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The effects of home exercise program on limb-girdle disease: a cohort study

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Abstract

This study aims to demonstrate the effectiveness of rehabilitation in patients with limb-girdle disease, who were given a home exercise program and called for follow-up in certain periods by observing the functional areas of the shoulder and pelvic groups every six months. Suitable statistical methods were conducted. Descriptive findings and continuous variables regarding the patients were presented. As a result of the analyses, no statistically significant difference was found only in pre-treatment and post-treatment wrist strength and trunk extension variables, but a statistically significant difference was found between the pre-treatment and post-treatment scores of all other types of strength. Significant differences were found between the pre-treatment and post-treatment cervical flexion strength in terms of general weakness and inability to walk, joint limitation and walking status. It is believed that the rehabilitation method to be applied to patients with limb-girdle muscular dystrophy (LGMD) detected in the early period will provide effective results and may form the basis for referring them to physicians and health professionals. It has been concluded that rehabilitation will help them lead a comfortable life.

Key words: *limb girdle, shoulder girdle, pelvic girdle, anatomy, effects of home exercise.*

Introduction

The human body needs the muscular system, which is positioned like a natural corset on the skeletal system, to ensure its life continuity and perform basic actions. The muscle groups have important functions in digestive activities, functioning of the internal organs, circulatory and metabolic activities, and all the processes of the endocrine system for the body to get the necessary energy from nutriments.

Neuromuscular diseases (NMD) are a heterogeneous group of rare, hereditary and nonhereditary diseases with peripheral or central involvement, involving the muscle and nerve tissue. One of the most important groups included in these diseases is the genetic muscle disorders (GMD) [11].

Since GMDs are rare, it has not been possible to establish a guideline specifying epidemiological studies in these diseases. The prevalence has been tried to be determined by the level of cases.

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Although the data are insufficient, with the development of specific diagnostic tests and the improved access to them, there has been an increase in the number of studies investigating the prevalence of genetic muscle disorders in recent years [13].

The hereditary forms can be subdivided into several diagnostic groups by distributing predominant muscle involvement and weakness: Duchenne and Becker, Emery-Dreifuss, myotonic dystrophy, facioscapulohumeral, oculopharyngeal; and limb-girdle, which is the most heterogeneous group [7]. Limb-girdle muscular dystrophy (LGMD) is a genetically heterogeneous disorder responsible for muscle atrophy and severe dystrophies. Despite the critical developments in the insight and information of pathomechanisms of LGMD, any definitive treatments do not exist. Current strategies are only based on improving of the signs of disorder and enhancing the quality of life without resolving any underlying cause [18].

The lack of medical treatment for the disease and the need for long-term care due to the worsening of the condition as it progresses makes the participation of the individual and their family, who have an important role in the patient's care, valuable. Muscular dystrophy can be a devastating diagnosis for children and their families in the long run. Understanding the genetic implications and prognosis is important in diagnosing the specific dystrophy type and informing the patient and the family [9].

The history of LGMD

Genetic diseases have existed throughout human history. In parallel with developments in medicine and technology, diseases were diagnosed using clinical courses, laboratory findings, imaging reports and scientific methods. Throughout history, people have tried to identify these diseases that affect life, hinder the social interactions of individuals, require continuous healthcare services, and burden the health economics, and to find solutions to them.

Researchers have reported that muscle dystrophy R2 (LGMDR2), which occurs in the functional areas of the shoulder and hip girdle connecting the upper and lower limbs to the axial skeleton, is caused by inherited muscular dystrophies DYSF gene mutations [14]. First, in 1954, John Walton and Frederick Nattrass defined LGMD as a separate entity from the X-linked dystrophinopathies like other muscular dystrophies [15]. With the increasing development of molecular genetics research, pathogenesis more precise classification and understanding are possible. Until today, more than 30 different subtypes of LGMD have been identified, most of which are inherited autosomal recessive conditions. Advances in the application of new omics technologies and the generation of large-scale biomedical data will help better understand disease mechanisms in LGMD. They should ultimately help to accelerate the development of novel and more effective therapeutic approaches [15].

The anatomy of LGMD

Learning the functional anatomy of the muscles in the areas affected by limb-girdle disease provides significant convenience in diagnosis and treatment. Therefore, researchers have found valuable information about the shoulder and pelvic girdle muscles [2,10] (Fig. 1).

The perineal muscles in the functional area of the pelvic girdle are located in the region between the external genitalia and the anus; these muscles control the urinary and stool functions. They extend the pelvic joint, making the limb straight when bent. Although they are the strongest muscles making this movement, they are generally used in exercises that require strength, such as sitting, standing up and climbing stairs. They are not used while standing and are rarely used when walking on level surfaces; therefore, paralysis of these muscles is no different than walking on flat ground. The functional muscles of the pelvic region have the most important role in the movement, regular walking, lateral flexion and rotation. By connecting the upper and lower limbs with the trunk; they simultaneously ensure the trunk mobility to be stabilized and separated between the pelvic girdles, they increase joint mobility and reduce the energy required for walking [2,5] (Fig. 1).

