Original Article

Demographic and clinical features and rehabilitation outcomes of patients with Friedreich ataxia: A retrospective study

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ABSTRACT

Objectives: This study aims to investigate the demographic and clinical features, and rehabilitation outcomes of Friedreich Ataxia patients in the inpatient setting.

Patients and methods: Between January 2000 and December 2012, a total of 37 Friedreich Ataxia patients (26 males, 11 females; mean age 29.2±9.6 years; range, 16 to 53 years) were included in the study. Demographic and clinical features of the patients and rehabilitation outcomes were reviewed retrospectively. Duration of rehabilitation, age at onset of disease, first onset of extremity, were determined. The Functional Ambulation Category (FAC) and Hoffer Ambulation Scale levels were used to assess the ambulation levels of patients. The Barthel Index scores were used to assess daily living activities of the patients before and after the treatment.

Results: The mean rehabilitation duration was 4.7±1.4 weeks. There was cardiac involvement in nine (24.3%) patients and obstructive/restrictive pulmonary disease in five patients (13.5%). A total of 33 patients (89.2%) had severe ataxia, four (10.8%) had mild ataxia, 33 (89%) had dysarthria, 18 (48.6%) had scoliosis, 16 (43.2%) had kyphose, 15 (40.5%) had pes cavus, five (13%) had optic atrophy, and two (6%) had nystagmus of all patients. At the end of the rehabilitation program, all patients were discharged with functionally improved levels based on the FAC, Hoffer Ambulation Scale (p<0.05), and Barthel Index (p<0.001), indicating a statistically significant difference.

Conclusion: An appropriate and comprehensive rehabilitation program improves the ambulation level and independence level of daily activities in patients with Friedreich ataxia, which highlights the importance of rehabilitation in these patients.

Keywords: Friedreich ataxia; inpatient rehabilitation; rehabilitation outcome.

Friedreich ataxia (FA) is a recessively inherited autosomal neurodegenerative disease, which onsets in childhood, adolescence, or early adulthood period.[1] Respective gene was analyzed in 1996 and coded as frataxin.[1] Frataxin has an important role on mitochondrial iron homeostasis. Frataxin deficiency causes iron and free radical accumulation in mitochondria and an increase in oxidative stress, which this results in cellular damage and death. High-energy consuming cardiac muscles, pancreas islet cells, and neural system are more sensitive to iron overload. As a result, a degeneration occurs in the dorsal root ganglion cells, posterior cord, spinocerebellar and corticospinal tract, and cerebellar dentate nucleus.[2] The prevalence is estimated to be 3 to 4 in 100,000.[3] The initial symptom onset often occurs in the puberty period, and the estimated life expectancy is about 40 to 50 years.[3] Men and women are considered to be equally affected.[4] Ataxia, hearing loss, scoliosis, foot deformity, hypertrophic cardiomyopathy, and diabetes mellitus can be accompanied.[4] As the nature of the disease is progressive, most of the patients become dependent on wheel chairs within 10 to 15 years.[3] Yet, there is no pharmacological treatment, although attempts are still made to decelerate the deterioration and to develop the treatment methods.[4,5] A majority of the cardiomyopathy patients are lost in third or fourth decade of lie due to cardiac arrhythmia or congestive failure. Kyphoscoliosis and restrictive pulmonary function disorder are also other reasons which are associated with mortality.[6]

Ataxia has a progressive characteristic and initially begins from the lower extremities and spreads to the body and upper extremities. Neurological symptoms such as dysarthria, loss of deep tendon reflexes, loss of proprioception is accompanied by movement incoordination.[7] In cranial nerve examination, decrease in visual acuity, horizontal nystagmus, hearing loss, dysarthria, and dysphagia may be present. Rapid and repeating movements are slowed down. Intentional tremor may be observed. Walking is wide-based due to the coexistence of the posterior column involvement, spasticity and cerebellar ataxia. Although asymmetrical involvement is not rare, the involvement is usually symmetrical. There is a mild dementia at the full-blown disease. Optic atrophy associated with the vision loss is not rare.[8]

Rehabilitation methods such as physical and occupational therapy, specific coordination exercises, speech therapy, posture and gait training are used in the rehabilitation of the patients with FA.[9] Although there are a lot of genetic researches, experimental treatments and articles related with the neurological findings, orthopedic deformities and surgical interventions, cardiac pathologies for the patients with FA in the literature, there is a limited number of studies on the rehabilitation outcomes of these patients. In our study, we aimed to investigate the demographic and clinical features, functional levels, and rehabilitation outcomes of the FA patients in the inpatient setting in our hospital.

