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NATIONAL INSTITUTE FOR HEALTH AND CARE EXCELLENCE

Health and social care directorate

Quality standards and indicators

Briefing paper

Quality standard topic: Idiopathic Pulmonary Fibrosis

Output: Prioritised quality improvement areas for development.

Date of Quality Standards Advisory Committee meeting: 19 May 2014

Contents

1 Introduction

This briefing paper presents a structured overview of potential quality improvement areas for idiopathic pulmonary fibrosis. It provides the Committee with a basis for discussing and prioritising quality improvement areas in order to develop draft quality statements and measures for public consultation.

1.1 Structure

The structure of this briefing paper includes a brief overview of the topic followed by a summary of each of the suggested quality improvement areas followed with supporting information.

Where relevant, guideline recommendations selected from the key development sources below are presented to aid the Committee when considering specific aspects for which statements and measures should be considered.

1.2 Development sources

The key development sources referenced in this briefing paper are:

- Idiopathic pulmonary fibrosis: The diagnosis and management of suspected idiopathic pulmonary fibrosis NICE clinical guideline 163 (2013)
- British Thoracic Society BTS Guideline on Pulmonary rehabilitation in adults

Key policy documents, reports and national audits
Relevant national policy documents, reports and audits will be used to inform the development of the quality standard.

- British Thoracic Society (ongoing audit) BTS Lung Disease Registry Programme – idiopathic pulmonary fibrosis.
- British Thoracic Society (2014 – undergoing consultation) BTS quality standards for pulmonary rehabilitation.
- Department of Health (2013) Improving quality of life for people with long term conditions.
- NHS England (2013) NHS standard contract for respiratory: interstitial lung disease (adult).
- NHS Lung Improvement (2013) Improving the quality and safety of home oxygen services: the case for spread.

2 Overview

2.1 Focus of quality standard

This quality standard will cover the diagnosis and management of idiopathic pulmonary fibrosis in adults, from the initial suspicion of the disease to referral, supportive care and treatment.

2.2 Definition

Idiopathic pulmonary fibrosis is a chronic, progressive fibrotic interstitial lung disease (ILD) of unknown origin. It is a difficult disease to diagnose and often requires the collaborative expertise of a consultant respiratory physician, radiologist and histopathologist to reach a consensus diagnosis. Most people with idiopathic pulmonary fibrosis experience symptoms of breathlessness, which may initially be only on exertion. Cough, with or without sputum, is a common symptom. Over time, these symptoms are associated with a decline in lung function, reduced quality of life and ultimately death.

2.3 Incidence and prevalence

IPF is rare in people younger than 45 and the median age of presentation is 70 years. The prevalence is around 15 to 25 per 100,000 and increases with age. The average hospital with a catchment of 500,000 will have 40 to 45 new cases a year and a GP surgery of 10,000 patients will have 2 to 3 new cases every three years. Around two-thirds of people with IPF are smokers and IPF often co-exists with chronic obstructive pulmonary disease (COPD).

The median survival for people with idiopathic pulmonary fibrosis in the UK is approximately 3 years from the time of diagnosis. However, about 20% of people with the disease survive for more than 5 years. The rate of disease progression can vary greatly. A person's prognosis is difficult to estimate at the time of diagnosis and may only become apparent after a period of careful follow-up.

2.4 Management

Specific pharmacological therapies for IPF are limited but the last decade has seen more trials of new drugs which have had a variable impact on clinical practice. A number of difficulties arise when undertaking clinical trials in IPF in terms of defining precise, diagnostic inclusion criteria and clinically meaningful end-points. However, such trials are the only way by which promising new treatments will come to benefit patients. Furthermore, it is only by performing rigorous clinical trials that it has become evident that drugs once widely used to treat IPF may in fact have been harmful. The limitations of current pharmacological therapies for IPF highlight the importance of other forms of treatment including lung transplantation and best

supportive care such as oxygen therapy, pulmonary rehabilitation and palliation of symptoms.

2.5 National Outcome Frameworks

The tables below show the outcomes, overarching indicators and improvement areas from the frameworks that the quality standard could contribute to achieving.

Table 1 NHS Outcomes Framework 2014–15

Domain	Overarching indicators and improvement areas
1 Preventing people from dying prematurely	*Overarching indicator* 1a Potential Years of Life Lost (PYLL) from causes considered amenable to healthcare i Adults 1b Life expectancy at 75 i Males ii Females
2 Enhancing quality of life for people with long-term conditions	*Overarching indicator* 2 Health-related quality of life for people with long-term conditions** *Improvement areas* Ensuring people feel supported to manage their condition 2.1 Proportion of people feeling supported to manage their condition Reducing time spent in hospital by people with long-term conditions
4 Ensuring that people have a positive experience of care	*Overarching indicator* 4a Patient experience of primary care i GP services ii GP Out-of-hours services 4b Patient experience of hospital care *Improvement areas* Improving hospitals' responsiveness to personal needs 4.2 Responsiveness to in-patients' personal needs Improving people's experience of accident and emergency services 4.3 Patient experience of A&E services
Alignment across the health and social care system ** Indicator complementary with Adult Social Care Outcomes Framework (ASCOF)	

Table 2 The Adult Social Care Outcomes Framework 2013–14

Domain	Overarching and outcome measures
1 Enhancing quality of life for people with care and support needs	***Overarching measure*** 1A Social care-related quality of life* ***Outcome measures*** **People manage their own support as much as they wish, so that are in control of what, how and when support is delivered to match their needs.** 1B Proportion of people who use services who have control over their daily life **Carers can balance their caring roles and maintain their desired quality of life.** 1D Carer-reported quality of life
3 Ensuring that people have a positive experience of care and support	***Overarching measure*** **People who use social care and their carers are satisfied with their experience of care and support services** 3A Overall satisfaction of people who use services with their care and support 3B Overall satisfaction of carers with social services. 3E Improving people's experience of integrated care*
Aligning across the health and care system * Indicator complementary with the NHS Outcomes Framework	

Table 3 Public health outcomes framework for England, 2013–2016

Domain	Objectives and indicators
4 Healthcare public health and preventing premature mortality	***Objective*** Reduced numbers of people living with preventable ill health and people dying prematurely, while reducing the gap between communities ***Indicators*** Mortality from respiratory diseases
Aligning across the health and care system * Indicator shared with the NHS Outcomes Framework	

3 Summary of suggestions

3.1 Responses

14 stakeholders responded to the 2-week engagement exercise 19/03/2014 – 02/04/2014.

Stakeholders were asked to suggest up to 5 areas for quality improvement. Specialist committee members were also invited to provide suggestions. The responses have been merged and summarised in table 1 for further consideration by the Committee.

Full detail on the suggestions is provided in appendix 3 for information.

Table 1 Summary of suggested quality improvement areas

Suggested area for improvement	Stakeholder
Diagnosis • Awareness of clinical features of IPF • Multidisciplinary team (MDT) • Radiology • Finding a cause	APF, ARNS, BI, BLF, GORDS, IM, SCM1, SCM2, SCM3, SCM4, SHSFT
Information and support • Patients • Carers	ARNS, BI, BLF, IM, SCM1, SCM2, UKCPA

Management
• Pulmonary rehabilitation
• Best supportive care
  • Symptom control
  • Palliative care
• Pharmacological interventions
• Lung transplantation
• Oxygen assessment
	APF, ARNS, BI, BLF, IM, SCM1, SCM2, SCM4, SHNHST, UKCPA

Review / follow up	SHNHST, SCM3
Additional areas • Data collection • Clinical trials	SHNHST, ARNS SCM4, BLF

Stakeholder organisations who submitted suggestions are listed in table 2.

Table 2 Stakeholder details (abbreviations)

Abbreviation	Full name
APF	Action for Pulmonary Fibrosis
ARNS	Acute Respiratory Nurse Specialists/ Royal Brompton Hospital and Harefield NHS Trust
BI	Boehringer Ingelheim
BLF	British Lung Foundation
GORDS	Group of Occupational Respiratory Disease Specialists
IM	InterMune
NHSE	NHS England
RCN	Royal College of Nursing
SCM1	Specialist Committee Member 1
SCM2	Specialist Committee Member 2
SCM3	Specialist Committee Member 3
SCM4	Specialist Committee Member 4
SHNHST	Sheffield Teaching Hospitals NHS Foundation Trust
UKCPA	UK Clinical Pharmacy Association

4 Suggested improvement areas

4.1 Diagnosis

4.1.1 Summary of suggestions

Awareness of clinical features of IPF

Stakeholders suggested that there needs to be a higher level of awareness of IPF and symptoms among the public and in primary care or non-specialist secondary care as early diagnosis and intervention can delay disease progression and improve quality of life.

MDT

Stakeholders suggested that diagnosis is being made by inappropriate clinicians, such as GPs, prior to tests being completed.

Stakeholders suggested that a timely interstitial lung disease (ILD) MDT consensus diagnosis has been shown to improve the diagnostic accuracy of IPF and can expedite treatment/early diagnosis having a significant impact on outcomes and quality of life.

Stakeholders suggested that ILD MDTs need a defined composition.

A stakeholder commented there is an urgent need to develop the role of the CNS and support workers as an integral part of the MDT.

Radiology

Stakeholders suggested that access to, or links with, regional and national radiology experts/panels/networks with subspeciality in ILD are needed for accurate diagnosis.

Stakeholders suggested that standardisation of CT (computed tomography) protocols/techniques in diagnosis/follow up of IPF are needed to ensure appropriate decisions on management are made.

A stakeholder commented there is a need for guidance on when to undertake specific radiological tests and there is a need for access to specialist imaging opinion at all stages.

Finding a cause

Stakeholders commented that because treatment is prescribed depending on the stage of progress of the disease, quick and accurate diagnosis has a significant impact on outcomes.

A stakeholder commented it is vital to differentiate IPF from fibrosis with a cause which has an identical radiological appearance.

A stakeholder commented that IPF can be a diagnosis of elimination. They commented that NICE guidelines do not highlight possible causes of IPF and that further guidelines would be helpful.

A stakeholder suggested that occupational and environmental histories should be taken to assist with diagnosis.

4.1.2 Selected recommendations from development source

Table 3 presents recommendations that have been provisionally selected from the development source which may support potential statement development. These are presented in full below to inform the Committee's discussion.

