

Diseases of the Chest Wall and Respiratory Muscle Disorders: Diagnosis and Management

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The alteration of the lung volume, in the form of reduction of total lung capacity (TLC), is regarded to restrictive lung diseases; as a rule the diseases of the chest wall and muscles, neuromuscular apparatus stipulate it; also, there are parenchymal ailments that may adjust the respiratory function of the lungs. The former ones lead to the decrease of TLC and refer to extrapulmonary diseases, and vice versa, the latter ones cause elevated TLC, like in COPD and asthma.



Definition and Background

When the thoracic cage is affected by disease, lung volume is reduced. These changes in the anatomic structure of the chest can have severe consequences, including atelectasis, ventilation-perfusion mismatch, and hypoxia.

Chest wall and muscle diseases include the following:

- [Myasthenia gravis](#)
- [Amyotrophic lateral sclerosis](#)
- Myopathy, which affects neuromuscular structures

Other thoracic and extrathoracic factors that may be found in patients with reduced total lung capacity (TLC) (eg, in [obesity](#)) include [obesity hypoventilation syndrome](#), kyphoscoliosis, and ascites.

Etiology

There are several causative factors of extrapulmonary conditions associated with a reduction in TLC.

Spinal problems: Kyphoscoliosis, lateral curvature, anteroposterior angulation, kyphosis, or a combination of these conditions. The severity and advance of the disease are estimated by the scope of the Cobb angle, which is an angle made up of two limbs of a convex prime curvature of the spine. An angle larger than 100° indicates respiratory failure.

Neuromuscular disorders are responsible for the dysfunction of an integral part of the respiratory system, as the respiratory muscles enable the act of breathing.

The level at which the respiratory pump can be affected may vary, but can include the following:

- [Central nervous system](#)
- Peripheral nervous system
- Spinal cord
- Neuromuscular junction
- Respiratory muscle

Obesity is a major cause of restricted lung volume, as there is a direct relationship between body mass index and TLC and thus the development of obesity hypoventilation syndrome and obstructive sleep apnea. Hereditary factors also play a pivotal role (eg, myasthenia gravis).

Epidemiology

A mild form of kyphoscoliosis affects 1/1,000 persons in the United States, while 1/10,000 Americans experience serious deformities of the spine. Currently, there are between 36,000 and 60,000 patients with myasthenia gravis in the United States. Based on the Centers for Disease Control and Prevention data, over 30% of the American population has been diagnosed with varying degrees of obesity. All these factors are major contributors to TLC reduction.

There is **no gender or race predisposition to chest wall and muscle diseases**. Spinal problems leading to TLC decrease are seen more frequently in younger patients (< 35 years of age), while obesity hypoventilation syndrome tends to occur in patients over the age of 50.

Presentation

History

Patients with chest wall and muscle diseases experience a number of symptoms, including the following:

- Breathlessness
- Pain in the back and chest due to spinal deformities
- Muscle weakness
- Snoring, which disrupts sleeping patterns
- Severe fatigue

In addition, recurrent respiratory infections may occur in these patients as a result of the reduction in TLC and chronic hypoxia.

Physical Examination

Respiratory diseases are easily recognized on physical examination in patients with advanced stages of kyphoscoliosis and obesity. In cases of neuromuscular disease, patients may experience accessory muscle involvement, superficial breathing, and paradoxical or irregular breathing.

Auscultation may reveal **systolic murmur, wheezes, and crackles upon inspiration; percussion produces hyper-resonance.** The pathology of the spine may lead to obvious deformities of the back, chest, shoulders, and sternum. In addition, clubbing may be seen in these patients.

Serious complications stemming from the reduction of TLC may trigger hepatomegaly and ascites as well as peripheral edema.

Differential Diagnosis

- [Acute respiratory distress syndrome](#)
- [Chronic obstructive pulmonary disease \(COPD\)](#)
- [Acute bronchitis](#) and [chronic bronchitis](#)
- [Emphysema](#)
- [Eosinophilic pneumonia](#)
- [Lymphocytic interstitial pneumonia](#)
- [Hypersensitivity pneumonitis](#)
- Occupational diseases: [Coal worker's pneumoconiosis](#), [asbestosis](#)
- [Idiopathic pulmonary fibrosis](#)
- Interstitial (nonidiopathic) pulmonary fibrosis
- Pulmonary eosinophilia
- [Sarcoidosis](#)
- [Silicosis](#)
- Lung transplantation
- [Obesity](#)

Diagnosis

Diseases of the chest wall where the muscles are not involved include **kyphosis that is idiopathic or secondary.** Neuromuscular diseases such as polio or muscular dystrophy often cause secondary kyphoscoliosis. As well, fibrothorax, massive pleural effusion, massive obesity, ankylosing spondylitis, and thoracoplasty are other contributing factors to a reduction in TLC and respiratory distress.