PATIENTS AND METHODS

An approval of our study was obtained from our hospital local ethics committee. A written informed consent was obtained from each patient. The study was conducted in accordance with the principles of the Declaration of Helsinki.

Our hospital is one of the largest national rehabilitation centers in Turkey. Friedreich ataxia patients, who were clinically diagnosed based on the Harding diagnostic criteria[10,11] by a neurologist, applied to our hospital for rehabilitation. Harding diagnostic criteria were as follows: Primary: Age of onset before 25 years/autosomal recessive inheritance/progressive limb and gait ataxia/absent tendon reflexes in the legs/extensor plantar response/axonal sensory neuropathy/dysarthria (if five years passed from onset II). Secondary: scoliosis/pyramidal weakness in lower limbs/absent reflexes in upper limbs/distal loss of position and vibration sense/cardiomyopathy III). Additional:

nystagmus/optic atrophy/deafness/distal amyotrophy (weakness and wasting)/pes cavus/diabetes. However, there are significant variations about the onset, clinical findings, and the progression rate of the disease.[11,12]

A total of 37 FA patients, (26 males, 11 females; mean age 29.2±9.6 years; range, 16 to 53 years) who received inpatient rehabilitation in our hospital between January 2000 and December 2012 years were included in the study. Archive files of patients were reviewed. Demographic and clinical features and pre- and post-rehabilitation functional levels were retrospectively analyzed. Duration of rehabilitation, age (years), educational level, age at onset of disease (years), and first involvement limb of FA were determined for all patients before and after treatment.

As well as the bowels and bladder of patients, daily living activities such as feeding, bathing, grooming, transferring, mobility, toilet, and stair use were assessed with the Barthel Index.[13] The Barthel Index is a well-documented instrument which is currently in use for measuring the outcomes of a rehabilitation and meets adequate validity and reliability.[14] Total maximum score is 100 in this index.

The Functional Ambulation Category (FAC)[15,16] and Hoffer Ambulation Scale[17,18] were used to evaluate functional ambulation levels before and after treatment. The FAC has six categories ranging from 0 (non-functional ambulation) to 5 (independent). The intermediary categories include the quantify levels of assistance, supervision, and independent yet limited mobility. Ambulation levels were categorized by the Hoffer Ambulation Scale as non-ambulation, non-functional (therapeutic) ambulation, household, community and normal ambulation. Although the validity and reliability studies of the FAC and Hoffer Ambulation Scales for FA are not available in the literature, they are widely used scales for the rehabilitation patients and were previously used in many studies.[15-18]

None of the patients were receiving pharmacological treatment for FA. Rehabilitation of each patient was planned according to the clinical presentation of the disease. During the rehabilitation program, balance and coordination exercises, stretching and strengthening exercises, range of motion (ROM) exercises, cardiopulmonary exercises, posture exercises and ambulation training, postural biofeedback, spasticity management, occupational therapy, speech therapy, scoliosis and breathing exercises were used. A patient-specific rehabilitation program was implemented two

hours per day during hospitalization. For ambulation, lower limb orthosis and walking aids (i.e., crutch, cane, or walker) were given to the patients who were able to ambulate functionally.

Statistical analysis

Statistical analysis was performed using the SPSS for Windows version 11.0 software (SPSS Inc., Chicago, IL, USA). Analyses were completed by transferring the data into the IBM SPSS Statistics 23.0 extension bundle (IBM Corp., Armonk, NY, USA). Categorical variables were expressed in number and percentage, while descriptive statistics were expressed in mean ± standard deviation (SD), and median (min-max) values for the numeric variables.

