Pulmonary Function Testing in Neuromuscular Disorders

Girish D. Sharma, MD
Department of Pediatrics, Rush University Medical Center, Chicago, Illinois

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ABSTRACT
This is a summary of the presentation on pulmonary function testing in neuromuscular disorders presented as part of the program on pulmonary management of pediatric patients with neuromuscular disorders at the 30th annual Carrell-Krusen Neuromuscular Symposium on February 20, 2008. Pediatrics 2009;123:S219–S221

NEUROMUSCULAR DISORDERS SUCH as Duchenne muscular dystrophy are associated with gradual loss of muscle function over time. Involvement of respiratory muscles results in ineffective cough and decreased ventilation and leads to atelectasis and respiratory insufficiency.1–3 A combination of reduced lung compliance caused by generalized and widespread microatelectasis and chest wall deformity caused by increased chest wall compliance4 results in increased work of breathing and chronic respiratory insufficiency. Furthermore, musculoskeletal deformities such as kyphoscoliosis contribute to restrictive lung disease.

Pulmonary function testing in patients with neuromuscular disorders helps to evaluate the respiratory status of patients at the time of diagnosis, monitor their progress and course, evaluate them for possible surgery, and gives an overall idea of the prognosis.

CLINICALLY USEFUL TESTS FOR PATIENTS WITH NEUROMUSCULAR DISORDERS

Lung volumes:
- total lung capacity (TLC)
- residual volume (RV)

Spirometry:
- peak flow rate (PFR)
- cough peak flow (CPF)
- forced vital capacity (FVC)
- forced expiratory volume in 1 second (FEV₁)
- maximum insufflation capacity (MIC)

Respiratory muscle strength:
- maximal expiratory pressure (MEP)
- maximal inspiratory pressure (MIP)

Other tests, including laboratory tests:
- oxyhemoglobin saturation by pulse oximetry
- capnography: end-tidal CO₂ measurement
- arterial or venous blood gas profile

DESCRIPTION OF TESTS

Lung Volumes
Lung volume (how big the lungs are) at any instant depends on the mechanics of the lungs and chest wall and the activity of muscles of inspiration and expiration. Other factors that affect lung volume are height, weight, age, gender, and race. TLC is the lung volume when the lung is fully inflated and requires measurement of functional residual capacity (FRC). TLC is set by the lung’s inward recoil and the strength of contraction of the chest muscles.
Hence, it will be affected by muscle weakness caused by neuromuscular disorders, and a low TLC is the hallmark of a restrictive ventilatory defect. The restrictive lung disorders are categorized according to their cause:

- increased lung recoil (interstitial fibrosis);
- chest wall abnormality (kyphoscoliosis); and
- neuromuscular weakness (Duchenne muscular dystrophy, spinal muscular dystrophy).

The increased lung recoil is associated with low values for FRC and RV. RV, the volume of gas that remains in the lungs at the end of a maximal exhalation, is determined by the ability of expiratory muscles to compress the chest wall inward. Hence, muscle weakness caused by involvement of expiratory muscles results in elevated values of RV and may be one of the earliest and only findings. In neuromuscular weakness, the TLC is low, FRC is normal, and RV is high. The FRC and RV may be normal or low in chest wall deformities. Restrictive lung disease can be classified according to the reduction in percentage predicted values for TLC as follows:

- 80% to 120% predicted: normal;
- 70% to 80% predicted: mild restriction;
- 60% to 70% predicted: moderate restriction; and
- less than 60% predicted: severe restriction.

**Spirometry**
A spirometer is used to track the changes in lung volume during a forced exhalation from TLC to RV. Spirometry is represented graphically by the flow-volume loop (Fig 1 www.pediatrics.org/content/vol123/Supplement_4.).

**Peak Flow Rate**
Muscle weakness caused by neuromuscular disorders results in reduced values for peak flow which is measured by peak flow meter. PFR is effort dependent. The use of CPF can minimize effort-related variation. Thus, for patients with neuromuscular weakness, CPF measured by peak flow meter is a better and more reliable measurement of expiratory muscle strength. This inexpensive and sturdy device can be used to monitor muscle strength at home.

For assisted CPF measurements, the patient air-stacks as deeply as possible and abdominal thrust is applied. The CPF is measured by using a peak flow meter. The difference between assisted and unassisted CPF can be used to measure glottic integrity.

**Forced Vital Capacity**
During an FVC maneuver, the patient takes a maximum breath and fills his or her lungs to TLC and then exhales to the maximum. The total volume of air expelled during forced exhalation after maximum inspiration is the FVC. The values are reduced in patients with neuromuscular disease. Measurements of vital capacity while the patient is sitting, laying supine, laying on his or her side, and wearing a thoracolumbar stabilizing device is practiced by some physicians and may give more realistic information. Similarly, vital capacity measured with the patient in a recumbent position may be helpful. A >7% decrease in vital capacity from a sitting to a supine position may indicate that diaphragmatic weakness is out of proportion to chest wall muscle weakness.

FEV1 is reduced in proportion to FVC; thus, FEV1/FVC remains normal. A reduced FEV1/FVC value of <70% predicted suggests an obstructive process.

MIC* is the measure of maximum volume of air that the glottis can hold in the lungs by “air stacking” consecutively delivered from a volume-cycled ventilator and manual resuscitator. The MIC/vital capacity difference is a direct function of glottic integrity and can be used to measure the strength of bulbar muscles.

**Respiratory Muscle Strength**
MIP and MEP are measured while a patient inhaled or exhaled maximally against a closed shutter. They are the most sensitive indicators of decreased respiratory muscle strength.

**RECOMMENDATIONS**
According to an American Thoracic Society consensus statement, “patients should visit a physician specializing in pediatric respiratory care twice yearly after confinement to a wheelchair, fall in vital capacity below 80% predicted, and/or age 12 years old.”

Children should have at least 1 visit with a physician who specializes in pediatric respiratory care early in the course of disease (4–6 years of age) and before confinement to a wheelchair to have baseline pulmonary function testing performed.

Individuals who require mechanically assisted airway-clearance therapy or mechanically assisted airway ventilation should be evaluated by a pulmonologist every 3 to 6 months or as indicated for routine follow-up.

Objective evaluation at each clinic visit should include oxyhemoglobin saturation by pulse oximetry, spirometric measurements of FVC, FEV1, and maximum midexpiratory flow rate, maximum inspiratory and expiratory pressures, and CPF rate.

Awake carbon dioxide should be evaluated at least annually in conjunction with spirometry. When available, capnography is ideal for this purpose. If capnography is not available, a venous or capillary blood sample should be obtained to assess for the presence of alveolar hypoventilation.

Additional measures of pulmonary function and gas exchange may be useful, including lung volumes, assisted CPF, and MIC.

Annual laboratory studies for patients who require a wheelchair for ambulation should include a complete blood count, serum bicarbonate concentration, and chest radiography.

**FUTURE POSSIBILITIES**
Because measurement of static pressure at mouth during a maximum effort requires aptitude, cooperation, motiva-
tion, and coordination, a low value for MIP or MEP could result because of poor performance and technique. Measurement of pressure developed during maximal sniff is easy even in small children, and measurement of cough gastric pressure has been used to measure expiratory muscle strength. Similarly, magnetic stimulation of the phrenic nerve to quantify diaphragmatic strength has been used. A significant correlation has been found between FVC and transdiaphragmatic pressure during sniff.

We have used an impulse oscillometry system to measure airway obstruction and reversibility in patients with neuromuscular disorders who were unable to perform conventional spirometry as a result of muscle weakness.

REFERENCES
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