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Advanced chronic lung disease: need for an active interdisciplinary approach

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Summary

Modern treatment of patients with advanced lung disease includes not only pulmonary therapy but also integrative treatment of co-morbidities directly or indirectly linked to the underlying lung disease. Moreover, impairment of heath related quality of life is frequent in these patients and influences their physical and psychological "well-being". Therefore, a multidisciplinary team approach gives the opportunity to appropriately deal with all these issues. It has been shown that treating patients with advanced lung disease in specialised centres has a positive impact on survival, especially for patients with CF and IPF. Referral to the intensive care unit is not generally contraindicated and mortality depends on the underlying lung disease and co-morbidities. Nowadays, lung transplantation is a valuable therapeutic option in advanced lung disease and should be considered early especially in IPF patients.

Key words: advanced lung diseases; lung transplantation; cystic fibrosis; pulmonary fibrosis; multidisciplinary team; co-morbidities

Introduction

"End-stage lung disease" refers to any chronic lung disorder that limits daily living and impairs health-related quality of life. In general, it leads to progressive deterioration of lung function by objective measurements and usually premature mortality. There are five major disease categories of end-stage lung disease: 1) obstructive lung diseases, 2) restrictive lung diseases, 3) pulmonary vascular diseases, 4) neuromuscular diseases and 5) carcinoma (representing the largest group) (table 1). The other term often used in the literature is "advanced lung disease". This term is more restricted to chronic, non-neoplastic lung diseases that also lead to irreversible pulmonary deterioration. Reduced functional reserves of the lung are the common features of advanced lung disease causing dyspnoea and exhaustion during daily activity. Often, these restrictions in daily life lead to depression and anxiety and finally they may compromise heath-related quality of life. Internationally, the number of centres specialised in treating advanced lung diseases in an interdisciplinary set-

Table 1

End-stage lung diseases.

Obstructive lung diseases
Chronic obstructive pulmonary disease (COPD) due to smoking
Chronic obstructive pulmonary disease (COPD)due to alpha-1-AT-deficiency
Cystic fibrosis (CF)
Bronchiectasis
Post-bronchiolitis-syndrome
...
Restrictive lung diseases
Idiopathic pulmonary fibrosis (IPF)
Sarcoidosis
Post-TBC-syndrome
Pleural disease, eg. asbestosis
Rheumatological diseases
...

Pulmonary vascular diseases
Primary or secondary pulmonary hypertension (PPH)
Veno-occlusive disease
...
Neuromuscular diseases
ALS
Post-polio-syndrome
M. Duchenne
...
Carcinoma
Primary pulmonary carcinoma
Carcinoma with pulmonary involvement

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ting is increasing; and often works in close collaboration with a lung transplant program. Lung transplantation can be the ultimate therapy for a highly selected subgroup of these patients.

In the following paper specific aspects of advanced lung disease will be reviewed including co-morbidities, outcome in the intensive care unit, criteria for lung transplantation and goals of the multidisciplinary team approach. This review focuses on advanced lung disease due to 1) chronic obstructive lung disease (COPD), 2) cystic fibrosis (CF) and 3) idiopathic pulmonary fibrosis (IPF). Other important aspects, such as palliative care, ethical aspects of end-of life care and treatment strategies of symptom relief are not covered here but have been reviewed in another recent publication [1, 2].

Co-morbidities in advanced lung disease

Nowadays a multidisciplinary management of co-morbidity in advanced lung disease patients is becoming increasingly important and has a direct impact on survival [3]. In general, most advanced lung disease patients suffer from several additional complications, which are directly or indirectly linked to the underlying disease (table 2). Pulmonary complications in advanced lung disease patients include infections, bronchogenic carcinoma, secondary pulmonary hypertension with or without cor pulmonale, pneumothorax and pulmonary embolism. In addition, non-pulmonary co-morbidity is frequent such as cardiovascular complications, treatment related complications, osteoporosis, deconditioning, malnutrition, non-pulmonary infections (eg, due to treatment), gastrointestinal complications and psychosocial problems.

