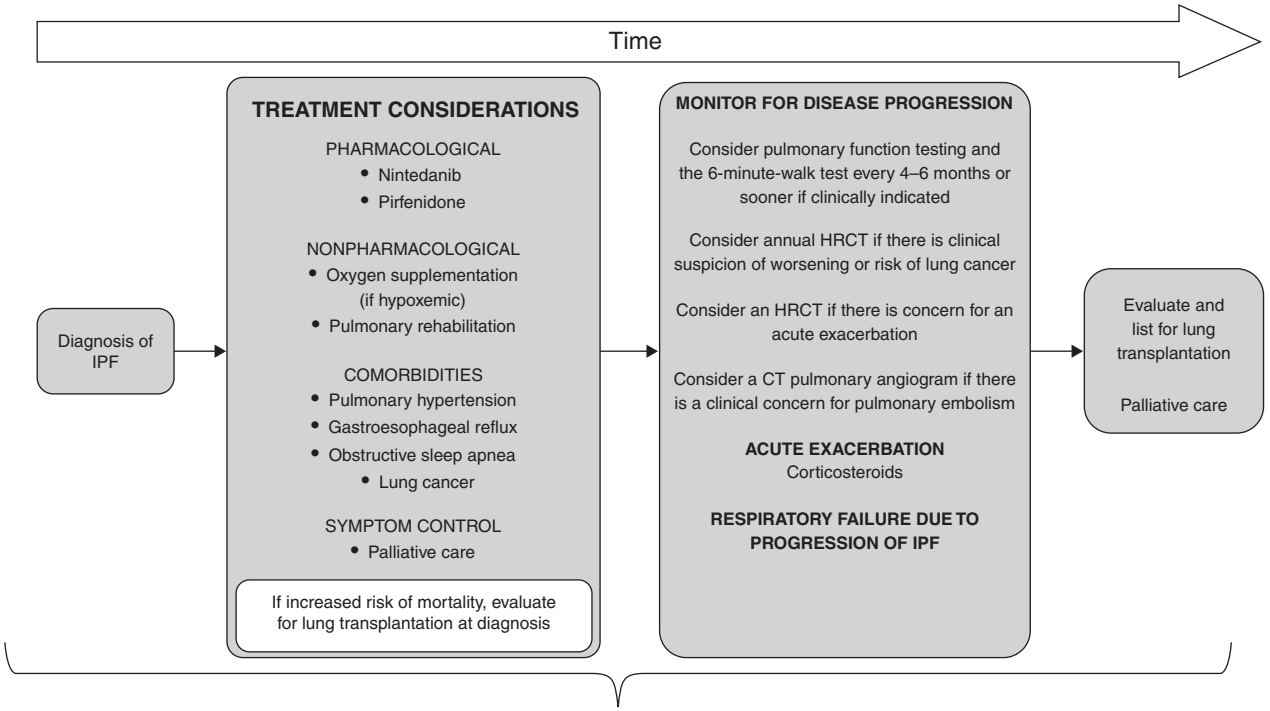

Figure 11. Schematic pathway for clinical management of patients with idiopathic pulmonary fibrosis (IPF), developed using consensus by discussion. Treatment considerations should include both pharmacological (nintedanib and pirfenidone) and nonpharmacological (oxygen supplementation and/or pulmonary rehabilitation) therapies. Patients should be evaluated and treated for existing comorbidities, including pulmonary hypertension, gastroesophageal reflux, obstructive sleep apnea, and lung cancer. Patients may benefit from involvement of palliative care to help with symptom management (cough, dyspnea, and/or anxiety). Patient values and preferences should be explored. Patients at increased risk of mortality should be referred for lung transplantation at diagnosis. Patients should be evaluated every 3–6 months or more often for disease progression. Acute exacerbations may be treated with corticosteroids. Mechanical ventilation is not recommended for the majority of patients with respiratory failure. Adapted from Reference 1. CT = computed tomography; HRCT = high-resolution computed tomography.

Table 4. Definition of Progressive Pulmonary Fibrosis

Definition of PPF
In a patient with ILD of known or unknown etiology other than IPF who has radiological evidence of pulmonary fibrosis, PPF is defined as at least two of the following three criteria occurring within the past year with no alternative explanation*:
1 Worsening respiratory symptoms
2 Physiological evidence of disease progression (either of the following): a. Absolute decline in FVC ⩾5% predicted within 1 yr of follow-up b. Absolute decline in D_{LCO} (corrected for Hb) ⩾10% predicted within 1 yr of follow-up
3 Radiological evidence of disease progression (one or more of the following): a. Increased extent or severity of traction bronchiectasis and bronchiolectasis b. New ground-glass opacity with traction bronchiectasis c. New fine reticulation d. Increased extent or increased coarseness of reticular abnormality e. New or increased honeycombing f. Increased lobar volume loss

Definition of abbreviations: ILD = interstitial lung disease; IPF = idiopathic pulmonary fibrosis; PPF = progressive pulmonary fibrosis.
*Although it is critical to exclude alternative explanations of worsening features for all patients with suspected progression, this is particularly important in patients with worsening respiratory symptoms and/or decline in D_{LCO} given the lower specificity of these features for PPF compared with FVC and chest computed tomography.

(Figure 13). These may include increased traction bronchiectasis and bronchiolectasis, new ground-glass opacity with traction bronchiectasis, new fine reticulation, increased coarseness of reticular abnormality, new or increased honeycombing, and increased lobar volume loss.

In IPF, progression is usually manifested by increased extent of the UIP pattern, in both transverse and coronal planes (133–135). The size and number of honeycomb cysts often increase as the disease progresses. Progression of traction bronchiectasis and bronchiolectasis is a strong independent predictor of mortality in IPF (136). In ILDs other than IPF, however, the pattern of progression is variable and may include evolution of ground-glass abnormality to reticular abnormality (134, 137), evolution of reticular abnormality to honeycombing (137), and/or increase in traction bronchiectasis/bronchiolectasis.