Original Investigation / Özgün Araştırma

Upper Extremity Functions in Spina Bifida

Spina Bifida'da Üst Ekstremite Fonksiyonları

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ABSTRACT

Objective: In this study it is aimed to investigate the upper extremity functions of children with meningomyelocele (MMC) compared to those of the healthy children

Methods: Twenty-three patients with MMC (MMC group) and 14 healthy children (control group) whose ages were ranging from 7 to 12 were included in the study. The functions of dominant and non-dominant upper extremities of both groups were evaluated by Jebsen-Taylor Hand Function Test (JTT).

Results: The mean ages of MMC group and control group were similar (p=0.476). Sixteen of the patients with MMC had an additional shunted hydrocephalus. The mean timing scores of all 7 activities at JTT of both dominant and non-dominant upper extremity of the MMC group were found to be significantly longer than those of the control group (p<0.05 for all parameters). The mean timing scores of the patients with hydrocephalus to perform 5 of the 7 task by the dominant and 6 by the non-dominant were significantly longer than those of the patients without hydrocephalus (p<0.05 for all parameters). The mean timing scores of the MMC patients without hydrocephalus to perform 4 of the 7 task by the dominant and 4 by the non-dominant were significantly longer than those of the normal control subjects (p<0.05 for all parameters).

Conclusion: The results of this study revealed that the upper extremity functions of children with MMC were incapable in compare to normal children and causes other than hydrocephalus might have negative effects on their hand functioning.

Keywords: Meningomyelocele, occupational therapy, upper extremity

ÖZET

Amaç: Bu çalışmada meningomiyelosel (MMS)'li çocuklarda üst ekstremite fonksiyonlarının araştırılması ve sağlıklı çocuklarla karşılaştırılması amaçlandı.

Yöntemler: Çalışmaya yaşları 7 ile 12 arasında değişen 23 MMS'li hasta (MMS grup) ile 14 sağlıklı çocuk (kontrol grup) dahil edildi. Her iki grubun dominant ve non-dominant üst ekstremite fonksiyonları Jebsen-Taylor El Fonksiyon Testi (JTEFT) ile değerlendirildi.

Bulgular: MMS grubu ve kontrol grubunun yaş ortalaması benzer idi (p=0.476). MMS'li hastaların 16'sında hidrosefali mevcuttu. MMS grubunun dominant ve non-dominant üst ekstremiteye ait JTEFT ile değerlendirilen tüm aktivitelerin ortalama gerçekleştirme sürelerinin kontrol grubuna oranla istatistiksel olarak anlamlı derecede uzun olduğu saptandı (tüm parametreler için p<0.05). Hidrosefalisi olan hastalarda dominant ekstremitede 7 aktivitenin 5'inde, non-dominant ekstremitede ise 6 aktivitede gerçekleştirme süre ortalamalarının hidrosefalisi olmayan hastalara oranla istatistiksel olarak anlamlı derecede uzun olduğu saptandı (tüm parametreler için p<0.05). Hidrosefalisi olmayan hastalarda dominant ve non-dominant ekstremitede 7 aktivitenin 4'ünde ortalama aktivite gerçekleştirme sürelerinin kontrol grubuna oranla istatistiksel olarak anlamlı derecede uzun olduğu saptandı (tüm parametreler için p<0.05).

Sonuçlar: Bu çalışma sonuçları MMS'li çocukların üst ekstremite fonksiyonlarının normal çocuklar ile karşılaştırıldığında yetersiz olduğunu ve hidrosefalinin el fonksiyonlarına olumsuz etkileri olabildiğini ortaya koymaktadır.

Anahtar sözcükler: Meningomiyelosel, iş-uğraşı terapisi, üst ekstremite

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Introduction

Spinal dysraphism, named also as spina bifida or neural tube defect, is a generalized term used for the incomplete development of the spinal cord. Meningomyelocele (MMC) which is the most common form and generally used as a synonym of spina bifida, has been known as the second most physically handicapping condition after cerebral palsy among children (1,2,3).

MMC is a complex syndrome causing various neurological clinical symptoms, the most common of which are the paralysis of lower extremities, neurogenic bladder and bowel dysfunctions. Lower extremity and spinal deformities are the most common features of MMC cases. Additionally, children with MMC may also have different neurological conditions such as hydrocephalus, syringomyelia, tethered cord, and the Arnold Chiari malformation (4-8). It is well known that all these neurological complications increase the morbidity and mortality of the MMC patients. It has also been reported that these complications may negatively affect the upper extremity functions and activities of daily living (ADLs) of MMC patients (8-10).