Table 3 Specific areas for quality improvement

Suggested quality improvement area	Selected source guidance recommendations
Awareness of clinical features of IPF	NICE CG163 Idiopathic pulmonary fibrosis Recommendation 1.1.1 (KPI) identifies the clinical features but does not recommend increasing awareness among the public or specific clinical groups.
MDT	NICE CG163 Idiopathic pulmonary fibrosis 1.2.1, 1.2.2, 1.2.3, 1.2.4 (1.2.2 KPI)
Radiology	NICE CG163 Idiopathic pulmonary fibrosis does not reference local/regional experts/panel or refer to standardisation of CT protocols/techniques. NICE CG163 Idiopathic pulmonary fibrosis 1.2.1, 1.2.2 and 1.2.3 refer to diagnostic radiology.
Finding a cause	NICE CG163 Idiopathic pulmonary fibrosis 1.2.1 (detailed history), 1.2.4, 1.2.5 & 1.2.6 identify biopsies and when these should be considered.

Awareness of clinical features of IPF

NICE CG163 Idiopathic pulmonary fibrosis – Recommendation 1.1.1(Key priority for implementation)

1.1.1 Be aware of idiopathic pulmonary fibrosis when assessing a patient with the clinical features listed below and when considering requesting a chest X-ray or referring to a specialist:

- age over 45 years
- persistent breathlessness on exertion
- persistent cough
- bilateral inspiratory crackles when listening to the chest
- clubbing of the fingers
- normal spirometry or impaired spirometry usually with a restrictive pattern but sometimes with an obstructive pattern

MDT

NICE CG163 Idiopathic pulmonary fibrosis Recommendations 1.2.1, 1.2.2,1.2.3 and 1.2.4 (1.2.2 key priority for implementation)

1.2.1 Assess everyone with suspected idiopathic pulmonary fibrosis by:

- taking a detailed history, carrying out a clinical examination (see recommendation 1.1.1 for clinical features) and performing blood tests to help exclude alternative diagnoses, including lung diseases associated with environmental and occupational exposure, with connective tissue diseases and with drugs
- **and** performing lung function testing (spirometry and gas transfer)
- **and** reviewing results of chest X-ray
- **and** performing CT of the thorax (including high-resolution images).

1.2.2 Diagnose idiopathic pulmonary fibrosis only with the consensus of the multidisciplinary team (listed in table 1), based on:

- the clinical features, lung function and radiological findings (see recommendation 1.2.1)
- pathology when indicated (see recommendation 1.2.4).

1.2.3 At each stage of the diagnostic care pathway the multidisciplinary team should consist of a minimum of the healthcare professionals listed in table 1 [of CG163], all of whom should have expertise in interstitial lung disease.

Table 1 [of CG163] Minimum composition of multidisciplinary team involved in diagnosing idiopathic pulmonary fibrosis

Stage of diagnostic care pathway	**Multidisciplinary team composition (all healthcare professionals should have expertise in interstitial lung disease)**
After clinical evaluation, baseline lung function and CT	Consultant respiratory physician Consultant radiologist Interstitial lung disease specialist nurse Multidisciplinary team coordinator
When considering performing bronchoalveolar lavage, and/or transbronchial biopsy or surgical lung biopsy Only some patients will have bronchoalveolar lavage or transbronchial biopsy but they may be being considered for surgical lung biopsy	Consultant respiratory physician Consultant radiologist Consultant histopathologist Thoracic surgeon as appropriate Interstitial lung disease specialist nurse Multidisciplinary team coordinator
When considering results of bronchoalveolar lavage, transbronchial biopsy or surgical lung biopsy	Consultant respiratory physician Consultant radiologist Consultant histopathologist Interstitial lung disease specialist nurse Multidisciplinary team coordinator
See chapter 6.5 (Multidisciplinary Team) in full guideline for more information on the expertise of the multidisciplinary team.	

If a confident diagnosis cannot be made

1.2.4 If the multidisciplinary team cannot make a confident diagnosis from clinical features, lung function and radiological findings, consider:

- bronchoalveolar lavage or transbronchial biopsy and/or
- surgical lung biopsy, with the agreement of the thoracic surgeon.

Radiology

See NICE CG163 Idiopathic pulmonary fibrosis Recommendations 1.2.1, 1.2.2 and 1.2.3 above.

Finding a cause

See NICE CG163 Idiopathic pulmonary fibrosis Recommendation1.2.1 above regarding a detailed clinical history.

NICE CG163 Idiopathic pulmonary fibrosis Recommendations 1.2.4, 1.2.5 and 1.2.6

If a confident diagnosis cannot be made:

1.2.4 If the multidisciplinary team cannot make a confident diagnosis from clinical features, lung function and radiological findings, consider:

- bronchoalveolar lavage or transbronchial biopsy and/or
- surgical lung biopsy, with the agreement of the thoracic surgeon.

1.2.5 Discuss with the person who may have idiopathic pulmonary fibrosis:

- the potential benefits of having a confident diagnosis compared with the uncertainty of not having a confident diagnosis **and**
- the increased likelihood of obtaining a confident diagnosis with surgical biopsy compared with bronchoalveolar lavage or transbronchial biopsy **and**
- the increased risks of surgical biopsy compared with bronchoalveolar lavage or transbronchial biopsy.

1.2.6 When considering bronchoalveolar lavage, transbronchial biopsy or surgical lung biopsy take into account:

- the likely differential diagnoses **and**
- the person's clinical condition, including any comorbidities.

4.1.3 Current UK practice

Awareness of clinical features of IPF

No information on current levels of awareness was identified.

MDT

No data on current UK practice has been found. However, as recently as 2013 NHS England stated in the NHS standard contract for respiratory: interstitial lung disease (adult) that growing evidence points to the importance of combined multi-disciplinary team (MDT) input for assigning correct diagnoses and initiating appropriate therapy in individuals with ILD. Misdiagnosis contributes to increased morbidity and mortality in this patient group.

Radiology

No data on current UK practice has been found.

Finding a cause

No data on current UK practice has been found.

4.2 Information and support

Patients

Stakeholders suggested that providing patients with information at the appropriate times about their condition and how it should be managed leads to them feeling less isolated, less anxious about how their illness will progress, improves patient experience, symptoms, improve their wellbeing and access to social services.

Stakeholders commented information should be tailored to individuals and their needs and that an ILD specialist nurse should provide accurate information (verbal and written) at all stages.

Stakeholders suggested that patients may need support to get the best from medication, and that information and support will empower them to make informed decisions about possible treatments. Reference was made by a stakeholder to NICE clinical guideline CG76 on medicines adherence.

Carers

A stakeholder commented that carers should have an assessment of emotional, psychological and social needs and if needed receive tailored interventions identified by a care plan to address their needs.

Stakeholders suggested that services/support at time of diagnosis are important to carers.

A stakeholder commented that bureaucracy/lack of funding can mean service users/their family do not receive full/adequate information.

A stakeholder suggested an interstitial lung disease specialist nurse should provide accurate information (verbal and written) at all stages

4.2.1 Selected recommendations from development source

Table 4 presents recommendations that have been provisionally selected from the development sources which may support potential statement development. These are presented in full below to inform the Committee's discussion.

Table 4 Specific areas for quality improvement

Suggested quality improvement area	Selected source guidance recommendations
Patients	NICE CG163 Idiopathic pulmonary fibrosis 1.3.1, 1.3.3 and 1.3.4 (1.3.1 and 1.3.3 KPI)
Carers	NICE CG163 Idiopathic pulmonary fibrosis 1.3.1, 1.3.3

Patients

NICE CG163 Idiopathic pulmonary fibrosis Recommendations 1.3.1, 1.3.3 and 1.3.4 (1.3.1 & 1.3.3 key priorities for implementation)

1.3.1 The consultant respiratory physician or interstitial lung disease specialist nurse should provide accurate and clear information (verbal and written) to people with idiopathic pulmonary fibrosis, and their families and carers with the person's consent. This should include information about investigations, diagnosis and management.

1.3.3 An interstitial lung disease specialist nurse should be available at all stages of the care pathway to provide information and support to people with idiopathic pulmonary fibrosis and their families and carers with the person's consent.

1.3.4 Offer advice, support and treatment to aid smoking cessation to all people with idiopathic pulmonary fibrosis who also smoke, in line with Smoking cessation services (NICE public health guidance 10).

Carers

See NICE CG163 Idiopathic pulmonary fibrosis Recommendations 1.3.1 and 1.3.3 above.

4.2.2 Current UK practice

A study on palliative care for people with non-malignant lung disease stated that ILD patients and carers report a lack of information sharing, and although they may know that the disease is terminal, they have a poor understanding of prognosis or what may occur at end of life. [1] [2]

[1] Bawah S, Higginson IJ, Ross JR, et al. 'I wish I knew more…' – the end-of-life planning and information needs for end0stage fibrotic interstitial lung disease: views of patients, carers and health professionals. *BMJ Support Palliat Care* 2013; 3: 84-90

4.3 Management

Pulmonary Rehabilitation

Stakeholders suggested IPF pulmonary rehabilitation, tailored to the patient's need, should be considered because it can improve disease management and quality of life (improving exercise capacity and confidence, reducing dependency and social isolation).

A stakeholder commented access to pulmonary rehabilitation varies across the country and this is traditionally focused on COPD.

Best supportive care

- **Symptom control**

  A stakeholder commented that coughing can be very distressing and difficult to manage. Reflux is recognised as contributory factor to cough and disease progression and is an IPF outcome. Best supportive care for symptom control is needed.

  A stakeholder suggested smoking cessation services are needed to reduce disease progression.

- **Palliative care**

  Stakeholders suggested there is evidence patients are being referred to palliative care too late. Effective palliative care is needed at various stages. Palliative specialists can provide measures to alleviate symptoms and help plan end of life care to improve IPF patients' quality of life. Respiratory teams should liaise with palliative care teams. A stakeholder suggested access to palliative care is varied throughout the country and local planning protocols are needed for palliative care referral early in diagnosis and to discuss advanced care planning.

Disease modifying pharmacological interventions

A stakeholder suggested it would be useful to stipulate the validity of pulmonary lung function tests in relation to starting treatment. They stated that some audits show some patients have had delays in the pulmonary lung function tests qualifying them to start treatment and this may also affect the interpretation of repeat pulmonary lung function tests used to decide discontinuation of treatment.

[2] Bawah S, Higginson IJ, Ross JR, et al. 'When it's really bad, I'd make a trade with the devil…'- the specialist palliative care needs for end-stage fibrotic interstitial lung disease: views of patients, carers and health professionals. *Palliat Med*, in press.

A stakeholder commented that timely initiation of treatment optimises the period within which effective treatment can improve outcomes therefore referral, diagnosis and initiation of treatment needs to be done promptly.

Stakeholders commented there are no drugs which can cure IPF but that drug treatment can help in the management of the condition.

A stakeholder suggested close monitoring of side effects and efficacy of novel agents including immunosuppressants through specialist nursing teams is needed.

