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Evaluation of respiratory status in patients with myopathy

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Abstract:

Myopathic diseases can lead to significant respiratory disorders due to the involvement of skeletal muscles, including respiratory muscles. These disorders not only affect the quality of life of patients, but are also one of the main causes of mortality in this group. This study aimed to evaluate pulmonary function and its associated factors in patients with myopathy. This is a prospective cross-sectional study that was performed in 2020-2021 on 44 patients with myopathy. The participant's demographic information and features of disease, including age, sex, type of disease, age of onset and duration of disease, and mobility status of the patient (using Walker or wheelchair) were collected by a checklist. The slow inspiratory vital capacity (VC) and FEV1 were obtained. Data analysis was performed with SPSS version 24 software using t-test, Mann–Whitney, Pearson, and Spearman correlation tests. The sitting and lying FVC were significantly higher in patients who had no drug usage (P = 0.007 and P = 0.005, respectively), no sputum (0.003 and 0.002, respectively), no dysphagia (P = 0.002 and P = 0.001, respectively), and no coughs while eating (P = 0.022 and P = 0.006). There were significant inverse relationships between age and sitting FVC (r = -0.638) and lying FVC (r = -0.497), and age had a significant direct relationship with changes in the FVC (r = 0.652) (P < 0.001 for all). Furthermore, we observed a significantly inverse relationship between age of disease onset and sitting FVC (r = -0.491, P = 0.001) and significantly direct relationship between age of disease onset and lying FVC (r = 0.447, P = 0.002) and FVC changes (r = 0.371, P = 0.013). Assessments of respiratory functions, especially through spirometry in both sitting and supine positions, and attention to clinical symptoms such as dysphagia and COUGH, ARE OF GREAT clinical importance in the management of myopathy patients. These assessments can help in the early diagnosis of respiratory failure and timely interventions.

Keywords:

Myopathy, respiratory function, spirometry

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Introduction

Neuromuscular disorders are a heterogeneous group of neurologic diseases that affect a number of neural structures, including motor nerves, neuromuscular junctions, and muscles.[1] Myopathy is a general term referring to any disease that affects the muscles that control voluntary movement in the body.[2] Patients with myopathy could experience muscle weakness due to a dysfunction of the muscle fibers.[3] Common symptoms include weakness of the proximal muscles, body aches, and fatigue. The etiologies of myopathy include genetic, idiopathic, inflammatory, degenerative, drug-induced or toxin-induced.[4]

Respiratory dysfunction is a major problem in patients with neuromuscular disorders. Weakness in the respiratory muscles (such as the diaphragm and intercostal muscles) is one of the main reasons for this issue.[5] The disorder initiates mildly and gradually progresses to severe respiratory distress. Muscle weakness reduces ventilation, resulting in increased carbon dioxide and

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decreased arterial blood oxygenation.[6] Disorders such as kyphoscoliosis and other chest wall deformities could contribute to a worse condition in these patients. Laryngeal and pharyngeal disorders can lead to reduced respiratory cilia movements, and as a result, the risk of pneumonia is higher in these patients. In some myopathies, it affects the respiratory center and causes respiratory disorders.[7] Symptoms include decreased orthopedic respiratory capacity, sleep disturbances, and headache following awakening.[8] Another cause of respiratory disorders in patients with myopathies could occur due to diaphragm paralysis. Unilateral paralysis of the diaphragm may be asymptomatic, but these patients may experience shortness of breath while sleeping in supine position or during exercise.[9]

Prompt diagnosis of respiratory malfunctions in these patients is very important. Early symptoms may be very vague, such as morning fatigue, sleep disturbances, and mood swings. These symptoms are related to hypoxia during sleep.[10] Decreased nighttime ventilation usually occurs before daytime respiratory distress and can lead to respiratory failure and Cor Pulmonale syndrome.[9]

Based on the recommendation of Consensus Care Guidelines for Congenital Muscular Dystrophies, spirometry in the sitting position could be a valuable diagnostic test. It also recommends evaluating sleep disorders if the FVC is reduced by more than 20%.[11,12]

Forced vital capacity (FVC) and first second of forced expiration (FEV1) are commonly used to assess respiratory muscle strength in neuromuscular patients. FVC and FEV1 are significantly decreased in patients with progressive diseases.[13] Respiratory muscle weakness is also evaluated by measuring the difference between FVC in the supine and sitting positions. This index is more accurate than single FVC measurements.[14]

Given the significant morbidity and mortality associated with respiratory failure in myopathies, early and accurate assessment of pulmonary function is paramount for proactive management. While spirometric measurements, particularly Forced vital capacity (FVC) in both sitting and supine positions, are established as key indicators of respiratory muscle strength, there is a need to better elucidate the specific clinical and demographic factors that correlate with respiratory decline in this heterogeneous patient population. This study, therefore, aims to comprehensively evaluate respiratory status via spirometry in patients with various myopathies. The findings from this study are intended to enhance clinical vigilance, contribute to the development of more effective monitoring protocols, and ultimately guide timely interventions to improve outcomes for patients with neuromuscular disorders.

