Abstract

Chronic fibrotic interstitial lung disease (ILD) forms a substantial proportion of disabling lung diseases and leads to significant morbidity and mortality. The mortality of these patients when admitted to the Intensive Care Unit with acute respiratory worsening requiring mechanical ventilation can reach up to 90%. Indian law does not allow the physician to make the final decision about mechanical ventilation, we are forced to follow the wishes of the family despite knowing the extremely poor outcome of aggressive intervention and invasive ventilation. Patients more often become ventilator dependent and do not gain much regarding the quality of life with mechanical ventilation. Hence, there is a desperate need for palliative support for these patients with advance care planning to reduce the suffering of these patients toward the end of life. The article describes various methods by which the decision making process of mechanical ventilation could be made simpler and acceptable to the patient and the families of fibrotic Interstitial lung disease patients and also the dilemma faced by chest physician in India with virtually no prior end of life planning and no clear guidelines on ventilation when it comes to palliation of patients with advanced ILD.

Keywords: Interstitial lung disease, invasive ventilation, palliative

Introduction

Chronic interstitial lung disease (ILD) forms a substantial proportion of disabling chronic lung diseases and leads to significant morbidity and mortality. There is a desperate need for palliative support for this group of the patient as survival is sometimes similar to that of advance malignancy. These patients have very poor survival when admitted to the Intensive Care Unit (ICU) with acute respiratory worsening requiring mechanical ventilation. Similarly, the very small fraction of patients who do survive the ICU stay would require prolong rehabilitation and could be left with significant morbidity in the form of oxygen dependency, poor mobility, and tracheostomy. Therefore, decision-making process for ventilating or palliating without invasive ventilation ILD patients is very crucial and equally difficult as well.

The Grim Reality

The decision-making process becomes even more complicated in Indian Scenario for variety of reasons as follows: (1) most of the ILD patients in India would have consulted number of chest physicians and may not have a single chest physician who would have had a long-term follow-up and a professional relationship with the patient. Similarly, it is highly unlikely that the palliative care team is involved in such cases, (2) the patient might not be admitted to the hospital of their usual care and not under the care of their usual chest physician, (3) in case the patient is unable to make the decision, there are usually too many family stakeholders involved in the decision-making process (4) Indian law does not allow the treating physician to make the final decision unlike in many Western countries and we are forced to follow the wishes of the families despite knowing the extremely poor outcome of aggressive intervention and (5) there is a considerable cost involved in managing such critically ill patients which can become a limiting factor in due course of time.

The process of this crucial decision-making actually starts in the clinic during the routine consultation of an ILD patient. It is important to emphasize the family on proper recordkeeping of the lung function results, baseline

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Therefore, deterioration is identified in IPF patients.\[9\] In ventilated patients it was 68%. However, 92% with IPF admitted to the ICU the hospital mortality rate was higher than the other types of ILD. In a series of 38 patients ILDs and although statistically insignificant, the mortality in Acute Physiology and Chronic Health Evaluation (APACHE) rate reaching up to 70%. Fernandez- perez\[4\] reported the much worse outcome in nonsurgical patients with a mortality of daily living. Fernandez- perez et al. reported 44% of survival in cases of acute worsening in ILD but nearly 45% of the enrolled patients were postoperative cases and he reported the much worse outcome in nonsurgical patients with a mortality rate reaching up to 70%.\[10\] Other determinants of poor outcome were higher degree of pulmonary hypertension and higher Acute Physiology and Chronic Health Evaluation (APACHE) III scores. This study had a good mixture of various types of ILDs and although statistically insignificant, the mortality in idiopathic pulmonary fibrosis (IPF) patient’s category was higher than the other types of ILD. In a series of 38 patients with IPF admitted to the ICU the hospital mortality rate was 61% and in ventilated patients it was 68%. However, 92% of the survivors died in a median span of 2 months after the hospital discharge.\[15\] The mortality increases further to over 90% when there is no underlying cause for acute deterioration is identified in IPF patients.\[6,7\] A study in India showed that honeycombing, precapillary pulmonary hypertension, and traction bronchiectasis were independently associated with increased ICU mortality.\[10\] Therefore, the current literature suggest that diagnosis of IPF, acute exacerbation of unknown etiology, pulmonary hypertension, and higher APACHE scoring will help to identify the large subset of patients who might not get benefited with mechanical ventilation from the ones who are more likely to survive.

