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Pulmonary Review

Early Onset Scoliosis: A Pulmonary Perspective

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Abstract

Early-onset scoliosis impairs lung function, often severely as it progresses. This review depicts current understanding of the changes in respiratory function resulting from early-onset scoliosis and how pulmonologists and their assessment tools affect clinical treatment decisions by spine surgeons.

Keywords: Early; Scoliosis; Pulmonary

Introduction

Early-onset scoliosis (EOS) can produce chronic respiratory disease that worsens as deformities of the spine and thorax progress [1]. Management of respiratory disease in children with EOS often begins before spine treatment starts and extends well beyond the final surgical procedure. Before referral to a specialty spine center, children with EOS may experience recurrent respiratory infections, failure to thrive, and nighttime hypoxemia. Ideally at the time of referral, all respiratory conditions that affect lung function, such as asthma, tonsil hypertrophy, and pneumonia, have been diagnosed and treated so that evaluation of pulmonary functions reflects primarily the impact of the spine and thoracic cage deformity.

Importantly, features based on 2-dimensional imaging of the spine do not correlate well with respiratory functional measures in children with EOS. For example, the Cobb angle correlates poorly with indices of breathing during sleep, vital capacity, and symmetry of left and right lung function by lung perfusion scan testing [2-4]. Both structural changes and respiratory functional changes evolve with time but may do so at different rates. The results of functional assessments of breathing complement the structural evaluation of the spine and thorax in deciding when and how best to intervene with surgical and nonsurgical options (eg, casting and bracing).

Respiratory Effects of EOS

Early-onset scoliosis produces restrictive lung disease by constraining and deforming intrathoracic space, increasing rigidity of the chest wall, and impairing respiratory muscle function (Fig. 1). Lung function depends on adequate lung volume and movement of both the thorax and the diaphragm. All of these features are compromised in children with EOS. Not all children with EOS have abnormal lung function. There is no specific degree of spine deformity that guarantees normal lung function, but thoracic curves of less than 30° are unlikely to produce symptoms in most children with EOS. Pulmonary studies of children specifically with mild early-onset spine and thoracic cage abnormalities have not been reported.

Spirometry is a pulmonary function test that measures forced vital capacity (FVC) in awake cooperative children who can follow directions and exert fully while breathing in and out with maximum force and velocity, usually starting after 5 years of age. Spirometry is noninvasive, cheap, and readily available, with established norms for children of different genders, ages, and ethnicities. Vital capacity is the most frequently used lung function to assess and monitor the consequences of restrictive spine and chest wall disease

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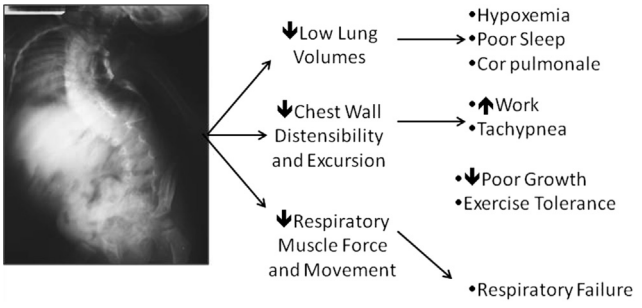

Fig. 1. Pathophysiology of early-onset scoliosis and clinical consequences.

because it is sensitive all 3 pathophysiologic processes of EOS: 1) reductions in lung volumes; 2) changes in chest wall stiffness; and 3) respiratory muscle weakness. Normal values are based on height, but in children with scoliosis and shortened height owing to spine curvature, height underestimates the real reduction in lung function measured by spirometry. Instead, arm span and/or ulnar length is used as a surrogate for height to predict normal lung function values [5,6]. In contrast to spirometry in the outpatient setting, lung volumes measured intraoperatively under anesthesia reflect passive lung inflation and deflation when respiratory muscle use is reduced or nonexistent [7]. Lung volumes can also be measured by computed tomography and 3-dimensional reconstruction techniques. However, these measurements are usually performed in the supine position at the end of tidal breathing and do not include dynamic breathing maneuvers [8]. Lung volume measures therefore depend on the circumstance under which they are measured and will not be the same across different measurement methodologies.

