The Effectiveness of Bobath Treatment Approach in Physiotherapy of Case with Ehlers-Danlos Syndrome

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Abstract

Ehlers-Danlos Syndrome (EDS) is one of the most frequent connective tissue diseases (1:5,000–1:10,000) and characterized by multiple clinical findings depending on connective tissue weakness in body system. Most of EDS patients consists of “hypermobility type (EDS-HT)” Physiotherapy program has an important place in the treatment of children with EDS-HT diagnosis and Bobath therapy aims to regulate muscle tone, provide postural alignment with special grip techniques and improve functions with the active participation of individual. The case with EDS-HT diagnosis is 5 year- old female child with 14 kg body weight, and 96.7 cm height. In 18th month, the case was directed to physiotherapy program towards hypotony and postural control problems in Private Yildiz Çocuk Special Education and Rehabilitation Center. The case was evaluated three times for three years. Motor functions of the case were evaluated using Gross Motor Function Measurement (GMFM) scale, the disability level of the case was determined using Gross Motor Function Classification Scale and the functional independence level of the case was evaluated using Functional Independence Measure for Children (WeeFIM) scale. The applied physiotherapy program was designed from the exercises used in Bobath treatment approach. According to subsequent evaluations made through GMFM and weeFIM for throughout 3 years when the case received physiotherapy, a significant increase was observed in the scores of all parameters of GMFM and weeFIM, and an increase at one level in disability level was observed in GMFCS evaluation.

Keywords: Ehlers-Danlos syndrome; Physiotherapy; Bobath concept

Introduction

Ehlers-Danlos Syndrome (EDS) is one of the most frequent connective tissue diseases (1:5,000–1:10,000) and characterized by multiple clinical findings depending on connective tissue weakness in body system [1]. Three main characteristics of this life-long disease are skin hyperexcitability, joint hypermobility and tissue fragility [2-4]. Significant problems observed in Ehlers-Danlos Syndrome includes orthopedic complications like joint pain, swell, dislocation, back pain, etc., and walking and hand function disorders, as well. Surgical interventions may be needed to heal dislocated joints, however this approach may generally not be sufficient to remove the problem in treatment. Physiotherapy is required in clinic recovery [5].

Most of EDS patients consists of “hypermobility type (EDS-HT)” [2]. EDS-HT is one of 6 types of EDS in classification system designed according to clinical, biochemical and molecular findings and suggested by Beighton et al. According to this classification system, EDS is classified in 11 sub-groups and EDS-HT is classified as EDS type III [2]. EDS-HT patients complaints from serious joint hypermobility related with frequently repeated joint dislocations [1]. Instability in nearly all joints is reported in high incidence in shoulder, knee, hip, chin, ankle and fingers [1]. Other symptoms of EDS-HT includes muscle cramps, tendinitis, chronic pain, repeated headache and fatigue [6].

Hypermobility treatment generally consists of physiotherapy, local steroid injections, resting and surgery when needed [7]. On the other hand, physiotherapy is used in order to increase condition and body muscle strength, improve proprioception and preserve normal joint spaces of all joints [8]. Physiotherapy program has an important place in the treatment of children with EDS-HT diagnosis, and hypotonic and motor growth retardation [9]. Physiotherapy (exercise treatment, traction, joint stabilization methods, proprioceptive growth, passive mobilization and general instructions) is the basis of alternating modalities [7].

Bobath concept is an important approach in physiotherapy concepts. International Bobath Instructors Training Association (IBITA) defines Bobath Concept as the approach towards solving the problem in the evaluation and treatment of individuals with function, movement and postural control disorder depending on a lesion in central nervous system. In addition, Bobath Concept aims to diagnosing and analyzing problems with the participation in functional activities and everyday life activities, as well as determining movement components and underlying defects [10]. Bobath therapy aims to regulate muscle tone, provide postural alignment with special grip techniques and improve functions with the active participation of individual [11]. The facilitation of normal movement components and their special implementation according to task, which includes strategies preserving muscles and joint uniformity, are critical elements of Bobath Concept [12].
In line with those effects of Bobath concept, this study aims to examine the clinic effectiveness of Bobath treatment approach in a total treatment program of a case with EDS-HT diagnosis.

**Case Report**

The study case (Z.Ş.) is 5 year-old female child (birth date: 05.10.2007) with 14 kg body weight, and 96.7 cm height. She was born as the second child of her family with the weight of 3150 grams. The case, who was diagnosed as EDS-HT on 05.10.2007, was regularly monitored by orthopedist and traumatology specialist. After birth, plastic molds were suggested for her hands and the family was informed about accurate position and rehabilitation program of the child. In 18th month, the case was directed to physiotherapy program towards hypotony and postural control problems in Private Yıldız Pediatric Special Education and Rehabilitation Center, and was planned as case presentation due to its rare incidence. Written approval regarding the research of the case was taken from her parents. Within the scope of this study, the case was evaluated 3 times on 01.01.2010, 01.01.2011 and 01.01.2012. Severe hypotonic, postural control deficiency and balance loss were observed in the first evaluation. On the other hand, postural analysis determined hallux valgus and pronation deformities in feet (Figures 1 and 2). Following evaluations were respectively made in the case.