For the numeric variables, the Shapiro-Wilk normalization test was used and it was observed that not all of the numeric variables were normally distributed. As numeric variables did not conform with the normal distribution, non-parametric statistical methods were used. Our FAC and Hoffer Ambulation Scale values were divided into two groups as non-ambulated/non-functionally ambulated and ambulated. The relationship between two independent categorical variables was analyzed using the chi-square test, and the relationship between two dependent categorical variables was analyzed using the McNemar's test. The differences between two dependent numeric variables were analyzed by the Wilcoxon analysis. The differences between two independent groups were investigated by the Mann-Whitney U test. The correlations between two independent numeric variables were interpreted by the Spearman's Rho coefficient. A p value of <0.05 was considered statistically significant.

The power of the study was determined via the G*Power 3.1 software program (Heinrich Heine University, Dusseldorf, Germany) bundle. When evaluating the power analysis, the Barthel Index results, the study's one of the most basic hypothesis, were used. Based on these results, the effect size was calculated as 0.77. With the 0.05 significance and 95% confidence interval, the power of 37 participants was found to be 98%.

RESULTS

Demographic findings including age (years), age at first symptom onset (years), rehabilitation duration (weeks) and sex (female/male), marital status (married/single/widow), and educational level (elementary/middle school/high school/college) of patients are shown in Table 1.

Furthermore, in all patients, the initial symptoms were ataxia and gait instability. The first involvement was the lower limb distal in 32 patients (86.5%) and lower limb proximal in five patients (13.5%).

Comorbidities are shown in Table 2. Cardiac involvement was the most seen systemic involvement with a rate of 9 (24.3%). Also, 12 (32.4%) of the patients had other systemic diseases such as obstructive/restrictive pulmonary disease, five (13.5%) had glomerulonephritis, hemolytic anemia, vasculitis, diabetes, and also eight (21.6%) of the patients had a previous fracture history (upper extremity fractures n=3; lower extremity fractures n=5).

Urodynamic analyses were applied for the patients with neurogenic bladder in five patients (13.5%). Three (8.1%) of the patients were recommended using permanent urinary catheter, one (2.7%) of

Table 1. Demographic findings of the patients with Friedreich ataxia

Demographic findings	n	%	Mean±SD	Median	Min-Max
Age (years)			29.2±9.6	27	16-53
Age at first symptom onset (years)			15.8±6.5	14	5-35
Rehabilitation duration (weeks)			4.7±1.4	4	2-8
Gender					
Female	11	30			
Male	26	70			
Marital status					
Married	12	33			
Single	23	62			
Widow	2	5			
Educational level					
Elementary school	12	32			
Middle school	11	30			
High school	13	35			
College	1	3			

SD: Standard deviation; Min: Minimum; Max: Maximum.

Table 2. Comorbid diseases observed in our patients with Friedrich ataxia

Comorbid diseases	n	%
Cardiac problems		
Minimal mitral regurgitation + tricuspid regurgitation	2	5.4
Minimal mitral regurgitation	1	2.7
Cardiomyopathy	3	8.1
Congestive cardiac failure	3	8.1
Visual problems		
Optic atrophy	5	13
Nystagmus	2	6
Auditory problems		
Hearing loss	4	11
Speech disorder		
Dysarthria	33	89
Aphasia	1	3
Swallowing disorder		
Dysphagia	2	5.4
Cognitive disorder		
Cognitive dysfunction	7	18.9
Deformities and fracture		
Pes cavus	15	40.5
Scoliosis	18	48.6
Kyphosis	16	43.2
Pes equinovarus	1	2.7
Genu recurvatum	13	35.1
Fracture	8	21.6
Mallet finger	4	10.8
Neurogenic bladder		
Hyperreflexic synergistic bladder	4	10.8
Hyperreflexic dyssynergistic bladder	1	2.7

the patients was recommended applying a clean intermittent catheterization program, and one (2.7%) of patients was recommended urinating spontaneously.