Arms and hands are known to be the most developed neuromuscular organs of human beings and to have great importance in fulfilling ADLs. Functional usage of upper extremities is necessary to perform activities requiring strength and coordination such as grasping, holding, manipulating and feeling objects. Upper extremity functionality is extremely important not only for activities such as personal hygiene, self-care, dressing, eating but also for communication and mobilization. In terms of ADLs, upper exremity functions being already important in healthy individuals are even more important in patients with neurological diseases leading to paralyses of lower extremities.

Although many studies have investigated the lower extremity deformities, gait abnormalities, spinal deformities, bladder and bowel dysfunctions in patients with MMC, relatively very little research is available concerning upper extremity dysfunctions resulting from MMC (11-21). During 1970's three clinical studies by Grimm, Anderson and Sand et al it was suggested for the first time those children with MMC might have impaired hand function. Later upper extremity dysfunction was confirmed by other investigators (22-29). Poor upper extremity functioning was mostly explained by hydrocephalus, cerebellar dysfunction due to Chiari malformation, and cervical spinal lesions in the literature (22,27,30,31). However in 1997, Muen et al. compared the hand functions of patients with MMC and shunted hydrocephalus to patients with isolated shunted hydrocephalus and to normal subjects (31). They reported that MMC patients had weaker power in the small hand muscles, and poorer fine motor control and coordination than both normal and hydrocephalus subjects. This study prop up that causes other than hydrocephalus might negatively influence upper extremity functioning in patients with MMC.

The aim of this prospective study was to investigate the upper extremity functions of children with MMC compared to healthy children, and to determine the factors affecting upper extremity function in this patient population.

Methods

This study involved 23 patients with MMC fulfilling the inclusion criteria of the study (MMC group) out of 40 applications to Kocaeli University Department of Physical Medicine and Rehabilitation from December 2006 to September 2007 and a control group of 14 healthy children (control group).

The medical history of each MMC patient was recorded by a specialist of Physical Medicine and Rehabilitation and their systemic, musculo-skeletal and neurological examinations were performed. Patients were excluded from the study if they were younger than 7 and older than 12 years of age, were illiterate, had cognitive function disorder or inadequate cognitive functions. Patients with upper motor neuron deficits like spasticity, sensory impairment, cerebellar dysfunction, positive contracture and/or insufficient body balance (who were unable to maintain the supported sitting posture for at least half an hour) were also excluded from the study. Inclusion to the control group required healthy children aged between 7-12 who could read and write. Ethical approval for the study was received from the ethical comitee of Kocaeli University Faculty of Medicine. Informed consent was obtained from every child and his/her parents.

The demographic properties of the patients such as age, dominant hand, presence of hydrocephalus, level of the lesion were examined and recorded. The level of the lesion was characterized according to the criteria of International Myelodysplasia Study Group. Hand functions of both the MMC and control groups were evaluated by Jebsen – Taylor Hand Function Test (JTT).

Patients were asked to perform the standardized 7 functions described in JTT including writing, simulated page turning, lifting small objects, simulated feeding, stacking, lifting large-lightweight objects and lifting large-heavy objects. All the examinations were conducted on a laboratory table. The subjects were

so positioned that they could only sit straight on an adjustable, comfortable chair and face the table with adequate lightining. The height of the chair was adjusted so that the child's forearm was parallel to the surface of the table. The test to be done was explained and demostrated to the subject by the occupational therapist before the experiment to make certain the subject understands the instructions completely. The same test materials were used and the tests were applied by the same occupational therapist both to MMC and control groups. The patients were instructed to perform 7 defined tasks as rapidly and accurately as possible according to written standardized instructions in the testing set. Each duty was repeated first with the non-dominant hand and later with the dominant one. Each test was timed in seconds by an electronic Digital Readout Stopwatch and dominant and non-dominant hand JTT timings were recorded for analysis.

Statistical analysis was performed using SPSS 12 programme for Windows. Demographical data were expressed using mean and standard error of means. The dominant and non-dominant hand JTT results of MMC and control groups were compared using Mann Whitney-U Test. Results were considered significant when $p<0.05$.

Results

Seventeen of the 40 MMC patients were excluded from the study. The reasons for exclusion were inappropriate age in 9 patients, illiteracy in 4, spasticity in 1, insufficient body balance in 2, and cognitive disorder leading to insufficient communication in 1.