![Figure 1: Hallux valgus deformity in feet.](image1)

![Figure 2: Pronation deformity in feet.](image2)

**Motor function evaluation**

Motor functions of the case (consists of 5 parts as; lying and rolling; sitting; crawling and kneeling; standing; walking; running and jumping) were evaluated using Gross Motor Function Measurement (GMFM) scale [13]. Percentage scores of the case were obtained from each 5 parts, and total score was obtained by calculating the average of percentage scores of each parts. Table 1 shows the evaluation results of GMFM scale repeated three times in one year interval.

| Year | A: Bedding and Rolling | B: Sitting | C: Crawling and Over knees | D: Standing | E: Walking, Running and Jumping | Total |
|------|------------------------|------------|---------------------------|-------------|-------------------------------|-------|
| I    | 32                     | 32         | 12                        | 2           | 0                            | 78    |
| II   | 47                     | 50         | 27                        | 15          | 2                            | 141   |
| III  | 51                     | 60         | 42                        | 37          | 45                           | 235   |

Table 1: The results of GMFM motor functions evaluation of the case.

**Evaluation of disability level**

The disability level of the case was determined using Gross Motor Function Classification Scale (GMFCS), which is applied making classification between 1-5 according to different age groups [14,15]. According to this classification system, disability levels of children are evaluated via their motor functions and indoor and social life limitations [15]. Table 2 shows evaluation results of the case.

| Age | Level |
|-----|-------|
| 2-4 | III   |
| 2-4 | II    |
| 4-6 | II    |

Table 2: Results of GMFCS disability level evaluation of the case.

**Evaluation of functional independence level**

The functional independence level of the case was evaluated using Functional Independence Measure for Children (WeeFIM) scale (the scale consists of 6 parts and 18 items as self-care, sphincter control, mobility-transfer, locomotion, communication, social communication. Each item was scaled up to 7 scores according to independence level) [16]. Table 3 shows weeFIM values regarding the functional independence level of the case.

|       | I. Assessment (01.01.2010) | II. Assessment (01.01.2011) | III. Assessment (01.01.2012) |
|-------|----------------------------|-----------------------------|------------------------------|
| Self-care | 6                          | 6                           | 13                           |
| Sphincter Control | 2                          | 3                           | 8                            |
| Mobility | 2                          | 4                           | 8                            |
| Locomotion | 2                          | 5                           | 8                            |
| Communication | 2                          | 5                           | 14                           |
| Social Networking | 3                          | 9                           | 21                           |
| Total     | 17                         | 33                          | 72                           |

Table 3: The results of weeFIM functional independence evaluation of the case.
The applied physiotherapy program was designed from the exercises used in Bobath treatment approach, which included supine and facedown balance education; improving back and hip extensors knee flexors and abdominal muscles; sitting and balance education; standing with assistive devices; ambulation with and without assistive devices and with obstacles; transferring weight in standing and stairs; ascending and descending stairs; and sense-body awareness education.

**Discussion**

According to subsequent evaluations made through GMFM and weeFIM for throughout 3 years when the case received physiotherapy, a significant increase was observed in the scores of all parameters of GMFM and weeFIM, and an increase at one level in disability level was observed in GMFCS evaluation.

Among GMFM parameters, the results of especially standing and walking, running and jumping parameters are remarkable. The scores obtained from three evaluations of standing parameter are 2, 15 and 37, respectively. Walking, running and jumping parameter obtained 0, 2 and 45 scores, respectively for the years. The increase in these two parameters is remarkable. The low scores obtained in standing and walking, running and jumping parameters in the first evaluation indicate deficiency in motor functions, which may result from hypotony and joint hypermobility observed in the case with Ehlers-Danlos Syndrome. Thanks to the physiotherapy program applied for three years, the improvement in motor functions, which reflected to GMFM scores, can be related with the exercises of Bobath treatment approach.

When the development of the case was examined according to GMFCS, a one level increase was observed according to the system whose focal point is "grading the level representing the skills and limitations of gross motor functions of child". In this system, the classification of present performance in gross motor functions are important, rather than the quality of movement or recovery [15]. Since the case could be able to walk short distance by using walker in the first evaluation, she was classified as level III. In her second evaluation determined as Level II a year after, the case could walk again by using walker but longer distances. In the last evaluation of the case, she was classified as Level II, since she could not realize activities like ascending stairs independently, or jumping, in spite of walking independently. In addition, a progress of one level, which is obtained in one year, is important in gross motor functions. This result may be caused by the Bobath treatment approach including stabilization and postural control exercises in physiotherapy program, and independence gained through aging.