Muscles in the functional area of the shoulder girdle contribute to pulling and lifting movements. The forces of the forearm, which are under the influence of the active site of the shoulder girdle, hold, stretch and work. Muscles under the power of the functional area of the pelvic girdle work by crouching down, getting up and walking, with the support of the forces affecting the knee joint and ankle joints (Fig. 2).

The muscles of the shoulder girdle

Deltoid muscle is divided into three parts: the clavicular part, the acromion part and the spinal part.

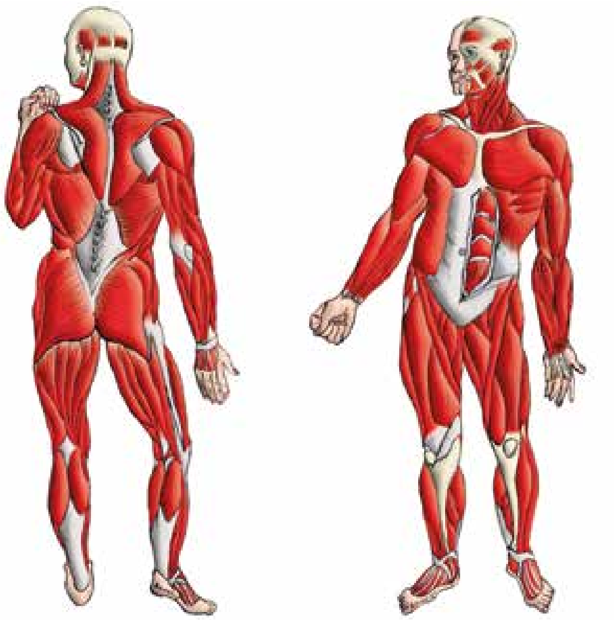

Fig. 1. Muscular anatomy.
This drawing was drawn by anatomy painter Prof. Dr. Vatan Kavak.

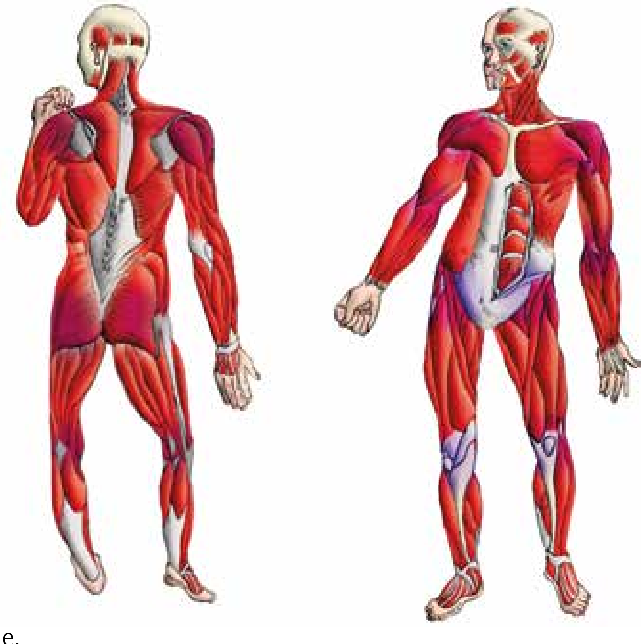

Fig. 2. Limb-girdle.
This drawing was drawn by Anatomy Painter Prof. Dr. Vatan Kavak.

It is responsible for internal rotation, abduction, adduction and external rotation movements.

Musculus supraspinatus starts from the fossa supraspinatus and inserts on the tuberculum majus humeri. Its most important task is to begin and continue shoulder abduction.

Musculus infraspinatus arises from the fossa infraspinatus and inserts on the tuberculum majus humeri. It is responsible for external rotation.

Musculus teres minor arises from the margo lateralis scapulae and inserts on the tuberculum majus humeri. It is responsible for external rotation and partial adduction.

Musculus subscapularis originates from the fossa subscapularis and attaches to the tuberculum minus humeri. It is responsible for internal rotation.

Musculus teres major starts from the margo lateralis scapulae and is attached below the tuberculum minus humeri. It performs functions such as internal rotation, extension and adduction of the arm.

The muscles of the pelvic girdle

Anterior pelvic muscles

Musculus iliopsoas has two divisions: one *musculus iliacus* – originates from the iliac fossa, and the other, *musculus psoas major* – stems from the lateral parts of the lumbar vertebrae. The muscle leaves the pelvic cavity through the lacuna musculorum (the space below the inguinal ligament) and inserts onto the lesser trochanter of the femur. Since this muscle reduces tension during stretching, it has a spiritual effect and a physical effect on the individual. Its most important task is hip flexion; it also provides balance, movement, upright posture, and supports breathing by connecting to the diaphragm.

