

PEDIATRICS®

OFFICIAL JOURNAL OF THE AMERICAN ACADEMY OF PEDIATRICS

Pulmonary Function Testing in Neuromuscular Disorders

Girish D. Sharma

Pediatrics 2009;123;S219-S221

DOI: 10.1542/peds.2008-2952D

The online version of this article, along with updated information and services, is located on the World Wide Web at:

http://www.pediatrics.org/cgi/content/full/123/Supplement_4/S219

PEDIATRICS is the official journal of the American Academy of Pediatrics. A monthly publication, it has been published continuously since 1948. PEDIATRICS is owned, published, and trademarked by the American Academy of Pediatrics, 141 Northwest Point Boulevard, Elk Grove Village, Illinois, 60007. Copyright © 2009 by the American Academy of Pediatrics. All rights reserved. Print ISSN: 0031-4005. Online ISSN: 1098-4275.

American Academy of Pediatrics

DEDICATED TO THE HEALTH OF ALL CHILDREN™



Pulmonary Function Testing in Neuromuscular Disorders

Girish D. Sharma, MD

Department of Pediatrics, Rush University Medical Center, Chicago, Illinois

The author has indicated he has no financial relationships relevant to this article to disclose.

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.

www.pediatrics.org/cgi/doi/10.1542/peds.2008-2952D

doi:10.1542/peds.2008-2952D

Abbreviations

TLC—total lung capacity
 RV—residual volume
 PFR—peak flow rate
 CPF—cough peak flow
 FVC—forced vital capacity
 FEV₁—forced expiratory volume in 1 second
 MIC—maximum insufflation capacity
 MEP—maximal expiratory pressure
 MIP—maximal inspiratory pressure
 FRC—functional residual capacity

Accepted for publication Jan 5, 2009

Address correspondence to Girish D. Sharma, MD, Rush University Medical Center, Department of Pediatrics, 1653 W Congress Pkwy, Suite 452 Pavilion, Chicago, IL 60612. E-mail: girish_d.sharma@rush.edu.

PEDIATRICS (ISSN Numbers: Print, 0031-4005; Online, 1098-4275). Copyright © 2009 by the American Academy of Pediatrics

The figure for this article appears online at: www.pediatrics.org/content/vol123/Supplement_4

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.⁵ 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.

MIC⁶ 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 inhales or exhales 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.”¹

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.⁷ 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.⁸

REFERENCES

1. Finder JD, Birnkrant D, Carl J, et al. Respiratory care of the patients with Duchenne muscular dystrophy: ATS consensus statement. *Am J Respir Crit Care Med.* 2004;170(4):456–465
2. Wang CH, Finkel RS, Bertini ES, et al. Consensus statement for standard of care in spinal muscular atrophy. *J Child Neurol.* 2007;22(8):1027–1049
3. Gozal D. Pulmonary manifestations of neuromuscular disease with special reference to Duchenne muscular dystrophy and spinal muscular atrophy. *Pediatr Pulmonol.* 2000;29(2):141–150
4. Papastamelos C, Panitch HB, Allen JL. Chest wall compliance in infants and children with neuromuscular disease. *Am J Respir Crit Care Med.* 1996;154(4 pt 1):1045–1048
5. Bach JR. Mechanical insufflation-exsufflation: comparison of peak expiratory flows with manually assisted and unassisted coughing techniques. *Chest.* 1993;104(5):1553–1562
6. Kang S, Bach JR. Maximum insufflation capacity. *Chest.* 2000;118(1):61–65
7. Nicot F, Hart N, Forin V, et al. Respiratory muscle testing: a valuable tool for children with neuromuscular disorders. *Am J Respir Crit Care Med.* 2006;174(1):67–74
8. Sharma GD, Cooper M, Arora H, et al. Forced oscillatory (IOS) assessment of pulmonary function and bronchodilator response in children with neuromuscular disorder [abstract]. *Am J Respir Crit Care Med.* 2003;167(7):A512

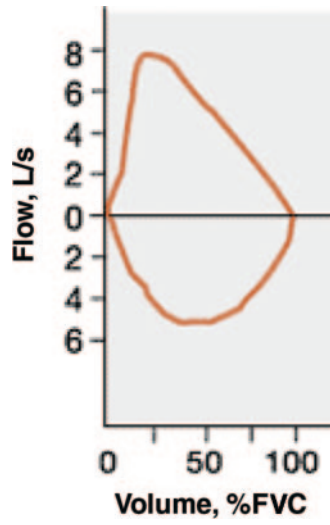


FIGURE 1
Flow-volume loop. FVC indicates forced vital capacity.

Pulmonary Function Testing in Neuromuscular Disorders

Girish D. Sharma

Pediatrics 2009;123;S219-S221

DOI: 10.1542/peds.2008-2952D

Updated Information & Services	including high-resolution figures, can be found at: http://www.pediatrics.org/cgi/content/full/123/Supplement_4/S219
References	This article cites 8 articles, 4 of which you can access for free at: http://www.pediatrics.org/cgi/content/full/123/Supplement_4/S219#BIBL
Subspecialty Collections	This article, along with others on similar topics, appears in the following collection(s): Respiratory Tract http://www.pediatrics.org/cgi/collection/respiratory_tract
Permissions & Licensing	Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at: http://www.pediatrics.org/misc/Permissions.shtml
Reprints	Information about ordering reprints can be found online: http://www.pediatrics.org/misc/reprints.shtml

American Academy of Pediatrics

DEDICATED TO THE HEALTH OF ALL CHILDREN™