Infections

A recent study shows that, in addition to bacterial infections, viral infections are frequent causes of severe COPD-exacerbation leading to prolongation of hospital stay, and leading to greater impairment of lung function than those with non-infectious exacerbations [4]. In COPD-patients with combined bacterial and viral exacerbation the FEV_1 fall was greater (20.3% vs 3.6%, $p < 0.03$) and symptom count was higher ($p < 0.02$) than those with a bacterial pathogen alone [5]. In CF, viral respiratory infections are associated with an increased short and long term morbidity, prolonged hospitalisation, persistent decrease of pulmonary function, increased use of antibiotics, and more frequent exacerbations at follow-up [6]. There is a clear association between viral infection and subsequent bacterial colonisation in CF: in up to 68% new bacterial colonisation is found during the viral season and new bacterial colonisation predominantly occurs within 3 weeks after a viral upper respiratory tract infection and moreover up to 85% of new pseudomonal colonisations followed a viral upper respiratory tract infection within 3 weeks [6].

Furthermore, there is an increase of atypical mycobacterial infection in advanced lung disease, especially in COPD, non-CF bronchiectasis and CF (up to 20% of CF patients are co-infected with atypical mycobacteria) [7]. Moreover, even infection with typical tuberculosis is frequent in some patients with advanced lung disease. The incidence of tuberculosis in patients with IPF is more than five times higher compared to the general population. The presentation is often atypical, mimicking lung cancer or bacterial pneumonia [8]. Another rare but often severe infectious

Table 2
Co-morbidities in advanced lung disease.

Pulmonary complications
Bacterial, viral and fungal infection
Bronchogenic carcinoma
Cor pulmonale
Pneumothorax
Pulmonary embolism
Cardiovascular complications
Coronary artery disease
Left ventricular failure
Hypertension
Stroke
Complications related to treatment (such as)
Systemic steroids
Immunosuppressants
New agents (IPF, PAH)
Other medical complications
Non-pulmonary infections
Gastrointestinal complications
– Related to underlying disease (like portal hypertension in CF)
– Gastro-esophageal reflux, peptic ulcer, diverticular disease
Osteoporosis
Deconditioning
Malnutrition / cachexia
Other issues
Psychosocial issues
Ethical issues

complication, especially in cavitary and bullous disease, is mycetoma due to *Aspergillus fumigatus* [9].

Due to the wide differential diagnosis in cases of infection with different treatment consequences, there is often a need for an invasive diagnostic approach (bronchoscopy) and consequent treatment of the infection.

Bronchogenic carcinomas

Bronchogenic carcinomas are frequent complications in advanced lung disease. Patients with COPD have an increased risk for bronchogenic carcinoma (up to 20-fold); and already in the early 1980's Turner-Warwick et al. documented a 14-fold increased risk for bronchogenic carcinomas in patients with IPF [10]. Symptoms of bronchogenic carcinoma are usually non-specific and there are often only subtle changes on chest radiographs, particularly in the presence of pre-existing structural abnormalities of the lung. In consequence, a high index of suspicion should be maintained in the management of this patient group. An established diagnosis of bronchogenic carcinoma in advanced lung disease has a major impact on the therapeutic approach in these patients. Fortunately, to date, in selected patients with carcinoma in the presence of severe COPD the tumour can still be resected due to increased experience with surgery from lung volume reduction surgery (LVRS) [11].

Secondary pulmonary hypertension and cor pulmonale

Cor pulmonale, defined as "right ventricular enlargement with or without right ventricular failure, due to a primary pulmonary process" is another important co-morbidity in advanced lung disease. There are multiple causes of an increase in pulmonary artery pressure: 1) vascular obliteration due to loss of lung tissue (emphysema), fibrosis (IPF, bronchiolitis) or luminal occlusion (pulmonary emboli); 2) chronic hypoxaemia causing vasoconstriction of pulmonary arteries and a release of various mediators of the vascular endothelium (eg, endothelin 1); 3) changes of intrathoracic pressure; 4) elevation of pulmonary venous pressure; and 5) increase of pulmonary blood volume [3, 12]. Up to 40% of COPD patients with a FEV_1 below 1 litre suffer from pulmonary arterial hypertension and right ventricular dysfunction. This is a poor prognostic indicator with an increased 4-year mortality rate of 73% [12]. Similarly, patients with IPF and pulmonary arterial hypertension have a poorer prognosis. Recently published data found pulmonary arterial hypertension in 32% of IPF patients and was associated with an increased one-year mortality rate (28.0% vs 5.5%, respectively; $p = 0.002$) [13]. There was a linear correlation between the degree of pulmonary arterial hypertension and mortality. Pulmonary arterial hypertension may be an important adjunct in monitoring disease progression, triaging for transplantation, and guiding therapy. Only a few small studies exist with regard to pulmonary arterial hypertension in CF. Fraser et al. documented pulmonary arterial hypertension without evidence of cor pulmonale in 7 of 18 adult CF patients [14]. In this study, linear regression analysis revealed a significant correlation of pulmonary arterial hypertension with FEV_1, SaO_2 during wakefulness and during sleep and after 6 min exercise. In the multivariate analysis, awake SaO_2 showed to be the strongest predictor for pulmonary arterial hypertension. A five year clinical follow-up of the original cohort revealed that mortality was significantly higher in those with pulmonary arterial hypertension compared to those without ($p = 0.0129$).

