Pulmonary Function Testing in Children

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Pulmonary function tests (PFTs) are useful and important methods to evaluate children with or suspected of having lung disease. Although PFTs results do not establish a specific diagnosis, they provide helpful information for the diagnosis, classification, control, and management of respiratory illnesses in children. Therefore, measurement of lung function has become an essential component in the management of pulmonary diseases. In addition, the National Asthma Education and Prevention Program (NAEPP) and the Global Initiative for Asthma (GINA) recommend the use of PFTs in the assessment, management and long-term monitoring of patients with asthma.

As noted, PFTs do not make the diagnosis of a pulmonary disease, but they assist in the identification, assessment and management of:

• Obstructive lung diseases such as asthma, cystic fibrosis (CF), the consequences of chronic lung disease of prematurity (CLD)
• Restrictive lung diseases (chest wall abnormalities such as scoliosis or pectus excavatum; neuromuscular diseases [Duchenne muscular dystrophy]; pulmonary fibrosis; and obesity)
• Dynamic airway obstruction (vocal cord dysfunction or airway malacia)
• Fixed airway obstruction (neck tumor or mediastinal masses)
• Pulmonary hemorrhage

There are several indications for PFTs in children:
• To define the type and severity of the pulmonary physiologic abnormality
• To monitor the course of pulmonary diseases and lung function impairment
• To determine the effectiveness of therapy and guide treatment changes
• To follow pulmonary side effects of certain therapies (chemotherapy or radiation therapy)
• To assist in the preoperative planning of anesthesia and in anticipating the need for postoperative oxygen and/or assisted ventilation

At the same time, there are relative contraindications for PFTs in children:
• Young age or inability to follow directions
• Hemoptysis of unknown origin
• Recent pneumothorax
• Unstable cardiovascular status
• Thoracic, abdominal, cerebral aneurysm
• Recent eye surgery
• Nausea, vomiting, pain
• Recent thoracic or abdominal surgery

PFTs are patient effort- and technique-dependent. For example, short or incomplete exhalations, variable technique, suboptimal maximal effort will affect results by underestimating lung function. Therefore, they require maximal patient effort for a good quality test. In addition, they need to meet the American Thoracic Society (ATS) standards to obtain meaningful results. These standards are set as to what is an acceptable blow and for repeatability (how closely repeated efforts should match). Essentially, PFT results should be acceptable and repeatable. Most children can perform a valid spirometry study by 6 years of age.

PFTs measure specific parameters of lung function, namely inspiratory and expiratory flow rates and lung volumes. In some children, PFTs may detect abnormal changes in the lungs before they are appreciated on physical examination or chest X-ray. PFTs actually consist of several different or separate tests, each test examining lung function in a different way. PFTs consist of:

• Spirometry
  - Pre- and post-bronchodilator
  - Positional (upright versus supine)
• Lung volume (LV)
• Infant PFTs, which measure spirometry and lung volumes in infants
• Diffusion capacity (DLCO)
• Fractional exhaled nitric oxide (FeNO)
• Challenge studies:
  - Methacholine
  - Exercise
  - Cold air
• Exercise physiology with gas exchange

• Six-minute walk
• Resting metabolic rate
• Maximal respiratory pressures (PI Max and PE Max)
• Ventilatory drive study

Children’s Hospital of Wisconsin has three state-of-the art pulmonary function laboratories to perform this complex testing in children.

Categories of lung function

1. Spirometry

Spirometry measures inspiratory and expiratory flow rates and compares the child’s absolute values (in liters) with predicted values. The predicted values are based on height, gender, race and age using appropriate reference equations. In the end, there is a percentage of what is predicted for each child compared to other similar children.

Spirometry suggests the lung function abnormality is obstructive, restrictive or mixed. Obstructive disease is associated with a decreased FEV1, normal FVC, and decreased FEV1/FVC ratio, whereas restrictive disease is associated with a symmetrically decreased FEV1 and FVC with a normal ratio.
If a patient is diagnosed with an obstructive disorder, the reversibility of the obstruction should be assessed by measuring FEV1 after inhalation of a bronchodilator (for example albuterol). A post-bronchodilator increase in FEV1 of >12% predicted (or more than 25% predicted in FEF25-75%) and 200 mL (in children 12 years and older) would suggest a reversible obstructive lung defect, for example supporting the diagnosis of asthma. However, lack of response to a bronchodilator should not preclude the use of it, if clinically indicated.

It is also essential to assess flow volume loops. They provide very important information. They can help in the diagnosis of obstructive or restrictive defects; they are able to identify variable intrathoracic obstruction, variable extrathoracic obstruction (vocal cord dysfunction) or fixed obstruction (tumors compressing airways). Specifically, small airway obstruction as seen in asthma causes a concave appearance or “scooping” near residual volume.

2. Lung volumes (LV)

LV measure the volume of all air-containing spaces in the thorax using either plethysmography or gas dilution. Lung volumes are subdivided into fractions related to normal physiological function. Each subdivision is called a volume and a combination of two or more volumes is called a capacity.

