Study of the effect of sensory dynamic orthosis on postural control in a case of nemaline myopathy

Estudio del efecto de la órtesis sensorial dinámica en el control postural en un caso de miopatía nemalínica

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Received: September 12, 2018
Accepted: March 19, 2019

ABSTRACT

Background: the analysis and assessment of balance and postural control parameters are carried out using standardized system frameworks (Timed up, Go Test, Tinetti or Berg), which require physical and cognitive abilities that are not always present in certain patients with disabilities. Stabilometry and posturography are objective and applicable assessment methods. *Case presentation:* we present a case of a 7-year-old girl diagnosed with congenital etiology nemaline myopathy type III, whose aim is to assess both the immediate and long-term impact of the sensory dynamic orthosis (SDO®) on balance control following a ten-week period. The child was assessed using a pressure platform (Podoprint Namrol). The evaluations were performed in sitting position with and without the SDO® (eyes open and eyes closed). *Results:* shorter displacements of the center of pressure in the medial-lateral axis were collected in the first clinical evaluation wearing the SDO® (variability with eyes open 77.78 %; with eyes closed 76.31 %) while in the second clinical evaluation without the SDO® 52.77% variability (eyes open) and 84.21 % (eyes closed). Furthermore, there was less displacement of center of pressure (COP) in the anterior-posterior axis with the SDO® 19.07 % variability EO (Eyes Open) and 15.75 % EC (Eyes Closed) if compared to the first evaluation. *Conclusion:* the exclusive use of the sensory dynamic orthosis in this case, it does not seem to produce improvement in a long-term balance control, hence it should be complemented with additional therapies.

Keywords: nemaline myopathy, postural balance, physiotherapy, neurology.

RESUMEN

Introducción: *el análisis y la evaluación del control postural se llevan a cabo utilizando escalas estandarizadas (Timed up, Go Test, Tinetti o Berg), que requieren habilidades físicas y cognitivas que no siempre se presentan en ciertos pacientes con discapacidad. La estabilometría y la posturografía son métodos de evaluación objetivos y aplicables.* Presentación del caso: *niña de 7 años diagnosticada con etiología congénita miopatía nemalínica tipo III, cuyo objetivo es evaluar el impacto inmediato y a largo plazo de la órtesis dinámica sensorial (ODS®) en el control postural después de un periodo de 10 semanas valorada con una plataforma de presión (Podoprint® Namrol). Se realizó en posición sentada con y sin ODS® (ojos abiertos y ojos cerrados).* Resultados: *se obtuvieron menores desplazamientos del centro de presión en el eje medial-lateral en la primera valoración clínica con ODS® (variabilidad con ojos abiertos 77,78 %, con los ojos cerrados 76,31 %) y en la segunda valora-*

ción sin la ODS® de 52,77 % variabilidad (ojos abiertos) y 84,21 % (ojos cerrados). Por otro lado, hubo un menor desplazamiento del Centro de Presiones (COP) en el eje antero-posterior con ODS® con un 19.07% de variabilidad (OA) y un 15,75% (OC) si lo comparamos con la primera valoración. Conclusiones: *el uso exclusivo de la órtesis sensorial dinámica en este caso, no produce mejoría en el control postural a largo plazo, por lo que debe complementarse con terapias adicionales.*

Palabras clave: *mielopatia nemanílica, control postural, Fisioterapia, neurología.*

BACKGROUND

Nowadays, there is a large number of diseases classified as *rare*. They need scientific, theoretical and practical research to learn about their causes, effects and prognosis among others. Many of these diseases, of neurological, muscular, orthopedic origin not only affect balance and postural control, but also day to day activities performance, conditioning standards of living. Due to the low number of individuals who present a certain *rare disease* and the lack of scientific research, there is no such an effective protocol that helps improve and facilitate the functionality of patients who suffer from these deficiencies.

Sensory dynamic orthosis are designed to help enhance the quality of the user's physical abilities. Through clinical observation, people who present an abnormal postural tone have the ability to achieve greater control over their movements when adequate sensory feedback is given.