Methods and Material

Study design and setting

This is a prospective cross-sectional study that was performed in 2020-2021 in Al-Zahra Hospital affiliated to Isfahan University of Medical Sciences.

Study participants and sampling

The current study was conducted on patients with myopathy that referred to our medical center. The inclusion criteria were previously diagnosed myopathy by expert neurologists and signing the written informed consent to participate in this study. Patients with the following criteria were excluded: Having underlying respiratory disease, including asthma, chronic obstructive pulmonary disease (COPD), or parenchymal pulmonary disease, being subjected to pharmacological intervention for acute respiratory pathology, and inability to perform spirometry.

A total number of 44 patients were recruited based on the mentioned criteria by easy sampling. The participant's demographic information and features of disease, including age, sex, type of disease, age of onset and duration of disease, and mobility status of the patient (using Walker or wheelchair) were collected by a checklist. We also gathered data regarding history of cough and history of shortness of breath.

Participants were asked to fill Epworth questionnaire (validity of which was previously determined in Iran by Amra and colleagues in 2018[15]), and the pulmonary tests were performed. The spirometry was conducted in both supine and sitting positions based on ATSIERS criteria.

Slow inspiratory vital capacity (VC) and FEV1 were obtained. Measured values were expressed as percentages of predicted values. Blood gas measurements were also performed by using a venous sample obtained with the patient resting in the sitting position and breathing room air.

Data analysis

The obtained data were entered into the Statistical Package for Social Sciences SPSS (version 24, SPSS Inc., Chicago, IL). Quantitative data were reported as mean ± standard deviation, and qualitative data as frequency distribution (percentage). Comparisons between groups were performed based on the mean of FVC using *t*-test (Mann–Whitney) in two groups, and analysis of variance (Kruskal–Wallis) in more than two groups was conducted. Chi-2 test was used to investigate the relationship between qualitative variables.

Ethical consideration

Data confidentiality was considered in this study.

Results

In this study, data from 44 patients with myopathy were analyzed. The study population consisted of 16 females (36.3%) and 28 males (63.7%) with the mean age of 39.0 ± 19.02 years (ranging from 15 to 85 years). The mean body mass index (BMI) in patients was 25.15 ± 4.52 kg/m^2 (ranging from 14.69 to 39.51 kg/m^2). 31 patients (70.5%) had no previous medical diseases and 34 patients (77.3%) had no histories of smoking. The most commonly used drugs among patients were Aspirin, Valsartan, L'Carnitine, Metformin, Amlopres, Omeprazole, Metoral, Levothyroxine, and Atorvastatin. The mean disease duration was 10.7 ± 7.05 years, and the mean age at the time of disease diagnosis was 32.0 ± 22.03 years. Analysis of blood gases demonstrated that the mean amounts of blood pH, HCO3, and PCO2 were 7.36 ± 0.04, 23.29 ± 6.61, and 38.70 ± 11.38, respectively.

Patients' Epworth test scores are shown in Table 1. Based on these data, 24 patients (54.5%) had low scores, and the mean score of patients was 4.64 ± 4.52.

As indicated in Table 2, there were no significant differences between mean scores of sitting FVC (75.45 ± 22.06) and lying FVC (69.90 ± 23.42) (P = 0.293).

FVC changes of 30 patients (68.2%) were less than 10%. Data indicated that the FVC changes were between 10-20% in 13.6% of patients and more than 20%in 18.2% of patients. The mean FVC changes were 16.94 ± 10.31, respectively [Table 3].

We also assessed the relationships between demographic and patients' characteristics with sitting FVC, lying FVC, and changes in the FVC. Based on our findings, the sitting and lying FVC were significantly higher in patients who had no drug usage (P = 0.007 and P = 0.005, respectively), no sputum (0.003 and 0.002, respectively), no dysphagia (P = 0.002 and P = 0.001, respectively), and no coughs among eating (P = 0.022 and P = 0.006). The changes in the FVC were significantly lower in patients with the mentioned conditions [Table 4].