The most common:
- Tidal volume (TV) — the volume of gas inhaled and exhaled with each breath
- Vital capacity (VC) — the maximum volume that can be exhaled after a maximal inspiration
- Total lung capacity (TLC) — the total amount of gas after a maximal inspiration
- Residual volume (RV) — the amount of gas left in the lungs after maximal exhalation
- Functional residual capacity (FRC) — the amount of gas left in the lungs at the end of exhalation

Measurements of lung volumes require a plethysmograph or “body box” and usually are only done in a dedicated PFT lab. They are particularly helpful confirming restrictive lung defects (decreased TLC), but they also are helpful assessing patients with obstructive lung defects (increased RV or FRC).

Clinical indications may include:
- Interstitial lung disease
- Pulmonary fibrosis
- Neuroumscular diseases (Duchenne muscular dystrophy)
- Chest wall pathology (scoliosis)
- Asthma
- Cystic fibrosis

3. Diffusion capacity for carbon monoxide (DLCO)

DLCO provides information about the rate at which oxygen is transferred from the lungs to the pulmonary circulation. It is a measure of impedance to gas flow across the alveolar capillary membrane. DLCO value varies directly with lung size, but exercise, hemoglobin and evenness of ventilation perfusion affect results as well. Smoking may decrease DLCO.

DCLO is increased in left-to-right shunts, hepato-renal syndrome, pulmonary arteriovenous malformations, intra-alveolar/pulmonary hemorrhage and polycythemia. DLCO is low in interstitial lung diseases, pulmonary fibrosis, collagen vascular diseases, hypersensitivity pneumonitis, histiocytosis X, drug-induced lung disease (amiodarone, methotrexate, bleomycin) and anemia. But it also can be reduced in congestive heart failure, alveolar proteinosis, bronchial obstruction, bronchiolitis obliterans, pulmonary vascular obstruction (pulmonary embolism) and pneumonectomy.

For the same reasons described in LV, DLCO is performed only in specialized centers.

4. Fractional exhaled nitric oxide (FeNO)

FeNO is a test that measures the nitric oxide (NO) production by lung epithelium. This NO production can be elevated in a patient with eosinophilic airway inflammation (asthma) and atopy. FeNO is mostly used in the diagnosis and management of asthma.

Patients with elevated FeNO values are likely to respond to inhaled corticosteroids (ICS). It is also helpful to assess adherence to ICS (poor compliance or ineffective technique may be present in patients with unchanged or increased FeNO values who have been prescribed ICS).

5. Infant pulmonary function test

A very common question is “how young can a child be to perform PFTs?” As noted, most children by the age of 6 years can perform spirometry, other tests at older ages. However, newer technologies enable clinicians to obtain this information in infants. Utilizing the raised volume rapid thoraco-abdominal compression technique (RVRTC), we are able to perform PFTs in infants who are least 5 kg and less than 36 inches tall. They also should be at least 57 weeks post-conceptual age. Given that this test requires sedation, expensive equipment and a high level of training by the respiratory therapists, it is limited to specialized centers.

6. Challenge studies

- Methacholine
- Exercise
- Cold air
Challenge studies are bronchoprovocation tests using different techniques that try to elicit airway reactivity in a child who is suspected to have asthma. The type of challenge study depends on the clinical history. For example, an exercise challenge test may be useful in the evaluation of adolescent with exercise-induced symptoms (exercise-induced asthma, exercise-induced vocal cord dysfunction). The child runs on a treadmill using a standardized protocol; a test is considered positive if there is a >10% decrease in FEV1 or a >12.5% decrease in PEFR. A methacholine challenge is used when the diagnosis of asthma is not clear, either based on history or normal spirometry at baseline. Administration of methacholine produces bronchoconstriction; a test is considered positive if there is a 20% decline in FEV1. A normal (negative) test makes the diagnosis of asthma less likely.

Conclusions:

- PFTs assist in the diagnosis of lung disease by describing and quantifying the impairment in function.
- They can also helpful to assess the response to treatment.
- PFTs consist of several different or separate tests – each test examines lung function in a different way.
- Providers should be familiar with the benefits and limitations of each test available.

References

To refer a patient
Visit chw.org/refer or call (414) 266-2460 or toll-free (800) 266-0366
To make an appointment
Call Central Scheduling at (414) 607-5280 or toll-free (877) 607-5280
For more information
chw.org/pulmonary

Specialty Spotlight
Fetal Concerns Center of Wisconsin

The Fetal Concerns Center of Wisconsin is a cooperative effort between Children’s Hospital of Wisconsin and Froedtert & The Medical College of Wisconsin. The center is made up of a multidisciplinary team of specialists who are experts in over 75 medical specialties, including:

- Maternal fetal medicine
- Fetal surgery
- Fetal cardiology
- Neonatology
- Fetal imaging
- Genetics

These uniquely trained specialists lead the Fetal Concerns Center, and many are ranked among the Best Doctors in America®. In addition to the core team of physicians, additional specialists are involved on a case-by-case basis. Our care team also includes fetal care coordinators and fetal heart coordinators who work closely with families to coordinate their care and to be a single point of contact throughout the entire process.