Musculus psoas minor is a small muscle that lies medial to the poses major. It arises from the lateral parts of the last thoracic and the first lumbar vertebrae and inserts on the iliopubic eminence. It plays an important role in flexing the lumbar vertebrae.

Posterior pelvic muscles

Musculus gluteus maximus arises from the posterior part of the gluteal surface and dorsal surface of the sacral bone and inserts on the gluteal tuberosity. The muscle performs the movements of the hip joint (extensio et rotatio externa femoris).

Musculus gluteus medius arises from the middle part of the gluteal surface and inserts on the greater trochanter of the femur. The muscle produces abductio femoris in the hip joint; it also produces rotatio interna femoris (contractions of anterior fibres) or rotatio externa femoris (contractions of posterior fibres).

Musculus gluteus minimus arises from the anterior part of the gluteal surface and inserts on the greater trochanter of the femur. The muscle produces abductio femoris in the hip joint; it also produces rotatio interna femoris (contractions of anterior fibres) or rotatio externa femoris (contractions of posterior fibres).

Musculus tensor fasciae latae arises from the spina iliaca anterior superior and inserts on the iliotibial tract (thickening of the fascia lata). The muscle tenses the fascia lata and produces flexio femoris in the hip joint.

Musculus piriformis arises from the pelvic surface of the sacral bone, leaves the pelvic cavity through foramen ischiadicum majus and inserts on the greater trochanter of the femur. The muscle produces abductio et rotatio externa femoris.

Musculus obturatorius internus arises from the inner surface of the obturator membrane, leaves the pelvic cavity through foramen ischiadicum minus and is inserted near the greater trochanter of the femur. The muscle produces rotatio externa femoris.

The two gemelli muscles – musculus gemellus superior and *musculus gemellus inferior* lie above and below the obturator internus muscle. The upper one arises from spina ischiadica, the lower one – from tuber ischiadicum; both muscles are inserted near the greater trochanter of the femur and produce rotatio externa femoris.

Musculus quadratus femoris arises from tuber ischiadicum and inserts on the intertrochanteric crest of the femur; the muscle produces rotatio externa femoris.

Musculus obturatorius externus arises from the outer surface of the obturator membrane; it lies deep and is covered by muscles of the medial compartment of the thigh. The muscle is attached near the greater trochanter of the femur and produces rotatio externa femoris (Fig. 1).

The clinical course of LGMD

This group of diseases is usually non-syndromic and clinical involvement is typically limited to skeletal muscle [17]. The clinical course of patients with LGMD varies depending on criteria such as the age of onset, the degree of muscle weakness, and the progression rate of the disease. Although the exact triggering causes are unknown, symptoms may appear at almost any age; however, it is a disease that often starts at a younger age and generally has a dramatic clinical course. Due to the body region it involves it has a process that affects the actions of walking and balancing in general and increases the possibility of bed confinement in the long term. Developing conditions such as muscle atrophy, loss of strength and walking, balance problems, pain, musculoskeletal deformities, heart and respiratory problems, fatigue and psychological problems cause the patient to become desperate, especially when these conditions become permanent in their lives;

they feel like nobody will help them and they start keeping themselves isolated from vital and social activities, which, in time, results in many psychological problems such as depression [4-6].

At the European Neuromuscular Center Workshop held in 1995, more precise criteria for diagnosing and classifying of LGMD were determined. The different subtypes were classified according to their genetic characteristics. A medical rehabilitation program provides positive changes in the course of the disease, an increase in the quality of life and a longer life span by protecting the physical and psychological functions of the patient and even by increasing the physical and mental capacity [12]. There are delays in applying to specialist physicians and getting a diagnosis in centres providing primary healthcare services. LGMD is overlooked by physicians and health professionals who do not have a good grasp of the literature since it is a disease that can easily be confused with other neuromuscular disorders. As the prognosis varies from person to person, delayed diagnosis or misdiagnosis in patients with LGMD is one of the important points that should be considered. Studies on LGMD are based more on medical treatments rather than rehabilitation.

Material and methods

The aim and importance of the study

In this study, patients who applied to the Diyarbakir Gazi Yaşargil Training and Research Hospital Department of Physical Medicine and Rehabilitation Neuromuscular Disease Center and were diagnosed with LGMD as a result of genetic analysis were included. All patients were followed up every six months between 2013 and 2021 and completed at least five years of follow-up. Since LGMD varies from patient to patient, giving them a personalized home exercise program based on the level of the person after every follow-up visit and providing information such as the duration of the exercises was aimed to determine the effectiveness of home exercise and the status of these patients after at least five years. No such long-term study designed for LGMD patients was found in the literature review. This reveals the importance and authenticity of the study. Exercise-based studies have usually been conducted in patients with different accompanying muscular disorders; therefore, the inclusion of patients with the same diagnosis in our study has added to the importance of the study.