At the present time, just a few studies have investigated about Sensory Dynamic Orthosis, hence here lies the interest in conducting a clinical study whose objective is to assess the effect of this type of therapy and announce its existence and utilization.

Nemaline myopathy is a congenital myopathy that consists of accumulations of muscle proteins. It shows thin filaments in skeletal muscle fibers and represents around 20 % of congenital myopathies. Nemaline myopathy is characterized by weakness, hypotonia, and weak or absent tendon reflexes, causing a deficit of postural control and therefore limitations in day to day activities[1-6]. Sensory dynamic orthosis (SDO®) are designed to help enhance the qualities of the patient's physical abilities, contributing to joint alignment, increasing sensory and proprioceptive stimulus and improving control and postural schema.

Generally, the analysis and assessment of balance and postural control parameters are carried out using standardized system frameworks (Timed up, Go Test, Tinetti or Berg), which require physical and cognitive abilities that are not always presented in certain disabled patients. Stabilometry and posturography are objective and applicable assessment methods[7–9].

Stabilometry allows to define objectively the average position of the corporal center of pressure (COP) as well as short movements around this stance. This analysis provides a series of variables that are related to these oscillations. The execution of this test in different conditions allows, at the same time, the involvement of different body systems and their influence on postural control[8-9].

Furthermore, the interest of stabilometry in the rehabilitation treatment of balance alterations is based on objective information, which is truly useful for personal patients' program rehabilitation designing.

The aim of this clinical study is to determine the effect of the sensory dynamic orthosis on a postural stability sitting position in a nemaline myopathy case.

CASE PRESENTATION

Clinical study of a 7-year-old girl diagnosed with nemaline myopathy type III, due to a mutation of the ACTA1 gene with chronic respiratory failure (CRF), atelectasis in right middle lobe, right lower lobe and left lower lobe. Cesarean delivery at 38 weeks' gestation. Postnatal resuscitation is needed. Birthweight 2884 g, length 48 cm and head circumference 35 cm. In the first Apgar test a score of 5/9 was collected, whereas in the second the score was 7/9. As for the respiratory and cardiovascular exploration, a respiratory muscle weakness was noticed because of an unusual weak level of crying.

Description

The infant presents weakness, hypotonia and generalized muscular atrophy. Muscular restrictions are shown, especially at the level of the spine musculature, lower limbs and pelvis, equinovarus foot deformity. Flexor postural pattern with scoliosis, pectus excavatum at the left thoracic wall and pelvic tilt. Noninvasive mechanical ventilation is needed and BiPAP (continuous positive airway pressure) assistance at home. Pharmacological treatment with Mestinon® has been started. The infant receives three physiotherapy sessions per week for an hour and a half each.

Valuation methodology

Postural stability with eyes open (EO) and eyes closed (EC) is assessed in sitting position using Podoprint® (Namrol Group, Barcelona, Spain), first without the sensory dynamic orthosis and immediately right after with the sensory dynamic orthosis. A second assessment similar to the former is conducted after a ten-week period. The performance of the test in different conditions allows to analyze the participation of different body systems and their influence on the posture. In this case, there is generally less postural control and more variability with closed eyes.

The patient was set on the pressure platform, placed on a table at a sufficient height so that the feet would not touch the floor. The hands were supported on top of the thighs. Every measure lasted for 30 seconds. In each of them the patient wore splints on each foot in order to prevent plantar flexion. The first measurements were registered without the sensory dynamic orthosis worn in sitting position (figure 1). The patient wore anti-equine splints of both feet in every measure.

The following variables related to postural measurement were collected: center of pressure (CoP) (mm) medial-lateral and anterior-posterior deviation, CoP (m/s) medial-lateral and anterior-posterior speed deviation, Romberg coefficient (%) and CoP surface (mm²). The Romberg coefficient is obtained by dividing the surfaces of the ellipses recorded with eyes closed (EC) and with eyes open (EO), multiplied by one hundred [RC = (EC / EO) * 100].

