Improving Access and Guideline Adherence in Pulmonary Care in Patients With Duchenne Muscular Dystrophy

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BACKGROUND: Duchenne muscular dystrophy (DMD) is a devastating, progressive neuromuscular disease that results in cardiopulmonary failure and death. In 2018, the DMD Care Considerations guidelines were updated to improve the multidisciplinary approach to care and promote early respiratory management. We sought to evaluate the impact of a multidisciplinary clinic on access to pulmonary care and adherence to respiratory care guidelines. METHODS: Utilizing retrospective data, we assessed for pulmonary care between 2016–2019 and congruence with guidelines from March 2018–February 2019. Using a standardized visit protocol, subjects were monitored for adherence to pulmonary function testing (PFT) and polysomnography (PSG) recommendations. RESULTS: Of the 84 subjects with DMD, only 51.2% had prior pulmonary involvement, and approximately one-third were seen in the year prior to clinic onset. Only 23% of subjects with a pulmonary referral completed this visit. After clinic initiation, the average age of a subject's first pulmonary contact decreased from 11.8 y to 7.9 y ($P < .001$), and 45% of the 77 unique clinic subjects had no previous pulmonary encounter. Adherence to PFT guidelines increased in both ambulatory (8.7% to 86.1%) and non-ambulatory subjects (25.9% to 90.1%). Approximately 79% of subjects seen in clinic either completed or had an order for PSG in the last 12 months. CONCLUSIONS: Development of a multispecialty clinic expanded access to pulmonary care and evaluation in subjects with DMD. Continued care in this clinic will allow a better understanding of barriers to access and the opportunity to monitor long-term pulmonary health. *Key words: Duchenne muscular dystrophy; pulmonology; lung function testing; spirometry; neuromuscular disorders; polysomnography.* [Respir Care 2022;67(3):347–352.

Introduction

Duchenne muscular dystrophy (DMD), an X-linked recessive disorder caused by mutations in the dystrophin gene, is the most common neuromuscular disorder in childhood and occurs in approximately 1:3,000–5,000 male births.[1] Mutations in the dystrophin gene lead to severely reduced or absent dystrophin protein, resulting in destabilization of the scaffolding network linking muscle to the extracellular matrix[2] and ultimately increased oxidative stress, muscle fiber necrosis, and fibrofatty replacement. Currently, there is no cure for DMD; and without medical intervention, patients usually die in the second or third decade of life.[3] Life expectancy has increased over the past 10–15 years due to early initiation of corticosteroids and noninvasive respiratory support.[4] However, respiratory insufficiency remains one of the major causes of morbidity and mortality in the DMD population.[5]

The gradual loss of respiratory function in DMD often begins in the latter half of the first decade of life with progression to restrictive pulmonary disease, hypoventilation, inability to clear airway secretions, and increased risk for pulmonary infections.[6] Early use of respiratory interventions to improve lung recruitment, assist with cough, and counter hypoventilation has been shown to reduce morbidity and improve survival.[7-9] Unfortunately, patients with DMD may not perceive the loss of respiratory muscle strength and subsequent ineffective cough until they progress to respiratory failure.[10] Timely and early initiation of support to counter respiratory failure improves both quality of life and overall survival.[11-13]

In 2018, the DMD Care Considerations Working Group steering committee updated their care guidelines with an emphasis on multidisciplinary care and early lung function testing.[14] Assessments and interventions for respiratory care were outlined according to disease stage (ambulatory,

early non-ambulatory, late non-ambulatory). The updated respiratory standard of care includes pulmonary function testing (PFT) for all patients at 5–6 y of age and every 6–12 months thereafter. Lung function testing should also include maximum inspiratory pressure (P_{Imax}), maximum expiratory pressure (P_{Emax}), cough peak flow (CPF), and capnography. This early testing is critical as clinical respiratory decline and loss of ambulation often manifest in the second decade, but "silent" respiratory muscle weakness without obvious symptomology can begin much earlier.[15] Additionally, patients with DMD have a multitude of subsequent comorbidities requiring involvement of multiple subspecialty providers. This leads to numerous visits to medical centers and clinics with potential to decrease clinic visit adherence. Initiation of a comprehensive, multidisciplinary clinic with patients being seen by multiple providers in a single visit can relieve this management burden.