Ethical aspect of the study

The study was approved by the Ethics Committee of the University of Health Sciences and the Ethics Committee of Diyarbakır Gazi Yaşargil Training and Research Hospital (Serial no. 808/Date: 02.07.2021).

Works done before starting the research and during the research process

The study was started with 71 patients, and it lasted for eight years. However, some patients withdrew from the study during the process. This is common in cohort studies. Forty-nine of our patients, diagnosed with LGMD according to the results of the genetic analysis, completed the initial and final stages and regularly came for follow-up. In the last step, the process was conducted with 49 people. At each follow-up visit, the patients were examined elaborately, necessary laboratory tests were requested, and consultation was requested from the relevant units due to systemic involvement. At each visit, the motor, walking, daily life activities, depression-anxiety and patient satisfaction scales were applied to the patients; the results of the scales were recorded, and the same person performed all these procedures. The records of the genetic consequences of all patients included in the study for definitive diagnosis are in our archive.

In the Home Exercise Program, the following exercises were given to the patients

In the exercise program, the proper exercises from the following were selected according to the individual: joint range of motion exercises, strengthening exercises, gait training exercises, posture exercises, stretching exercises, breathing exercises, relaxation exercises and programs for device implementation.

Data analysis

Assumptions of normality and homogeneity of variances

For the normality assumption, the skewness and kurtosis values of the continuous variables were checked. Since the values were between –1.5 and

+1.5, it was revealed that all of the continuous variables met the normality assumption and therefore, it was possible to use parametric tests. In addition, for the homogeneity of variances, another assumption, the *p*-value, which is the significance value, was checked. Accordingly, it was determined that the variances were homogeneous ($p > 0.05$).

The variables used for the analysis

Grading for the evaluation of the Oxford Scale:

Scale	Evaluation of the Oxford Scale
0/5	No shrinkage
1/5	There is visible/palpable muscle contraction but no movement
2/5	Motion disappeared with gravity
3/5	Movement against gravity only
4/5	Movement against gravity with a resistance
5/5	Movement against gravity with full resistance

The Medical Research Council Muscle Grading System (MRC) and Functional Ambulation Categories (FAC) scales were used.

Statistical analysis

Frequency and percentage values were reported for the discontinuous descriptive variables. For continuous variables, mean and standard deviation values were reported. In comparing the two groups, Student's *t*-test was used for the significance of the differences between the averages. Correlations between the measurements were analysed using Pearson's correlation analysis. The paired-samples *t*-test was conducted to measure the differences between before the treatment and after the treatment. A *p*-value of < 0.05 was considered statistically significant for the tests used.

Results

According to Table I, the gender ratio of the participants was equal, the majority of them were married, and the first complaint was a general weakness of the foot. It was observed that the current complaints of the patients changed compared to the first follow-up visit and that the complaint of general weakness turned into an inability to walk. It was observed that the educational status of the participants mainly was at the primary-middle school level, the majority of them were unemployed, and the majority of them inherited the disease from their families. It was determined that in most of the patients, there was consanguinity between the parents, they had a joint limitation, and they could walk. Lumbar lordosis and posture disorder were detected in all patients at the first visit. In the first control evaluation, scoliosis was detected in very few patients. While almost one-third of the patients were able to walk independently before the treatment, the number of people who could walk independently decreased depending on the time after treatment.

According to Table II, the mean age of the participants was approximately 35, the mean weight was approximately 66, and the mean height was approximately 163 cm. It was determined that the mean age of diagnosis was 32, the mean age at of onset of complaints was 15, the total number of siblings was 8, and the age at loss of independent ambulation was approximately 29. When the anthropometric information of the patients was examined, it was seen that most of them were at the ideal weight, and very few of them were below or above the ideal weight.

According to Table III, which shows information about the health status of the patients, it was established that the hip strength before the treatment was the lowest score, and the cervical extension strength score was the highest. It was established that the other disease values were normal both before and after the treatment.

According to the data obtained from Table IV, it was determined that the knee and ankle joint range of motion values, which are thought to affect ambulation, were limited, and the joint range of motion values of other disease conditions were normal.

According to Table V, which shows the difference between the health status of the patients before and after physical therapy, it was found that there was no statistically significant difference only in terms of pre-treatment and post-treatment wrist strength and trunk extension strength ($p > 0.05$). A statistically significant difference was found between the pre-treatment and post-treatment scores of all other types of strength since post-treatment scores were lower than pre-treatment scores (*$p < 0.05$, **$p < 0.01$).