Lung transplantation

Stakeholders suggested that patients are not being referred for transplantation early enough (they can become too old/ill for transplantation to be considered) and that many patients are not referred within 3-6 months as stated in the NICE CG163 idiopathic pulmonary fibrosis.

Stakeholders commented that the referral needs to be done early so assessment can take place at the appropriate stage of progression.

Oxygen assessment

Stakeholders suggested that patients often do not receive oxygen for exercise because they do not desaturate on a 6-minute walk test, but that they have problems on exertion. Quality of life and level of independence is significantly improved by access to the appropriate levels/type of ambulatory oxygen.

A stakeholder commented that the quality of assessment for ambulatory oxygen is variable leading to patients having delayed/inappropriate assessment. This can be focused on COPD.

A stakeholder suggested that, to avoid waste, patients need to be individually assessed prior to the oxygen being supplied.

4.3.1 Selected recommendations from development source

Table 5 presents recommendations that have been provisionally selected from the development sources which may support potential statement development. These are presented in full below to inform the Committee’s discussion.

Table 5 Specific areas for quality improvement

Suggested quality improvement area	Selected source guidance recommendations
Pulmonary rehabilitation	NICE CG163 Idiopathic pulmonary fibrosis 1.5.1, 1.5.2, 1.5.3, 1.5.4 (1.5.1 KPI) BTS Guideline on Pulmonary rehabilitation in adults (ii3)
Best supportive care • Symptom control	NICE CG163 Idiopathic pulmonary fibrosis 1.5.5, 1.5.6, 1.5.7, 1.5.8, 1.5.9, 1.6.1, 1.6.2 (1.5.5, 1.5.6, 1.6.1 KPI)
Best supportive care • Palliative care	NICE CG163 Idiopathic pulmonary fibrosis 1.5.5, 1.5.7 & 1.5.10 access to full range of services offered by palliative care teams and collaboration (1.5 end of life care/ referral to palliative care & 1.6.1 consider referral to palliative care)
Disease-modifying pharmacological interventions	NICE CG163 Idiopathic pulmonary fibrosis 1.5.11, 1.5.12, 1.5.13, 1.5.14, 1.5.15 (1.5.11 and 1.5.12 KPI) Does not specify the information highlighted by the stakeholders
Lung transplantation	NICE CG163 Idiopathic pulmonary fibrosis 1.5.16, 1.5.17 (1.5.17 KPI)
Oxygen assessment	NICE CG163 Idiopathic pulmonary fibrosis 1.5.6, 1.5.7, 1.5.8

Pulmonary rehabilitation

NICE CG163 Idiopathic pulmonary fibrosis – Recommendations 1.5.1, 1.5.2, 1.5.3, 1.5.4 (key priority for implementation 1.5.1)

1.5.1 Assess people with idiopathic pulmonary fibrosis for pulmonary rehabilitation at the time of diagnosis. Assessment may include a 6-minute walk test (distance walked and oxygen saturation measured by pulse oximetry) and a quality-of life assessment.

1.5.2 Repeat the assessment for pulmonary rehabilitation for people with idiopathic pulmonary fibrosis at 6-month or 12-month intervals.

1.5.3 If appropriate after each assessment, offer pulmonary rehabilitation including exercise and educational components tailored to the needs of people with idiopathic pulmonary fibrosis in general.

1.5.4 Pulmonary rehabilitation should be tailored to the individual needs of each person with idiopathic pulmonary fibrosis. Sessions should be held somewhere that is easy for people with idiopathic pulmonary fibrosis to get to and has good access for people with disabilities.

BTS Guideline on Pulmonary rehabilitation in adults (ii3)

Pulmonary rehabilitation in people with other chronic respiratory diseases

Interstitial lung diseases

Good practice points

- The benefits of exercise and the recommendation of incorporating exercise activities into a healthy lifestyle should be discussed with all patients with interstitial lung disease (ILD). Such discussion needs to be tailored to realistic achievability for that person's condition.

- If healthcare professionals consider referring certain patients with stable ILD who are limited by breathlessness in ADL to pulmonary rehabilitation, they should discuss with the patient the likely benefits.

- Patients with idiopathic pulmonary fibrosis (IPF) have a potential for significant desaturation during exercise related activities.

Best supportive care

- **Symptom control**

NICE CG163 Idiopathic pulmonary fibrosis – Recommendations 1.5.5, 1.5.6, 1.5.7, 1.5.8, 1.5.9 (key priorities for implementation: 1.5.5 & 1.5.6)

1.5.5 Offer best supportive care to people with idiopathic pulmonary fibrosis from the point of diagnosis. Best supportive care should be tailored to disease severity, rate of progression, and the person's preference, and should include if appropriate:

- information and support (see recommendation 1.3.1)

- symptom relief

- management of comorbidities

- withdrawal of therapies suspected to be ineffective or causing harm

- end of life care.

1.5.6 If the person is breathless on exertion consider assessment for:

- the causes of breathlessness and degree of hypoxia **and**
- ambulatory oxygen therapy and long-term oxygen therapy **and/or**
- pulmonary rehabilitation.

1.5.7 If the person is breathless at rest consider:

- assessment for the causes of breathlessness and degree of hypoxia **and**
- assessment for additional ambulatory oxygen therapy and long-term oxygen therapy **and**
- the person's psychosocial needs and offering referral to relevant services such as palliative care services **and**
- pharmacological symptom relief with benzodiazepines and/or opioids.

1.5.8 Assess the oxygen needs of people who have been hospitalised with idiopathic pulmonary fibrosis before they are discharged.

1.5.9 If the person has a cough consider:

- treatment for causes other than idiopathic pulmonary fibrosis (such as gastrooesophageal reflux disease, post-nasal drip)
- treating with opioids if the cough is debilitating
- discussing treatment with thalidomide [footnote in guideline states ‘At the time of publication (June 2013), thalidomide did not have a UK marketing authorisation for this indication [etc]’] with a consultant respiratory physician with expertise in interstitial lung disease if the cough is intractable.

<u>NICE CG163 Idiopathic pulmonary fibrosis – Recommendations 1.6.1 and 1.6.2 (key priority for implementation 1.6.1)</u>

1.6.1 In follow-up appointments for people with idiopathic pulmonary fibrosis:

- assess lung function
- assess for oxygen therapy
- assess for pulmonary rehabilitation

- offer smoking cessation advice, in line with Smoking cessation services (NICE public health guidance 10)
- identify exacerbations and previous respiratory hospital admissions
- consider referral for assessment for lung transplantation in people who do not have absolute contraindications (see recommendations 1.5.16 and 1.5.17)
- consider psychosocial needs and referral to relevant services as appropriate
- consider referral to palliative care services
- assess for comorbidities (which may include anxiety, bronchiectasis, depression, diabetes, dyspepsia, ischaemic heart disease, lung cancer and pulmonary hypertension).

1.6.2 Consider follow-up of people with idiopathic pulmonary fibrosis:

- every 3 months or sooner if they are showing rapid disease progression or rapid deterioration of symptoms **or**
- every 6 months or sooner if they have steadily progressing disease **or**
- initially every 6 months if they have stable disease and then annually if they have stable disease after 1 year.

Best supportive care

- **Palliative care**

See recommendations 1.5.5 & 1.5.7 and 1.6.1 above

NICE CG163 Idiopathic pulmonary fibrosis – Recommendation 1.5.10

1.5.10 Ensure people with idiopathic pulmonary fibrosis, and their families and carers have access to the full range of services offered by palliative care teams. Ensure there is collaboration between the healthcare professionals involved in the person's care, community services and the palliative care team.

Disease-modifying pharmacological interventions

NICE CG163 states: ‘There is no conclusive evidence to support the use of any drugs to increase the survival of people with idiopathic pulmonary fibrosis.’ CG163 contains the following recommendations on disease-modifying pharmacological interventions:

NICE CG163 Idiopathic pulmonary fibrosis – Recommendations 1.5.11, 1.5.12, 1.5.13, 1.5.14, 1.5.15 (key priorities for implementation 1.5.11 & 1.5.12)

1.5.11 For guidance on pirfenidone for the management of idiopathic pulmonary fibrosis, refer to Pirfenidone for the treatment of idiopathic pulmonary fibrosis (NICE technology appraisal guidance 282).

1.5.12 Do not use any of the drugs below, either alone or in combination, to modify disease progression in idiopathic pulmonary fibrosis:

- ambrisentan
- azathioprine
- bosentan
- co-trimoxazole
- mycophenolate mofetil
- prednisolone
- sildenafil
- warfarin.

1.5.13 Advise the person that oral N-acetylcysteine[2] is used for managing idiopathic pulmonary fibrosis, but its benefits are uncertain.

1.5.14 If people with idiopathic pulmonary fibrosis are already using prednisolone or azathioprine, discuss the potential risks and benefits of discontinuing, continuing or altering therapy.

1.5.15 Manage any comorbidities according to best practice. For gastro-oesophageal reflux disease, see Managing dyspepsia in adults in primary care (NICE clinical guideline 17).

Lung transplantation

NICE CG163 Idiopathic pulmonary fibrosis – Recommendation 1.5.16 &1.5.17 (key priority for implementation 1.5.17)

1.5.16 Discuss lung transplantation as a treatment option for people with idiopathic pulmonary fibrosis who do not have absolute contraindications. Discussions should:

- take place between 3 and 6 months after diagnosis or sooner if clinically indicated
- be supported by an interstitial lung disease specialist nurse
- include the risks and benefits of lung transplantation

- involve the person's family and carers with the person's consent.

(See recommendations 1.5.5 – 1.5.10 about best supportive care.)

1.5.17 Refer people with idiopathic pulmonary fibrosis for lung transplantation assessment if they wish to explore lung transplantation and if there are no absolute contraindications. Ask the transplant centre for an initial response within 4 weeks.

Oxygen assessment

NICE CG163 Idiopathic pulmonary fibrosis – see recommendations 1.5.6, 1.5.7, 1.5.8 above

4.3.2 Current UK practice

Pulmonary rehabilitation

No data on current practice could be found.

Best supportive care

- **Symptom control**

  No data on current practice could be found.

- **Palliative care**

A study on palliative care for people with non-malignant lung disease found that despite the poor prognosis and symptom burden of non-malignant lung disease, there is inequitable access to specialist palliative care services, often with no formal process for identifying patients at EOL. [3]

Disease-modifying pharmacological interventions

No data on current practice could be found.

Lung transplantation

Pulmonary Fibrosis: rate of disease progression as a trigger for referral for lung transplantation

This was a single centre retrospective review of patients with pulmonary fibrosis who were assessed for lung transplantation over a 5 year period between 1999 – 2004 and was published in 2007.