Pulmonary embolism

The clinical diagnosis of acute pulmonary embolism in advanced lung disease is difficult to establish due to the underlying lung disease. Nevertheless, pulmonary embolism is a frequent complication in advanced lung disease: Tillie-Leblond et al. found a 25% prevalence of pulmonary embolism in COPD-patients hospitalized for severe exacerbation of unknown origin [15]. The risk for pulmonary embolism was higher in patients with a history of previous thromboembolic disease, malignant disease, and decrease in $PaCO_2$ from baseline. In IPF patients awaiting lung transplantation, the incidence of pulmonary embolism increases to 22% [16]. Nevertheless, the major reason for death was a more rapid progression of IPF. In case of pulmonary embolism anticoagulation is applied in a similar manner as for patients without advanced lung disease (INR between 2–3).

Secondary spontaneous pneumothorax

In most advanced lung disease secondary spontaneous pneumothorax can occur with a clear relationship to clinical severity. Patients with pneumothorax often have worse outcomes [17]. As with pulmonary embolism diagnosis of a pneumothorax can be difficult. Acute onset of breathlessness and chest pain are the clinical key feature. Conventional radiology is often indistinct: The pleural line may be difficult to visualise because the lung appears hyperlucent, resulting in minimal difference in the radiodensity of the lung and the pneumothorax. Particularly in bullous lung disease pneumothorax may be difficult to distinguish from a large, thin-walled, air-contained bulla. It is important to mention, that even a small pneumothorax can result in severe respiratory impairment. There is often a need for a computerised tomography to distinguish between pneumothorax and pulmonary embolism [17]. The correct treatment of a pneumothorax is largely debated. In general a small pneumothorax (less than 15% involvement of the hemithorax) without sign of tension (no mediastinal shift to the contralateral side) can be treated without chest-drain [17]. The first episode of a large pneumothorax can be treated as well with chest tube drainage. When necessary a talk pleurodesis or with a surgical procedure via videothoracoscopy can be performed (for further consideration see reference [17] and [18]). The second episode of pneumothorax needs generally a surgical procedure.

Cardiovascular complications

The majority of COPD patients and IPF patients are current or ex-smokers and cardiovascular disease is frequent. In a population-based cohort of new COPD patients of Saskatchewan (Canada), 5648 COPD patients with 23426 patient's years (PY) were investigated [19]. COPD patients had significant higher cardiovascular morbidity and mortality rates compared to the general population. In addition, increased hospitalisation rates were reported for cardiovascular disease than for COPD exacerbation (177 per 1000 PY versus 101 per 1000 PY, respectively). Heart failure was the most common cause for hospitalisation in cardiovascular disease (59 per 1000 PY), followed by ischaemic heart disease (42 per 1000 PY), cerebrovascular disease (21 per 1000PY), cardiomyopathy (21 per 1000PY) and less frequently, right heart failure (4 per 1000PY). Moreover, cardiovascular disease was the major cause of death in this study cohort (41 per 1000PY versus 16 per 1000PY death due to COPD).

Table 3

Major side effects of immunosuppressive agents.

