

Strength and Functional Measurement for Patients with Muscular Dystrophy

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1. Introduction

Progressive muscle weakness is the major symptom of patients with muscular dystrophy.

The aims of the chapter are to introduce the strength decrease pattern and functional assessment, and to exam the advantages and disadvantages of these measurements applied to various types of muscular dystrophy. Three parts of the measurement for muscular dystrophy are included: the strength decrease pattern, the common general functional scales, and the disease specific scale.

This chapter places emphasis on patients with more weakness in proximal than distal parts. The most common type of proximal muscular dystrophy is Duchenne muscular dystrophy (DMD). Due to rapid deterioration, DMD can be seen as a severe form of muscular dystrophy. Other types of proximal muscular dystrophies have a slower rate of disease progression compared to DMD, such as Beck muscular dystrophy (BMD), limb girdle muscular dystrophy (LGMD), facioscapulohumeral muscular dystrophy (FSHD) and others.

2. Strength measurement

Muscle strength can be assessed by many methods, such as manual muscle testing (MMT) and using the quantitative methods by instrument. Common instruments include the handheld dynamometer (HHD), and the isokinetic dynamometry or other fixation instruments.

For MMT method, the Medical Research Council (MRC) Scale is the most often used system, with the procedures detecting the magnitude of strength by grading muscle strength from 0 to 5. The MRC scale is an ordinal scale, with grades 0-5 also named as "Zero, Trace, Poor, Fair, Good, and Normal". Grade 0 (Zero) cannot be palpated in muscle contraction. Grade 1 (Trace) has some evidence of slight muscle contraction but the strength is too weak to move the joint. Grade 2 (Poor) strength can move the joint (full range of motion) when gravity is eliminated during the test. The Poor grade is sub-graded as Poor minus (2-), Poor, and Poor plus (2+). The 2- indicates strength able to move the joint but unable to complete the range

of motion. The 2+ indicates strength to complete the range of motion and also the ability to resist slight force made by the rater. Grade 3 (Fair) strength can be tested at the antigravity position. Similar to Poor grade, the Fair grade is sub-graded as Fair minus (3-: cannot complete the range of motion), Fair (3: can complete the range of motion), and Fair plus (3+: can against gravity with minimal resistance). Grade 4 (Good) completes the range of motion against gravity with moderate resistance, and Grade 5 (Normal) can resist with strong resistance. We recommend using the MMT method to measure the strength decrease pattern for patients with muscular dystrophy especially in clinical applications. The MMT grading system can clearly provide information as to whether patients can move their body in an antigravity position, and even if the strength is very weak, the strength can be discriminated by grade Poor, Trace or Zero. The weakness strength of patients with muscular dystrophy may be unable to be measured by some instrumentation, as most of these are designed to be measured in an antigravity position and the resistance is added during the measurement. The grading system is also graded (recorded) as different symbol methods. For example, the Kendall system ranked the grade from 0 to 10: it leaves the 0 as Zero, and 10 as Normal, and transforms the strength to a percentage, with a range from 0 to 100 %; the 100% being the grade of "Normal" strength. The percentages of Normal grade strength can be used for calculating the mean strength from many muscles. (Kendall et al., 1993)

For isokinetic dynamometer or other fixation methods in strength measurement, complicated procedures are often not practicable due to the expense and time required to prepare the instruments. Although inconvenient, isokinetic dynamometry has been considered to be the gold standard for assessing dynamic muscle strength and provides much information of various muscle performance characteristics (Mark et al., 2004). The isokinetic strength has been studied in patients with mild or moderate strength impairment (Kilmer et al., 1994; Tiffreau et al., 2007). The patient with DMD with severe progressive muscle weakness highlights the method's limitations for assessment of very weak strength (Bäckman, 1988).