[3] Partridge MR, Khatri A, Sutton L, et al. Palliative care services for those with chronic lung disease. *Chron respire Dis* 2009; 6(1): 13-17

Between March 1999 and March 2004, 129 patients with pulmonary fibrosis underwent formal transplant assessment. Sixty-nine were accepted and listed for lung transplantation. Of these, 17 were transplanted, 37 died while waiting, 4 were removed from the list and 11 were still waiting at the conclusion of the study.

The study concluded that:

- The rate of disease progression appears to be a more sensitive indicator for transplantation referral than any single physiological measure of disease severity and should act as an important trigger for early transplant referral.
- Lung transplantation is the only treatment modality that provides an actuarial survival advantage in this population.
- The progressive nature of this disease and the short interval between diagnosis and death make this therapeutic option available only to a limited number of younger patients.
- Patients accepted onto the active waiting list will wait on average 12–18 months for a suitable donor organ in the UK.
- Patients with pulmonary fibrosis have the highest waiting list mortality of all patients awaiting lung transplantation
- Timely assessment is important in all patients under consideration for lung transplantation, none more so than those with pulmonary fibrosis in whom the window of opportunity for transplant may be as little as 22 months.
- Referral criteria suggest that all patients under the age of 65 years who are symptomatic and have failed to respond to steroid and immunosuppressive therapy should be considered for transplantation.

Oxygen assessment

NHS Lung Improvement (2013) Improving the quality and safety of home oxygen services: the case for spread

Home oxygen therapy is provided to about 85,000 people in England, costing approximately £110 million a year. Home oxygen service assessment and review (HOS-AR) is variable as patients in many local areas do not receive a quality assured clinical assessment and a review of their ongoing need for long term home oxygen.

The variation in provision of HOS-AR increases the potential for poor quality care and waste and it has been estimated that 24% to 43% of home oxygen prescribed in England is not used or provides no clinical benefit.

4.4 Review and follow up

4.4.1 Summary of suggestions

Clear follow up regimens

A stakeholder commented that adequate medical resource with clear follow up regimens is needed.

4.4.2 Selected recommendations from development source

Table 6 presents recommendations that have been provisionally selected from the development sources which may support potential statement development. These are presented in full below to inform the Committee's discussion.

Table 6 Specific areas for quality improvement

Suggested quality improvement area	Selected source guidance recommendations
Clear follow up regimens	NICE CG163 Idiopathic pulmonary fibrosis 1.6.1 and 1.6.2

Clear follow up regimens

NICE CG163 Idiopathic pulmonary fibrosis – Recommendations 1.6.1 and 1.6.2

4.4.3 Current UK practice

No data on current UK practice has been found.

4.5 Additional Areas

4.5.1 Summary of suggestions

The improvement areas below were suggested as part of the stakeholder engagement exercise. However these were felt either to be outside the remit of the quality standard referral and the development source (NICE guidance) or require further discussion by the Committee to establish potential for statement development.

There will be an opportunity for the QSAC to discuss these areas at the end of the session on 19 May 2014.

Data collection

A stakeholder suggested that detailed and accurate data collection with submission of data to central databases (national registry) will assist in understanding populations, effects of treatment etc. This may lead to increased study and inform public health policy and national guidelines.

British Thoracic Society (ongoing audit) BTS Lung Disease Registry Programme – idiopathic pulmonary fibrosis.

NICE clinical audit tool – Idiopathic pulmonary fibrosis

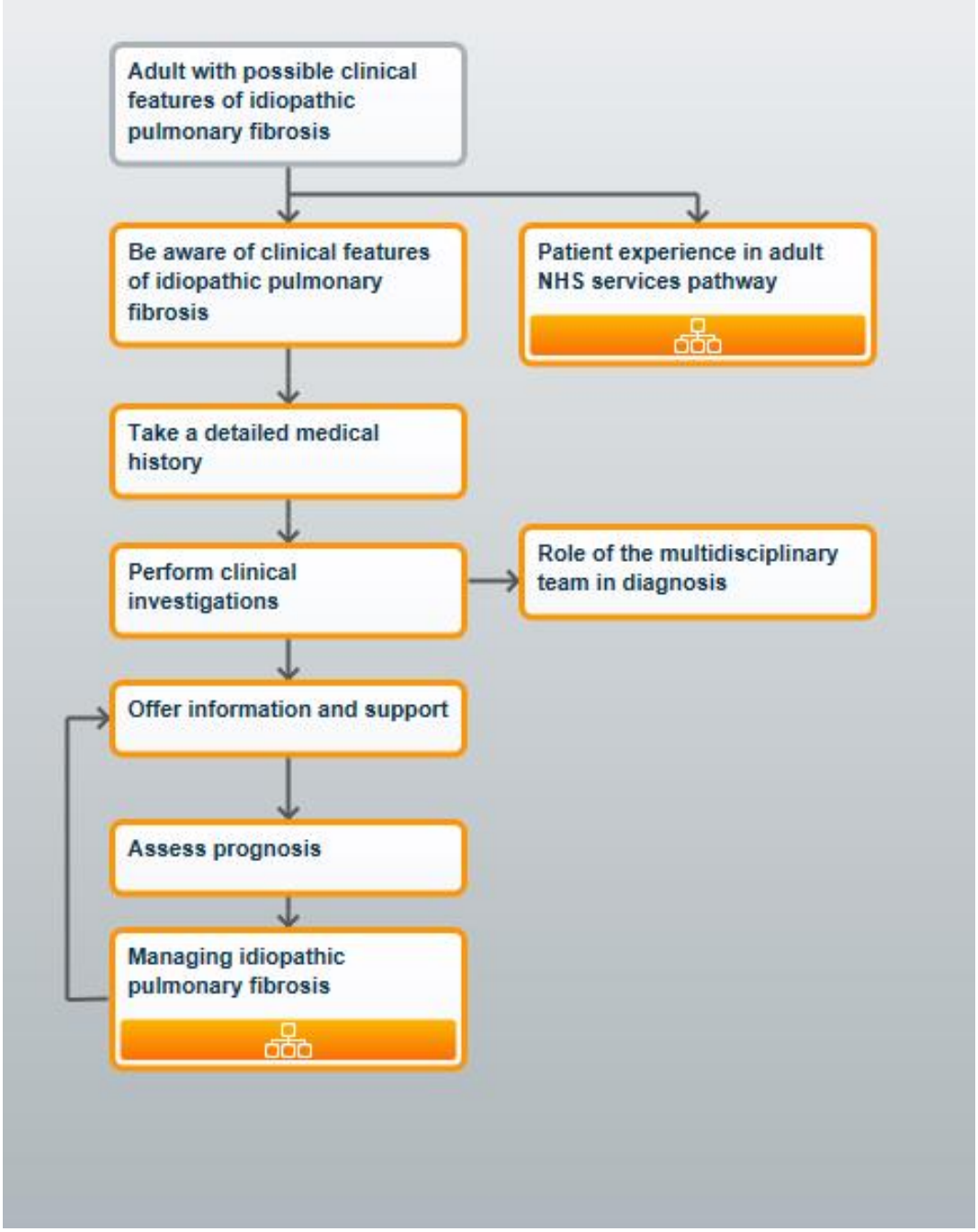
Clinical trials

A stakeholder commented it is necessary to optimise registry entry for consideration of clinical trials.

A stakeholder suggested the most appropriate treatment needs to be given even if this is through a clinical trial.

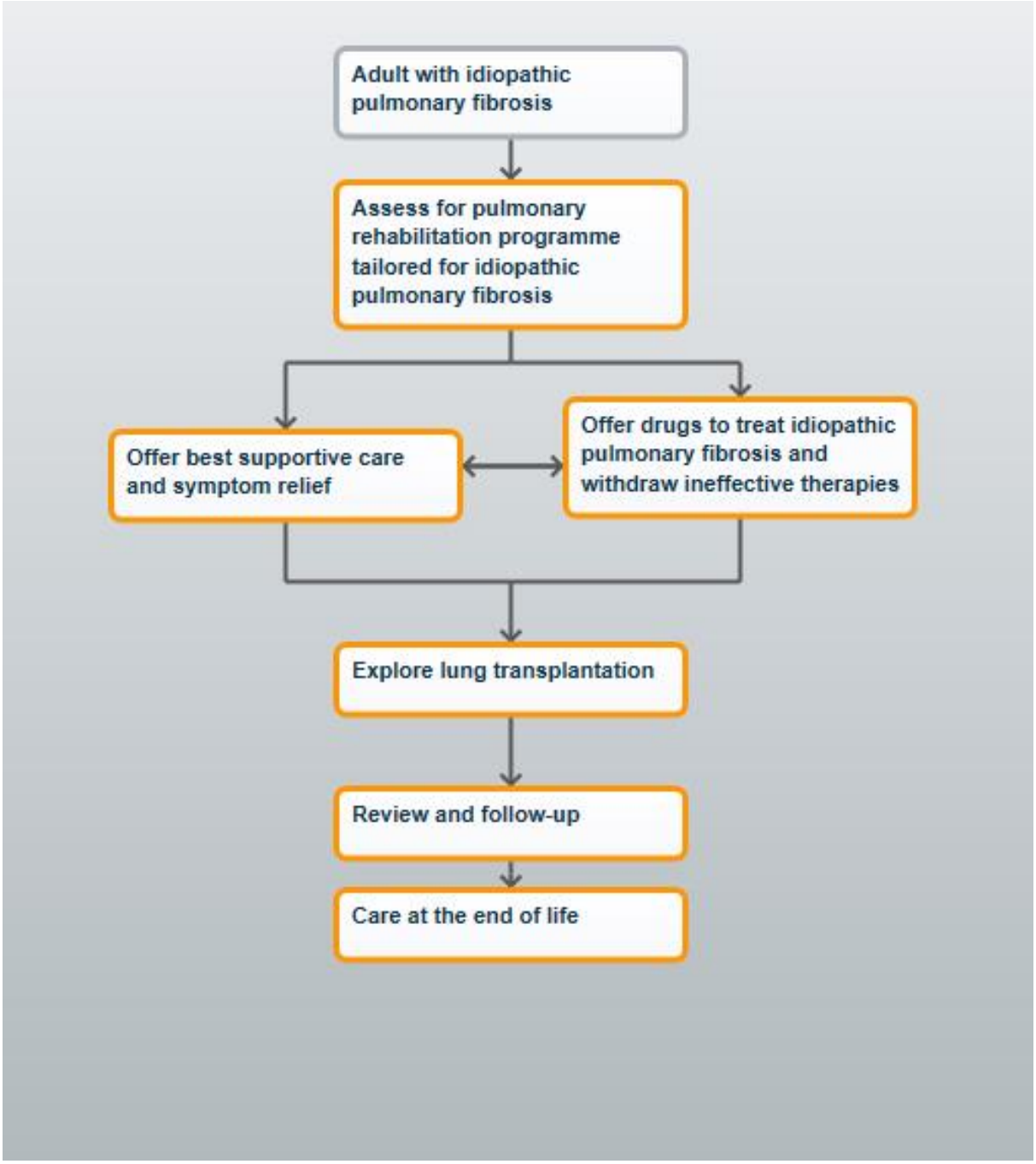
Appendix 1: Additional information

Idiopathic pulmonary fibrosis overview



http://pathways.nice.org.uk/pathways/idiopathic-pulmonary-fibrosis

Managing idiopathic fibrosis



http://pathways.nice.org.uk/pathways/idiopathic-pulmonary-fibrosis

Appendix 2: Key priorities for implementation (NICE CG163 Idiopathic pulmonary fibrosis)

Recommendations that are key priorities for implementation in the source guideline and which have been referred to in the main body of this report are highlighted in grey.