According to Table VI, which shows the correlation results between continuous variables, it was found that the age of the patient, the age of diagnosis, the age of onset of complaints, and the types of gait strength were negatively correlated; and the types of gait strength were positively, moderately and highly correlated among themselves. The correlation between the types of gait strength was an expected

Table I. Socio-demographic characteristics of the participants (n = 49)

Variables	*n*	%
Gender		
Female	24	49
Male	25	51
Marital status		
Single	34	69.4
Married	15	30.6
First complaint		
General weakness	39	24.4
Toe-walking	10	
Current complaints		
General weakness	28	57.1
Inability to walk	21	42.9
Educational status		
Primary – Middle school	30	61.2
High school	15	30.6
Higher education and above	4	8.2
Job status		
Unemployed	35	71.4
Employed	14	28.6
Positive family history for LGMD		
Yes	37	75.6
No	12	24.4
Consanguinity between the parents		
Yes	39	79.6
No	10	20.4
Joint limitation		
Yes	37	75.5
No	12	24.5

Variables	*n*	%
Inability to walk		
Able	31	63.3
Unable	18	36.7
Posture disorder		
Yes	49	100
No	0	0
Not Changed		
Presence of lumbar lordosis		
Yes	49	100
No	0	0
Presence of scoliosis in the first evaluation		
Yes	13	26.5
No	36	73.5
Pre-treatment FAC 1		
Non-functional ambulation	14	28.6
Dependent for physical assistance level II	1	2.0
Dependent for physical assistance level I	2	4.1
Dependent for supervision	5	10.2
Independent – level surfaces only	12	24.5
Independent	15	30.6
Post-treatment FAC 2		
Non-functional ambulation	20	40.8
Dependent for physical assistance level II	0	0
Dependent for physical assistance level I	1	2.0
Dependent for supervision	8	16.3
Independent – level surfaces only	15	30.6
Independent	5	10.2

Table II. Some demographic and anthropometric characteristics of the participants (n = 49)

Variables	Mean	SD
Age	34.53	11.782
Weight	65.83	15.815
Height	162.61	11.344
Age of diagnosis	29.32	12.188
Age of onset of complaints	15.48	5.799
Total number of siblings	8.22	3.050
Age at loss of independent ambulation	29.06	4.058
Body mass index	*n*	%
Below the ideal weight (< 18.49)	4	10.5
Ideal weight (18.5-24.99)	22	57.9
Above the ideal weight (25-25.99)	4	10.5
Overweight (> 30)	8	21.1

Table III. Health status of the participants (n = 49)

Variables	Mean	SD
Cervical_Flex_strength_1	3.5714	0.82285
Cerv_flex2	3.3469	0.83044
Cervical_Ext_strength_1	4.0102	0.90421
Cerv_ext2	3.8163	0.95030
Trunk_Flex_strength_1	3.2245	1.06586
Trunk_flex2	2.8776	1.15709
Trunk_Ext_strength_1	3.1735	1.08758
Trunk_ext2	3.0102	1.13876
Shoulder_strength_1	2.7755	1.12760
Shoulder_strength_2	2.2959	1.15433
Elbow_strength_1	3.3469	1.10964
Elbow_strength_2	3.1327	1.06446
Wrist_strength_1	3.9388	0.92214
Wrist_strength_2	3.8571	0.91287
Hip_strength_1	2.5510	1.18693
Hip_strength_2	2.2653	1.15516
Knee_strength_1	3.0306	1.24753
Knee_strength_2	2.8265	1.25645
Ankle_strength_1	2.8776	1.29715
Ankle_strength_2	2.5408	1.36105

Table IV. Joint range of motion of the participants (n = 49)

Variables	*n*	%
Shoulder_JRM_1		
Normal	49	100
Limited	0	0
Shoulder_JRM_2		
Normal	49	100
Limited	0	0
Elbow_JRM_1		
Normal	41	83.7
Limited	8	16.3
Elbow_JRM_2		
Normal	38	77.6
Limited	11	22.4
Wrist_JRM_1		
Normal	49	100
Limited	0	0
Wrist_JRM_2		
Normal	49	100
Limited	0	0
Hip_JRM_1		
Normal	49	100
Limited	0	0
Hip_JRM_2		
Normal	49	100
Limited	0	0
Knee_JRM_1		
Normal	44	89.8
Limited	5	10.2
Knee_JRM_2		
Normal	39	79.6
Limited	10	20.4