Based on the data presented in Table 5, there were significant inverse relationships between age and sitting FVC (r = -0.638) and lying FVC (r = -0.497), and age had significant direct relationship with changes in the FVC (r = 0.652) (P < 0.001 for all). Furthermore, we observed significantly inverse relationship between age of disease onset and sitting FVC (r = -0.491, P = 0.001) and significantly direct relationship between age of disease onset and lying FVC (r = 0.447, P = 0.002) and FVC changes (r = 0.371, P = 0.013). There were also significant direct relationships between disease duration and FVC changes (r = 0.394, P = 0.008) and HCO3 and lying FVC (r = 0.341, P = 0.024). No other significant relationships were observed. These data are shown in Table 5.

Table 1: Evaluation of Epworth score

Chance of dozing	Number	Percentage
0 (never)	8	18.2
1-8 (low)	26	59.1
9-16 (moderate)	10	22.7
>17 (high)	0	0
Mean±SD	4.64±4.52	
Max-Min	16-0	

Table 2: Comparison of FVC in supine and sitting positions

Variable	Mean	SD	Min	Max	*P* (Mann–Whitney)
Supine FVC	69.90	23.42	21.00	109.00	0.293
Sitting FVC	75.45	22.06	33.00	109.00	

Table 3: Frequency of ΔFVC

ΔFVC	Percentage	Number
Less than 10%	68.2	30
10-20%	13.6	6
More than 20%	18.2	8
Total	100.0	44
Mean±SD	16.94±10.31	
Min-Max	111-(16-)	

Discussion

In the current study, we assessed spirometry data of 44 patients. Based on our findings, the amounts of FVC were higher in patients who had no drug usage, no sputum, no dysphagia, and no coughs while eating. The changes in the FVC were significantly lower in patients with the mentioned conditions. Furthermore, we observed that there were significant inverse relationships between age and sitting and lying FVC, and age had significant direct relationship with changes in the FVC. We also observed significantly inverse relationship between age of disease onset and sitting and lying FVC and FVC changes. These data show the importance of respiratory function evaluations in patients with myopathies.

As mentioned earlier, progressive respiratory failure in myopathies is an important cause of morbidity and mortality related to the early onset of muscle weakness affecting respiratory muscles, related thoracic and spinal deformities, increasing stiffness and contractures of the ribcage, and decreased respiratory compliance. Respiratory problems can lead to symptoms such as morning headaches and daytime fatigue, reduce the frequency of recurrent respiratory infections, and, once initiated, are used long term to manage progressive respiratory failure.

Table 4: Relationships between patient's data and FVC in sitting and lying positions

Variable		Number	ΔFVC			FVC sitting			FVC supine		
			Mean	SD	*P* (Mann–Whitney)	Mean	SD	*P* (Mann–Whitney)	Mean	SD	*P* (Mann–Whitney)
Sex	female	16	4.25	3.45	0.095	79.75	13.70	0.608	76.56	15.23	0.213
	male	28	13.78	20.41		73.00	25.57		66.10	26.53	
Smoking	yes	10	9.20	6.71	0.168	67.40	18.06	0.196	63.00	18.15	0.249
	no	34	10.64	19.01		77.82	22.80		71.94	26.62	
Taking a medicine	yes	13	21.23	17.25	0.003	56.53	18.51	0.007	47.53	16.39	0.005
	no	31	5.74	14.81		83.38	18.46		79.29	19.27	
Sputum	no	41	7.97	13.93	0.010	79.14	20.24	0.003	72.92	21.07	0.002
	yes	3	42.33	25.40		38.66	9.81		28.66	13.27	
Dyspnea	no	24	5.00	9.61	0.079	74.66	23.36	0.868	71.16	23.30	0.832
	yes	20	16.70	21.43		76.40	20.95		68.40	24.09	
Cough	no	40	8.40	14.05	0.490	77.00	20.86	0.178	71.60	21.63	0.222
	yes	4	29.50	31.75		60.00	31.71		53.00	36.95	
Dysphagia	no	40	7.85	14.08	0.008	78.85	19.98	0.002	73.65	20.82	0.001
	yes	4	35.00	25.40		41.50	9.81		32.50	13.27	
Cough among eating	no	38	7.00	13.93	<0.001	78.47	20.44	0.022	74.10	21.28	0.006
	yes	6	31.33	20.48		56.33	24.20		43.33	16.68	
Using wheelchair	no	34	7.29	14.32	0.186	76.66	21.92	0.145	72.38	22.34	0.354
	yes	10	10.60	21.63		71.00	23.16		61.50	26.26	