Awareness of clinical features of idiopathic pulmonary fibrosis

- Be aware of idiopathic pulmonary fibrosis when assessing a patient with the clinical features listed below and when considering requesting a chest X-ray or referring to a specialist:
    - age over 45 years
    - persistent breathlessness on exertion
    - persistent cough
    - bilateral inspiratory crackles when listening to the chest
    - clubbing of the fingers
    - normal spirometry or impaired spirometry usually with a restrictive pattern but sometimes with an obstructive pattern.

(recommendation 1.1.1)

Diagnosis

Diagnose idiopathic pulmonary fibrosis only with the consensus of the multidisciplinary team (listed in table 1), based on:

- the clinical features, lung function and radiological findings (see recommendation 1.2.1)
- pathology when indicated (see recommendation 1.2.4).

(Recommendation 1.2.2)

At each stage of the diagnostic care pathway the multidisciplinary team should consist of a minimum of the healthcare professionals listed in table 1, all of whom should have expertise in interstitial lung disease. (Recommendation 1.2.3)

Table 1 Minimum composition of multidisciplinary team involved in diagnosing idiopathic pulmonary fibrosis

Stage of diagnostic care pathway	Multidisciplinary team composition (all healthcare professionals should have expertise in interstitial lung disease)
After clinical evaluation, baseline lung function and CT	Consultant respiratory physician Consultant radiologist Interstitial lung disease specialist nurse Multidisciplinary team coordinator
When considering performing bronchoalveolar lavage, and/or transbronchial biopsy or surgical lung biopsy Only some patients will have bronchoalveolar lavage or transbronchial biopsy but they may be being considered for surgical lung biopsy	Consultant respiratory physician Consultant radiologist Consultant histopathologist Thoracic surgeon as appropriate Interstitial lung disease specialist nurse Multidisciplinary team coordinator
When considering results of bronchoalveolar lavage, transbronchial biopsy or surgical lung biopsy	Consultant respiratory physician Consultant radiologist Consultant histopathologist Interstitial lung disease specialist nurse Multidisciplinary team coordinator
See chapter 6.5 (Multidisciplinary Team) in full guideline for more information on the expertise of the multidisciplinary team.	

Information and Support

The consultant respiratory physician or interstitial lung disease specialist nurse should provide accurate and clear information (verbal and written) to people with idiopathic pulmonary fibrosis, and their families and carers with the person's consent. This should include information about investigations, diagnosis and management. (Recommendation 1.3.1)

NICE has produced guidance on the components of good patient experience in adult NHS services. Follow the recommendations in Patient experience in adult NHS services (NICE clinical guideline 138). (Recommendation 1.3.2)

An interstitial lung disease specialist nurse should be available at all stages of the care pathway to provide information and support to people with idiopathic pulmonary fibrosis and their families and carers with the person's consent. (Recommendation 1.3.3)

Offer advice, support and treatment to aid smoking cessation to all people with idiopathic pulmonary fibrosis who also smoke, in line with Smoking cessation services (NICE public health guidance 10). (Recommendation 1.3.4)

Pulmonary rehabilitation

Assess people with idiopathic pulmonary fibrosis for pulmonary rehabilitation at the time of diagnosis. Assessment may include a 6-minute walk test (distance walked and oxygen saturation measured by pulse oximetry) and a quality-of life assessment. (Recommendation 1.5.1)

Repeat the assessment for pulmonary rehabilitation for people with idiopathic pulmonary fibrosis at 6-month or 12-month intervals. (Recommendation 1.5.2)

If appropriate after each assessment, offer pulmonary rehabilitation including exercise and educational components tailored to the needs of people with idiopathic pulmonary fibrosis in general. (Recommendation 1.5.3)

Pulmonary rehabilitation should be tailored to the individual needs of each person with idiopathic pulmonary fibrosis. Sessions should be held somewhere that is easy for people with idiopathic pulmonary fibrosis to get to and has good access for people with disabilities. (Recommendation 1.5.4)

Best supportive care

Offer best supportive care to people with idiopathic pulmonary fibrosis from the point of diagnosis. Best supportive care should be tailored to disease severity, rate of progression, and the person's preference, and should include if appropriate:

- information and support (see recommendation 1.3.1)
- symptom relief
- management of comorbidities
- withdrawal of therapies suspected to be ineffective or causing harm
- end of life care.

(Recommendation 1.5.5)

If the person is breathless on exertion consider assessment for:

- the causes of breathlessness and degree of hypoxia **and**
- ambulatory oxygen therapy and long-term oxygen therapy **and/or**
- pulmonary rehabilitation.

(Recommendation 1.5.6)

If the person is breathless at rest consider:

- assessment for the causes of breathlessness and degree of hypoxia **and**
- assessment for additional ambulatory oxygen therapy and long-term oxygen therapy **and**
- the person's psychosocial needs and offering referral to relevant services such as palliative care services **and**
- pharmacological symptom relief with benzodiazepines and/or opioids.

(Recommendation 1.5.7)

Assess the oxygen needs of people who have been hospitalised with idiopathic pulmonary fibrosis before they are discharged. (Recommendation 1.5.8)

If the person has a cough consider:

- treatment for causes other than idiopathic pulmonary fibrosis (such as gastrooesophageal reflux disease, post-nasal drip)
- treating with opioids if the cough is debilitating
- discussing treatment with thalidomide[1] with a consultant respiratory physician with expertise in interstitial lung disease if the cough is intractable.

(Recommendation 1.5.9)

Ensure people with idiopathic pulmonary fibrosis, and their families and carers have access to the full range of services offered by palliative care teams.

Ensure there is collaboration between the healthcare professionals involved in the person's care, community services and the palliative care team. (Recommendation 1.5.10)

Disease-modifying pharmacological interventions

For guidance on pirfenidone for the management of idiopathic pulmonary fibrosis, refer to Pirfenidone for the treatment of idiopathic pulmonary fibrosis (NICE technology appraisal guidance 282). (recommendation 1.5.11)

Do not use any of the drugs below, either alone or in combination, to modify disease progression in idiopathic pulmonary fibrosis:

- ambrisentan
- azathioprine
- bosentan
- co-trimoxazole
- mycophenolate mofetil
- prednisolone

- sildenafil
- warfarin.

(Recommendation 1.5.12)

Advise the person that oral N-acetylcysteine[2] is used for managing idiopathic pulmonary fibrosis, but its benefits are uncertain. (Recommendation 1.5.13)

If people with idiopathic pulmonary fibrosis are already using prednisolone or azathioprine, discuss the potential risks and benefits of discontinuing, continuing or altering therapy. (Recommendation 1.5.14)

Manage any comorbidities according to best practice. For gastro-oesophageal reflux disease, see Managing dyspepsia in adults in primary care (NICE clinical guideline 17). (Recommendation 1.5.15)

Lung transplantation

Discuss lung transplantation as a treatment option for people with idiopathic pulmonary fibrosis who do not have absolute contraindications. Discussions should:

- take place between 3 and 6 months after diagnosis or sooner if clinically indicated
- be supported by an interstitial lung disease specialist nurse
- include the risks and benefits of lung transplantation
- involve the person's family and carers with the person's consent.

(See recommendations 1.5.5 – 1.5.10 about best supportive care.) (Recommendation 1.5.16)

Refer people with idiopathic pulmonary fibrosis for lung transplantation assessment if they wish to explore lung transplantation and if there are no absolute contraindications. Ask the transplant centre for an initial response within 4 weeks. (Recommendation 1.5.17)

Review and follow-up

In follow-up appointments for people with idiopathic pulmonary fibrosis:

- assess lung function
- assess for oxygen therapy
- assess for pulmonary rehabilitation

- offer smoking cessation advice, in line with Smoking cessation services (NICE public health guidance 10)
- identify exacerbations and previous respiratory hospital admissions
- consider referral for assessment for lung transplantation in people who do not have absolute contraindications (see recommendations 1.5.16 and 1.5.17)
- consider psychosocial needs and referral to relevant services as appropriate
- consider referral to palliative care services
- assess for comorbidities (which may include anxiety, bronchiectasis, depression, diabetes, dyspepsia, ischaemic heart disease, lung cancer and pulmonary hypertension).

(Recommendation 1.6.1)

Consider follow-up of people with idiopathic pulmonary fibrosis:

- every 3 months or sooner if they are showing rapid disease progression or rapid deterioration of symptoms **or**
- every 6 months or sooner if they have steadily progressing disease **or**
- initially every 6 months if they have stable disease and then annually if they have stable disease after 1 year.