On neuromusculoskeletal system examination, only four (11%) of the patients had limited ROM in their lower limbs, including the hip and ankles. Their upper extremity muscle strengths were normal, although muscle weakness was more apparent in their lower extremities. Based on the manual lower extremity muscle testing, the muscle strength was 2-3/5 in 13 patients (35.1%). Inability was observed in the cerebellar tests of 35 patients (94.6%). Severe ataxia in 33 patients (89.2%), mild ataxia in four patients (10.2%), and intentional tremor in seven patients (18.9%) were observed.

Most of the ambulated patients were able to ambulate without any assistive orthosis. Only 11 (29.7%) of these patients were using an assistive device to ambulate. Three patients (8.1%) were using ankle-foot orthosis (AFO), two patients (5.4%) were using knee-ankle-foot orthosis (KAFO), and six patients (16.2%) were using a device to prevent the knee hyperextension.

At the end of the rehabilitation program, all of the patients were discharged with functionally better levels, based on the FAC, Hoffer Ambulation Scale, and Barthel Index (Table 3). After the rehabilitation, a statistically significant difference was found in the FAC, Hoffer Ambulation Scale (p<0.05), and Barthel Index (p<0.001) (Table 4).

Relationship between variables and correlation analyses

When the correlation between age and Barthel Index values was analyzed, a negative correlation was identified (p=0.032, r=-0.354). Functional level became worse with ageing. Furthermore, the mean age of the non-ambulating patients with severe ataxia

Table 3. Barthel Index scores, and Functional Ambulation Category, Hoffer Ambulation Scale scores in patients with Friedreich Ataxia at pre- and post-treatment

Functional assesment	Pre-treatment				Post-treatment			
	n	%	Mean±SD	Min-Max	n	%	Mean±SD	Min-Max
Barthel Index			46.8±8.8	35-65			54.2±10.3	40-70
Functional Ambulation Category level								
0	11	29.7			10	20.7		
1	8	21.6			2	5.4		
2	2	5.4			2	5.4		
3	15	40.5			9	24.3		
4	1	2.7			13	35.1		
5	0	0			1	2.7		
Hoffer Ambulation Scale level								
Normal ambulation	0	0			1	2.7		
Community ambulation	1	2.7			17	45.9		
Household ambulation	21	56.8			10	27		
Non-functional ambulation	15	40.5			9	24.3		
Non-ambulation	0	0			0	0		

SD: Standard deviation; Min: Minimum; Max: Maximum.

Table 4. Comparison of Barthel Index scores, and Functional Ambulation Category, Hoffer Ambulation Scale scores in patients with Friedreich Ataxia at pre- and post-treatment

Rehabilitation Outcome	Pre-treatment				Post-treatment				
	n	%	Mean±SD	Min-Max	n	%	Mean±SD	Min-Max	*p*
Barthel Index			46.8±8.8	35-65			54.2±10.3	40-70	<0.001*
Functional Ambulation Category									0.016*
Non-ambulated/non-functional ambulated	21	56.8			14	37.8			
Ambulated	16	43.2			23	62.2			
Hoffer Ambulation Scale									0.031*
Non-ambulated/non-functional ambulated	15	40.5			9	24.3			
Ambulated	22	59.5			28	75.7			

SD: Standard deviation; Min: Minimum; Max: Maximum; * p<0.05 statistically significant.

was significantly higher than the ambulating patients. However, no relationship between age at the first onset of symptom and ataxia levels and FAC; Hoffer Ambulation Scale levels (p>0.05), and Barthel Index scores (p=0.772, r=-0.049) was found.

We also evaluated the relationship between gender and rehabilitation outcomes. As a result of chi-square analysis, a statistically significant relationship was found between gender and FAC before and after the treatment. In this respect, it was found that the functional levels and ambulation of the males were significantly higher than the females' (p<0.05). It was observed that the male patients showed better functional outcomes.