The demographical data of the MMC and the control groups are given in Table 1. No statistically significant difference was found between the MMC and control groups with respect to age, gender, and dominant hand ($p>0.05$ for all parameters).

With respect to International Myelodysplasia Study Group criteria the level of the lesion of the MMC group was found to be lumbar in 19(82.6%), thoracic in 3(13,0%), and lumbosacral in 1(4.3%) patients. Sixteen (%69,6) of the 23 patients in the MMC group had hydrocephalus. All of them were treated by a shunt operation. Seven of the MMC patients had no additional hydrocephalus or other associated neurologic problems.

Table 2 shows the JTT performance timing results of the dominant and non-dominant hands of the MMC and control groups. The mean timing scores of the MMC group to perform the tasks by the dominant and non-dominant hands were significantly longer than those of the control group ($p < 0.05$ for all parameters).

The comparison of JTT performance timing results of the MMC patients with and without hydrocephalus was given in table 3. The mean timing scores of the patients with hydrocephalus to perform writing, simulated feeding, stacking, lifting large light-weight objects, lifting large heavy objects by the dominant and writing, simulated page turning, lifting small objects, simulated feeding, stacking, lifting large heavy objects by the non-dominant hands were significantly longer than those of the patients without hydrocephalus ($p < 0.05$ for all parameters).

The comparison of JTT performance timing results of the patients without hydrocephalus to normal control subjects was given in table 4. The mean timing scores of the MMC patients without hydrocephalus to perform simulated page turning, lifting small objects, lifting large light-weight objects, lifting large heavy objects by the dominant and simulated page turning, lifting small objects, stacking, lifting large light-weight objects by the non-dominant hands were significantly longer than those of the normal control subjects ($p < 0.05$ for all parameters).

Discussion

The aim of this study was to investigate the upper extremity functions of children with MMC in comparison to normal, healthy children, to characterize the upper extremity functional deficits, and to determine the underlying factors of upper extremity dysfunctions.

Table 1. Demographical data of the MMC and control groups.

	MMC Group (n= 23)	Control Group (n=14)	p
Age	9.0±2.0	9.0±2.4	0.476
Gender	12 (%52) female 11 (%48) male	9 (%64) female 5 (%36) male	0.216
Dominant hand	18 (%78) right 5 (%22) left	10 (%71) right 4 (%29) left	0.274

Table 2. Comparison of MMC and control groups JTT performance time results.

	MMC Group (n=23)	Control Group (n=14)	p
Dominant Hand			
Writing	69,6±9,5	18,1±2,1	<0,001
Simulated page turning	15,5±1,5	6,1±0,6	<0,001
Lifting small objects	16,1±1,7	5,9±0,3	<0,001
Simulated feeding	38,1±7,9	11,1±0,7	0,002
Stacking	10,3±1,7	3,7±0,3	0,001
Lifting large-lightweight objects	13,6±2,7	4,1±0,2	0,002
Lifting large-heavy objects	18,1±5,6	4,7±0,4	0,026
Non Dominant Hand			
Writing	90,7±10,3	39,5±4,3	<0,001
Simulated page turning	22,0±3,4	7,8±0,7	<0,001
Lifting small objects	25,5±5,9	6,5±0,2	0,004
Simulated feeding	42,0±8,0	13,4±0,9	0,002
Stacking	14,8±5,0	4,2±0,4	0,046
Lifting large-lightweight objects	17,2±4,5	4,7±0,4	0,011
Lifting large-heavy objects	22,1±6,0	5,6±0,5	0,012

Table 3. Comparison of MMC patients with and without hydrocephalus JTT performance time results.

	MMC Group with hydrocephalus (n=16)	MMC Group without hydrocephalus (n=7)	p
Dominant Hand			
Writing	85,6±11,2	33,4±7,5	0,005
Simulated page turning	17,1±1,7	11,7±2,5	0.107
Lifting small objects	18,1±2,2	11,4±1,1	0,088
Simulated feeding	47,3±10,4	17,0±4,6	0,015
Stacking	12,5±2,1	5,4±0,8	0,017
Lifting large-lightweight objects	16,3±3,7	7,3±1,2	0,029
Lifting large-heavy objects	22,4±7,9	8,4±1,6	0,038
Non Dominant Hand			
Writing	106,7±12,4	53,9±8,6	0,009
Simulated page turning	25,9±4,6	13,0±1,6	0.010
Lifting small objects	31,6±8,1	11,7±1,1	0,021
Simulated feeding	51,6±10,6	20,1±4,7	0,019
Stacking	18,6±7,0	6,3±0,7	0,023
Lifting large-lightweight objects	20,7±6,3	9,3±1,6	0,060
Lifting large-heavy objects	28,0±8,3	8,9±1,6	0,014

Table 4. Comparison of MMC patients without hydrocephalus and control groups JTT performance time results.

