Managing end stage disease in ILDs

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AIMS

• To define the end-stages of ILDs
• To define palliative care
• To outline the principals of end-stage ILDs care
• To state the components of complex care and the care providers in end-stage ILDs

SUMMARY

Interstitial lung diseases (ILDs), can be fatal diseases due to the devastation of the lung parenchyma and its replacement by fibrous tissue. The end-stage of ILDs might be thus defined as the terminal phase of the disease when improvement cannot be expected either spontaneously or through any treatment and the disease course leads inevitably to death. ILDs, which mainly lead to end-stage and death, are mainly of fibroproliferative nature, either primarily, or secondary (idiopathic pulmonary fibrosis (IPF), chronic hypersensitivity pneumonitis (HP), sarcoidosis stage IV, with extensive fibrotic involvement, or ILDs associated with connective tissue diseases (CTD- ILDs).

We must realize that the end-stage of ILDs does not mean an end to patient care, on the contrary it is the beginning of a new type of care. First we must state that all therapeutic options for stopping the disease course and prolonging the patient´s life have been drawn. The patient and his/her closest relatives should be tactfully and empathetically informed that therapy will be withdrawn and the patient should be informed about future care, which is based on the principals of the best palliative care.

We must realize that palliative care will need to be offered to most of our IPF patients and a substantial percentage of patients with other fibroproliferative ILDs (chronic HP, CTD-ILDs) at some point in during the course of the disease. In some cases it will be part of the care plan from the beginning. When we look at Figure 1, we can clearly see that only ½ of the patients with IPF in the Czech Republic are treated with antifibrotic treatment, the rest of these patients receive only palliative care.
Figure 1. Patients with IPF who met the criteria for antifibrotic treatment in the Czech Republic in 2015.

- **Without antifibrotic treatment (N = 104)**
- **With antifibrotic treatment (N = 104)**
- **Not known (N = 1)**

(Chart showing the distribution of patients with IPF in the Czech Republic in 2015, with nearly equal numbers for those with and without antifibrotic treatment, and a very small number for those not known.)
From the Figure 2, you can recognize that IPF leads to death in all patients irrespective of the timeliness of the diagnosis and treatment.

Figure 2. Long-term mortality of IPF patients in the Czech Republic based on the timeliness of diagnosis.
The comprehensive care for IPF patients consists of 3 pillars:

- Disease-centered management
  - Pharmacological approaches
  - Nonpharmacological approaches
- Symptom-centered management
- Education and self-management

Provider-patient partnership enabling patients to:

- Set realistic goals
- Remain in control of her/his care
- Prepare for the future

Palliative care should be an integral part and routine component of the care of all patients with IPF[1].

**How is palliative care defined by WHO?**

Palliative care is an approach that improves the quality of life of patients and their families facing the problems associated with life-threatening illness, through the prevention and relief of suffering by means of early identification and impeccable assessment and treatment of pain and other physical, psychosocial, and spiritual problems. This definition of palliative care was prepared for cancer patients, nevertheless it is well suited for end-stage ILDs [2].

**Palliative care goals set by WHO:**

- provide relief from pain and other distressing symptoms
- affirms life and regards dying as a normal process
- intends neither to hasten or postpone death
- integrates the psychological and spiritual aspects of patient care
- offers a support system to help:
  - patients live as actively as possible until death
  - the family cope during the patient’s illness and in their own bereavement
- uses a team approach to address the needs of patients and their families, including bereavement counselling
- enhances quality of life, and perhaps also positively influences the course of the illness
- is applicable early in the course of illness, in conjunction with other therapies that are intended to prolong life
The end-stages of ILDs pose many problems for patients that are difficult to cope with despite help from care-providers and family members. Examples of these problems are presented in the list below [3]:

- Impaired QoL
- Dyspnea
- Coughing
- Medication side effects
- Impaired quality of sleep
- Fatigue and exhaustion affecting daily activities
- Necessity to plan ahead
- Employment becomes impossible
- Cost of medical care
- Decreased libido and inability to engage in sexual activity
- Reduced social activity
- Fear of death

The needs IPF patients are well described and addressed in several studies and also stated in European IPF patients charter, which is utilized by healthcare policy makers [4,5]. Some studies have investigated the symptoms and challenges, not only in the patients, but also in the family members who cared for them. It is not surprising that caregivers also suffered from many problems, including fatigue, stress, poor sleep, worry, and guilt [6].

Real world palliative care for IPF patients was described in a study by the ILD Center in London (by Bajwah) and in a paper by Duck (2015), both of which described nursing care for patients on pirfenidone [7,8].

Physicians dealing with end-stage ILDs patients must accept that the focus of their care and concern should be directed towards the patient and his/her symptoms not the disease itself. That means that physicians should, at every patient visit, evaluate all symptoms and try to alleviate them as much as possible. To this end, physicians have pharmacological and non-pharmacological treatment as well as social and psychological support [9,1].