We evaluated the relationship between ambulation levels and ataxia levels of the patients. There was a significant relationship between pre- and post-treatment FAC, Hoffer Ambulation Scale levels. In patients with mild ataxia, ambulation levels also improved.

In addition, there was a statistically significant relationship between kyphosis, scoliosis, and ambulation condition of patients. Accordingly, in the non-ambulated patients, kyphosis and scoliosis rates were significantly higher than the patients who were ambulated (kyphosis p=0.018 scoliosis p=0.009). In the patients with kyphosis and scoliosis, functional levels were found to be worse.

However, no significant relationship was found between pes cavus and functional improvement (p>0.05). Also, no significant relationship was found between the rehabilitation duration (p>0.05), cardiac involvement (p>0.05), and functional level.

DISCUSSION

Although there are many genetic researches, experimental treatments, and articles related with

neurological findings, orthopedic deformities and surgical interventions, cardiac pathologies for the patients with FA in the literature, there is a limited number of studies on the rehabilitation outcomes of these patients. Therefore, we are unable to find enough literature data to discuss our outcomes. As far as we researched, our study is one of the first researches, which presents the comprehensive rehabilitation outcomes of the patients with FA.

Reetz et al.[19] found that the mean age of the patients was 33.9±13.8 years. They reported that the first onset age of the disease was 15.7±10.4 years. The mean age of the patients in the study of Dulgeroglu et al.[6] was 23.1±9.2 years. In the study of Stephenson et al.,[20] the authors found that the mean age was 29.4±9.0 years, and first onset age of the disease was 19.5±8.1 years. In the Ciancarelli et al.'s[21] study, the mean age of the patients was 32.5±10.8 years. In our study, the mean age was 29.2±9.6 years, and first onset age of the disease was 15.8±6.5 years, consistent with previous studies.

Reetz et al.[19] assessed 592 patients in the European FA Consortium and, of these patients, 54% females and 46% were males. There were 33.4% female and 66.7% male patients in the study of Ciancarelli et al.[21] There were 41.4% female and 58.6% male patients in the study of Milne et al.[22] In our study, there were 30% female and 70% male patients. In the study of Dulgeroglu et al.,[6] there were 30% female and 70% male patients. Although our rates are consistent with the other studies conducted in Turkey, this is not a prevalence study. In the literature, it has been stated that the female/male ratio is close to the each other.[8]

In the Reetz et al.'s[19] study, it was shown that 354 (60%) of the patients were early-onset (≤14 years), 137 (23%) of the patients were mid-onset (15-24 years old), and 100 (17%) of the patients were late-onset (≥25 years). Followed by the mid-onset patients, early-onset

patients had a more progressive disease, while the disease progress was slower in late-onset patients. Among our patients, the number of patients with late-onset was very low. In addition, our study is not a progression study. However, when the current results were examined, we found no relationship between the age at first onset of symptom and FAC, Hoffer Ambulation Scale levels (p>0.05), and Barthel Index scores (p=0.772, r=-0.049), showing the functional levels.

Dulgeroglu et al.[6] found cardiac problems in four patients (13.3%); Reetz et al.[19] found cardiomyopathy in 27 patients (5%).[19] In our study, we found cardiac problems in nine patients (24.3%). We believe that this is caused by early referral of the patients to the physicians for their cardiac complaints, and it may be the result of the developments of early diagnostic methods.

Pandolfo[4] reported that it could be observed optic atrophy in 30% of patients and sensorineural hearing loss in 20% of patients. We also observed optic atrophy in 13% and hearing loss in 11% of our patients. Accompanying visual and auditory diseases were lower in our patients.

In the Cochrane databases of Kearney et al.,[23] they found that the mean rate of incidence of pes cavus was 10%. Dulgeroglu et al.[6] found dysarthria in 56.6%, nystagmus in 33.3%, scoliosis in 56.6%, and pes cavus in 56.6% of patients. Reetz et al.[19] found scoliosis in 25% of patients. In our study, we found dysarthria in 89%, nystagmus in 6%, pes cavus in 40.5%, and scoliosis in 48.6% of patients. For the patients with scoliosis, we were unable to reach enough data with regard to the scoliosis type and the angle; however, we found that orthosis or surgery was recommended to none of the patients In the light of these data, it was assumed that their scoliosis angles were small.