Table 5: Correlations between FVC in sitting and lying positions and different variables

Variable	FVCΔ		FVC sitting		FVC supine	
	P	*R*	*P*	*R*	*P*	*R*
Age	<0.001	0.652	<0.001	-0.638	<0.001	-0.497
BMI	0.159	0.216	0.875	0.024	0.430	0.122
Age of onset	0.013	0.371	0.001	-0.491	0.002	0.447
Duration of disease	0.008	0.394	0.423	-0.124	0.818	0.036
PH	0.662	-0.068	0.624	0.076	0.966	-0.007
Hco_3	0.284	0.165	0.078	0.268	0.024	0.341
PCO_2	0.376	-0.138	0.7315	0.053	0.996	-0.001
Epworth score	0.058	0.288	0.199	-0.197	0.182	-0.205

Different studies have assessed the respiratory functions in similar conditions. A study was performed in 2011 by Durmus and colleagues on 47 patients with myopathies. Based on the results of this study, respiratory muscle weakness was an important issue in all cases, and patients with higher ages had significantly lower respiratory functions.[16] Another study was conducted by Foley and others in 2013. By assessing the data of 145 patients from 10 neuromuscular centers, they reported that the vital capacity of the patients decreases 2.3-2.6% per year and in patients with Ullrich congenital muscular dystrophy, the nocturnal non-invasive ventilation was initiated by 11.3 years. It was also shown that age had a significantly inverse relationship with respiratory functions.[17] These results were in line with the findings of our study.

An important point of this study was that we evaluated the respiratory functions in patients via spirometry and reported different correlations between the variables and patient's characteristics. We observed significantly reduced FVC in patients with higher disease onset age and older patients. researches evaluated the respiratory function and sleep disorders in patients with centronuclear myopathies. It was reported that age of disease onset had significant inverse relationship with respiratory functions in patients.[18,19] These results were also consistent with the findings of the present study.

Another important point of this study was that we evaluated the Epworth score in patients. This also showed that the majority of patients had medium scores. Although we did not observe a significant decrease in FVC between sitting and supine positions in participants, this lack of difference does not imply lack of diaphragmatic involvement in these individuals. Rather, it is reasonable to assume that the diaphragmatic contribution to respiratory compromise is not as disproportionate as to effect on spirometry but rather correlates more with the overall skeletal muscle weakness in the patient. In 2015, Meilleur and colleagues assessed the clinical outcome measures in collagen VI-and laminin alpha2-related congenital muscular dystrophies. They mentioned that there were no significant differences between the sitting and lying FVC among patients, but most cases suffer from skeletal muscle weakness.[20] These data were in line with our findings. Sinha 2021 also reported in his study that the effect of sitting and lying positions on pulmonary function tests varies across study populations, which makes it difficult to establish an optimal position for performing pulmonary function tests. However, supine lying position may be considered in those disease conditions where adopting a sitting position may not be possible for the subject.[21]

According to our results Δ FVC was significantly higher in patient's whit cough among eating, dysphasia, and

sputum. So it seems that visiting patients regularly and following the symptoms could be the most beneficial intervention. We also found that FVC pp sitting correlated inversely (i.e. decreased) with age, duration of diseases, and age of disease onset; on the other hand, FVC supine correlated inversely with age and age of disease onset, and Hco3 (VBG), which suggests supine Spirometry could predict hypo ventilation better than sitting spirometry. We attribute this at least in part to the fact that supine position could reduce the FVC.

These findings could have high clinical importance. We believe that early evaluations of patients with myopathies regarding the respiratory functions could be very valuable. This study possesses several notable strengths. Firstly, it addresses an under-investigated area by prospectively evaluating respiratory function in a specific cohort of patients with myopathy, providing valuable clinical insights. The comprehensive assessment protocol, which included spirometry in both sitting and supine positions, venous blood gas analysis, and the use of the validated Epworth Sleepiness Scale, offers a multifaceted view of the patients' respiratory status.

Despite its contributions, this study is subject to certain limitations. The most significant limitation is the relatively small sample size (n = 44), which may limit the statistical power and the generalizability of the findings to the broader population of patients with myopathy. The utilization of a convenience sampling method, as opposed to random sampling, introduces the potential for selection bias.