(Recommendation 1.6.2)

Appendix 3: Suggestions from stakeholder engagement exercise

ID	Stakeholder	Suggested key area for quality improvement	Why is this important?	Why is this a key area for quality improvement?	Supporting information
4.1	SCM2	Key area for quality improvement 1 There should be a higher level of awareness of IPF and its symptoms amongst the public and Primary Care	IPF is a rapidly fatal disease where early intervention may delay disease progression and improve quality of life	The incidence of this condition is rising. There is poor general knowledge of the condition. There are treatments that are now licenced in mild to moderate disease that patients may benefit from if they are diagnosed.	GribbinThorax. 2006;61(11):980-5 Vancheri ERJ 2010 35(3):496-504
4.1	SCM 4	Increase knowledge of disease in primary and non-specialist secondary care	Early diagnosis early referral		
4.1	SCM1	Key area for quality improvement 1	Diagnosis of IPF	Too many patients are being told they have IPF before tests are complete, and sometimes by very inappropriate people such as GPs.	
4.1	SCM2	Key area for quality improvement 2 People who may have Idiopathic Pulmonary Fibrosis are discussed at a specialist Interstitial Lung Disease MDT	We know that the diagnosis of IPF can be difficult. A multidisciplinary consensus diagnosis has been shown to improve the diagnostic accuracy of IPF. In addition access to multimodality treatment (eg medications, rehabilitation, palliative care, transplant) can be expedited.	ILD MDT's have been shown to improve the accuracy of IPF diagnosis. In addition the MDT can provide a pathway for patients to access diagnostic tests and treatments. Many patients with IPF are not discussed at an ILD MDT	Lamas DJ. Am J RespCrit Care M 2011 184 (7) 842-847 Thorax 2008:63 Supp 4 Flaherty KR et al AJRCCM 2004;170:904-10 AJRCCM2011:183;788-824
4.1	Boehringer Ingelheim	Referral to specialist for diagnosis of IPF	IPF can be hard to diagnose because its main symptoms are similar to those of other lung conditions. Diagnosis of IPF should be confirmed only with the consensus of a multi-disciplinary group	Patients feel physicians do not truly understand IPF and are too quick to blame symptoms on smoking or COPD. They feel frustrated by primary care physicians not being able to diagnose IPF and that a diagnosis of COPD is often given	http://www.nhs.uk/Conditions/pulmonary-fibrosis/Pages/Diagnosis.aspx Accessed March 24th 2014 http://www.blf.org.uk/News/Detail/BLF-launches-new-charter-for-people-affected-by-IPF BLF IPF Patient Charter, Accessed

ID	Stakeholder	Suggested key area for quality improvement	Why is this important?	Why is this a key area for quality improvement?	Supporting information
				without proper evaluation of their condition. A trigger for diagnosis to specialist care is often the presence of auscultating lung crackles. Diagnosis of IPF is currently variable depending on where patients live and depending on the knowledge of the diagnosing clinician. Radiologists nationally have a two week referral guidance on their report forms for urgent referral to a lung cancer MDT. If something similar existed for IPF it would shorten the time taken for patients to be seen by an IPF MDT and could improve the standard of patient care in terms of diagnosis confirmation and appropriate treatment.	March 24th 2014 NICE Guideline CG163
4.1	British Lung Foundation	**Early and accurate diagnosis involving prompt referral through agreed pathways**	Due to the speed and nature of the disease, the quick and accurate diagnosis of IPF by referral to a multi-disciplinary team at an early stage has a significant impact on patient outcomes.	Early diagnosis means earlier treatment and support; however, patients and clinicians inform us that opportunities for diagnosis are frequently missed. Furthermore, IPF is sometimes misdiagnosed, and as a result patients can be given the wrong treatments.	"By the time diagnosis is made, average survival is little better than that for inoperable lung cancer." Katerina M Antoniou et al. 'Early diagnosis of IPF: time for a primary-care case-finding initiative?' The Lancet Respiratory Medicine, January 2014
4.1	InterMune	Timely referral to, and accurate diagnosis by a specialist multi-disciplinary team	Patients with idiopathic pulmonary fibrosis (IPF) should receive timely and accurate diagnosis and care, involving an appropriately skilled, specialist multidisciplinary team (British Lung Foundation IPF Patient Charter).	The median survival from diagnosis of IPF is about three years, a prognosis which is worse than many cancers. In addition, every IPF patient has a different unpredictable rate of decline so it is not possible to predict whether patients will have a	NICE clinical guideline 163: Idiopathic pulmonary fibrosis BLF 'IPF Patient Charter'. http://www.blf.org.uk/News/Detail/BLF-launches-new-charter-for-people-affected-by-IPF

ID	Stakeholder	Suggested key area for quality improvement	Why is this important?	Why is this a key area for quality improvement?	Supporting information
			NICE CG163 recognises that IPF is a difficult disease to diagnose and whilst GPs are recommended to assess people complaining of a persistent cough, breathlessness and respiratory cackles for IPF to speed up treatment and improve each patient's life (NICE Press Release NICE CG163) it is also important for them to refer suspect patients promptly to a specialist centre. Specialist centres offer the collaborative expertise of a consultant respiratory physician, radiologist and histopathologist often required to reach a consensus diagnosis of IPF.	slow or fast decline (Kim, Raghu). Therefore the earlier the diagnosis the better it is for the patient in terms of prognosis and quality of life. Delayed referral to a specialist centre decreases the survival time (Lamas).	Kim DS et al. Proc Am Thorac Soc 2006; 3:285-292 Raghu G et al. Am J Respir Crtit Care Med 2011; 183:788-824 Lamas D et al. Am J Respir Crit Care Med 2011; 184:842–847
4.1	Association Respiratory Nurse Specialists /Royal Brompton Hospital & Harefield NHS Trust	Key area for quality improvement 1 Diagnosing IPF	Accurately diagnosing IPF is essential to enable access to optimal therapeutic interventions in a timely manner	NHS England recognises that the relative rarity of ILDs means that diagnosis is difficult – input from a specialist multi disciplinary team is often required. Having established diagnosis highly specialised treatment may be indicated which requires administration and sometimes ongoing close monitoring by experts in a dedicated specialist centre	Please see NHS England/A14/s/c – this NHS Commissioning Board Concensus statement recognises that to deliver a high quality service patients with IPF need to have access to a specialist centre. The ATS / ERS / JRS /ALAT statement on evidence based guidelines for IPF state that the accuracy of diagnosis of IPF increases with specialist muti disciplinary team (MDT) discussion http://www.thoracic.org/statements/resources/interstitial-lung-disease/ipf0311.pdf

ID	Stakeholder	Suggested key area for quality improvement	Why is this important?	Why is this a key area for quality improvement?	Supporting information
4.1	Association Respiratory Nurse Specialists /Royal Brompton Hospital & Harefield NHS Trust	Key area for quality improvement 5 Best supportive care	There is no curative treatment for IPF other than lung transplantation for which many patients are not eligible. Many patients have rapidly progressive disease with a prognosis worse than many cancers yet they do not have access to the same support infrastructure as do the users of cancer services	NICE clinical guideline 138 recognises that there are essential components of good patient experience in Adult NHS services. There are a paucity of IPF nurse specialists in the UK yet they play a pivotal role in providing clear and accurate information both verbal and written to people diagnosed with IPF and their families. NICE guidance recognises that a disease specific specialist nurse should be there at each stage of the patient pathway to provide ongoing information and support and re evaluating care needs to enable appropriate and timely input from the interdisciplinary team	Please see NICE clinical guidance 163 which highlights the need for information and support, symptom relief; management of comorbidities and prompt withdrawal of therapies suspected to be ineffective or causing harm. The guideline also highlights the importance of end of life care for patients with IPF. Given the increasing number of referral to ILD specialist centres there is an urgent need to develop the role of the clinical nurse specialist and support workers as an integral part of the MDT.
4.1	Action for Pulmonary Fibrosis	Timely and Accurate Diagnosis	The limited treatments available to patients diagnosed with IPF are prescribed depending on the level of progress of the disease, so it is vital that a correct diagnosis is made as soon as possible after the patient presents with the symptoms.	Many patients are incorrectly diagnosed with less serious diseases prior to the correct diagnosis being made, thus delaying the opportunity for appropriate management of the disease.	Reports from individual patients and patient groups across the country indicate that many patients face delays in treatment caused by incorrect initial diagnosis.
4.1	SCM4	Defined criteria for ILD MDT composition to ensure adequate specialist qualification	Establish accurate diagnosis at an early stage		
4.1	SCM3	**Access to or links with regional/national radiology experts or 'panels'** with	The differentiation between UIP/IPF and non-UIP/IPF fibrosing lung disease is a key diagnostic step in the management of fibrosing interstitial lung diseases; an accurate	Local experience and expertise in the radiological (CT) diagnosis of IPF/UIP is subject to variability and depends on the experience of the radiologist. Access to or links with	1) ATS/ERS Statement: update of the International Multidisciplinary Classification of the Idiopathic Interstitial Pneumonia. Am J Respir Crit Care Med 2013;188:733-748

ID	Stakeholder	Suggested key area for quality improvement	Why is this important?	Why is this a key area for quality improvement?	Supporting information
		subspeciality training in the imaging of ILD. NB Radiologists in expert panels must be able to demonstrate that they have i) subspeciality training and ii) regularly report CT studies in patients with ILD (e.g. as part of an ILD MDM team)	diagnosis of IPF/UIP is of clinical/therapeutic and prognostic significance. HRCT has an important role in diagnosis but 30-50% of cases have 'atypical' appearances or are discordant with clinical/pathological features.	regional/national radiology experts in ILD is likely to be of significant benefit.	2) Raghu et al. Am J Respir Crit Care Med. 2011;183:788-824 3) Thomeer et al. Eur Respir J. 2008;31:585-591 4) Flaherty KR et al. AmJ Respir Crit Care Med. 2007;175:1054-1060
4.1	SCM3	**Standardisation of CT protocols/techniques in the diagnosis and/or follow-up of IPF** (e.g. thin-section [1-2 mm], reconstructed with a high-spatial-frequency bone algorithm, in full suspended inspiration and without iv contrast injection.	Perceived abnormalities on CT in patients with IPF (specifically, increased ground-glass opacification) are, not infrequently, related to technical factors (i.e. poor inspiratory effort, injection of iv contrast) and may not indicate disease progression.	Inappropriate management decisions may be made purely on the basis of 'faulty' imaging!	
4.1	SCM3	**Indications for imaging (chest x-ray, CT) in specific scenarios** in patients with suspected or an established diagnosis of IPF	Guidance on when (and when not) to undertake specific radiological tests is crucial in the management of patients with suspected or established IPF	Guidance on appropriate use of imaging tests will benefit management decisions and will have an impact on keeping the radiation burden as low as reasonably achievable.	