In a proactive effort to provide optimal care in congruence with new guidelines, we established a multidisciplinary DMD clinic with providers from pediatric cardiology, pediatric neurology, pediatric pulmonary, respiratory therapy, pediatric palliative care, nutrition, and physical therapy with the expressed goal of improved care coordination among multiple subspecialties. Since nearly 30–40% of the patients with DMD at our institution who were followed by either pediatric cardiology or pediatric neurology had no previous exposure to pulmonary providers, we hypothesized that this newly created multidisciplinary DMD clinic would result in a 20% increase in the proportion of patients meeting pulmonary standard-of-care guidelines.

QUICK LOOK

Current knowledge

Cardiopulmonary failure is a leading cause of morbidity in patients with Duchenne muscular dystrophy (DMD), and early intervention has been shown to improve survival. Recent care guidelines emphasize early disease recognition with a multidisciplinary approach. Instituting a dedicated, multispecialty clinic for patients with DMD may allow for improved pulmonary care access and increased pulmonary disease detection.

What this paper contributes to our knowledge

Utilization of a multidisciplinary clinic increased adherence to standardized care guidelines for subjects with DMD by improving rates of spirometry and polysomnography. The clinic can also serve as an early entry point for pulmonary care prior to the onset of respiratory symptoms.

Methods

Electronic health records (Epic Systems, Verona, Wisconsin) were queried for subjects under the age of 25 with a diagnosis of DMD who were evaluated at our institution by pediatric neurology, pediatric cardiology, or pediatric pulmonology between 2016–2018. Patients who received their primary DMD care at other centers but participated in research trials at Vanderbilt University Medical Center were excluded. Patients who subsequently moved or transferred centers were also excluded. Diagnosis of DMD was determined by ICD-10 code of G71.01 with subsequent chart analysis to confirm diagnosis via genetic testing or muscle biopsy. The study and protocol were approved by the Vanderbilt Institutional Review Board.

Prior to the creation of the DMD clinic, patients at our institution were independently followed by multiple providers, with involvement of cardiology and pulmonology usually resulting from ambulatory referrals from neurology. Pulmonary care was not standardized and often dictated by the individual provider. Additionally, appointments with subspecialists were rarely coordinated, resulting in multiple visits, increased transportation burden, and fragmented care.

Starting in March 2019, prospective data were collected from the DMD cohort at first evaluation in the clinic and compared to the implemented standard of care (Table 1). At time of first visit, subjects were categorized as

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Table 1. Standard-of-Care Recommendations Based on Clinical Stage

Ambulatory	Early Non-Ambulatory	Late Non-Ambulatory
Annual lung function testing	Bi-annual lung function testing	Bi-annual lung function testing
Annual capnography	Bi-annual capnography	Bi-annual capnography
Polysomnogram if symptoms of SDB	Polysomnogram if symptoms of SDB	Polysomnogram if symptoms of SDB
End-tidal CO_2 measurement at each visit	End-tidal CO_2 measurement at each visit	End-tidal CO_2 measurement at each visit
Annual cough peak flow	Bi-annual cough peak flow	Bi-annual cough peak flow
Cough assist use when sick	Daily cough assist use and as needed when sick	
	Nocturnal assisted ventilation as needed	Addition of daytime assisted ventilation as needed

SDB = sleep-disordered breathing
Adapted from 2018 Duchenne Muscular Dystrophy Care Considerations and Certified Duchenne Care Centers requirements.