Some symptoms can be reduced or eliminated by means of pharmacological treatment (breathlessness, cough, pain, depression, anxiety, comorbidities), while other symptoms and complaints require non-pharmacological treatments and management [10].
Nonpharmacological treatment options

- **Rehabilitation**
  - Standard of care for chronic lung diseases
  - Effectiveness in alleviating symptoms
  - Reducing the duration of hospital stays
  - Increasing exercise tolerance
  - Maximizing functional ability
  - In IPF, statistically significant improvement in 6MWT and dyspnea

- **Oxygen therapy**
  - For patients showing daytime hypoxemia, either resting or exercise-induced
  - Helps to improve also nocturnal hypoxemia
  - Improves overall QoL
  - Can be used during exercise training

- **Transplantation**

Specific solutions to concrete symptoms and the recommended steps to solve them [1,11]:

**Dyspnea**

- Regularly assess for and educate patients about dyspnea
- Refer patients for pulmonary rehabilitation and provide oxygen therapy if hypoxemic
- Evaluate for co-morbidities that can contribute to dyspnea (e.g. pulmonary hypertension, sleep-disorders that affect breathing, muscle weakness, psychosocial factors, and increased weight).
- Consider sildenafil and opioid therapy

**Cough**

- Regularly assess for and educate patients about cough
- Evaluate for co-morbidities that can contribute to cough (e.g. GERD, upper airway cough syndrome, asthma)
- Consider antitussives (e.g. benzonatate, codeine)
- In severe cases, consider oral corticosteroids
Fatigue and Deconditioning

- Regularly assess for and educate patients about fatigue and deconditioning.
- Refer patients for pulmonary rehabilitation and provide oxygen therapy if hypoxemic
- Evaluate for co-morbidities that can contribute to fatigue and deconditioning (e.g. obesity, sleep disorders that breathing, hypoxemia)

Depression and Anxiety

- Regularly assess for and educate patients about depression and anxiety
- Refer patients for psychosocial support through counseling or patient support groups
- Evaluate for co-morbidities that can contribute to depression and anxiety (e.g. dyspnea, fatigue and deconditioning, polypharmacy)
- Consider pulmonary rehabilitation
- Consider pharmacological therapy in patients unresponsive to conservative therapy.

Psychological and social support during the inevitable end-stages of ILDs is very helpful. This included not only professional psychological and social interventions, but also interaction with and use of patient advocacy groups and organizations with their websites and e-mail contacts. The final stages of ILD are incredibly difficult times for both the patient and care-givers. Care-providers are critical in helping patients cope with these times and with such difficult decisions as DNR directives if the patient’s condition begins to steadily worsen [12].

Conclusions and take home message

- The complexities of end-stage ILDs will ultimately become a concern for all patients at some point after the diagnosis
- Palliative care should be a part of a complete treatment plan from the very beginning
- Remember that the challenges induced by end-stage ILD concerns the patient’s entire family
- Treatment should include the use all modalities available - pharmacological and nonpharmacological treatment, psychological, and social support
- Be honest, but try not to diminish the patient’s hope
REFERENCES

   *This is very instructive paper with complex view on care for IPF patients, including concrete advices*

   *This paper shows the main problems which influence overall quality of life in IPF patients*

   *Real-world very practical study on the needs of IPF patients and their carers*

   *Important document showing for the first time the IPF patients rights and needs- passed already to helathcare policymakers*

   *Noteworthy abstract showing suffering not only of IPF patients but also of their carers*

   *Abstract dealing with specialised palliative care in advanced ILDs- experience from Royal Brompton Hospital*

   *Deals with nursing care for the IPF patients on pifrenidone*

   *Description of supportive care for IPF patients- its tools and position in the disease management*

   *Comprehensive review of nonpharmacological management of patients with IPF*

    *Describes management of chronic cough in patients with ILDs*

    *Dealing with end-of-life decisions and terminal phases of life in patients with IPF*
EVALUATION

1. What does the term end-stage ILD mean?
   a. Terminal phase of ILDs where improvement cannot be expected either spontaneously or as a result of any treatment and the disease course leads inevitably to death
   b. Terminal phase of ILDs, it does not lead inevitably to death, sometimes treatment can be converted to active and causal and the life of the patient can be prolonged

Answer A

2. What are the means of palliative care for end-stage ILDs patients?
   a. Pharmacological treatment, non-pharmacological treatment, and psychologic and social support
   b. Special pharmacological treatment targeted to alleviate the symptoms of the disease
   c. Only nonpharmacological treatment, i.e. rehabilitation, and psychologic and social support

Answer A

3. Who should be involved in complex palliative care for patients?
   a. Specialized nurses and social workers
   b. Home care services
   c. Physicians (ILD- experts, general practitioners), nurses (optimally specialized), family members, psychologists, social workers, physiotherapists, and patients themselves
   d. ILD- experts and general practitioners

Answer C

4. What symptoms should physicians regularly evaluate?
   a. Dyspnea, cough, and pain
   b. The physicians should not bother patients by asking about symptoms, the patient will tell about them spontaneously if they appear
   c. Dyspnea, cough, fatigue and deconditioning, sleep disorders, depression and anxiety

Answer C

5. When and in whom should palliative care be started?
   a. Palliative care should be offered only to ILDs patients in the end-stage of the disease
   b. All patients with ILDs should be offered palliative care from the beginning
   c. Palliative care should be part of the care for all ILDs patients, focused on alleviating the symptoms of the disease or treatment of adverse events

Answer C