Neuromuscular diseases such as weakness of the respiratory muscle may also emerge because of myopathy or myositis, quadriplegia, or phrenic neuropathy. These, coupled with infectious or metabolic factors, can lead to the development of respiratory

dysfunction and consequent failure.

Laboratory Studies

Biochemical blood examination reveals an elevated creatine kinase level that indicates myositis, leading to muscle weakness and restricted TLC.

Complete blood count indicating polycythemia and an elevated hematocrit level is seen in obese patients.

Thyroid Function

A decline in the function of the thyroid gland (hypothyroidism) contributes to the emergence of obesity and obesity hypoventilation syndrome.

Pulmonary function testing (PFT)

This test includes spirometry, measurements, estimation of lung volume, diffusing capacity, and arterial blood gas. PFT helps assess the efficacy of administered therapy.

Ventilatory failure in neuromuscular diseases is indicated by a fall in maximal inspiratory pressure below 30 cm of water; a decreased vital capacity and functional residual capacity, with preserved residual volume, are characteristic of chronic muscular diseases.

Arterial blood gas analysis reveals hypercapnia in OHS, which is defined as high PaCO₂ (> 45 mm Hg).

Electrodiagnostic

Repetitive nerve stimulation, stimulation of a muscle at 2–3 Hz, may reveal the competence of the muscular workout. Single-fiber electromyography estimates neuromuscular block, jitter, and fiber density of the muscles.

Management

The management of neuromuscular diseases aims to **minimize the influence of affected secretion clearance and equip the body to be more resistant to respiratory infections**. Noninvasive positive-pressure ventilation via a nasal or oronasal mask is effective in patients with respiratory failure and seriously affected gas exchange.

Permanent tracheostomy and ventilator assistance with a portable ventilator are useful in those patients where noninvasive positive-pressure ventilation has not been effective. Both methods smooth the symptoms of dyspnea and pulmonary hypertension and improve gas exchange, thus reducing these patients' hospitalization rate.

Weight loss also leads to a noticeable improvement in pulmonary function test findings. A polysomnographic study is required to study nocturnal hypoventilation or upper airway obstruction.

Mechanical ventilation (invasive ventilation with a tracheotomy tube) can be administered in advanced cases when there are copious secretions or there is an absence of control over upper airway functioning.

Pharmacotherapy

When TLC is reduced, medication is used in cases of obesity hypoventilation syndrome as

well as severe obesity. Medroxyprogesterone acetate leads to a decrease in PaCO₂ and an elevation in PaO₂; acetazolamide stimulates the decline of serum bicarbonate levels and reduces the PaCO₂ level; beta 2 agonists such as albuterol, metaproterenol, and atrovent affect beta 2 receptors in the bronchial smooth muscle, providing bronchospasm relief.

Medication administered in parenchymal disease associated with TLC reduction is not used in extra-pulmonary conditions, as the mechanism of the disease development and pathogenesis are completely different.

Surgical Intervention

Surgery may be performed in patients with pleural disorders associated with a restriction in TLC. **Decortication of the lung, or the removal of the restrictive surface layer, may be performed in cases of trapped lung and chronic empyema due to chest wall deformity. This procedure improves FVC and FEV1.**

Surgical approaches that help to relieve obesity hypoventilation syndrome in obese patients may be successfully applied to chest wall and muscle diseases:

- **Vertical banded gastroplasty:** Restricts stomach volume
- **Adjustable gastric banding**
- **Roux-en-Y gastric bypass:** The most common procedure. It provides short- and long-term effects regarding safety, efficiency, and durability; performed laparoscopically
- **Biliopancreatic diversion** and biliopancreatic diversion with duodenal switch leading to malabsorption

References

[The Epidemiology of Myasthenia Gravis](#) via medscape.com

[Change in prevalence of restrictive lung impairment in the U.S. population and associated risk factors: the National Health and Nutrition Examination Survey \(NHANES\) 1988-1994 and 2007-2010](#) via ncbi.nlm.nih.gov

[Restrictive Lung Disease Medication](#) via emedicine.medscape.com

[Respiratory involvement in inherited primary muscle conditions](#) via ncbi.nlm.nih.gov

[Neuromuscular and Other Diseases of the Chest Wall](#) via thoracickey.com

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