	MMC Group without hydrocephalus (n=7)	Control Group (n=14)	p
Dominant Hand			
Writing	33,4±7,5	18,1±2,1	0,067
Simulated page turning	11,7±2,5	6,1±0,6	0.008
Lifting small objects	11,4±1,1	5,9±0,3	<0,001
Simulated feeding	17,0±4,6	11,1±0,7	0,547
Stacking	5,4±0,8	3,7±0,3	0,101
Lifting large-lightweight objects	7,3±1,2	4,1±0,2	0,004
Lifting large-heavy objects	8,4±1,6	4,7±0,4	0,019
Non Dominant Hand			
Writing	53,9±8,6	39,5±4,3	0,191
Simulated page turning	13,0±1,6	7,8±0,7	0.013
Lifting small objects	11,7±1,1	6,5±0,2	<0,001
Simulated feeding	20,1±4,7	13,4±0,9	0,330
Stacking	6,3±0,7	4,2±0,4	0,020
Lifting large-lightweight objects	9,3±1,6	4,7±0,4	0,003
Lifting large-heavy objects	8,9±1,6	5,6±0,5	0,086

The dominant and non-dominant hand functions of 23 patients with MMC were evaluated by JTT and compared to the hand functions of 14 healthy children. JTT was standardized by researchers Jebsen and Taylor in 1969 and the clinical validity and reliability of the test have been proven by many clinical studies (25,32-36). This test comprises 7 timed motor tasks which require speed, dexterity, and strength. In our study, it has been detected that subjects with MMC performed all 7 tasks in a significantly longer mean duration of time by both dominant and non-dominant hands compared to normal healthy subjects. JTT was used by many other investigators in order to evaluate the hand functions of MMC patients (25,26,32) and poor upper extremity functioning was obtained in all these studies. Several explanations have been accused for poor upper extremity functions in patients with MMC. Hydrocephalus had been the most common implicated cause of upper extremity impairment by most of these studies (22,25-27). High level of spinal cord lesion, cerebellar dysfunction due to Chiari malformation, mental retardation, cerebral palsy were also been proposed to account for poor upper extremity functions in patients with MMC (25,31,37,38). As our exclusion criteria enclosed cognitive dysfunctions, upper motor neuron findings, and cerebellar involvement, and cervical spinal lesion these factors were not been expected to be confounders in our patient population. In our study patients with shunted hyrocephalus were found to have poorer hand functions compared to both healthy controls and MMC patients with no additional hydrocephalus. Therefore this study also confirmed adverse effects of hydrocephalus on upper extremity functions. However the study also proposed that factors other than hydrocephalus, cerebellar dysfunction, cerebral palsy, mental retardation, high spinal lesion might be responsible from poor hand functioning in this patient population, because MMC patients with no additional hydrocephalus, IQ deficit, cerebral, cerebellar, or cervical pathology also showed poorer hand function than normal subjects. The upper extremity dysfunction in these children might be explained by motor learning deficits resulting from the compensatory usage of upper extremities in many ADLs to provide balance and to support lower extremities. Learned non-use phenomenon due to paucity of experience might be the main cause of poor hand functioning in these patients. Moreover this might be a worsening factor in the MMC patients with hydrocephalus or other associated neurological complications.

The results of this study revealed that the upper extremity functions of children with MMC were incapable in compare to normal children and causes other than hydrocephalus might have negative effects on their hand functioning. The present medical rehabilitation programs of MMC patients generally focus on exercise and physical

therapy interventions aiming to improve lower extremity dysfunctions/deformities and upper extremities are often neglected (39,40). As poor upper extremity functioning was shown by many studies in the literature and the present study, occupational therapeutic procedures aiming to improve upper extremity functioning must be an important part of the therapeutic process in patients with MMC (8-10,22-24). This will contribute to the independence of patients in their self-care, ADLs, and to socialization.

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