ambulatory if they were able to walk, even with assistance, and non-ambulatory if they were dependent on a wheelchair for mobility. Completion of yearly lung function testing and bi-annual testing, including spirometry, P_{Imax}, P_{Emax}, CPF, and end-tidal capnography, was assessed at each visit for ambulatory and non-ambulatory subjects, respectively. All lung function testing was performed by a dedicated respiratory therapist. Screening for symptomatic sleep-disordered breathing (daytime sleepiness, napping during the day, change in school performance or behavior, gasps/pauses in breathing) was performed at each visit. Ambulatory subjects with positive screen were referred for polysomnography (PSG), and non-ambulatory subjects were referred when symptomatic or when testing demonstrated decreased lung function (based on criteria outlined by 2018 guidelines). All PSGs for patients with DMD at our institution are level 1 sleep studies. Subjects with lung function testing below the guideline thresholds or abnormal PSG were considered for initiation of lung recruitment (ie, insufflation-exsufflation devices) and assisted ventilation (bi-level positive airway pressure with backup rate).

Statistical Analysis

Study data were stored and managed in REDCap (Vanderbilt University, Nashville, Tennessee). Independent t test was used to compare the average of first pulmonary diagnosis. Chi-square test was performed to assess difference between preclinic and postclinic adherence to guidelines for both PFTs and PSGs. Statistical analysis was performed using Microsoft Excel (Microsoft, Redmond, Washington).

Results

Demographics

A total of 84 subjects were identified via the electronic medical record with a diagnosis of DMD and seen by one of the 3 subspecialties (pulmonary, cardiology, or neurology) at least once in the 3 y prior to initiation of the clinic. During the period of 2016–2018, there were 5 deaths at an average age of 18.4 y. All subjects in our cohort resided at home with caregivers. Two subjects required home health nursing.

Prior to Clinic Initiation

Prior to clinic onset, 52 (62%) of the 84 subjects were referred to pediatric pulmonology. Of these, 77% were never seen by a pulmonary provider. We found that 43 subjects (51.2%) had any lifetime contact with a pulmonary provider (adult or pediatric). The average age of these subjects at time of first contact with pulmonary was 11.8 y.

Only 31 subjects (36.9%) were seen by a pulmonary provider in the year (March 2018–February 2019) preceding initiation of the clinic. Of these subjects, 5 (16.1%) were ambulatory and 26 (83.8%) were non-ambulatory. Only 40% of ambulatory subjects were meeting recommended guidelines of at least one annual PFT. Of the 26 non-ambulatory subjects, 6 (23.1%) would be considered in agreement with standard of care with at least 2 PFTs within the year. Approximately 60% of ambulatory subjects had a PSG either ordered or completed compared with 38% of non-ambulatory subjects.

After Clinic Initiation

From the period between March 2019–April 2021, a total of 77 unique subjects were seen in the multidisciplinary clinic over 168 visits. The clinic was the first pulmonary contact in 45.2% (38) of the subjects (Table 2). The average age of subjects at first clinic visit was 13.3 y. However, there were 10 new DMD diagnoses between March 2019–April 2021 with an average age of 7.9 y at time of first visit compared to 11.8 y prior to clinic initiation. There were also 2 subject deaths during this period with an average age of 15.3 y.

Of the ambulatory subjects seen in clinic, 8.7% had any lung function testing in the prior year (Table 3). After

Table 2. Subject Demographics

	Prior to Clinic Initiation (2016–2018)	Since Clinic Initiation (2019–2021)
Total subjects	84	94
Any pulmonary contact	43 (51.2%)	83 (88.3%)
Any cardiology contact	71 (84.5%)	86 (91.4%)
Any neurology contact	81 (97.6%)	88 (93.6%)
Age at first pulmonary visit, y	11.8	13.3 (New diagnosis: 7.9)
Deaths, *n*	5	2

Data are presented as *n* (%) unless otherwise noted.

initiation of the clinic, this increased to 86.1% of subjects, with the majority of those who did not complete PFT being too young to adequately or accurately perform the maneuvers. Similarly, whereas 8.7% of ambulatory subjects had a PSG completed or ordered prior to their first visit, after initiation of the clinic 63.9% of ambulatory subjects in clinic had a PSG completed or ordered.