Dermatological and soft tissue
Skin thinning and purpura
Alopecia
Hirsutism
Cancer
Cardiovascular
Hypertension
Premature atherosclerotic disease
Influence on lipid metabolism
Gastrointestinal
Gastritis / oesophagitis
Peptic ulcer disease
Pancreatitis
Hepatitis
Visceral perforation
Renal
Renal insufficiency
Hypo / hyperkalaemia
Fluid volume shifts
Infectious disease
Increased risk:
Bacterial infections
Viral infections
Fungal infections
Opportunistic infections

Bone / muscle
Osteoporosis
Avascular necrosis
Myopathy
Endocrine
Diabetes mellitus
Adrenal insufficiency
Neuropsychiatric
Euphoria / dysphoria
Depression
Psychosis
Insomnia
Genitourinary and reproductive
Amenorrhoea/ Infertility
Intrauterine growth retardation

Pulmonary cachexia and deconditioning

Deconditioning is an additional frequent co-morbidity, not only in COPD patients. A cross-sectional study of lung transplant candidates showed decreased peripheral muscle force, especially in CF patients [20]. In order to improve muscle force, active pulmonary rehabilitation should be instituted early to improve muscle force. In view of poor nutritional status in some patients (especially in CF), exercise training should be combined with nutritional support.

Complications related to treatment

Advanced lung disease patients often complain of complications related to treatment. Systemic steroids, widely used in these patients, are associated with substantial side effects, including osteoporosis, arterial hypertension, diabetes mellitus, myopathy and cataracts. They have a direct influence on humoral and cellular immunity [21]. All other immunosuppressive agents used in advanced lung disease are associated with several pulmonary and non-pulmonary toxicities (see table 3). Therefore, their use must be balanced with the possible serious side effects.

Other co-morbidities

Other co-morbidities as direct or indirect consequences of advanced lung disease are frequent, such as osteoporosis, non-pulmonary infections (due to treatment) and gastrointestinal complications. In a recent study in patients awaiting lung transplantation, osteoporosis was present in 60% and osteopenia in an additional 31% [22]. Body mass index and glucocorticoid use turned out to be the only independent risk factors for osteoporosis; neither age or gender, underlying disease or FEV_1%, exhibited significance as risk factors. However, the predictive value of body mass index and glucocorticoid use was relatively poor (adjusted R^2 between 0.30 and 0.37). In addition, in advanced lung disease, patients are compromised regarding their daily life with increasing disability, resulting in anxiety, dependency, poor coping strategies, and depression in both, the patients and their family. Psychosocial co-morbidities play an important role, not only with regard to end-of-life issues and terminal care.

In conclusion, co-morbidities are frequent in advanced lung disease. They may be due to the underlying disease process, due to the presence of common risk factors in this population such as smoking, or due to the treatment itself.

Advanced lung disease and intensive care

Several concerns exist with regard to treatment of advanced lung disease patients in an intensive care unit (ICU). The prognosis of this group of patients requiring admission to ICU is commonly believed to be poor. Especially in COPD patients, it was an agreement that admission to ICU was accompanied with low survival. Several studies reported in-hospital mortality rates varying between 20–80% due to differences of study cohorts. Data on long-term survival after hospital discharge is limited. One of the studies with the longest follow-up, published in 2002, reported a mortality rate of 64% at 3 years [23]. In this study, 74 COPD patients with admissions to a tertiary referral ICU were investigated. Minimum follow-up was 3 years, mean FEV_1 0.74±0.3 litre, reflecting severe obstructive lung disease and mean age 66±9 years. Eighty-five percent of patients needed mechanical ventilation for a median of 2 days (range 1 to 17 days). Median of ICU-stay was 3 days (range 2 to 17 days). Up to 80% of patients survived to hospital discharge. Hospital mortality correlated with initial $PaCO_2$ at admission and APACHE II score. Mortality at 6 months, 1, 2, and 3 years after discharge was 41%, 49%, 58%, and 64%, respectively. No independent predictors of long term mortality were found. The authors concluded that patients with COPD requiring intensive care admission have a realistic chance of surviving to hospital discharge, but in the longer term the mortality rate is high with nearly two thirds having died at 3 years. However, this outcome is similar to the background mortality of patients with equivalent severe underlying respiratory disease. Although physiological abnormalities at presentation may help to predict hospital outcome, no predictors of survival in the longer term were identified. In particular, the need for mechanical ventilation did not distinguish long term survivors from non-survivors. A recent study found equal results in COPD patients [24]. Mortality rates following admission to ICU at 6 months and 1, 3, and 5 years were 39%, 43%, 61%, and 76%, respectively. Hospital mortality correlated with age, previous mechanical ventilation, albumin level at ICU admission, long-term use of oral corticosteroids, and APACHE II score. As well as in the previous study, no signifi-

cant factors predicting 5-year survival were found. Interestingly, pre-admission quality of life was besides of age and APACHE II score the third significant predictor for 6-year mortality in a multicentre cohort study [25].