For HHD, it is a convenient, portable and inexpensive device for assessing isometric strength in a clinical setting. The rater handholds the device and presses it against the force that subjects exert with maximal effort. The make test and break test are two methods for HHD. In the make test, the rater resists the patient's maximal isometric contraction, whereas in the break test the rater overcomes the force of the patient produced in eccentric contraction (Bohannon, 1988; Stratford & Balsor, 1994). Both methods have their advantage and disadvantages. To measure the weakness strength of patients with muscular dystrophy, we suggest using the make method. The HHD has been studied in DMD; the strength measured by a force transducer and the data presented in Newtons or kilograms has been seen as real compared to the MMT method where the strength record is in ratio-level parametric data (Scott & Mawson, 2006; Brussock et al., 1992; Stuberg & Metcalf, 1988) However, the main disadvantage of the HHD is the unsure reliability of some muscle strength on the tester when stabilizing the dynamometer (Bohannon, 1999).

3. Strength decrease pattern of various types of muscular dystrophy

3.1 Methods of strength measurement

We previously measured the strength decrease pattern of some common types of muscular dystrophy, such as Duchenne muscular dystrophy (DMD), limb girdle muscular dystrophy

(LGMD) and facioscapulohumeral muscular dystrophy (FSHD) (Lue et al., 1992; Lue & Chen, 2000a; Lue & Chen, 2000b). Patients had been diagnosis by two qualified neurologists and followed up for at least two years. Before the strength measurement, they did not receive medication or a strengthening program for improving muscle strength.

The manual muscle test was used by well trained physical therapists. Thirty-two muscle groups were examined on both sides; the muscle groups included neck and trunk muscles, and upper and lower extremities. The neck and trunk muscles included neck flexors/extensor and the trunk flexors/extensors. In the upper extremities, the shoulder (flexors, extensors, and abductors), elbow (flexors and extensors) and wrist (flexors and extensors) muscle strength were measured. In the lower extremities, the hip (flexors, extensors, and abductors), knee (flexors and extensors) and ankle (dorsi- and plantar-flexors) muscle strength were measured. To calculate the mean strength, we used Kendall's percentage method (Kendall et al., 1993).

3.2 Natural strength decrease pattern of patients with DMD

DMD is a quick deterioration muscular dystrophy, with the strength decrease in a linear pattern positively correlated with age. For every year increment in age, the average strength decreases by about 3.9 percent of normal strength. About half of normal strength will be retained at the age of 12 years. The lower extremities are weaker than the upper extremities. The proximal parts are weaker than distal parts; the weakness of the elbow and wrist extensors is more dominant than that of the flexors. In the lower extremities, hip and knee extensors are weaker than hip and knee flexors. If the strengths of agonist and antagonist muscles of a joint are significantly different, the part of the stronger side becomes shorter and joint contracture easily develops. Therefore, in the upper extremities, elbow and wrist flexion contracture is easily found. Routine active or passive range of motion exercise for patients to maintain the full range of motion is a very important part of any rehabilitation program. Similar to the upper extremities, in the lower extremities, hip flexor contracture is commonly found in early stages of DMD; after the patients are unable to walk, the knee joints may develop severe flexion contracture as the joints are not routinely performing the (normal) range of motion exercises. At the end of life of a patient with DMD, the strength of finger flexors can manage some activities, even though at the age of twenty. Therefore, we recommend using computer games as finger exercises or a leisure activity for patients with DMD, and the keyboard may or may not need modification.

3.3 Natural strength decrease pattern of patients with LGMD

LGMD is also named limb girdle muscular dystrophy syndrome, which is combined with various types of limb girdle muscular dystrophy. Therefore, the strength decrease pattern has greater variation than other types of muscular dystrophy. The speed of muscle strength decrease will become slower than the onset after long disease duration. The strength decrease patterns do not fit well in a linear regression model (R^2 only 0.074), and fit better in an inverse regression model (R^2 equal to 0.154), with the equation as follows: mean muscle strength = $0.61 + (0.63/\text{disease duration})$. No significantly stronger strength in flexor than extensor muscles for extremities are found in patients with LGMD. The limitation of this study needs to be mentioned, as these patients with LGMD may or may not have only one type of LGMD. For more precise study in the future, the gene deficit should be confirmed and including the same type of LGMD for strength study is essential.