Variables	*n*	%
Ankle_JRM_1		
Normal	43	87.8
Limited	6	12.2
Ankle_JRM_2		
Normal	39	79.6
Limited	10	20.4
Cervical_Flex_JRM_1		
Normal	49	100
Limited	0	0
Cervical_Flex_JRM_2	49	100
Normal	0	0
Limited	49	100
Cervical_Ext_JRM_1	0	0
Normal	49	100
Limited	0	0
Cervical_Ext_JRM_2	49	100
Normal	0	0
Limited	49	100
Lumbar_Flex_JRM_1	0	0
Normal	49	100
Limited	0	0
Lumbar_Flex_JRM_2	49	100
Normal	0	0
Limited	49	100
Lumbar_Ext_JRM_1	0	0
Normal	49	100
Limited	0	0
Lumbar_Ext_JRM_2	49	100
Normal	0	0
Limited	49	100

situation. Although the post-treatment recovery was statistically different from the pre-treatment, this difference was not big. In other words, the treatment did not eliminate the disease but only reduced the severity and effect.

According to Table VII, there was a statistically significant difference between the cervical flexion strength before and after the treatment regarding the current complaint of general weakness and inability to walk ($p < 0.01$).

There was a statistically significant difference in patients with joint limitation in terms of cervical flexion strength both before and after the treatment compared to patients without joint limitation ($p < 0.05$).

According to walking status, it was determined that there was a statistically significant difference between those who were able to walk and those who were not able to walk in terms of cervical flexion strength before and after the treatment ($p < 0.01$).

Discussion

Studies present the prevalence-incidence, case reports, and general concept regarding limb-girdle muscular dystrophy in the literature. Conducting this study on LGMD diagnosed by genetic analysis in the eight years between 2013 and 2021 added a long-term approach to the study. The patients, who were

Table V. Comparison of the health status of the participants before and after physical therapy (n = 49)

Pairs	Variables	Mean	SD	Difference mean	SD	t	p
Pair 1	Cervical_Flex_strength_1 Cervical_ Flex_strength_2	3.5714 3.3469	0.82285 0.83044	0.22449	0.45713	3.438	0.001*
Pair 2	Cervical_Ext_strength_1 Cervical_ Ext_strength_2	4.0102 3.8163	0.90421 0.95030	0.19388	0.47671	2.847	0.006*
Pair 3	Trunk_Flex_strength_1 Trunk_Flex_strength_2	3.2245 2.8776	1.06586 1.15709	0.34694	0.54203	4.481	0.000**
Pair 4	Trunk_Ext_strength_1 Trunk_Ext_strength_2	3.1735 3.0102	1.08758 1.13876	0.16327	0.59850	1.910	0.062
Pair 5	Shoulder_strength_1 Shoulder_strength_2	2.7755 2.2959	1.12760 1.15433	0.47959	0.58594	5.730	0.000**
Pair 6	Elbow_strength_1 Elbow_strength_2	3.3469 3.1327	1.10964 1.06446	0.21429	0.45644	3.286	0.002*
Pair 7	Wrist_strength_1 Wrist_strenght_2	3.9388 3.8571	0.92214 0.91287	0.08163	0.34380	1.662	0.103
Pair 8	Hip_strength_1 Hip_strength_2	2.5510 2.2653	1.18693 1.15516	0.28571	0.47871	4.178	0.000**
Pair 9	Knee_strength_1 Knee_strength_2	3.0306 2.8265	1.24753 1.25645	0.20408	0.43203	3.307	0.002*
Pair 10	Ankle_strength_1 Ankle_ strength_2	2.8776 2.5408	1.29715 1.36105	0.33673	0.66448	3.547	0.001*

** < 0.05, ** < 0.01*

followed up according to the genetic test results and given a home exercise program, underwent physical examination every six months, and the effective monitoring of their clinical course increased the reliability of the findings of this study.

The overall prevalence of LGMD is 2 to 10 per 100,000 people [16]. The estimated incidence of LGMD is 1 to 6 per 100,000 people [3,8]. Obtaining a sufficient number of patients diagnosed with LGMD (n = 49), whose incidence is relatively low in populations compared to other diseases, made the research meaningful in a heuristic framework. The fact that some of the patients evaluated within the scope of the study did not show up for the follow-up examination at regular periods was the most important limitation in terms of the research. Still, these patients were not included in the study.

The findings obtained in the study showed parallelism with the results in the literature. It was determined that the gender ratio of the participants was equal, the majority of them were married, and the first complaint was the general weakness of the foot. It was observed that the current complaints of the patients changed compared to the first follow-up visit and that the complaint of general weakness turned into an inability to walk. The participants' educational status was mainly at the primary-middle school level. The majority of them were unemployed, and the majority of them inherited the disease from their families [1]. The mean age of diagnosis was 32, the mean age of onset of complaints was 15; and the fact that there was a long time between the mean age of onset of complaints and the mean age of diagnosis shows that it is important to consider this in the differential diagnosis since it is a rare disease. It was determined that the total number of siblings was eight, and the age at loss of independent ambulation was 29. When the anthropometric information of the patients was examined, it was observed that most of them were at the ideal weight, and very few of them were below or above the ideal weight. While almost one-third of the patients were able to walk independently before the treatment, the number of people who could walk independently decreased depending on the time after treatment. The age of onset and course of the disease were consistent with the case reports in the literature [1].