ID	Stakeholder	Suggested key area for quality improvement	Why is this important?	Why is this a key area for quality improvement?	Supporting information
		including: i) Diagnosis ii) Follow-up iii) 'Discordant' decline (i.e. worsening symptoms but stable physiology)			
4.1	SCM4	Access to specialist imaging opinion at all stages	Establish accurate diagnosis at an early stage	? use imaging networks	
4.1	Group of Occupational Respiratory Disease Specialists (GORDS)	Diagnosis	It is vital to differentiate idiopathic fibrosis, from fibrosis with an identical radiological appearance that has a cause. Few patients with possible IPF have a biopsy, making diagnosis dependant on a detailed clinical assessment. Radiological UIP pattern disease can also be caused by connective tissue disease, drug therapy, and occupational/environmental exposures. IPF is therefore a diagnosis of exclusion. Since IPF is difficult to treat, every effort should be put in to not missing a cause that is driving the condition. In these cases, removing or treating the cause improves prognosis.	Finding a cause is important for patients, as it offers different management options that are established to improve prognosis. Examples include UIP pattern disease from connective tissue disease, drug therapy, chronic EAA, and hard metal lung disease. The current NICE IPF guidelines do not sufficiently highlight this, in marked contrast to similar guidelines from the American Thoracic and European Respiratory Societies, where the differential diagnosis of IPF is discussed in detail. Further guidelines in this area would be helpful, in terms of a standard set of questions to ask, and standard set of immunology to check, in order to exclude these diseases.	See Raghu G et al. An Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management. Am J Respir Crit Care Med Vol 183. pp 788–824, 2011
4.1	Sheffield Teaching Hospitals	Key area for quality improvement 6	All patients with Pulmonary Fibrosis should have an occupational and environmental history taken and	This is because IPF is a diagnosis made when all known causes have been excluded. Consequently,	This standard is supported by evidence reviews (many; for example; Raghu G et al Am J Resp

ID	Stakeholder	Suggested key area for quality improvement	Why is this important?	Why is this a key area for quality improvement?	Supporting information
	Foundation Trust		possible exposures to causes of fibrosis identified	occupational (e.g. workplace exposure to hard metals, asbestos) and environmental (e.g. extrinsic allergic alveolitis to moulds) exposures are important to deliberate and exclude. This is in the patient's best interest, as removal of the cause may be more beneficial that drug therapies. This should be done by (i) assessing all patients with an appropriate occupational history and (ii) having access where appropriate to such sub specialty interests. The GORDS group offer this nationally. GORDS website is found at; http://www.hsl.gov.uk/centres-of-excellence/centre-for-workplace-health/gords.aspx	Crit Care Med 2011;183:788-824.) This comprehensive review states that IPF is a diagnosis of exclusion, where domestic, occupational, connective tissue disorders and drug causes are all actively excluded.
4.2	UK Clinical Pharmacy Association	Key area for quality improvement 3 **Provision of Education and Support**	Patients should be provided with information about idiopathic pulmonary fibrosis - the condition and how it should be managed.	Local protocol's for the provision of Information & support, including information relating to available local services. May assist patients in feeling less isolated, and to 'put their disease into perspective'	Lindell KO, Olshansky E, Song M, et al. Impact of a disease-management program on symptom burden and health-related quality of life in patients with idiopathic pulmonary fibrosis and their care partners. Heart Lung 2010;39:304–14.
4.2	SCM2	Key area for quality improvement 3 Patients and carers should have access to information and support	Patients and carers have often never heard of this condition and are unprepared for the loss of independence and progressive symptoms.	Information and support provided by the doctor and ILD specialist nurse will improve patient experience, symptoms and access to social services	

ID	Stakeholder	Suggested key area for quality improvement	Why is this important?	Why is this a key area for quality improvement?	Supporting information
4.2	Boehringer Ingelheim	Availability of appropriate support and information for patients diagnosed with IPF and best supportive care advice	Informing patients with evidence based information about their condition can reduce their anxiety about how their illness may progress. People with IPF should have their individual needs assessed and be given advice on how best to manage their symptoms.	There is a fine balance to providing information. Too much information at the start can be overwhelming; too little can leave the patient uncertain about how to deal with their future. When information is not provided patients will seek it themselves from unregulated sources such as the internet, which may increase their anxiety. Best supportive care should be tailored to disease severity, rate of progression, and the person's preference, and should include if appropriate: information and support around symptom relief, management of comorbidities, withdrawal of therapies suspected to be ineffective or causing harm, end of life care	NICE Guideline CG163
4.2	British Lung Foundation	**Up-to-date information and support for patients**	Patients, their families and carers should receive accurate information and support at every stage of IPF development. This would help them better manage the condition and significantly improve their well-being.	There is an IPF information deficit – patients have informed us that they lack access to high-quality information regarding their condition and treatment. Furthermore, for patients, their families and carers, there is a need for information on finances, social care and practical support. Peer support groups also have a vital role in helping patients and families in dealing with the disease	“Specific information needs which were highlighted included… post-diagnostic information packs for everyone who is newly diagnosed, explaining their rights, what care and treatment they are entitled to, signposting to specialist centres and where to find support.” BLF Round-table discussion with patients , carers and clinicians on IPF, March 2013
4.2	InterMune	Information and support	Providing patients with information and support enables them to actively	A growing body of evidence demonstrates that patients who are	NICE clinical guideline 163: Idiopathic pulmonary fibrosis

ID	Stakeholder	Suggested key area for quality improvement	Why is this important?	Why is this a key area for quality improvement?	Supporting information
			participate in their care and self-management and experience improvements in health related quality of life. NICE CG163 recognises the importance of this and recommends that the consultant respiratory physician or interstitial lung disease specialist nurse should provide accurate and clear information (verbal and written) to people with IPF, and their families and carers with the person's consent. This should include information about investigations, diagnosis and management. In addition they recommend that an interstitial lung disease specialist nurse should be available at all stages of the care pathway to provide information and support to people with idiopathic pulmonary fibrosis and their families and carers with the person's consent.	more actively involved in their health care experience better health outcomes and incur lower costs (Health Policy Brief). Providing information and support to patients will raise awareness of their condition and will ultimately empower them and their carers to feel in control of their wellbeing, and enable them to make informed decisions about their condition and possible treatments. (NICE CG76). With regards to treatments, it is likely that patients may need support to help them make the most effective use of their medicines (NICE CG76). This should happen when the initial decision to prescribe a medicine is taken and then reviewed regularly. Lack of support may lead to non-adherence which may limit the benefits of medicines, resulting in lack of improvement or deterioration in health and increased costs; medicines wastage and costs arising from increased demands for healthcare if health deteriorates. A recent study demonstrated that adherence and compliance can be achieved by specialist nurse and clinician review, support and education of the patient (Chaudhuri).	NICE clinical guideline 76: Medicines adherence NICE clinical guideline 138: Patient experience in adult NHS services: improving the experience of care for people using adult NHS services Health Policy Brief: Patient Engagement. Feb 2013 https://www.healthaffairs.org/healthpolicybriefs/brief.php?brief_id=86 Chaudhuri, N et al. Respir Med. 2014 Jan;108(1):224-6 BLF 'IPF Patient Charter'. http://www.blf.org.uk/News/Detail/BLF-launches-new-charter-for-people-affected-by-IPF

ID	Stakeholder	Suggested key area for quality improvement	Why is this important?	Why is this a key area for quality improvement?	Supporting information
4.2	Boehringer Ingelheim	Carers of people with IPF are offered an assessment of emotional, psychological and social needs and, if accepted, receive tailored interventions identified by a care plan to address those needs.	Carers frequently have negative experience of service provision and often feel they are not listened to or valued. Bureaucracy and lack of funding may contribute to service users and their families not obtaining full or adequate information. Services/support provided at the time of diagnosis are important to carers who want more information on what IPF is and what benefits and services are available.	The health status of patients has a dramatic effect on the health status of the carer. Carers feel they have to fight for services with the result that many individuals feel they received too little too late. Carer anxiety and depression is partly linked to functional incapacity of the patient. Patients and carers differ in how they perceive their own needs, in how they view, judge and evaluate the disease and how they cope with its progress	Experiences of providing care to people with long term conditions, Dr. Jennifer Harris, Social Policy Research Unit, University of York, York YO10 5DD. DH 1968 JH 07.03 http://www.york.ac.uk/inst/spru/pubs/pdf/CARERS%20REPORT.pdf Accessed on line March 24th 2014
4.2 & 4.3	Association Respiratory Nurse Specialists /Royal Brompton Hospital & Harefield NHS Trust	Key area for quality improvement 5 Best supportive care	There is no curative treatment for IPF other than lung transplantation for which many patients are not eligible. Many patients have rapidly progressive disease with a prognosis worse than many cancers yet they do not have access to the same support infrastructure as do the users of cancer services	NICE clinical guideline 138 recognises that there are essential components of good patient experience in Adult NHS services. There are a paucity of IPF nurse specialists in the UK yet they play a pivotal role in providing clear and accurate information both verbal and written to people diagnosed with IPF and their families. NICE guidance recognises that a disease specific specialist nurse should be there at each stage of the patient pathway to provide ongoing information and support and re evaluating care needs to enable appropriate and timely input from the interdisciplinary team	Please see NICE clinical guidance 163 which highlights the need for information and support, symptom relief; management of comorbidities and prompt withdrawal of therapies suspected to be ineffective or causing harm. The guideline also highlights the importance of end of life care for patients with IPF. Given the increasing number of referral to ILD specialist centres there is an urgent need to develop the role of the clinical nurse specialist and support workers as an integral part of the MDT.
4.3	SCM1	Key area for quality improvement 2	Pulmonary Rehab for IPF patients	Patients are being sent on COPD rehab courses. These are not always appropriate.	

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4.3	UK Clinical Pharmacy Association	Key area for quality improvement 2 **Pulmonary Rehabilitation**	Pulmonary rehabilitation is recommended by the British Thoracic Society (BTS)	Patients with interstitial lung disease often de-saturate considerably on exercise. Pulmonary rehabilitation can improve exercise and quality of life, although may not be sustained at 6 months.	BTS guidelines at: https://www.brit-thoracic.org.uk/document-library/clinical-information/pulmonary-rehabilitation/bts-guideline-for-pulmonary-rehabilitation/
4.3	SCM2	Key area for quality improvement 4 People with IPF have access to Pulmonary rehabilitation	IPF patients may rapidly lose confidence and reduce there levels of physical exertion as they become more breathless , leading to deconditioning, becoming more dependant and social isolation. Pulmonary rehabilitation can break this cycle and lead to improvements in exercise capacity and confidence.	Pulmonary rehabilitation has shown benefits in people with IPF but access to this treatment is limited.	NICE IPF guidelines 2013
4.3	Action for Pulmonary Fibrosis	Pulmonary Rehabilitation tailored to the patient	Effective management of the disease can help to delay its progress and improve quality of life for the patient. Effective Pulmonary Rehab is a significant contributor to improvement in management of the disease.	NICE has identified the need for referral to Pulmonary Rehab at the time of diagnosis. Currently, referral for patients is variable across the country and usually consists of attending a general course which may not meet their particular needs. Even when assessed, patients often have to wait too long to attend a course.	Pulmonary Rehabilitation is a key priority for implementation within the NICE Guideline 163 published in June 2013. It is recognised as an important aspect of Best Supportive Care within the NICE Guideline. Many patients report that they have not been referred to pulmonary rehab, have had to wait too long to gain a place or have found that the course does not address their particular needs.
4.3	Boehringer Ingelheim	Assessment for pulmonary rehabilitation for people with IPF	There is evidence that appropriate and effective pulmonary rehabilitation can drive significant improvements in the quality of life and health status of people with IPF. Pulmonary rehabilitation is recommended within the NICE guideline. Rehabilitation should be considered at diagnosis.	Services offering pulmonary rehabilitation are varied across the country. Sessions should be designed for people with IPF and tailored to patients' needs. The sessions should be a mixture of advice and exercise classes. The sessions should be easy for people	NICE Guideline CG163