3.4 Natural strength decrease pattern of patients with FSHD

Comparing the severity of the strength decrease, the strength decrease of patients with FSHD is the mildest compared to the strength decrease of patients with LGMD or DMD. The shoulder muscle strength is the weakest, followed by elbow muscle strength. The strengths of the trunk area and lower extremities are the best. A special pattern of the strength asymmetry is found in patients with FSHD, as the average right side muscle strength is weaker than the left side. Most of the subjects included in this study were right-handed. The dominant side may increase the use and lead to more prominence of strength decrease, therefore, in clinical applications for patients with FSHMD, too many strengthening programs or overload activity for the upper extremities may not suit such patients. The mechanism for asymmetry of strength found in patients with FSHD still requires further studies to be elucidated.

4. Functional measurements

4.1 The brooke and vignos scales

The common functional scales to rate the grade of disease severity are the Brooke Scale and the Vignos Scale. Both scales were firstly designed for DMD, and nowadays have been used in many neuromuscular diseases. The Brooke scale was designed to assess the upper extremity function. The grades of the Brooke scale range from 1 to 6; 1 means that the subject can elevate their arms full range to the head with the arms straight; while 2 means that the shoulder strength is insufficient to elevate their arms and the subject needs to flex the elbow to elevate the arms; in grades 3 and 4, the subject is unable to elevate the shoulders but can raise hands to the mouth with or without weight respectively; grade 5 refers to the subject being unable to raise hands to the mouth and only some hand movement exists, while grade 6 refers to no useful function of hands (Table 1).

Grade	Description
1	Starting with arms at the sides, the patient can abduct the arms in a full circle until they touch above the head
2	Can raise arms above head only by flexing the elbow (shortening the circumference of the movement) or using accessory muscles
3	Cannot raise hands above head, but can raise an 8-oz glass of water to the mouth
4	Can raise hands to the mouth, but cannot raise an 8-oz glass of water to the mouth
5	Cannot raise hands to the mouth, but can use hands to hold a pen or pick up pennies from the table
6	Cannot raise hands to the mouth and has no useful function of hands

Table 1. Grading system for the Brooke scale.

The Vignos scale was designed to assess the lower extremity function. The grades of the Vignos scale range from 1 to 10; 1 means that the subject can walk and climb stairs without assistance; 2 and 3 means that the strength is insufficient to walk upstairs without assistance as they need to use a rail for climbing stairs (grade 2: in a normal speed; grade 3: slowly);

grades 4 and 5 refer to subjects still having the ability to walk unassisted but unable to climb stairs (grade 4 also can rise from a chair but grade 5 cannot); grades 6 to 8 refer to patients using the long leg brace for walking or standing (grade 6: walk without assistance; grade 7: walk with assistance for balance; grade 8: cannot walk, only for standing); grade 9 refers to the subject being unable to stand, but can sit in a wheelchair; and the final grade 10 refers to the subject being confined to a bed (Table 2).

Grade	Description
1	Walks and climbs stairs without assistance
2	Walks and climbs stair with aid of railing
3	Walks and climbs stairs slowly with aid of railing (over 25 seconds for eight standard steps)
4	Walks unassisted and rises from chair but cannot climb stairs
5	Walks unassisted but cannot rise from chair or climb stairs
6	Walks only with assistance or walks independently with long leg braces
7	Walks in long leg braces but requires assistance for balance
8	Stands in long leg braces but unable to walk even with assistance
9	Is in a wheelchair
10	Is confined to a bed

Table 2. Grading system for the Vignos scale.

4.2 Timed tests

Some studies also record the time needed for some activities as a functional testing for patients with muscular dystrophy. The raters measure the time need for a person to complete the activity. The example of these common activities are climbing some steps of stairs, walking a fixed distance, sitting to standing from a chair, rising from the floor, dressing a cloth and cutting a square.

5. Advantages and disadvantage of the common functional scales of various types of muscular dystrophy

The Brooke and Vignos scales are easy to rate the severity of the patients, but the study found some disadvantages (Lue et al., 2009). We assessed the acceptability of the Brooke and Vignos scales in patients with DMD, BMD, FSHMD, and LGMD from a multi-center study. The patients with DMD were classified as severely progressive group, while the others (BMD, FSHD, and LGMD) were classified as slowly progressive group.