Spirometry also includes measures of airflow and hence airway obstruction. Airway obstruction occurs in up to 33% of children with EOS and does not improve with bronchodilator asthma therapy [9]. Bronchoscopy in these children often identifies a right or left mainstem bronchial narrowing owing to airway compression by the vertebra and mediastinal structures as a direct result of the spine deformity. The index to discern mainstem bronchial compression is the forced expiratory volume in 1 second (FEV1) divided by the FVC. This ratio should be greater than 80% to rule out obstruction, even when FVC is reduced by the restrictive skeletal process.

There has been concern that measures of FVC and FEV1 are too variable, which limits their clinical utility. The coefficient of variation for these values among normal children on a day-to-day basis averages 5% for FVC and 8% for FEV1. Changes greater than this degree of variability that can be ascribed to treatment are greater than 8% for FVC and greater than 15% for FEV1 [10]. Variability among children with lung diseases such as asthma and cystic fibrosis is greater than among normal children [10,11]. However, the major determinant of variability in spirometry is the experience of the child performing the test, and less so the child's age or disease [12]. First-time lung function testing, even with good technique, may overestimate the degree of pulmonary compromise resulting from EOS. Variability specifically among children with EOS performing spirometry has not yet been reported.

Low lung volumes owing to thoracic deformity predispose children with EOS to hypoxemia during sleep and during respiratory infections. Reduced resting lung volume leads to reduced airway caliber, which in turn leads to regions in the lung that ventilate poorly, producing hypoxemia. A study of hemoglobin levels in children with EOS reported that 23% of them had elevated levels (ie, mild polycythemia) reflecting intermittent hypoxemia [13]. Recurrent hypoxemia has been documented during sleep in up to 90% of children with EOS [4]. This occurs most often in rapid eye movement sleep, when muscle tone in the chest wall and upper airway normally falls. Breathing abnormalities during sleep among children with EOS, known as hypopnea, lead to poor sleep quality with frequent arousals, poor growth rates, recurrent pulmonary vasoconstriction, and when severe and/or prolonged, pulmonary hypertension [14]. Consequently, formal overnight studies of breathing studying all stages of sleep, including rapid eye movement sleep, are used increasingly to quantitate the frequency, duration, and severity of hypoxemia in children with EOS. These are most likely to be abnormal and require specific sleep treatments in children whose vital capacities are less than 40% of predicted norms [15].

Increased stiffness of the chest wall diminishes chest wall excursion and increases the work to breathe (Fig. 1). Children respond to this constraint by breathing shallowly and rapidly. Tachypnea with exercise precedes tachypnea at rest, but both are sensitive signs of restrictive chest wall disease. Increased respiratory work initially leads to exercise intolerance and, when severe, to reductions in daily activities such as climbing stairs or hills. Children adapt to increasing chest wall stiffness by transitioning from aerobic activities and sports to passive activities, such as computer games. Increased respiratory work also leads to increased caloric expenditure to breathe. How much this contributes to poor nutritional status is unclear. Children with EOS also tend to eat small frequent meals because larger meals exacerbate restrictive chest wall disease by restricting diaphragmatic motion. Reduced weight for age occurs in up to 50% of children with EOS on presentation [16]. Decreased chest wall movement owing to EOS also renders the accessory respiratory muscles less effective. Intercostal muscle contraction no longer results in chest wall excursion and children with EOS become increasingly reliant on their diaphragms to breathe.

Diaphragm function is also reduced in EOS, as manifested by reduced inspiratory force generation and diaphragmatic excursion [17,18]. Vital capacity is diminished in direct proportion to reduced respiratory muscle strength in children with EOS [18]. Normal diaphragmatic fibers are oriented cephalad to caudad, with muscle contraction