Goulipian et al.[24] demonstrated that rehabilitation and orthopedic shoes reduced the pain, fatigue, falling, and ankle sprain and improved the quality of life and ability to be independent. During the rehabilitation, they reduced the ankle problems and need to use walking aids of a male patient and increased the step length, walking speed and cadence. In our study, we also improved our FA patients' functional independence by giving them an AFO or a KAFO in addition to the orthopedic shoes, and reduced the joint problems.

In another study, Stephenson et al.[20] compared eight FA patients with eight healthy individuals. There were six male and two female patients. They found that the walking speeds of FA patients were significantly slower than the control group. They also found a negative correlation between the first onset FA age and walking parameters. They concluded that the lack of stability contributed to the gait disorders in adult FA patients. In addition, Reetz et al.[19] found the gait instability as 78%. We also found a significant relationship between ataxia level, showing the lack of the stability and FAC and Hoffer Ambulation Scale levels (p<0.05). Ambulation became worse in patients with severe ataxia. In this respect, our results are similar with the results of Stephenson et al.[20] However, we observed no relationship between the age at onset of first symptom and FAC, Hoffer Ambulation Scale levels (p>0.05), and Barthel Index scores (p=0.772 r=-0.049).

In the study of Ciancarelli et al.[21] study which was conducted in 24 FA patients, they reported that there were also cognitive function and mood disorders. They demonstrated that the rehabilitation should be added to the conventional neuromotor approaches. As it is a chronic disease, we also believe that FA patients should receive cognitive and psychosocial rehabilitation.

For the level 1, Dulgeroglu et al.[6] reported that functional community ambulation rate of the patients increased to 14 (46.6%) at discharge, while it was 11 (36.6%) at admission based on the Hoffer Ambulation Scale levels. For the level 2, they showed that the rate of patients who were household ambulation increased to 7 (23.3%) at discharge, while it was 4 (1.3%) at admission. In our study, according to the Hoffer Ambulation Scale levels, the rate of the functional community ambulation of the patients increased to 17 (45.9%) at discharge, while it was 1 (2.7%) at admission. The rate of the patients who were ambulating at household, decreased to 10 (27%) at discharge, while it was 21 (56.8%) at admission. Non-functional ambulation rate of the patients decreased from 15 (40.5%) to 9 (24.3%). Most of our patients improved to a functional community ambulation level (Table 3). Developments in rehabilitation methods and patient awareness about early starting of the rehabilitation program can be thought as the reasons.

Dulgeroglu et al.[6] found a significant relation between rehabilitation duration and functional outcomes. However, we were unable to find any significant correlation in our study (p=0.0361 r=-0.155). Dulgeroglu et al.[6] reported that the mean rehabilitation duration was 37 days. Our mean rehabilitation duration was 4.72 weeks (average 33 days). Rehabilitation

durations of both studies are similar. Thus, we believe that there is a need for more comprehensive studies, which would determine the optimum duration, and intensity and content of the rehabilitation program for the patients with FA.

Although Dulgeroglu et al.[6] did not find any significant difference among the patients whose ambulation levels changing in respect of kyphoscoliosis. We found a statistically significant relationship between kyphosis, scoliosis, and the ambulation state of the patients. Accordingly, kyphosis and scoliosis rates in the non-ambulating patients were significantly higher than kyphosis and scoliosis rates in the patients who were ambulating (kyphosis p=0.018 scoliosis p=0.009). In the patients with kyphosis and scoliosis, the functional levels were worse. We believe that the ambulation would be more difficult in case of vertebral orthopedic deformities, as kyphoscoliosis and the other deformities may disturb the biomechanical and postural balance. Also, kyphoscoliosis may compress the spinal cord or radicular nerve and it may cause neuromuscular defects.