In contrast to COPD patients, outcome in IPF patients admitted to ICU is poor. One retrospective study at the Mayo Clinic followed 38 IPF patients admitted to ICU between 1995 and 2000 [26]. Reasons for ICU-admission were progression of IPF in 40% and respiratory complications (pneumonia, pulmonary embolism, congestive heart failure, pneumothorax) in 42%. Fifty percent of patients required mechanical ventilation, 95% had at least single or multiple organ failure, and 28% sepsis. Sixty one percent of patients died during hospitalisation and an additional 32% during the first 2 months following discharge. One of the major problems in IPF is to differentiate between IPF-exacerbation with a worse outcome and treatable complications such as pneumonia with more favourable outcome.

Admission to ICU in CF patients, especially in adult CF patients, remains a controversial issue. In a retrospective multicentre study in France, survival of 42 adult CF patients admitted to ICU between 2000 and 2003 was higher than previously reported (27). All patients (mean age 28±8 years) had severe lung disease (mean FEV_1 28 ± 12% predicted; mean $PaCO_2$ 47 ± 9 mm Hg). Overall ICU mortality was 14%. On admission, non-invasive ventilation was started in 57% of cases, and in 67% of patients continued successfully. One year after ICU discharge, 10 of 28 survivors have received a lung transplant. Among markers recognised for severity of CF disease, only annual loss of FEV_1 was associated with a poor outcome, and endotracheal intubation was identified as strong independent predictor for mortality. Although CF patients requiring ICU admission need to be selected carefully, ICU admission should be considered in these patients.

In conclusion, mortality in ICU is linked to the underlying lung disease, disease severity and additional co-morbidities. Mechanical ventilation for exacerbation in COPD and CF patients has an acceptable mortality rate, and COPD-exacerbations requiring ICU admission have no influence on long-term survival for this group of patients. Nevertheless, whenever possible non-invasive ventilation should be considered in most cases of advanced lung disease.

Advanced lung disease and lung transplantation

Advanced lung disease patients are possible candidates for lung transplantation (LTx). Nowadays, LTx has evolved from an experimental procedure to a viable therapeutic option in many countries. In Switzerland, the 1, 5 and 9 year survival rates in 2002 were 77% (95% CI 72–82), 64% (95% CI 57–71) and 56% (95% CI 45–67), respectively [28]. The 5-year survival rate of patients transplanted after 1998 is 72% (95% CI 64–80). Therefore, LTx should be considered in patients with advanced lung disease.

International guidelines exist with regard to patient selection criteria and referral for LTx [29]. Guidelines and contraindications for LTx are summarised in table 4 and 5. In general, referral for LTx assessment is recommended when patients have a predicted 2- to 3-year survival of less than 50%, or dyspnea of New York Heart Association (NYHA) class III or IV level, or both.

In COPD, when an increase in the BODE-index of 5 or more is diagnosed, referral to a lung transplant centre is recommended (Bode-index is a combination of the four factors: B = body-mass index, O = degree of airflow obstruction, D = dyspnoea, E = exercise capacity, measured by the six-minute–walk test [30]).

In CF and non-CF bronchiectasis a FEV_1 below 30% or a rapid decline in FEV_1, in particular in young female patients and patients with CF related diabetes should lead to referral.

Patients with restrictive lung disease, primarily with IPF, show often rapid disease progression. Patients with histologically confirmed usual interstitial pneumonia (UIP) should be evaluated for LTx at time of diagnosis even if they have only a mild impairment in pulmonary function [29].

In pulmonary vascular disease dyspnoea with a persistent NYHA class III or IV on maximal medical therapy is the most important factor for LTx-evaluation. Since these patients often benefit from the modern medical treatment options of pulmonary arterial hypertension such as endothelin-antagonists they should be referred to a specialised clinic. In chronic thrombo-embolic disease thrombendarterectomy can be an option and should always be considered first in these patients. Lung volume reduction surgery (LVRS) in COPD is another option to delay or bridge to LTx. LVRS can significantly improve symptoms and lung function in selected patients who are initially potential candidates for LTx and can allow postponing LTx for up to 5 years [31].