Of the non-ambulatory subjects seen in clinic, there was an increase from 25.9% to 90.1% of subjects meeting the guidelines of bi-annual PFT. The percentage of non-ambulatory subjects with capnography measurements rose from 3.5% to 93.1%. Similarly, the proportion of non-ambulatory subjects with a PSG completed or ordered increased from 18.5% to 84.0%.

Additional measurements, such as P_{Imax}, P_{Emax}, and CPF were measured as part of routine spirometry for non-ambulatory subjects. Eighty (61.1%) of the non-ambulatory patient visits had P_{Imax} and P_{Emax} performed, whereas 99 (75.6%) had a completed CPF.

Discussion

This study highlights a strategy for improving access to pulmonary care and increased pulmonary function monitoring. With the adoption and initiation of the 2018 care guidelines, a multidisciplinary clinic was established in effort to reduce perceived barriers to care and foster a collaborative environment when caring for these patients. By combining the pulmonary visit with other subspecialty visits, we were able to capture a higher percentage of subjects followed at our institution with a substantial decrease in age at first contact for new diagnoses.

Table 3. Pulmonary Function Testing and Polysomnography in Last 12 Months at Time of Clinic Visit

	Ambulatory		Non-Ambulatory	
	n/*n*	%	*n*/*n*	%
First visit	23/77	29.9	54/77	70.1
Total clinic visits	36/168	21.6	131/168	78.4
Prior to clinic, PFT x1	2/23	8.7	14/54	25.9
After clinic start, PFT x1	31/36	86.1	118/131	90.1
Prior to clinic, PFTx2			5/54	9.3
After clinic start, PFTx2			110/131	84.0
Prior to clinic, P_{Imax}/P_{Emax} x1			7/54	13.0
After clinic start, P_{Imax}/P_{Emax} x1			108/131	82.4
Prior to clinic, P_{Imax}/P_{Emax} x2			0/54	0
After clinic start, P_{Imax}/P_{Emax} x2			65/131	40.5
Prior to clinic, CPF x1			4/54	7.4
After clinic start, CPF x1			117/131	89.3
Prior to clinic, CPF x2			0/54	0
After clinic start, CPF x2			65/131	49.6
Prior to clinic, PSG performed or ordered	2/23	8.7	10/54	18.5
After clinic start, PSG performed or ordered	23/36	63.9	110/131	84.0

P values for all pre/post clinic comparisons are < .001.
PFT = pulmonary function testing
P_{Imax} = maximum inspiratory pressure
P_{Emax} = maximum expiratory pressure
CPF = cough peak flow
PSG = polysomnography/polysomnogram

Since implementation of the clinic, both pulmonary and cardiology saw an increase in the percentage of patients with DMD seen by a provider (Table 2). Pulmonary experienced the largest increase (51.2% to 88.3%). The percentage of subjects seen by neurology showed a slight decrease (97.6% to 93.6%) due to a number of factors including patients with frequent cancellations/rescheduling and hesitancy due to infection control during the global pandemic.

Prior to clinic, both lack of pulmonary referral and follow-up were driving factors for lack of pulmonary evaluation and management. Despite 62% of subjects having a pulmonary referral, approximately three-quarters of those had not been seen by a pulmonary provider. This referral pattern is not unique to our institution as members of the Canadian Pediatric Neuromuscular Group reported less than a quarter of patients have a routine referral to pediatric pulmonology.[16] The creation of the multidisciplinary clinic reduced the need for a discrete ambulatory referral, eliminating a potential barrier to pulmonary care.

In the year prior to the clinic onset, only slightly more than a third of subjects with DMD followed at our institution were seen by a pulmonary provider, which is consistent with previous studies. Andrews et al[17] found that, over the course of a decade, less than half of subjects with DMD in a multi-institution research network had bi-annual visits with a pulmonary provider, and $< 32\%$ of their population had pulmonary evaluation in the final year of the study. Our bimonthly clinic served as the first pulmonary contact for 45% of our total subjects with DMD population. The average age of first contact with pulmonary increased from 11.8 y to 13.3 y due to capturing of older subjects with no previous pulmonary encounters, whereas the average age of initial pulmonary evaluation for new diagnoses decreased to 7.9 y.