Croarkin et al.[12] investigated the correlation between unsupported independent walking and supported walking, by using walking parameters and FA Rating Scale scores in a study, which were conducted over 14 children and 24 adolescent. The mean age was 13±2.3 years, and the mean first onset age of the disease was 8±3.1 years. They found that 15 patients (39.5%) were walking with an assistance, while 23 patients (60.5%) were able to ambulate without any support or assistance. They found no significant difference between the independent groups who were walking without any support and the group who were walking with an assistance with regard to the age and first onset age of the disease. Considering the age and ataxia levels, we found that the ataxia level was more serious in non-ambulating patients and ataxia level became worse with ageing. However, we found no relationship between the age at onset of first symptom of the disease and ataxia levels (p>0.05).

Croarkin et al.[12] found a significant correlation between the FA Rating Scale score and locomotor state. They reported that the FA Rating Scale scores could be a useful tool for determining the patients who would be able to walk without any support. We found that there was a significant relationship between pre- and post-treatment FAC, Hoffer Ambulation Scale levels (p<0.05). In this respect, the rate of the unsupported ambulation significantly increased in the patients with

mild ataxia after the treatment. We agree with the Croarkin et al.'s[12] opinion that ataxia evaluation is a useful method for identifying patients who would able to walk without a support.

In a retrospective study including 29 patients with FA, Milne et al.[22] evaluated the effects of receiving inpatient rehabilitation program for independence. To evaluate the efficiency of inpatient rehabilitation, they used the Functional Independent Measurement (FIM). They also mentioned that the effects of the rehabilitation on the quality of life and disease severity should be investigated. They demonstrated that the inpatient rehabilitation was extremely important, as it increased the independence in the daily living activities and provided patients functional improvement. We evaluated the daily activities of the patients by using the Barthel Index. After the rehabilitation, we found a highly significant improvement in the Barthel Index (p<0.001).

The study of Milne et al.[22] is the only study that we can discuss the exercise methods, which are implemented during the inpatient rehabilitation program. In the literature, we are unable to find any studies, which evaluate the rehabilitation outcomes of the patients undergoing inpatient rehabilitation for FA, and our results are similar with the study results of Milne et al.[22] Similar to our treatments, Milne et al.,[22] implemented the patient-specific exercises such as stretching, balance retraining, pelvic and spinal control, spasticity management, hand function and cardiopulmonary exercises, communication and swallowing exercises and specialized equipment prescription for the rehabilitation of the 29 FA patients. Unlike our rehabilitation practices, they implemented aquatic physiotherapy and the most importantly they provided safety and risk management services to the patients with FA and developed home and community programs. Unfortunately, we cannot provide sufficient community rehabilitation services under the circumstances of Turkey. This service is critical for the patients' independent participation in social life. Unfortunately, this is the lack of our society and we believe that the social rehabilitation would be better in our country in the future.

In addition, Maring et al.[2] reported that the main goal of physical therapists should be to increase the individual participation, independent from the environmental events as much as possible by taking into account the progressive nature of the disease. We share the same opinion, and we found that the community ambulation levels increased after the rehabilitation.

Study limitation

Study limitation was that the selection of the inpatient rehabilitation participants. Only patients with increased awareness were admitted to the rehabilitation hospital and frequency of FA was, therefore, low. In the future studies, it should be planned to include more patients. In addition, our study is a retrospective study. Furthermore, to assess the rehabilitation outcomes, randomized-controlled, prospective studies should be planned. We believe that there is a need for more comprehensive studies, which would determine the optimum duration, intensity and content of the rehabilitation program in patients with FA.

In conclusion, there is no pharmacological treatment for slowing down the progression of FA. There is a need for comprehensive researches with regard to the content, intensity, and duration of rehabilitation program to be implemented and with respect to the sustained recovery after inpatient rehabilitation. However, as we demonstrated in our study, a convenient and comprehensive rehabilitation program increases the ambulation level and independence in daily activities of the patients. Therefore, for the patients with FA, inpatient rehabilitation is extremely important.

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