In most of the patients, it was observed that there was consanguinity between the parents, which indicates the importance of informing society about hereditary diseases. The majority of the patients had joint limitations, and they could walk. Lumbar lordosis and

Table VI. The correlation matrix between continuous variables (n = 49)

Cervical_Flex strength-1	Age	Age of diagnosis	Age of onset of complaints	Age at loss of independent ambulation	Cerv_flex2	Cervical_Ext_ strength_1	Cerv_ext2	Trunk_Flex_ strength_1	Trunk_flex2	Trunk_Ext_ strength_1	Trunk_ext2	Shoulder_ strength_1	Shoulder_ strength_2	Elbow_ strength_1	Elbow_ strength_2	Wrist_ strength_1	Wrist_ strength_2	Hip_ strength_1	Hip_ strength_2	Knee_ strength_1	Knee_ strength_2	Ankle_ strength_1	Ankle_ strength_2
Cervical_ Flex_ strength_1																							
Age	–0.677**																						
Age of diagnosis	–0.416**	0.781**																					
Age of onset of complaints	–0.042	0.256	0.363*																				
	–0.738**	0.504**	0.310*	–0.135																			
Cerv_flex_2	0.847**	–0.607**	–0.372**	0.068	–0.709**																		
Cervical_Ext_ strenkth_1	0.846**	–0.650**	–0.469**	–0.144	–0.694**	0.744**																	
Cerv_ext2	0.856**	–0.718**	–0.593**	–0.121	–0.736**	0.822**	0.869**																
Trunk_Flex_ strength_1	0.765**	–0.632**	–0.407**	–0.059	–0.688**	0.734**	0.754**	0.772**															
Trunk_fle x_2	0.764**	–0.642**	–0.483**	–0.076	–0.747**	0.793**	0.748**	0.832**	0.884**														
Trunk_Ext_ strength_1	0.748**	–0.644**	–0.415**	–0.078	–0.699**	0.728**	0.761**	0.757**	0.963**	0.857**													
Trunk_flex_2	0.727**	–0.607**	–0.423**	–0.058	–0.608**	0.745**	0.657**	0.753**	0.865**	0.922**	0.856**												
Shoulder strength_1	0.764**	–0.689**	–0.483**	–0.109	–0.773**	0.719**	0.748**	0.768**	0.771**	0.809**	0.780**	0.724**											
Shoulder_ strength_2	0.739**	–0.652**	–0.492**	–0.039	–0.692**	0.760**	0.716**	0.753**	0.800**	0.815**	0.805**	0.766**	0.868**										
Elbow_ strength_1	0.771**	–0.633**	–0.354*	0.028	–0.707**	0.658**	0.744**	0.684**	0.725**	0.748**	0.760**	0.681**	0.804**	0.756**									
Elbow_ strength_2	0.804**	–0.653**	–0.400**	0.031	–0.740**	0.748**	0.767**	0.745**	0.818**	0.792**	0.844**	0.746**	0.815**	0.849**	0.913**								
Wrist_ strength_1	0.775**	–0.616**	–0.328*	0.064	–0.695**	0.790**	0.775**	0.712**	0.693**	0.705**	0.717**	0.645**	0.738**	0.742**	0.795**	0.804**							
Wrist_ strength_2	0.790**	–0.667**	–0.398**	0.108	–0.705**	0.809**	0.759**	0.750**	0.740**	0.742**	0.760**	0.703**	0.788**	0.812**	0.790**	0.845**	0.930**						
Hip_ strength_1	0.764**	–0.549**	–0.330*	0.058	–0.819**	0.700**	0.723**	0.720**	0.806**	0.782**	0.820**	0.739**	0.826**	0.753**	0.809**	0.831**	0.717**	0.718**					
Hip_ strength_2	0.703**	–0.552**	–0.375**	–0.009	–0.810**	0.684**	0.676**	0.705**	0.822**	0.785**	0.858**	0.750**	0.822**	0.799**	0.739**	0.826**	0.710**	0.758**	0.917**				
Knee_ strength_1	0.759**	–0.589**	–0.442**	–0.024	–0.752**	0.713**	0.757**	0.769**	0.825**	0.804**	0.841**	0.770**	0.764**	0.768**	0.782**	0.836**	0.699**	0.699**	0.896**	0.876**			
Knee_ strength_2	0.743**	–0.614**	–0.507**	0.003	–0.767**	0.698**	0.735**	0.793**	0.831**	0.827**	0.853**	0.791**	0.795**	0.797**	0.769**	0.835**	0.665**	0.723**	0.907**	0.908**	0.940**		
Ankle_ strength_1	0.701**	–0.620**	–0.443**	0.007	–0.800**	0.611**	0.703**	0.700**	0.744**	0.784**	0.769**	0.688**	0.786**	0.706**	0.790**	0.782**	0.734**	0.706**	0.867**	0.849**	0.839**	0.859**	
Ankle_ strength_2	0.625**	–0.507**	–0.371**	0.009	–0.736**	0.577**	0.605**	0.610**	0.783**	0.784**	0.780**	0.729**	0.753**	0.778**	0.715**	0.740**	0.649**	0.659**	0.834**	0.874**	0.806**	0.842**	0.876**

