post SDR 2 year	44.4	4	42 (walker)	III
Post SDR 3 year	46.4	5	132 (walker)	III

spasticity, a functional status expressed by the total score of the GMFM was similar to CP peers classified on the GMFCS level III. Before the surgery, the GMFM total score was approximately 10 points below the development curve. The patient's GMFM total score increased significantly from preoperative 32.6 to 41.5, 44.4 and 46.4 1, 2 and 3 years' post-operatively, respectively. Our results are consistent with a few studies concerning functional improvements after the SDR procedure. Statistically significant changes in activity domain according to GMFM total score were observed 20 months after the operation.⁴³ Retrospective analysis of patients classified on GMFCS levels I-IV performed 18 months from the SDR showed a reduction in spasticity and an improvement in FAQ index.⁴⁴ In the presented case, during the annual follow-up appointments patient's activity assessed from family perspective in the FAQ-10 was consistently improved, from the level 2 before the SDR to level 5 3 years after. The evaluation of activity from the family perspective is particularly important due to the fact that the patients' functional status can influence quality of life of his or her family.¹⁶ We must remember that CP is a lifelong disorder.⁴⁵ Thus, all possible evidence based on a solution should be implemented in a clinical practice. Children with CP and their families should have the appropriate information about potential benefits or even defects results from the SDR procedure. Despite number of publications referring to this topic,^{15,16} limited proportion of patients fulfilling the eligibility criteria for surgery decide for the SDR. This is especially important due to the fact that there are only few patients in Polish population who underwent such procedure by now. The presented patient has participated in complex rehabilitation programs during his childhood. There is some controversy whether rehabilitation or the SDR alone can lead patient to functional improvement. Mäenpää et al. compared results of two similar groups of CP patients in terms of age, gender, spasticity in the lower limbs and the level of functional mobility who underwent different types of intervention. One group consisted of patients who had SDR and the process of rehabilitation treatment, the second group participated only in the treatment process based on an intensive rehabilitation. In both groups, there were significant functional improvements. However, no significant difference was observed between groups.³² In 2006 Engsborg et al. observed significant improvement in muscle strength and gross motor skills in a similarly designed study. However, the group where physiotherapy treatment was combined with SDR presented significantly better results.⁴³

Besides different mechanisms of action, results of SDR can be compared with for example an orthopedic surgery. Thomas et al. were seeking to reduce the consequences of spasticity in the 2-year follow-up analysis.³⁵ They observed significant changes in the passive mobility, energy expenditure and in the spasticity measured using Ashworth scale. Results did not differ between SDR and orthopedic surgery group.³⁵ At present, based on our results and according to other authors, it can be concluded that SDR procedure alone or combined with rehabilitation programs may have positive outcomes without or with small number of adverse effects.³³

It should be underlined that in a published in 2013 systematic review of evidenced therapies for children with

CP, the SDR procedure was graded 'do it', i.e. was included to group of scientifically proved interventions recommended in the spasticity treatment and care among those patients.⁴⁶

Results presented in this study may be helpful not only in predicting effects of the SDR but also in revising goals and selection of interventions for children with CP. The SDR is a procedure aimed to supplement, not replace the standard treatment program for patients with CP. The SDR has to facilitate the multi-profile treatment and the rehabilitation, intensifying their actions. According to the current standard of treatment of children with spastic form of CP, therapy should be multifaceted including a combination of rehabilitation, orthopedics, pharmacological treatment and surgery. That is why there is a need for further research seeking to explain the long-term results of the SDR in patients with CP, especially in the activity and participation domains according to the ICF classification.¹⁶

6. Conclusions

The SDR performed during childhood combined with a comprehensive rehabilitation may be beneficial and bring a long-term efficacy in the treatment of spasticity in patients with CP.

Conflict of interest

None declared.

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