The results showed that the Brooke and Vignos scales were easy to assess, and it took a little time to complete the tests, and the patients did not feel uncomfortable. The Brooke scale is acceptable to grade arm function of the severely progressive group; the DMD, each grade of the Brooke scale is distributed with the acceptable percentage (ranging from 7.1% to 33.3%). However, it is insufficient to discriminate differing levels of severity of the slowly progressive group (BMD, FSHMD, and LGMD). No subject was graded at 4, and only one was graded at 6. The floor effect was large in all types of the slowly progressive group (ranging from 20.0% to 61.9%), especially high in BMD.

In the Vignos scale, using the long leg brace to grade the lower limb function may be a major problem for this scale. Grades 6 to 8 are items using long leg braces for walking or standing; these grades are inapplicable, because some cases did not use long leg braces for walking or standing. The floor effect of the Vignos scale was also large in BMD (23.8%) and in FSHD (50.0%). Among the slowly progressive muscular dystrophies, the function of patients with FSHD was the best; they had better leg function and were less influenced in their daily living activities than other types of slowly progressive muscular dystrophy. Using the two scales in combination with other measures (or instead, to use a complicated instrument for various types of muscular dystrophy) to calculate their function is suggested.

6. The muscular dystrophy functional rating scale

The Muscular Dystrophy Functional Rating Scale (MDFRS) is a disease specific scale designed for various muscular dystrophies. The MDFRS was developed by Lue et al. in 2006. Four domains are included in MDFRS. It was developed in many stages: the preliminary pool of items, the admission of various types of muscular dystrophies and the reliability, validity and responsiveness studies (Lue et al. 2006). The results showed the MDFRS is a reliable and valid disease-specific measure of functional status for patients with muscular dystrophy. The internal consistency was excellent, with the value of the Cronbach' alpha ranging from 0.84 to 0.97. The test-retest reliability and the inter-rater reliability were high (ICC=0.99) for all domains. The MDFRS demonstrated moderate to high correlation with a range of functional rating scales. The confirmatory factor analysis supported a four-dimensional construct. The floor and ceiling effects were small and the responsiveness of various types of muscular dystrophies was well.

The MDFRS combines four domains to rate mobility, basic activities of daily living, arm function and impairment. The number of items of each domain is 9, 6, 7 and 11 respectively (Table 3). The scale offers much important information of muscular dystrophy such as the mobility ability, dependence of daily living, the arm function, and many impairment conditions. The arm function part of the MDFRS effectively conquers the disadvantages of the Brooke scale (Lue et al. 2006; Lue 2010).

Each item of MDFRS is scored on a 4-point scale (1-4), with 1 representing being unable to do the activity and is completely dependent; 2 needing assistance from another person, 3 is independent, without assistance from another person but movement or completion of an activity is slow, and 4 means no problem for the activity and can be done at normal speed. The impairment domain includes the items for measuring contractures and scoliosis, strengths, and respiratory function, and the scoring system was specially designed by the characteristics of the items.

In the mobility domain, the 9 items included measuring the ability of stair climbing, outdoor mobility, indoor mobility, transfers from bed to chair, wheelchair manipulation, standing from sitting, sitting from lying, rolling and changing body position in bed. The items of stair climbing, outdoor and indoor mobility can effectively rate the function of the initial stage of the disease, and the ability of the sitting from lying, rolling and changing body position in bed is needed to examine the condition of the patients with terminal stages of the disease, such as the patients with DMD.

Domains			
Mobility domain	Basic ADL domain	Arm function domain	Impairment domain
1 Stair climbing	1 Feeding	1 Managing objects over head	1 Severity of upper limb joint contracture
2 Outdoor mobility	2 Combing hair	2 Carrying objects	2 Severity of lower limb joint contractures
3 Indoor mobility	3 Brushing teeth	3 Cleaning table	3 Number of contracted joints in the upper limbs
4 Transfers from bed to chair	4 Dressing upper/lower parts of body	4 Writing	4 Number of contracted joints in the lower limbs
5 Wheelchair manipulation	5 Toileting	5 Turning books	5 Severity of neck contracture
6 Standing from sitting	6 Bathing	6 Picking up small objects	6 Strength of the neck
7 Sitting from lying		7 Managing objects over head	7 Strength of the trunk
8 Rolling			8 Scoliosis
9 Changing body position in bed			9 Orthopnea
			10 Sputum clearance
			11 Ventilator assisted

Table 3. Domains and Items of the Muscular Dystrophy Functional Rating Scale.