*** Correlation is significant at the 0.01 level (2-tailed), * Correlation is significant at the 0.05 level (2-tailed).*

Table VII. Comparison of flexion strength of patients before and after treatment with some related variables (n = 49)

	Pre-treatment cervical flexion strength		Post-treatment cervical flexion strength	
	x̄	SD	x̄	SD
Current complaint				
General weakness	3.98	0.700	3.75	0.700
Inability to walk	3.02	0.641	2.80	0.679
t	4.911		4.710	
p	0.000**		0.000**	
Joint limitation				
No	3.77	0.769	3.54	0.767
Yes	2.95	0.689	2.75	0.753
t	3.253		3.114	
p	0.002*		0.003*	
Walking status				
Not walking	2.88	0.676	2.66	0.685
Walking	3.96	0.618	3.74	0.670
t	5.689		5.571	
p	0.000**		0.000**	

*** Correlation is significant at the 0.01 level (2-tailed), * Correlation is significant at the 0.05 level (2-tailed).*

posture disorder were detected in all patients at the first visit. In the first control evaluation, scoliosis was detected in very few patients.

When the information regarding the health status of the patients was examined, it was observed that the hip strength score before the treatment was the lowest, and the cervical extension strength score was the highest. It was seen that the other disease values were normal both before and after the treatment.

It was determined that the knee and ankle joint range of motion values, which are thought to affect ambulation, were limited, and the joint range of motion values of other disease conditions was normal.

According to Table V, which shows the difference between the patients' health conditions before and after physical therapy, it was seen that there was no statistically significant difference only in terms of pre-treatment and post-treatment wrist strength and trunk extension strength. A statistically significant difference was found between the pre-treatment and post-treatment scores of all other types of strength since post-treatment scores were lower than pre-treatment scores.

According to the correlation results between continuous variables, the patient's age, the age of diagnosis, the age of onset of complaints, and the types of gait strength were negatively correlated; and the types of gait strength were positively, moderately and highly correlated among themselves. The correlation between the types of gait strength was an expected situation. Although the post-treatment recovery was statistically different from the pre-treatment, this difference was not big. As in the natural course of this disease, treatment did not eliminate the disease, but we thought it reduced its severity and effect.

The rarity of the disease was an important factor in the inability to establish a control group; however, the evidence on how effective exercise is in these patients remains unclear [19]. We believe that it will be more revealing to conduct such studies in the future by establishing a control group.

There was a statistically significant difference between the cervical flexion strength before and after the treatment in the current complaint of general weakness and inability to walk.

A statistically significant difference was found in patients with joint limitation in terms of cervical flexion strength both before and after the treatment compared to patients without joint limitation.

According to walking status, it was determined that there was a statistically significant difference between those who were able to walk and those who were not able to walk in terms of cervical flexion strength before and after the treatment.

In these patients, pain and fatigue increased when repetitive and overloading exercises were continued for a long time in each session. It was observed that the increase in muscle protein breakdown due to intense exercises was caused by the accumulation of lactic acid, which caused acidosis in the cell. We can state that exercising, particularly to strengthen the shoulder and pelvic functional muscles, with a rehabilitation team specialized in the anatomy and neuromuscular disease, in well-equipped sports centres supported by the social state, will bring effective results in terms of meeting the daily exercise needs of the patient [19].

Conclusions

In the study, variables such as general health status and muscle strength of patients diagnosed with LGMD, a disease with low prevalence and incidence compared to other diseases in the population, the prognosis is dramatic, and does not have a definitive treatment, were examined. It was observed that the findings obtained within the scope of the study were overall consistent with the literature and home exercises recommended by specialist physicians had a positive effect on the course of the disease. It is believed that the findings and inferences obtained may form the basis for referring the patients diagnosed with LGMD, which is difficult to diagnose and can be easily confused with other diseases, to specialist physicians and health professionals in the early period. The study has revealed that in patients diagnosed with LGMD, early diagnosis and home exercise programs may increase the quality of life and ensure a longer life span.

Disclosure

The authors report no conflict of interest.

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