It is important to emphasise the right time for LTx evaluation. Several factors determine this time point: underlying disease, disease progression, expected time on waiting list, and risk of death on waiting list (figure 1). Waiting time is based on many factors such as height and blood group. It tends to be longer for taller patients and for recipients with rare blood groups. The decision to refer patients to LTx centres should not be based on a single factor, because no simple, single-point determinant is sufficiently predictive of early mortality. Moreover, it is recommended to

Table 4
Time for referral to a LTx-centre.

Chronic obstructive pulmonary disease
BODE index exceeding 5
Cystic fibrosis and other causes of bronchiectasis
FEV_1 below 30% predicted or a rapid decline in FEV_1 – in particular in young female patients
Exacerbation of pulmonary disease requiring ICU stay
Increasing frequency of exacerbations requiring antibiotic therapy
Refractory and/or recurrent pneumothorax
Recurrent haemoptysis not controlled by embolisation
Idiopathic pulmonary fibrosis (IPF) and non-specific interstitial pneumonia (NSIP)
Histologic or radiographic evidence of UIP irrespective of vital capacity
Histological evidence of fibrotic NSIP
Pulmonary arterial hypertension
NYHA functional class III or IV, irrespective of ongoing therapy
Rapidly progressive disease
Sarcoidosis, lymphangioleiomyomatosis (LAM), pulmonary Langerhans cell histiocytosis (eosinophilic granuloma)
NYHA functional class III or IV

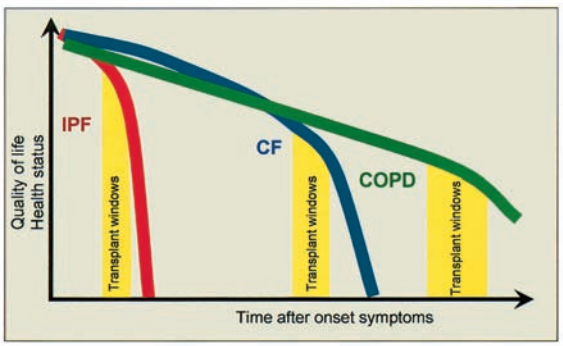
Table 5
Contraindications for LTx.

Absolute contraindications for LTx
Malignancy in the last 2 years, with the exception of cutaneous squamous and basal cell tumors. In general, a 5-year disease-free interval is prudent.
Untreatable advanced dysfunction of another major organ system (eg, heart, liver, or kidney).
Documented non-adherence or inability to follow through with medical therapy
Untreatable psychiatric or psychological condition associated with the inability to cooperate or comply with medical therapy.
Absence of a consistent or reliable social support system.
Substance addiction (eg, alcohol, tobacco, or narcotics) that is either active or within the last 6 months.
Relative contraindications for LTx
Age older than 65–70 years.
Critical or unstable clinical condition (eg, shock, mechanical ventilation or extra-corporeal membrane oxygenation).
Severely limited functional status with poor rehabilitation potential.
Colonisation with highly resistant or highly virulent bacteria, fungi, or mycobacteria.
Severe obesity defined as a body mass index (BMI) exceeding 30 kg/m^2.
Mechanical ventilation.

rely on a variety of clinical (eg, rate of infection, intensive care unit hospitalisation, oxygen need, weight loss, etc.), laboratory (e.g., PaO_2 and $PaCO_2$), and functional findings (eg, pulmonary function tests, echocardiography, exercise capacity, etc). As an example, in CF patients LTx should be considered early when severe complications like haemoptysis or recurrent pneumothoraces occur. In addition, malnutrition with decreasing body mass index, frequent hospitalisation due to infective exacerbation and chronic infections with multiresistant bacteria such as Burkholderia cepacia, are factors that should prompt to early referral for LTx evaluation.

In conclusion, advanced lung disease patients are potential candidates for LTx. Referral to a transplant centre depends on underlying lung disease, disease progression, and co-morbidities. Due to rapid progression of IPF, these patients should be considered for lung transplantation early on when they still have few symptoms and almost a normal pulmonary function.

Figure 1
The right time for LTx-evaluation.