In weeFIM parameters evaluating the case, improvements are observed in all self-care, sphincter control, mobility-transfer, locomotion, communication, social communication parameters. The improvement in the motor functions of the case with general hypotony and motor growth deficiency obtained through physiotherapy program caused significant increase in functional activities. The improvements in especially locomotion and mobility parameters are clearly obtained through exercises from Bobath approach [10], which aims to diagnose and analyze problems together with the participation to functional activities and everyday life activities. The case had improvements in self-care, communication, and social communication parameters with the help of pre-school supportive education programmed received special education center. This development in functional independence is the secondary gain of the independence gained through aging, as in GMFCS.

Literature does not include standard physiotherapy programs applied in EDS-HT disease or hypermobility. Basing on the results of the study applying Bobath treatment approach, this study suggests including Bobath approach in physiotherapy programs aiming to provide postural alignment and improve functions through active participation of individual [1].

This study, designed upon the absence of studies applying Bobath treatment approach in EDS-HT disease, is the first study to examine the effect of Bobath treatment approach, which was included in physiotherapy programs of cases with EDS-HT diagnosis, on treatment targets. In other words, Bobath treatment method, which is used in physiotherapy and rehabilitation, was applied on a new field with no specific protocol. The absence of a scoring system evaluating growth in EDS is the limitation of the present study.

In conclusion, significant increases were observed in the motor function level and independence in everyday life activities of the case with EDS-HT diagnosis, who was applied biopsychosocial treatment program including Bobath treatment approach together with pre-school supportive education and was monitored for three years in terms of physiotherapy. Future studies may be suggested to compare the results of Bobath concept and other physiotherapy approaches in EDS-HT disease, and therefore clinically become a guide by creating effective protocols.

**References**

1. Rombaut L, De Paepe A, Malfait F, Cools A, Calders P (2010) Joint position sense and vibratory perception sense in patients with Ehlers-Danlos syndrome type III (hypermobility type). Clin Rheumatol 29: 289-295.
2. Beighton P, De Paepe A, Steinmann B, Tsipouras P, Wenstrup RJ (1998) Ehlers-Danlos syndromes: revised nosology, Villefranche, 1997. Ehlers-Danlos National Foundation (USA) and Ehlers-Danlos Support Group (UK). Am J Med Genet 77: 31-37.
3. Jasiewicz B, Potaczek T, Tesiorowski M, Lokas K (2010) Spine deformities in patients with Ehlers-Danlos syndrome, type IV - late results of surgical treatment. Scioiosis 5: 26.
4. Indrami D, Helen W, Odiri O (2011) Pregnancy and Delivery in Ehlers-Danlos Syndrome (Hypermobility Type): Review of the Literature. Obstetrics and Gynecology International.
5. Le Talléc H, Lassalle A, Kheniou H, Durufle A, Plassat R, et al. (2006) [Two cases of rehabilitation in Ehler-Danlos syndrome]. Ann Readapt Med Phys 49: 81-84.
6. Rombaut L, Malfait F, Cools A, De Paepe A, Calders P (2010) Musculoskeletal complaints, physical activity and health-related quality of life among patients with the Ehlers-Danlos syndrome hypermobility type. Disability and Rehabilitation 32: 1339-1345.
7. Grahame R (2000) Heritable disorders of connective tissue. Baillieres Best Pract Res Clin Rheumatol 14: 345-361.
8. Murray KJ (2006) Hypermobility disorders in children and adolescents. Best Pract Res Clin Rheumatol 20: 329-351.
9. Malfait F, Wenstrup RJ, De Paepe A (2010) Clinical and genetic aspects of Ehlers-Danlos syndrome, classic type. Genet Med 12: 597-605.
10. www.ibita.org
11. Mayston MJ (2001) People with cerebral palsy: effects of and perspectives for therapy. Neural Plast 8: 51-69.
12. Kollen BJ, Lennon S, Lyons B, Wheatley-Smith L, Scheper M, et al. (2009) The effectiveness of the Bobath concept in stroke rehabilitation: what is the evidence? Stroke 40: e89-97.
13. Palisano R, Rosenbaum P, Walter S, Russell D, Wood E, et al. (1997) Development and reliability of a system to classify gross motor function in children with cerebral palsy. Dev Med Child Neurol 39: 214-223.

14. Palisano RJ, Hanna SE, Rosenbaum PL, Russell DJ, Walter SD, et al. (2000) Validation of a model of gross motor function for children with cerebral palsy. Phys Ther 80: 974-985.

15. Gânel MK (2011) Physiotherapy for children with cerebral palsy. In: Åeljka Petelin GadÅe (ed) Epilepsy in Children “Clinical and Social Aspects (1st ed). Croatia Intech Open Access Publisher, Croatia.

16. Ottenbacher KJ, Msall ME, Lyon N, Duffy LC, Ziviani J, et al. (2000) The WeeFIM instrument: its utility in detecting change in children with developmental disabilities. Arch Phys Med Rehabil 81: 1317-1326.