FIGURE 1. **The sensory dynamic orthosis worn in sitting position.**

The SDO® is prescribed by a physiotherapist from "Centro Landa Extremadura" after evaluating the postural attitude according to the patient's specific needs. This garment is custom-made, it favours postural control and muscle tone. It is designed to provide sensory and proprioceptive feedback and is made of cotton, Lycra and elastane. Its reinforcing panels adapt themselves to the patient's specific needs. It is breathable and non-invasive[10] (figure 2).

FIGURE 2. **The sensory dynamic orthosis.**

Intervention program

The objectives of the intervention program that was carried out in both clinical valuations were: to decrease muscle retractions and normalize muscle tone, to improve object manipulation and postural hygiene, to favour ribcage expansion, to mobilize secretions and to increase vital capacity.

Procedures: manual therapy techniques, Bobath approach, transfer facilitation, normal movement and waist dissociation, manual respiratory physiotherapy and with assisted ventilation (mobilize and expel secretions, increase ventilatory capacities).

RESULTS

Data collected in the first and second valuation, both with and without the SDO®, open and closed eyes is shown in table 1.

After having analyzed the results from each valuation, we could conclude that there was an average speed increase of the CoP in both axis in the first and second valuation with and without the SDO® (EO: -35.71 %/ -77.08 %; EC:-11.11/-97.72 %) respectively. As for the CoP displacement, there was less displacement in the medial-lateral axis wearing the SDO® in the initial valuation of 77.78 % variability (EO) and 76.31 % (EC) if compared without the SDO®.

Once the ten-week treatment period finished, we collected a lower CoP deviation in the medial-lateral axis without the SDO® of 52.77 % variability (EO) and 84.21 % (EC). Furthermore, less CoP displacement in the anterior-posterior axis with the SDO® of 19.07 % variability (EO) and 15.75 % (EC) was also collected if compared with the first valuation. However, higher values of average speed in both axis were registered, with and without the sensory dynamic orthosis. Additionally, the CoP surface experienced a reduction without the SDO® after the physiotherapy intervention of 51.72 % (EO) and 45.45 % variability (EC) if compared with the first valuation.

DISCUSSION

The results collected in the current study show that the use of the sensory dynamic orthosis could have a positive effect on postural stability in a patient diagnosed with nemaline myopathy. It is demonstrated that the use of the SDO® along with medication and the physiotherapy sessions given over the ten-week period have a positive impact, being more noticeable when the girl does not wear the SDO®. However, it needs to be taken into consideration that the prolonged use of the SDO® at home and the girl's tolerance have not been optimum.

With reference to the intervention period, Lorentzen et al.[11] assessed daily activities, functional abilities of upper and lower limbs and balance 12 weeks after training, while our study was over a ten-week period. It was

TABLE 1. **Stabilometric results comparison in both evaluations.**

		EO without SDO	EC without SDO	EO with SDO	EC with SDO	Variability EO without/with SDO (%)	Variability EC without/with SDO (%)
CoP average speed (mm/s)	1st evaluation	0.9	0.8	1.4	0.9	-35.71	-11.11
	2nd evaluation	1.1	1.1	4.8	48.2	-77.08	-97.72
Variability (%)		-18.18	-27.27	-70.83	-98.13		
M-L axis average deviation (mm)	1st evaluation	3.6	3.8	0.8	0.9	77.78	76.31
	2nd evaluation	1.7	0.6	11.1	17.3	-84.68	-96.53
Variability (%)		52.77	84.21	-92.80	-94.80		
M-L axis speed (mm/s)	1st evaluation	0.7	0.8	1.4	0.9	-50	-11.11
	2nd evaluation	1.1	1.0	4.4	4.2	-75	-76.19
Variability (%)		-36.36	-20	-31.82	-78.57		
A-P axis average deviation (mm)	1st evaluation	59.5	69.1	81.3	84.4	-26.81	-81.87
	2nd evaluation	62.3	69.0	65.8	71.1	-5.32	-0.97
Variability (%)		-4.49	0.14	19.07	15.75		
A-P axis speed (mm/s)	1st evaluation	0.4	0.3	0.4	0.3	0	0
	2nd evaluation	0.4	0.1	2.0	1.7	-80	-94.11
Variability (%)		0	66.67	-80	-82.35		
CoP surface (mm2)	1st evaluation	2.9	1.1	2.7	1.7	6.90	-35.29
	2nd evaluation	1.4	0.6	35.2	48.2	-96.02	-98.76
Variability (%)		51.72	45.45	-92.33	-96.47		
Romberg coefficient (%)	1st evaluation	37.93	37.93	62.96	62.96	39.76	39.76
	2nd evaluation	42.8	42.8	136.9	136.9	68.74	68.74
Variability (%)		11.38	11.38	54.01	54.01		

