Pulmonary Function Testing in Neuromuscular and Chest Wall Disorders

Eric J. Gartman, MD*

KEYWORDS

• Pulmonary function testing • Neuromuscular disease • Chest wall disorders

KEY POINTS

- Patients with neuromuscular and chest wall disorders may have no respiratory symptoms and limited signs of skeletal muscle weakness, but can have significant respiratory muscle weakness.
- A single testing modality may fail to elucidate true respiratory compromise, and often a combination of tests is recommended to fully evaluate these patients.
- Common tests performed on this population include measurement of flow rates, lung volumes, maximal pressures, and airway resistance.
- Most tests needed to evaluate these patients are available through a standard pulmonary function laboratory, but occasionally referral to a specialty laboratory may be required.

Neuromuscular diseases (NMDs) and chest wall disorders have a wide range of effects on the respiratory system, and unfortunately these lead to significant disability and/or progressive respiratory failure in many patients. Respiratory disorders can develop as a consequence of the multitude of functional limitations that may arise in these varied conditions, and are implicated as a leading cause of mortality for many patients.[1,2] Alterations in respiratory system physiology can arise from a combination of effects on the brain, spinal cord, peripheral nervous system, neuromuscular junction, skeleton, and musculature. Further, each disease may have a unique pattern of effect on the respiratory system, and may progress differently over time.

Given the consequential respiratory system morbidity in these conditions, it is imperative to have a thorough understanding of the physiologic effects of each disease (discussed in detail in other articles). In turn, one then may have a better understanding of how to investigate and interpret the battery of physiologic tests that have been developed. It is important to recognize that given the diversity of dysfunction that may arise from these disorders, the same test or parameter that is helpful in 1 condition may not prove as useful in another. Further, patients often may not present with typical respiratory symptoms and may need to have significant functional impairment or weakness before standard pulmonary function tests demonstrate clear abnormalities.[3–6]

This article focuses on the various testing modalities that have been studied in the evaluation of these patients (**Table 1**), with the goal of understanding that it is often a pattern of findings, not just the extent of dysfunction, that is the key to assessing and following these patients over time.[7] Although some effects on the respiratory system are more obvious (eg, muscle weakness,

Disclosure Statement: The author has no relationship with a commercial company that has a direct financial interest in subject matter or materials discussed in article or with a company making a competing product.
Department of Medicine, Division of Pulmonary, Critical Care, and Sleep Medicine, Warren Alpert Medical School of Brown University, Providence, RI, USA
* Providence VA Medical Center, 830 Chalkstone Avenue, Providence, RI 02908.
E-mail address: eric_gartman@brown.edu

Clin Chest Med 39 (2018) 325–334
https://doi.org/10.1016/j.ccm.2018.01.005
0272-5231/18/Published by Elsevier Inc.

Table 1
Pulmonary function tests performed in neuromuscular and chest wall disorders

Commonly Available	Availability Usually Limited to More Advanced Laboratories or Research Centers
Spirometry	Sniff nasal pressures
Lung volumes	Invasive catheter measurement of pressures
Static mouth pressures (MIP/MEP)	Phrenic nerve stimulation (eg, magnetic)
Maximal voluntary ventilation (MVV)	Impulse oscillometry
Positional Testing (eg, supine)	
Flow-volume loops	
Bronchoprovocation testing (eg, methacholine)	

decreased flows), others may secondarily arise and may lead to the deleterious outcomes that are observed (eg, poor cough, swallowing and upper airway dysfunction, skeletal and chest wall abnormalities, and sleep-disordered breathing).[8–11] There are no set guidelines to dictate the frequency of testing, and generally this is determined by the rapidity of clinical progression.[12]

SPIROMETRY

The most commonly utilized testing modality, likely because of its ubiquitous availability, is spirometry, with the hallmark parameter being the vital capacity (VC) (and less so the maximal voluntary ventilation [MVV]). The performance of spirometry is standardized and meant to follow outlined quality controls, which enables it to serve as a reliable and repeatable measurement.[13] However, it is important to recognize that patients with neuromuscular diseases may not be able to perform the maneuvers as intended, and adaptation of the standards or accommodation may be necessary.[14,15]

The value of spirometry when used alone to test for impairment has long been debated, because it has been shown that significant muscle weakness may be present even though the vital capacity is normal.[16,17] However, it also has been recognized that patients may have little clinically perceived respiratory dysfunction, but when formally assessed they are clearly impaired. In 2 studies of patients with motor neuron disease (mostly ALS) totaling 254 participants, significant declines in lung function were evident even though they expressed few or no symptoms (sometimes as low as 50% of predicted).[4,5]

Spirometry's most consistent role is in the longitudinal assessment of patients and in determining prognosis. In multiple neuromuscular conditions, it has been shown to be a predictor of disease disability scores, disease-related complications, need for assisted ventilation, and survival time.[4,5,18–20] Further, improvements in spirometric parameters have been used as evidence of treatment response in Parkinson's disease and myasthenia gravis.[21–23]

In patients with cervical spinal cord injury, it has been recognized that bronchial hyperresponsiveness can develop, likely because of loss of sympathetic innervation.[24–27] This can be investigated using a broncho-provocation challenge, most commonly conducted by having the patient inhale increasing doses of methacholine and repeating spirometry following each dose. A positive test is achieved by observing more than a 20% decline in the baseline forced expiratory volume in 1 second (FEV1).

Supine Positioning

One valuable maneuver in the evaluation of certain neuromuscular disorders is comparing the vital capacities between seated and supine positioning. With significant diaphragm weakness or paralysis, the vital capacity will be observed to fall in the supine position, and the extent of the decline will depend on both the severity of weakness and whether one or both diaphragms are affected. In unilateral disease, the vital capacity can decline 15% to 25% (with right-sided issues being more significant because of the weight of liver) and can fall 40% or more in bilateral disease.[28–30]

It should be recognized, however, that certain conditions lead to an increase in vital capacity in the supine position, often leading to confusion clinically. Conditions that leave the diaphragm neurologically intact but the chest wall and abdominal muscles impaired can lead to such a scenario (eg, high spinal cord injuries and large ventral hernias with loss of functional abdominal musculature). The decrease in vital capacity in the upright position results from increased abdominal compliance leading to diaphragmatic dysfunction caused by effects on the length-tension properties of the diaphragm and on the normal chest wall configuration afforded by the abdominal muscles' actions on the rib cage. This has led to the suggestion that reducing the abdominal compliance by binding the abdominal wall can improve upright inspiratory function in such patients.[11,31–38]