Chest Wall Diseases
Respiratory Pathophysiology

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KEYWORDS

• Chest wall • Respiratory failure • Kyphoscoliosis • Flail chest • Ankylosing spondylitis
• Hypoventilation

KEY POINTS

- Chest wall diseases produce restrictive pathophysiology with decreased lung volumes and reduced respiratory compliance, primarily owing to decreased distensibility of the chest wall.
- Patients with kyphoscoliosis may have significantly reduced chest wall compliance, whereas individuals with pectus excavatum or ankylosing spondylitis may have normal chest wall compliance.
- In obese individuals, the decreased respiratory compliance is largely due to a reduction in lung compliance.
- Respiratory failure may develop acutely as in flail chest or after a variable period, depending on the magnitude of the imposed elastic loads on the respiratory muscles.
- Sleep breathing abnormalities, age-related decreases in chest wall compliance, and associated lung dysfunction may also contribute to the development of respiratory failure.

INTRODUCTION

The chest wall is an essential part of the human ventilatory pump. It consists of the rib cage, abdomen, the spine and its joints, and the respiratory muscles with their nerves. The inspiratory muscles (diaphragm and intercostal inspiratory muscles) act to expand the chest wall and displace the abdomen outward. The increase in rib cage and the abdomen dimensions, alone or in combination, during inspiration are able to accommodate changes in lung volume. At all lung volumes, the elastic components of the lung promote passive inward recoil. Similar to the lung, the elastic components of the chest wall promote passive inward recoil at high lung volumes, 75% of vital capacity (VC) to total lung capacity (TLC). Unlike the lung, the elastic components of the chest wall promote passive outward recoil at low lung volumes, residual volume (RV) to 75% of the VC. Thus, at high lung volumes, chest wall expansion is due to contraction of the inspiratory muscles and not passive chest wall recoil.

In its normal state, the elastic components of the chest wall are easily stretched. Consequently, the chest wall has a relatively high compliance, equal to that of lung. Accordingly, the work of breathing is negligible when the respiratory muscles expand a healthy chest wall. However, the work of breathing may increase significantly when disorders stiffen the chest wall and decrease its compliance. These diverse disorders can affect the rib cage (kyphoscoliosis [KS], thoracoplasty, pectus excavatum, or flail chest), abdomen (obesity), spine (ankylosing spondylitis), and even skin (scleroderma, extensive burn scaring, massive subcutaneous emphysema). The common denominator

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is decreased chest wall compliance, leading to an increased work of breathing. In this review, we focus on the pathophysiologic characteristics and factors involved in the pathogenesis of respiratory failure in these disorders.

KYPHOSCOLIOSIS

KS refers to a group of spinal diseases characterized by excessive spinal curvature in the lateral plane (scoliosis) and sagittal plane (kyphosis) as well as by spinal axis rotation. It is categorized as idiopathic (affecting adolescents, mostly females) or secondary or paralytic (associated with neuromuscular disease).[1] The severity of the spinal deformity is assessed by measuring the Cobb angle on a radiograph of the spine.[2] This angle is formed by the intersection of 2 lines, one parallel to the top and the other parallel to the bottom vertebrae of the scoliotic or kyphotic curve (Fig. 1). The greater the Cobb angle, the more severe the deformity; angles of greater than 100° are associated with respiratory symptoms, and angles of greater than 120° with respiratory failure.[3–6]

Respiratory Mechanics

The pathophysiologic hallmark of KS is severe restrictive respiratory impairment related to reduced distensibility of the chest wall, especially when the Cobb angle is greater than 90°.[7–9] In this setting, TLC (Fig. 2) and VC may be reduced to 30% of predicted.[4,8,10]

Respiratory system compliance is reduced, primarily owing to a decrease in chest wall compliance and, to a lesser degree, a decrease in lung compliance resulting from microatelectasis.[11–13] The volume–pressure relationship of the respiratory system curve is shifted to the right, thus, requiring greater than normal pressures to inflate the lungs (Fig. 3). In addition, the straight portion of the pressure–volume curve where compliance is relatively constant in the normal individual, is diminished.[12] This truncated section of the curve is largely the result of a significantly decreased TLC. The stiff chest wall reduces the resting position of the respiratory system (functional residual capacity [FRC]) and tidal breathing occurs on a flatter portion of the respiratory system volume–pressure curve.[12] This factor leads to greater inspiratory effort for relatively small tidal breaths, which increases the work of breathing.[12] The increase in the oxygen cost of breathing may reach values 3 to 5 times greater than those measured in healthy individuals[7,9,14] and, thus, place patients at risk for respiratory muscle fatigue.[15]

The decrease in chest wall compliance promotes breathing with shallow tidal breaths, and the decrease in FRC leads to breathing at low lung volumes, both of which predispose to the development of atelectasis.[7,10,16,17] Although lung compliance may be diminished, it is not as severely decreased as the chest wall compliance.[11,14,18] Because chest wall compliance decreases with age,[19] respiratory mechanics invariably deteriorate with age, even in the absence of worsening of the spinal deformity. Changes in respiratory system compliance primarily reflect decreases in chest wall, not lung compliance. Cobb angles of up to 50° have a minimal effect on respiratory system compliance as opposed to those greater than 100° (Fig. 4), which may decrease compliance to levels seen in acute respiratory distress syndrome.[20]

Concurrent respiratory muscle weakness considerably increases the severity of restriction. In the paralytic type of KS, the degree of lung restriction is determined primarily by the degree of the respiratory muscle weakness rather than by the degree of the spinal curvature.[21,22] Generally in these patients, the resulting lung abnormalities lead to a greater loss in VC for a given degree of spinal deformity than seen in patients with the idiopathic form of KS.[21,22]

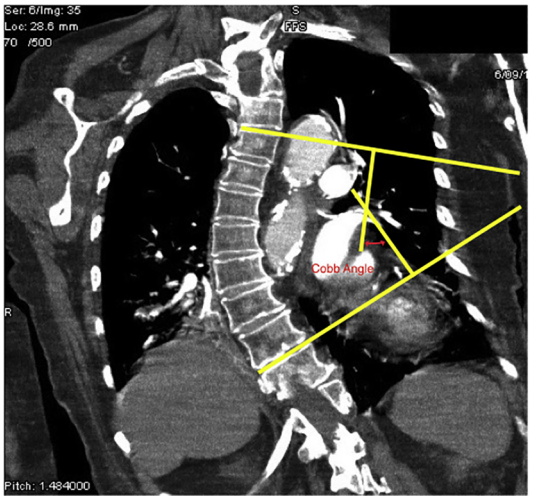

Fig. 1. Chest computed tomography scan (coronal view) of a patient with kyphoscoliosis. The angle formed from the 2 yellow lines drawn parallel to the vertebrae of the scoliotic curve is the Cobb angle.

Gas Exchange and Exercise Capacity

Hypoxemia is commonly found in patients with KS and correlates directly with VC and inversely with the angle of scoliosis. It is primarily due to ventilation–perfusion mismatch, and less often to