In the basic activity daily living domain, the 6 items included measuring the ability of feeding, combing hair, brushing teeth, dressing upper/lower parts of body, toileting and bathing. The bathing activity is the most difficult item, and the feeding and combing hair items are easy activities for patients with muscular dystrophy.

In the arm function domain, the 7 items included measuring the ability of managing objects overhead, carrying objects, clearing a table, writing, turning books, picking up small objects, and manipulating small objects. The items were designed to be more functional as needed for daily routine activities. The ability of managing objects over the head and carrying objects is useful to assess the better upper extremity function for patients with muscular dystrophy.

The part of impairment section of the MDFRS offers simple measurement methods for measuring the condition of the contracture and scoliosis, weakness of the head and trunk muscles to provide head control and sitting balance, and the condition of the pulmonary function. In the impairment domain, the 11 items included measuring the problem of severity of upper and lower limb joint contracture, the number of contracted joints in the upper and lower limbs, the severity of neck contracture, strength of the neck, strength of the trunk, severity of the scoliosis, and three respiratory problems such as orthopnea, sputum clearance ability, and the need to use a ventilator. These impairment items are all important symptoms and signs of the various types of muscular dystrophy, and decreasing the complication of contracture is the most important issue for management of such patients. At the end stage of the disease, vital respiratory care needs to be added, and the 3 items of impairment domain of the MDFRS could offer the general condition of the pulmonary function. Therefore, the assessment from the impairment domain could offer a lot of useful information for clinicians and caregivers to easily know the condition of the patients and provide better care for them at different stages of the disease.

The total scores of each domain sum up the scores of each item, therefore, the range of scores for 4 domains are 9-36 for the mobility domain, 6-24 for the basic activity of daily living domain, 7-28 for the arm domain, and 11-44 for the impairment domain respectively. The scores of each domain can be calculated as a percentage to represent the functional performance of a person compared to normal condition; the equation is as follows: (total scores-number of item) / full total scores and multiple 100. The % of mobility ability = (the sum of score from 9 item -9) / 36 *100; the % of basic activity of daily living ability = (the sum of score from 6 item -6) / 24 *100; the % of arm function ability = (the sum of score from 7 item -7) / 28 *100; and the % of impairment condition = (the sum of score from 11 item -11) / 44 *100.

7. Conclusion

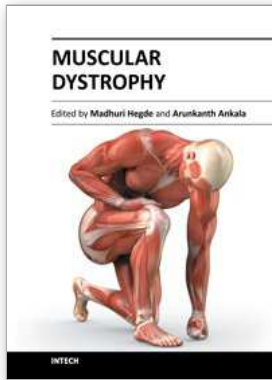
In conclusion, various types of muscular dystrophy present differing speeds of disease progression with decreasing muscular strength in different patterns. Due to some disadvantages of the Brooke and Vignos grading scales applied to patients with muscular dystrophy, clinical application of these scales should be used with caution, especially in patients with slowly progressive muscular dystrophy. We suggest that the applications can

be used in combination with MDFRS, which is a multi-domain instrument, a valid and reliable scale, capable of evaluating the various levels of functional status of different types of muscular dystrophy.

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With more than 30 different types and subtypes known and many more yet to be classified and characterized, muscular dystrophy is a highly heterogeneous group of inherited neuromuscular disorders. This book provides a comprehensive overview of the various types of muscular dystrophies, genes associated with each subtype, disease diagnosis, management as well as available treatment options. Though each different type and subtype of muscular dystrophy is associated with a different causative gene, the majority of them have overlapping clinical presentations, making molecular diagnosis inevitable for both disease diagnosis as well as patient management. This book discusses the currently available diagnostic approaches that have revolutionized clinical research. Pathophysiology of the different muscular dystrophies, multifaceted functions of the involved genes as well as efforts towards diagnosis and effective patient management, are also discussed. Adding value to the book are the included reports on ongoing studies that show a promise for future therapeutic strategies.

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