Following initiation of the clinic, there was a significant increase in the percentage of subjects (both ambulatory and non-ambulatory) who met the recommended standard of care for spirometry. The most common reason for failure in ambulatory subjects was a lack of understanding of testing procedure/maneuvers; most non-ambulatory failures were due to fatigue or disinterest in performing the testing. Both of these inherent limitations are well known[18] and documented in the DMD population.[19] However, scheduling lung function testing into structured clinic visits has enabled respiratory therapists to teach and reinforce good spirometric technique. These patients require vigilant symptomatic monitoring, especially those in the non-ambulatory phase when respiratory complication rates are increased.

The use of PSGs is a cornerstone of disease monitoring in patients with DMD. This becomes especially important due to lack of significant association between symptoms perception and presence of sleep-disordered breathing.[20] In our clinic, sleep studies were ordered or performed in conjunction with 79.2% of all subjects encounters. This represents a significant increase from prior to clinic initiation when the rate was 15.6%. Some encounters involved repeated sleep studies, which are recommended in DMD due to progression of clinical disease.[21] We included ambulatory subjects in our overall assessment of sleep studies to capture the bimodal distribution of sleep disorder breathing in patients with DMD[22] and to ensure that patients who were unable to perform lung function testing underwent objective assessment of their respiratory status.

Overall, we saw significant improvement in access to pulmonary care and overall adherence to national, standardized monitoring guidelines. Despite the perceived increase in ease in seeing multiple providers during a single visit, there remains a small subset of our cohort that would not be considered compliant with annual/bi-annual visits. It is unclear how the ongoing SARS-CoV-2 (COVID-19) pandemic has affected our patients' willingness to travel to our medical center for in-person assessments.

Limitations

There are several limitations of this study. The most striking limitation is the short time course of the DMD clinic, which has only operated for 24 months (and halted in-person visits for several months due to COVID-19). Additionally, infection prevention in the pulmonary function lab temporarily limited our ability to perform maximal pressures and CPF, although we were able to assess inspiratory function with negative inspiratory force.

There is an expectation that the remaining subjects with DMD will be seen in upcoming clinics, although a small subset has elected to see individual providers outside of the specified clinic. However, provider availability and clinic workspace remain an issue, especially given the multitude of clinical assessments these subjects undergo at each multidisciplinary visit. A systemic meta-review of the literature regarding implementation of clinical guidelines by Franke et al[23] found that limited time and personnel resources are frequently cited as barriers that negatively influence implementation. Expanded physician coverage and space may allow for increased volume per scheduled clinic day and decreased lag time between visits.

Given the inherent issues with mobility that worsen with age, barriers to transport remain a problem. This is compounded by the rural catchment area that surrounds our particular institution and becomes more significant when attempting to schedule, coordinate, and complete sleep studies. Whereas many sleep centers are reluctant to perform sleep studies on pediatric patients, there are fewer facilities in our region with the capability to handle patients with mobility and transfer issues. The infection prevention measures implemented by many sleep centers remain a barrier to not only scheduling but completion of PSGs.

Future Directions

As the clinic develops into a medical home for patients with DMD, we plan to explore the longitudinal impact across multiple disciplines. Quality-of-life surveys are completed at each visit with emphasis on assessing changes following initiation of lung recruitment or ventilation assistance. The consistent use of dedicated respiratory therapists for repeated spirometry attempts can improve patient technique, allowing for more accurate measurements and potentially more appropriate use of lung recruitment devices. We also plan to explore home spirometry for more frequent monitoring. Our standard practice of screening for sleep symptoms and tracking completed PSGs based on patient location will enable us to both predict and identify barriers to the management of sleep-disordered breathing.

Conclusions

The implementation of a multidisciplinary clinic has potential to improve access to pulmonary care and increase adherence to nationally recognized guidelines for patients with DMD. Additional investigation is needed to fully understand the clinical impact of a multidisciplinary clinic on respiratory outcomes and quality of life.

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