Critically Ill Patients with Interstitial Lung Disease

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KEYWORDS

• Intensive care unit • Interstitial lung disease • Idiopathic pulmonary fibrosis

KEY POINTS

- Clinically significant interstitial lung disease (ILD) impairs pulmonary mechanics and gas exchange, which affects mechanical ventilatory strategies.
- Low tidal volume ventilation, to minimize volutrauma, in patients with ILD who require mechanical ventilation is recommended, although this has been extrapolated from data in acute respiratory distress syndrome (ARDS).
- Exacerbations of idiopathic pulmonary fibrosis (IPF), which lead to respiratory failure, are devastating with poor prognosis and can be incited by pulmonary surgery. Short-term survival may provide a window for lung transplantation.
- For patients with suspected, undiagnosed ILD in the intensive care unit, evaluation by an experienced pulmonologist, review of previous imaging, detailed medication and occupational history, and consideration of surgical lung biopsy are recommended.

INTRODUCTION

Interstitial lung disease (ILD) is a clinical syndrome of various etiologies and histopathologic categorization that when clinically significant, impair respiratory function. Progressive inflammation and scarring of the pulmonary parenchyma generate degradation of pulmonary mechanics and/or gas exchange. Eventual respiratory failure is the driving factor in mortality in some forms of ILD.[1] Often patients with ILD require intensive care for respiratory failure, surgical or medical procedures, or nonrespiratory organ failure. Underling impairment in pulmonary function necessitates alteration of ventilatory strategies. Knowledge of ILD comorbidities and exacerbation syndromes is requisite for the intensivist to provide accurate prognosis and potentially limit futile care.

EPIDEMIOLOGY

Variability in exposure to risk factors for ILD (smoking, occupation, and age) leads to regional variability in phenotypes and incidence of ILD.[2] Accurate identification and reporting bias significantly limit precise estimates of incidence and prevalence.[2] Compared with chronic obstructive lung disease ILD is uncommon, which can lead to lack of familiarity in centers not experienced with the care and diagnosis of ILD. In the United States the estimated prevalence of ILD is 81 and 67 per 100,000 in males and females, respectively, which is derived from a population-based study from New Mexico.[3] This is in stark contrast to the prevalence of chronic obstructive lung disease at 6700 per 100,000 (6.7%).[3,4] Autopsy reports from the same study indicate preclinical or

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undiagnosed ILD in 1.8% of patients.[3] Seven percent of patients in the Framingham Heart Study were found to have interstitial abnormalities on chest computed tomography (CT), suggesting a large spectrum of undiagnosed subclinical ILD.[5]

In total there are more than 200 types of ILD.[2] ILD is separated into lung diseases of known causes, such as from occupation (pneumoconiosis), drug exposure, and other systemic disease[3] (rheumatologic syndromes), versus those that are idiopathic interstitial pneumonias. A full description of all of the types of ILD is outside the scope of this article, although some key clinical findings can be found in **Table 1**.

UNDERLYING PHYSIOLOGY

Pulmonary function tests (PFTs) and oxygen requirement measurements performed before intensive care unit (ICU) admission, should they be available, are invaluable. Generally, patients with ILD have restrictive physiology, which is typified by decreased forced vital capacity, total lung capacity, and increased ratio of forced expiratory volume in 1 second to forced vital capacity.[6] These changes on PFTs correlate with poor pulmonary compliance during mechanical ventilation, which impacts mechanical ventilatory strategy by elevating airway pressures needed to provide adequate tidal volumes (TVs). The PFTs may be pseudonormalized if concomitant emphysema coexists.[6]

Gas exchange is impaired almost universally in patients with clinically significant ILD,[6] often requiring ambulatory supplemental oxygen. In the ambulatory setting, gas exchange is measured by the lung's ability to take up carbon monoxide relative to the underlying hemoglobin content (diffusion capacity of lung for carbon monoxide [D_{LCO}]). D_{LCO} may be decreased out of proportion to the pulmonary mechanics if comorbid conditions of continued tobacco abuse, emphysema, or pulmonary hypertension are present. Ambulatory measurements of arterial blood gases are useful to determine baseline level of hypoxemia and acid-base status.

RESPIRATORY FAILURE

Acute or acute on chronic respiratory failure is a common presentation of patients with ILD to the ICU. Usually, this is caused by hypoxemic respiratory failure. General strategies for the treatment of this include exclusion of potentially reversible causes, such as pneumothorax, infection, pulmonary embolus, and left or right heart failure.

Outcomes

Most outcome data reported are from patients with idiopathic pulmonary fibrosis (IPF), the most common and most deadly ILD. The prognosis for patients with IPF who require mechanical ventilation for nonoperative respiratory failure is extremely poor, although mortality is not absolute.[7–11] Mallick[12] recently reviewed all reported available data of patients with IPF who were mechanically ventilated and found a hospital mortality of 87% and a 3-month mortality of 94%. This has led some to consider mechanical ventilation in patients with IPF with nonoperative respiratory failure futile,[8,11,12] although any survival window may potentially allow for lung transplantation.[9] A specific syndrome causing respiratory failure in IPF, acute exacerbation, is discussed later in this article.

Data from the poor outcomes associated with patients with IPF who require mechanical ventilation are often incorrectly assigned to all patients with ILD.[13] However, regardless of the cause of the ILD, a patient with severely degraded pulmonary compliance and/or gas exchange at baseline who is admitted with respiratory failure is likely to do poorly unless the cause of the respiratory failure is easily and quickly reversible. Respiratory failure caused by progressive worsening of ILD despite appropriate treatment is, in most cases, futile unless pulmonary transplantation is possible. Mechanical ventilatory and other organ support may be needed to allow for the appropriate diagnostic evaluation to ensure no reversible causes of deterioration are found. Critical care support for patients with respiratory failure from end-stage fibrosis that is not treatable pharmacologically or surgically should not be continued.

VENTILATORY STRATEGIES

Noninvasive Ventilation

Given reduced baseline pulmonary function and potentially disastrous outcome from endotracheal intubation, avoidance of intubation to prevent complications, such as ventilator-associated pneumonia, intensive care myopathy or neuropathy, and catheter-associated infections, is desirable whenever possible. Small, retrospective studies have reported some success with noninvasive positive pressure ventilation (NIV) with improved mortality compared with invasive mechanical ventilation (IMV) with survival in responders to NIV of around 40%.[14–17] As with most data in ILD, these studies are predominantly done with patients with IPF, because it is the most common ILD. Additionally, a selection bias for less