

Diseases of the Chest Wall and Respiratory Muscles Disorders — Diagnosis and Management

[See online here](#)

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 of Diseases of the Chest Wall and Respiratory Muscles Disorders

When the thoracic cage is affected the lung volume is reduced, these changes in the anatomic structure of the chest are fraught with severe consequences in the form of atelectasis, ventilation-perfusion mismatch, and hypoxia respectively.

Chest wall and muscles diseases encompass :

- [Myasthenia gravis](#)

- [ALS](#)
- Myopathy that affects neuromuscular structures.

Other thoracic and extrathoracic factors that may be found in the patients with reduced TLC, for example, in [obesity](#), there is [OHS \(obesity hypoventilation syndrome\)](#); kyphoscoliosis, and ascites.

Etiology of Diseases of the Chest Wall and Respiratory Muscles

There are several causative factors of extra-pulmonary conditions associated with the reduction of TLC:

Spine problems – kyphoscoliosis, lateral curvature, anteroposterior angulation, kyphosis or 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. If the figure is larger than 100°, it reflects respiratory failure.

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

The level at which the respiratory pump can be affected may vary from:

- [Central nervous system](#)
- Peripheral nervous system
- Spinal cord
- Neuromuscular junction
- Respiratory muscle
- Obesity is a major cause of the restricted lung volume as the correlation between BMI and TLC has direct dependence and stipulates the emergence of OHS and OSA.
- Hereditary factors play a pivotal role as well (Myasthenia Gravis).

Epidemiology of Diseases of the Chest Wall and Respiratory Muscles

Kyphoscoliosis as a mild form affects 1 per 1000 persons in the US, while 1 per 10,000 Americans is exposed to serious deformities of the spine. Nowadays, there are 53,000 and 59,900 patients with Myasthenia Gravis in the United States. Basing on the CDC data, about 35 % of the American population is diagnosed with different degree of obesity. All these factors are regarded as the major contributors to TLC reduction.

There is **no gender or race predisposition in the chest wall and muscle diseases**. Spinal problems leading to YLC decrease are more probable in the younger population – before 35 years old, while OHS tends to occur at the age of 50 years.

Presentation of Diseases of the Chest Wall and

Respiratory Muscles

History

First, the patients with the chest wall and muscle diseases complain about:

- Breathlessness
- Pain in the back and in the chest due to the spine deformities
- Weakness of the muscles
- Snoring growing into changing of sleeping pattern
- Severe fatigue exacerbating with an insignificant physical load

In addition, recurrent respiratory infections may occur in such patients as an after-effect of the reduction of TLC and chronic hypoxia.

Physical examination

This category of respiratory diseases is easily recognized owing to the specific habits of the patients, in particular, advanced stages of kyphoscoliosis and obesity. In cases of neuromuscular diseases, there might be the accessory muscles involvement, superficial breathing, paradoxical-irregular breathing.

Auscultation may reveal **systolic murmur, wheezes, and crackles upon inspiration; percussion produces hyper-resonance**. The pathology of the spine may lead to the obvious deformities of the back, chest, shoulders, and sternum. In addition, clubbing is the feature of severe hypoxia that may be found in this category of patients.

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

Differential Diagnosis of Diseases of the Chest Wall and Respiratory Muscles

- [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 of Diseases of the Chest Wall and Respiratory Muscles

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

Vice versa, neuromuscular diseases emerge like the weakness of the respiratory muscle owing to myopathy or myositis, quadriplegia, or phrenic neuropathy coupling with infectious or metabolic factors lead to the development of respiratory dysfunction and consequent failure.

Laboratory studies

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

CBC panpolycythemia, elevated hematocrit level in obese patients

Thyroid function studies

Decline in the function of the thyroid gland, hypothyroidism, causing the emergence of obesity and OHS consequently.

Pulmonary function testing (PFT)

This test includes spirometry, measurements, and estimation of lung volume, diffusing capacity, and arterial blood gas. PFT is informative in the assessment of the efficacy of the administered therapy.

Ventilatory failure in neuromuscular diseases is proved by the fall of maximal inspiratory pressure below 30 cm of water; a decreased vital capacity and FRC, with preserved RV, are characteristic of chronic muscular diseases.

Arterial blood gas analysis- OHS is represented by hypercapnia/ high PaCO₂ >45mmHg.

Electrodiagnostic Studies

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

Management of Diseases of the Chest Wall and Respiratory Muscles

The whole essence of the management of neuromuscular diseases predisposing TLC reduction is in the therapeutic prevention aimed to **minimize the influence of affected secretion clearance and fortifying the body general resistance 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 is useful in

those cases where noninvasive positive-pressure ventilation failed to assist. Both methods have a benign effect as they smooth the symptoms of dyspnea, pulmonary hypertension and improve gas exchange, thus reducing hospitalization rate.

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

Mechanical ventilation (invasive ventilation with a tracheotomy tube) administered in advanced cases of the chest wall and muscle diseases, when there are copious secretions, the absence of control over upper airway functioning.

Pharmacotherapy

In the TLC reduction conditions the medication that is used in OHS while massive obesity may be effective as well. Thus, Medroxyprogesterone acetate leads to a decrease in PaCO₂ and an elevation in PaO₂; Acetazolamide stipulates the decline of serum bicarbonate level and reducing PaCO₂ level; Beta2 Agonists: Albuterol, Metaproterenol, Atrovent -affect beta2 receptors in the bronchial smooth muscle, bronchospasm relief.

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

Surgical intervention

Surgery may be conducted in the case of a pleural disorder associated with the restriction. **Decortication may be administered regarding the trapped lung and chronic empyema due to chest wall deformity improving FVC and FEV1.**

Those surgical approaches that help to relieve the OHS in obese patients may be successfully applied to the chest wall and muscle diseases.

- **Vertical banded gastroplasty (VBG)** allows restricting the volume of the stomach.
- **Adjustable gastric banding (AGB)**
- **Roux-en-Y gastric bypass (RYGB)** is the most common procedure that provides short- and long-term after-effects regarding safety, efficiency, and durability; it is performed laparoscopically.
- **Biliopancreatic diversion (BPD)** and biliopancreatic diversion with duodenal switch (BPD-DS) 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

Legal Note: Unless otherwise stated, all rights reserved by Lecturio GmbH. For further legal regulations see our [legal information page](#).

Notes