The next and the most important step is to have discussion with the patient and family. If the patient has capacity and willingness to be a part of the discussion, then it is worth doing the discussion in front of the patient. However, in most instances, the patient might be too ill to make a decision. In that case, it is important to know the relationship of each of the family member present during the consultation and if they could bring forward one person among them to lead their views and questions. The patient and family should be informed at length about the possibility of prolong mechanical ventilation and the poor outcome, tracheostomy need, and risk of intercurrent infections if the patient is treated with mechanical ventilation. Furthermore, they might not be able to achieve their background mobility and may be left with morbidity such as oxygen dependence, tracheostomy, and that there is high post-discharge mortality.

It is a worthwhile exercise if the patient does have a known chest physician or general practitioner who has been following up the patient elsewhere then to involve him/her in the decision-making process. It is more likely that the patient and family will be able to make decision with lot more confidence if the primary chest physician is involved in the process and we could avoid overzealous mechanical ventilation.

The Eternal Dilemma

At times, the family may be unable to make the decision straightaway or the close family members may not be available. In these scenarios, we could possibly buy some time by keeping the patient on high-flow nasal cannula or noninvasive ventilation (NIV) until we have the decision. Gungor have shown in their paper that there are small proportion of patients who could benefit from NIV so long as they have low APACHE II score <20 and their NIV requirement is noncontinuous.\[11\] Another study has shown benefit in oxygenation status, imaging films, and reports which could help during an emergency to make a decision on escalation of care. It is imperative that the goals of the treatment are clearly discussed with the patient and the family, especially that the lung which is already completely scarred will never be revived back with the treatment and the current treatment is only to slow down the process of ILD and reduce symptoms. Disease-related education and involvement of either a nurse specialist or palliative care team has helped bringing down any overzealous nonrewarding treatment to terminally ill patients.\[1,2,3\] The need for educating the ILD patient about their disease and prognosis is demonstrated in multiple studies from the developed world.\[6,7\] With the growing level of literacy in India, patients might be interested in knowing what future holds for them and they might want to plan things accordingly. Furthermore, it is logical to think that the prognostic information is likely to be absorbed more easily in the clinic when discussed on several occasions rather than in a short period in the emergency department. Advance care planning is not a routine practice in India and physicians may hesitate to initiate such a discussion due to the fear of wrong perception by the family and patient. However, if such a discussion or questions related to prognosis are initiated by the ILD patient, then at least this opportunity should be taken up to discuss the treatment options and likely outcome during emergency and their view should be recorded in the notes. Some written information in the form of leaflet could be a good idea to enhance the patient education and could save the time for physician as well.

The Zero Hour

Once the patient has landed with acute worsening of ILD needing intensive care support, the first task is to risk stratify the patient. It is essential that every effort is made to gather maximum information about the patient from their previous records, investigation reports and to get information from the family about the recent functional status, exercise tolerance, oxygen use of the patient, and any dependency for the activity of daily living. Fernandez- perez et al. reported 44% of survival in cases of acute worsening in ILD but nearly 45% of the enrolled patients were postoperative cases and he reported the much worse outcome in nonsurgical patients with a mortality rate reaching up to 70%.\[10\] Other determinants of poor outcome were higher degree of pulmonary hypertension and higher Acute Physiology and Chronic Health Evaluation (APACHE) III scores. This study had a good mixture of various types of ILDs and although statistically insignificant, the mortality in idiopathic pulmonary fibrosis (IPF) patient’s category was higher than the other types of ILD. In a series of 38 patients with IPF admitted to the ICU the hospital mortality rate was 61% and in ventilated patients it was 68%. However, 92% of the survivors died in a median span of 2 months after the hospital discharge.\[15\] The mortality increases further to over 90% when there is no underlying cause for acute deterioration is identified in IPF patients.\[6,7\] Al Hammed reported 24 deaths out of 25 patients ventilated for unknown cause for IPF with acute respiratory failure. Gaudry et al. have recently reported a series of 27 patients with fibrosing ILD where the survival was slightly better as four patients managed to survive out of the hospital and two then subsequently received lung transplantation.\[8\] Lung transplantation still appears to be in very early stage in India. A large case series of 220 patients with IPF and nonIPF fibrotic ILD following acute respiratory worsening showed that patients with exacerbation as the underlying cause for respiratory failure have higher in-hospital mortality rate than the ones with infection or subacute progression.\[9\] Zafrani et al. showed that honeycombing, precapillary pulmonary hypertension, and traction bronchiectasis were independently associated with increased ICU mortality.\[10\] Therefore, the current literature suggest that diagnosis of IPF, acute exacerbation of unknown etiology, pulmonary hypertension, and higher APACHE scoring will help to identify the large subset of patients who might not get benefited with mechanical ventilation from the ones who are more likely to survive.
Although NIV is a useful bridge to buy time and avoid intubation, there is a growing problem related to NIV use in ICU from palliation point of view. A significant number of these patients eventually become NIV dependent. NIV is an uncomfortable treatment for the patient as it causes claustrophobia and can lead to nasal bridge ulceration which is extremely painful. There are constant issues related to feeding the patients and maintaining their oral hygiene as removing NIV for a short period for feeding can lead to rapid desaturation. Aerophagia related to NIV leads to abdominal bloating.

