Idiopathic Pulmonary Fibrosis: diagnosis and management

Review questions

Chapter	Review questions	Outcomes
Diagnosis	In suspected IPF what is the value of adding biopsy to clinical evaluation, PFTs, HRCT +/- bronchoaveolar lavage for confirming the diagnosis of IPF?	• Mortality • 1 and 3 year survival rates • Sensitivity • Specificity • Adverse events • Improvement in health-related quality of life
	In suspected IPF what is the value of adding multidisciplinary team (MDT) consensus to clinical assessment, PFTs and HRCT in the diagnosis of IPF?	• Mortality • 1 and 3 year survival rates • Sensitivity • Specificity • Inter-observer agreement • Improvement in health-related quality of life
	How and by whom is a MDT diagnostic consensus best achieved (i.e. constituency of the MDT, specialist clinics, networks)?	• Mortality • 1 and 3 year survival rates • Sensitivity • Specificity • Inter-observer agreement • Improvement in health-related quality of life
Prognosis	Do serial pulmonary function tests (resting spirometric, gas transfer measurement and oxygen saturation) predict prognosis of IPF?	• Mortality/survival (time to event) • Progression free survival • Acute exacerbation (time to event) ○ Respiratory hospitalisations (Surrogate outcome for acute exacerbation) • Eligibility for lung transplantation
	Does baseline sub-maximal exercise testing predict prognosis of IPF?	• Mortality/survival (time to event) • Progression free survival • Acute exacerbation (time to event) ○ Respiratory hospitalisations (Surrogate outcome for acute exacerbation) • Eligibility for lung transplant
	Does baseline echocardiography predict prognosis of IPF?	• Mortality/survival (time to event) • Progression free survival • Acute exacerbation (time to event) • Respiratory hospitalisations (Surrogate

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		outcome for acute exacerbation) • Eligibility for lung transplant
	Do baseline HRCT scores predict prognosis of IPF?	• Mortality/survival (time to event) • Progression free survival • Acute exacerbation (time to event) • Respiratory hospitalisations (Surrogate outcome for acute exacerbation) • Eligibility for lung transplant
Patient review and follow up	How often should a patient with confirmed diagnosis of IPF be reviewed?	• Change in percent predicted DLCO • Change in percent predicted forced vital capacity • Oxygen saturation at rest • Oxygen saturation on exertion • Distance walked on 6 min walk or incremental shuttle walk test • Eligibility for lung transplant
	In which healthcare setting and by whom should a review appointment for patients with confirmed IPF be conducted?	• Change in percent predicted DLCO • Change in percent predicted forced vital capacity • Oxygen saturation at rest • Oxygen saturation on exertion • Distance walked on 6 min walk or incremental shuttle walk test • Eligibility for lung transplant
Best supportive care	What is the clinical and cost effectiveness of best supportive care (palliation of cough, breathlessness and fatigue, and oxygen management) in the symptomatic relief of patients with IPF?	• Mortality • Hospitalisations due to IPF complications (including IPF exacerbations) • Improvement in cough and breathlessness • Improvement in psychosocial health (including depression) • Performance on sub-maximal walk test (distance walked and lowest SaO2) • Symptom relief
Psychosocial support	What is the specific type of psychosocial support and information for patients diagnosed with IPF?	• Dyspnoea • Improvement in psychosocial health (including depression) • Improvement in health-related quality of life
Pulmonary	What are the benefits of	• Mortality

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rehabilitation	pulmonary rehabilitation programmes for patients with confirmed IPF?	• 1 and 3 year survival rates • Dyspnoea • Hospitalisations due to IPF complications (including IPF exacerbations) • Improvement in cough and breathlessness • Improvement in health-related quality of life • Performance on sub-maximal walk test (distance walked and lowest SaO2) • Improvement in psychosocial health (including depression)
	What is the optimal course content, setting and duration for patients referred for pulmonary rehab programmes?	• Mortality • 1 and 3 year survival rates • Dyspnoea • Hospitalisations due to IPF complications (including IPF exacerbations) • Improvement in cough and breathlessness • Improvement in health-related quality of life • Performance on sub-maximal walk test (distance walked and lowest SaO2) • Improvement in psychosocial health (including depression)
Pharmacological interventions	Which drug should be initiated first, for how long, and what combination in the treatment of IPF? a. What is the clinical and cost effectiveness of pharmacological interventions to manage patients with suspected or confirmed IPF?	• Mortality • 1 and 3 year survival rates • Adverse events (please refer to AE table listed by GDG) • Dyspnoea • Gas transfer • Hospitalisations due to IPF complications, including IPF exacerbations • Improvement in health-related quality of life • Lung capacity • Performance on sub-maximal walk test (distance walked and lowest SaO2)
	Which measures can be taken to minimize the occurrence/severity of adverse events when undergoing pharmacological treatment for IPF?	• Mortality • 1 and 3 year survival rates • Adverse events (please refer to AE table listed by GDG) • Dyspnoea • Hospitalisations due to IPF complications, including IPF exacerbations • Improvement in health-related QoL • Performance on sub-maximal walk test

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		(distance walked and lowest SaO2)
Lung transplantation	What is the optimal timing to consider a patient with IPF for lung transplantation referral?	• Mortality or survival • Cross-over time • Hospitalisations due to IPF complications (including IPF exacerbations) • Improvement of health-related quality of life • Occurrence lung transplantation
Ventilation	In acute or acute-on chronic respiratory failure in patients with IPF, what is the value of non-invasive and invasive ventilation?	• Mortality (in hospital and post discharge) • Improvement of health-related quality of life • Hospital length of stay