Conclusion

There were significant inverse relationships between age and sitting and lying FVC and age had significant direct relationship with changes in the FVC. We also observed significantly inverse relationship between age of disease onset and sitting and lying FVC and FVC changes. These data show the importance of respiratory function evaluations in patients with myopathies. The results of this study can be used for the field of treatment and health system

It is suggested that periodic Assessments of respiratory function programs (including spirometry in both sitting and supine positions) be mandatory as a standard protocol for all patients with myopathy, regardless of the type of disease or the presence of overt respiratory symptoms.

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Conflicts of interest

There are no conflicts of interest.

References

1. Kumar P, Dwivedi P. Diagnosis of neuromuscular disorder. In: Computational Intelligence for Genomics Data. Academic Press; 2025. p. 225-40.
2. Michel C, Collins C. Pediatric neuromuscular disorders. Pediatr Clin 2020;67:45-57.
3. Latronico N, Rasulo FA, Eikermann M, Piva S. Critical illness weakness, polyneuropathy and myopathy: Diagnosis, treatment, and long-term outcomes. Crit Care 2023;27:439.
4. Shepherd S, Batra A, Lerner DP. Review of critical illness myopathy and neuropathy. Neurohospitalist 2017;7:41-8.
5. Patel N, Howard IM, Baydur A. Respiratory considerations in patients with neuromuscular disorders. Muscle Nerve 2023;68:122-41.
6. Triplett J, Kassardjian CD, Liewluck T, Tahir A, Lennon V, Kopecky S, *et al.*, editors. Cardiac and respiratory complications of necrotizing autoimmune myopathy. Mayo Clinic Proceedings. Elsevier; 2020.
7. Pfeffer G, Povitz M, Gibson GJ, Chinnery PF. Diagnosis of muscle diseases presenting with early respiratory failure. J Neurolog 2015;262:1101-14.
8. de Winter JM, Ottenheijm CA. Sarcomere dysfunction in nemaline myopathy. J Neuromuscul Dis 2017;4:99-113.
9. Boentert M, Wenninger S, Sansone VA. Respiratory involvement in neuromuscular disorders. Curr Opin Neurolog 2017;30:529-37.
10. Koo P, Sethi JM. Metabolic myopathies and the respiratory system. Clin Chest Med 2018;39:401-10.
11. Tasca G, Udd B. Hereditary myopathy with early respiratory failure (HMERF): Still rare, but common enough. Neuromuscul Disord 2018;28:268-76.
12. Carter JC, Sheehan DW, Prochoroff A, Birnkrant DJ. Muscular dystrophies. Clin Chest Med 2018;39:377-89.
13. El-Ganainy SO, El-Mallah A, Abdallah D, Khattab MM, El-Din MMM, El-Khatib AS. Elucidation of the mechanism of atorvastatin-induced myopathy in a rat model. Toxicology 2016;359:29-38.
14. Graham RJ, Muntoni F, Hughes I, Yum SW, Kuntz NL, Yang ML, *et al.* Mortality and respiratory support in X-linked myotubular myopathy: A RECENSUS retrospective analysis. Arch Dis Child 2020;105:332-8.
15. Amra B, Javani M, Soltaninejad F, Penzel T, Fietze I, Schoebel C, *et al.* Comparison of berlin questionnaire, STOP-bang, and Epworth Sleepiness scale for diagnosing obstructive sleep apnea in Persian patients. Int J Prevent Med 2018;9:28.
16. Durmus H, Laval S, Deymeer F, Parman Y, Kiyan E, Gokyigiti M, *et al.* Oculopharyngodistal myopathy is a distinct entity: Clinical and genetic features of 47 patients. Neurology 2011;76:227-35.
17. Foley AR, Quijano-Roy S, Collins J, Straub V, McCallum M, Deconinck N, *et al.* Natural history of pulmonary function in collagen VI-related myopathies. Brain 2013;136:3625-33.
18. Smith BK, Goddard M, Childers MK. Respiratory assessment in centronuclear myopathies. Muscle Nerve 2014;50:315-26.
19. van Kleef ES, van Doorn JL, Gaytant MA, de Weerd W, Vosse BA,

Wallgren-Pettersson C, *et al*. Respiratory muscle function in patients with nemaline myopathy. Neuromuscul Disord 2022;32:654-63.

20. Meilleur KG, Jain MS, Hynan LS, Shieh C-Y, Kim E, Waite M, *et al*. Results of a two-year pilot study of clinical outcome measures in collagen VI-and laminin alpha2-related congenital muscular dystrophies. Neuromuscul Disord 2015;25:43-54.
21. Sinha S, Sinha RK. Effects of body position on the outcome of pulmonary function tests in non-mechanically ventilated adults: A brief review. Indian J Forensic Med and Toxicol 2021;15:781-5.