CoP: center of pressure; Eo: eyesopen; eyes closed; SDO: the sensory dynamic orthosis; M-: medial-lateral; A-P: anterior-posterior;

demonstrated that continuity in postural control training after a certain number of sessions could lead to functional motor improvements in this type of motor disordered patients.

As for the SDO®, we conclude that its effectiveness depends on a range of factors, as well as its correct placement, the application time, the individual benefits and disadvantages and contiguous therapies, such as

physical therapy, occupational therapy and pharmacological therapy. In their study, Knox et al.[12] investigated the effect of the SDO® in a group of children diagnosed with infantile cerebral palsy who presented a series of motor disorders. The intervention protocol consisted in wearing the orthosis 4 hours every day for 4 weeks. The final results, collected on a series of rating scales similar to the Gross Motor Function Measure (GMFM) showed a significant benefit of the SDO® in these type of patients: more independent and stabilized sitting position, easier transfers, comfort, greater lower limb symmetry and involuntary movement reduction, thus matching with our study with regard to the improvement of trunk stability.

Conversely, a series of issues was noticed, such as a long time spent placing the SDO®, increasing difficulty in self-feeding and difficulty in wearing the SDO®, discomfort, difficulty when going to the bathroom and child growth development. The study of Gracies et al.[13], assessed acceptability, effects on swelling, resting posture, spasticity, and active and passive range of motion of individually tailored upper limb Lycra garments when worn for 3 hours by hemiplegic patients. The intervention protocol consisted in a valuation of the SDO® effects at the start and end of the session after 3 hours of application for two days. The results showed that the orthosis was well tolerated by all patients. It was indicated, among other results: (1) reduced swelling after 3 hours of use; (2) upper limb resting position improved; (3) active motion improvement; (4) passive motion was benefited by the SDO®; (5) proprioception improvement.

In his study, Nicholson et al.[14] assessed the effects of the SDO® on daily functional abilities, such as auto-care, mobility and social abilities. The results showed improvement in auto-care. The aim of Pasini et al.[15], study was to compare two types of interventions to improve postural control in sitting position in children with cerebral palsy. There were two intervention programs, they were at home and a perceptual-motor therapy program, twice a week. The results collected showed a positive effect on the CoP speed in the medial-lateral and anterior-posterior axis, unlike our study in which we gathered a greater CoP speed in both axis with and without the SDO® after the physiotherapy intervention.

CONCLUSION

The sensory dynamic orthosis had a positive immediate effect on our patient in the first valuation, but not in the second valuation, having in the latter a postural control improvement without the orthosis. Therefore, in our clinical study, the exclusive use of the SDO® seems not to be effective in improving postural control, so its application should be complemented with additional therapies.

ETHICAL RESPONSIBILITIES

Protection of people and animals. The procedures followed in this study are in accordance with the Declaration of Helsinki of the World Medical Association, in its 2016 update.

Confidentiality and informed consent. The father of the patient included in this study was informed by receiving an informed written consent to participate in the study, which he signed and gave to those responsible for the study.

Privacy. In this manuscript there are no personal data of the participant in the study.

Funding. The authors declare that they have not received any type of specific funding for this work.

Competing interests. The authors have declared that no competing interests exist.

Author contributions. All the authors declare to have contributed in the conception and the design of the study, the data collection and its later analysis and interpretation of the same ones and in the writing of the article.

SPECIAL THANKS

To colleagues and participants for the collaboration in this study.

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