Right time for LTx evaluation depends from several factors, such as underlying disease, disease progression, expected time on waiting list, and risk of death on waiting list

Advanced lung diseases and the multidisciplinary care team

There are several direct and indirect consequences of advanced lung disease which influence treatment of these patients; and there is an increasing request for specialised centres treating such patients. One of the major goals of caring for advanced lung disease patients is to optimise health related quality of life (HRQL) in every stage of the disease process. Moreover, quality of life is not only influenced by respiratory impairment with respiratory symptoms such as dyspnoea or cough, but by pulmonary complications and other co-morbidities. One of the most important factors influencing HRQL is the psychosocial functioning in daily life. Advanced lung disease patients are influenced by several psychosocial issues which are related to HRQL. Particularly, the inability to participate in normal daily activity influence HRQL. A dramatic change in relations and partnership and familial functioning with increasing dependency on other people is common. The goal of the multidisciplinary team approach in these patients is to deal with all this issues (figure 2). Not only chest physicians and respiratory nurses are needed to care for these patients; dieticians, physiotherapists, gastroenterologists and other specialists are equally important to deal with co-morbidities. Last but not least, with the increasing psychosocial issues psychiatrists and social workers play an important role in this multidisciplinary team.

The impact of specialised multidisciplinary centres for the management of these patients has not yet been evaluated. A randomised controlled trial evaluating the merits of such care centres would be methodologically difficult. Nonetheless, there is evidence, that patients treated in these specialised centres have improved survival. The establishment of a new adult CF centre in Cambridge in the early 90's provided a unique opportunity to perform outcome studies in CF patients [32]. There was a clear advantage in clinical outcome for patients receiving treatment in the specialised CF-centre. The maximum benefit was apparent with regard to nutrition and pulmonary disease severity, both of which are important determinants of prognosis. The authors concluded that it is crucial that paediatric patients receive centre care for their CF at the earliest possible age if they are to gain the impetus for prolonged survival in adulthood. Patients with CF have already requested their care be provided by centres, and it is now the clinical responsibility of all physicians to ensure that this care begins in childhood and is continued throughout adult life. In CF, the multidisciplinary team approach is now considered essential as documented in a European consensus statement [33].

In 1999, it was shown also for IPF patients that they have a significant survival benefit, when treated in a specialised interstitial lung disease clinics compared to IPF patients attending the general respiratory clinic [34]. Patients attending the IPF clinic had a median survival of >3714 days versus 1796 days for patients from the general clinic (CI 940–2652), p = 0.032. Age was an important determinant of outcome. There was no difference in survival of patients over 60 years of age. The authors stated that a dedicated multidisciplinary clinic may result in improved outcome in patients with IPF, particularly for patients younger than 60 years. This has implications that may facilitate the development of suitably powered therapeutic trials and may affect patient referral for transplantation.

In conclusion, the care of advanced lung disease patients in a multidisciplinary team may influence survival, especially in CF and IPF.

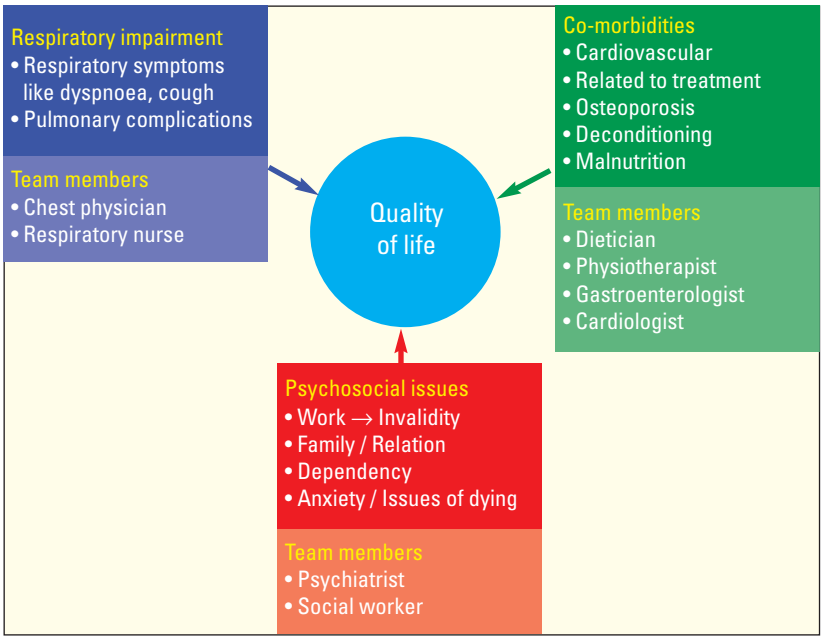


Figure 2

The multidisciplinary care team for advanced lung disease.

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