Although logically noninvasive and invasive ventilation should be looked at in similar ways when it comes to withdrawal as both are life-sustaining interventions, there are some differences. The withdrawal from NIV may not lead to immediate death as patients are breathing on their own most of the time. Therefore, withdrawal from NIV may not be as dramatic if the decision for nonescalation is taken before the patient becomes completely NIV dependent and oxygen requirement becomes too high. This would also allow the patient to be cared at home toward the end of their life if the family wishes so. The most binding rule in the medical practice is to follow the patient’s own wishes if the patient has the mental capacity to understand and give an informed consent; therefore, we are still allowed to withdraw the NIV if the patient gives the informed consent for the same. In the absence of the consent from patient, as of now, it does not seem to be clear whether nonrewarding NIV causing distress to the patient can be legally withdrawn and switched to oxygen alone. Most of the patients with ILD exacerbation remain in single-organ failure for a long time until they end up in superadded sepsis. They can very well be completely awake and in full senses, especially when they are on NIV. Therefore, it becomes very difficult for the family to take the decision for nonescalation. The American Thoracic Society definition of futility is “a life-sustaining intervention is futile if reasoning and experience indicate that the intervention would be highly unlikely to result in a meaningful survival for that patient.” If we go by this definition, then most of the ILD patients who are dependent completely on NIV should be offered withdrawal from NIV if the treatment is distressful and invasive ventilation is ruled out.

Once the patient is on invasive ventilation for few days and continues to show no improvement, the family may come back asking for withdrawal of the ventilator. Most places in the world would allow either withholding or withdrawing treatment following consent from the patient or next of kin and would not label this as killing the patient but allowing them to die as per their own wishes. Since Indian law has no clear stand on end-of-life issues except that suicide and abetment to suicide are punishable offenses withdrawal even with the expressed consent of the patient or next of kin can be misinterpreted post hoc. The physician and the family might then agree on having a joint decision for not escalating the organ support further which would be helpful in palliation of these patients.

To summarize

There is a lack of adequate palliative care support for end-stage lung disease, especially fibrotic lung diseases. Advance ILD patients should receive counseling in the chest outpatient department about the gravity of their disease and if possible advance care planning should be initiated and involvement of palliative care team would be beneficial. The prognosis of fibrotic ILD on invasive ventilation is extremely poor with worldwide mortality reaching up to 90%, particularly in patients with IPF and acute exacerbation with no obvious cause found when encountering critically ill ILD patient, a detailed discussion on outcome with a next of kin as well as discussion with the usual chest physician or general practitioner could be useful to avoid an overzealous treatment with mechanical ventilation. NIV could act as a bridge during this period and should have a written escalation plan in the notes with details of all discussions before the patient becomes completely NIV dependent. Acute palliative care services should be utilized while caring for end-stage lung disease patients. Clearly, we need more Indian data on the patient and family’s perceptions about determining ceiling of care and how best this can be achieved. We certainly need some refinement from Indian Law about withdrawal from noninvasive and invasive ventilation.

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Conflicts of interest
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