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				to get to, even if they have a disability.	
4.3	British Lung Foundation	**Personalised access to essential services**	To alleviate the symptoms of IPF, patients must have full access to essential services including ILD-specialist nurses, ambulatory and domiciliary oxygen, smoking cessation, and pulmonary rehabilitation.	Essential services like oxygen and pulmonary rehabilitation play a crucial role in increasing patient independence and quality of life. However, these services may not be locally available to patients, or inadequately tailored to their needs. The role of the ILD-specialist nurse is essential to coordinating care between specialist centres and locally commissioned services.	"To manage idiopathic pulmonary fibrosis, there is evidence to support a role for some types of best supportive care, such as smoking cessation, pulmonary rehabilitation, withdrawal of ineffective therapy, oxygen therapy and palliation of symptoms." NICE guidance CG163, June 2013
4.3	Sheffield Teaching Hospitals Foundation Trust	Key area for quality improvement 4	Pulmonary rehabilitation and oxygen therapy	Traditionally focussed on COPD there is a lack of understanding of ILD patient needs with suboptimal service/care delivery	
4.3	UK Clinical Pharmacy Association	Key area for quality improvement 5 **Symptomatic Management**	Cough can be one of the most distressing aspects of IPF for patients and can be very difficult to manage. Reflux is increasingly recognised as both a contributory factor to cough & disease progression & an outcome of IPF	All patients with progressive idiopathic fibrosis should receive best supportive care to improve their symptom control.	NICE TA282 http://thorax.bmj.com/content/early/2012/11/30/thoraxjnl-2012-202040.full.pdf
4.3	UK Clinical Pharmacy Association	Key area for quality improvement 1 **Access to palliative care**	There is evidence that patients with IPF are referred to palliative care too late as is advanced care planning (which is relevant to the CG recommendations around ventilation), and anecdotally that patients who are progressing slowly are taken off community teams	Local protocols for referring to palliative care services early in the diagnosis & discussing advanced care planning would be welcome	Qualitative studies demonstrate that palliative interventions need to be developed for patients with interstitial lung diseases. http://www.ncbi.nlm.nih.gov/pubmed/24644332 http://www.ncbi.nlm.nih.gov/pubmed/24644857

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			registers.		
4.3	SCM2	Key area for quality improvement 5 People with IPF have access to Palliative Care services	There is an inexorable decline in patients symptoms, with a median survival of 3 years. Symptoms such as breathlessness, cough & fatigue are common to other end of life conditions and measures to alleviate these symptoms from Palliative Care specialists and help patients plan end of life care will improve IPF patients quality of life.	Access to palliative services are patchy for patients with IPF, as these services are often prioritised for patients with cancer	NICE Guide for Commissioners on End of Life care for Adults – December 2011 QIPP (Quality, Innovation, Productivity & Prevention) – identifying people who are approaching the end of life; planning for their care NICE Quality Standard – End of Life Care for Adults – NHS Outcomes Framework 2012/13 4th National End of Life Report – October 2012 Adult Social Care Framework 2012 National Cancer Peer Review programme – Manual for Cancer Services – Specialist Palliative Care Measures 2012 NICE Improving Supportive and Palliative Care for Adults with Cancer 2004 Palliative Care Funding Review – Funding the Right Care & Support for Everyone – July 2011 Chronic Respiratory Disease, 2009, vol./is. 6/1(13-7), 1479-9723;1479- Partridge MR; Khatri A; Sutton L; Welham S; Ahmedzai SH
4.3	Action for Pulmonary Fibrosis	Referral to palliative care	Patients, their families and carers need effective palliative care to support them at various stages of the disease. Support in managing the changes taking place both physically and mentally can ease the burden for those involved.	Many patients are unaware of the valuable support that palliative care teams can provide. Often it is considered only as an end-of-life service when it should be introduced at an early stage after diagnosis.	Provision of effective palliative care is included as part of the Guideline for Best Supportive Care by NICE.
4.3	Boehringer	People in the later	End of life care helps people live as	People with serious diseases are	http://www.nhs.uk/planners/end-of-

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	Ingelheim	stages of IPF are assessed by their specialist team to identify and plan their palliative care needs	well as possible at the end of their life	often given choices for treatment during their disease process. Some patients may have many hospital admissions in a year. When the burdens of treating an illness outweigh the benefits, the goal of a patient's care may change from curing to comfort so they can enjoy the time remaining and achieve personal goals at the end of life. Services for palliative care are generally variable across the country	life-care/pages/hospital-care.aspx Accessed on line March 24th 2014 NICE Guideline CG 163
4.3	British Lung Foundation	**Integration with palliative care and end of life care**	In the later stages of disease progression, specialist respiratory teams should liaise and integrate their services with local palliative care and end of life care teams.	Palliative interventions do not halt disease progression but do improve quality of life. Patients have explained to us their struggles for timely access to palliative care services, and often these services have failed to meet their specific needs.	"Evidenced-based palliation is seldom applied, despite the high symptom burden and poor quality of life (QoL)." 'Interventions to improve symptoms and quality of life of patients with fibrotic interstitial lung disease: a systematic review of the literature'. Sabrina Bajwah et al, BMJ Thorax. December 2012
4.3	Sheffield Teaching Hospitals Foundation Trust	Key area for quality improvement 2	Enhanced palliative care	Done poorly in many centres. Specialist ILD nurses/care coordinators would be excellent service deliverers to this area with close working relationships to hospital and community palliative care teams	
4.3	UK Clinical Pharmacy Association	Key area for quality improvement 4 **Use of Lung Function Testing prior to Commencing**	NICE TA282 specifies that pirfenidone is recommended as an option for treating idiopathic pulmonary fibrosis if the forced vital capacity (FVC) between 50% and 80% predicted. Treatment should be	Audits in some centres have shown that some patients had quite a delay before the PFT 'qualifying' them for pirfenidone & actually starting treatment.	

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		Pirfenidone	discontinued if there is evidence of disease progression (a decline in per cent predicted FVC of 10% or more within any 12 month period). However it would also be useful to stipulate the 'validity' of the pulmonary function tests (PFTs) in relation to starting treatmen	This may affect the interpretation of repeat PFTs used for deciding on discontinuation after 12 months of treatment.	
4.3	Boehringer Ingelheim	People with IPF are offered medication in accordance with NICE guidance, as part of an individualised comprehensive management plan, and/or a lung transplantation depending upon eligibility criteria.	There are currently no drugs which can cure IPF but there are treatments that can help with symptoms.Lung transplantation may improve survival for people with IPF	Emerging evidence suggests that long standing treatments for IPF have no clinical merit, while other treatments are in development which may prove to be beneficial. Lung transplantation has been shown to improve survival in patients with IPF. In one study a single lung transplantation reduced the risk of death by 75% (95% CI 8%–86%, p = 0.03) compared with patients with IPF on the transplant waiting list. No cases of disease recurrence were reported in the donor lung after transplantation. Since IPF is a progressive disease and no treatment is known to prolong survival other than lung transplantation, patients should be referred for transplantation assessment soon after diagnosis. Data suggest however rates for referral for lung transplantation are double in the south of England compared to the north.	NICE Guideline CG163 Idiopathic pulmonary fibrosis: current understanding of the pathogenesis and the status of treatment Nasreen Khalil, Robert O'Connor CMAJ July 20, 2004 vol. 171 no. 2 doi: 10.1503/cmaj.1030055 http://www.cmaj.ca/content/171/2/153.abstract Accessed on line March 24th 2014

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4.3	InterMune	Prescribe pirfenidone in line with licence and NICE criteria.	Pirfenidone represents a first-in-class treatment, which has been shown to be effective in IPF by reducing decline in lung function and slowing disease progression (Noble, 2011). Pirfenidone is the only drug to have been granted a licence by the European Medicines Agency (EMA) for the treatment of adults with mild to moderate IPF and to have received a positive NICE technology appraisal.	Timely initiation of treatment (i.e. at diagnosis of IPF) optimises the 'window of opportunity' within which effective treatment can improve outcomes. To achieve this, suspected IPF patients need to be referred, diagnosed and initiated on treatment appropriately and promptly.	NICE TA282: Pirfenidone for treating idiopathic pulmonary fibrosis Pirfenidone SmPC: http://www.medicines.org.uk/emc/medicine/26942/SPC/Esbriet+267+mg+hard+capsules/
4.3	Association Respiratory Nurse Specialists /Royal Brompton Hospital & Harefield NHS Trust	Key area for quality improvement 2 Pharmacotherapy	IPF is associated with a poor prognosis – estimated survival from diagnosis to death is 3-5 years. The only curative treatment is lung transplantation. There is evidence that disease progression is slowed down in patients who are taking Pirfrnidone. Patients with IPF should have the opportunity to be assess for their suitability for this therapy	NICE technology Appraisal 282 recognises that Pirfenidone is effective in slowing disease progression in IPF in some patients. Given the cost of this therapy and side effect profile it is important that patients who will benefit from this therapy are appropriately identified by the specialist centre and that processes for monitoring the effect and impact of therapy are put in place	Please see NICE Clinical guideline 163 and NICE technology appraisal: Pirfenidone for treating IPF issued april 2013 which recommends Prifenidone for patients diagnosed with IPF who have an FVC between 50-80% predicted value at the commencement of therapy. Monitoring ensure that patients who are not continuing to benefit from therapy in that their FVC declines by 10% or more in a 12month period should be withdrawn from the programme.
4.3	Sheffield Teaching Hospitals Foundation Trust	Key area for quality improvement 3	Medication monitoring	Close monitoring of side-effects and efficacy of novel agents including immunosuppressant's through specialist nursing teams.	
4.3	SCM1	Key area for quality improvement 3	Referral for Lung Transplant	Patients are not being referred for lung transplant early enough. This	

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				results in IPF patients being too ill or too old.	
4.3	Action for Pulmonary Fibrosis	Timely referral for lung transplantation assessment	If people wish to be referred and provided there are no absolute contraindications, it is important that they have initial discussions early so that the assessment can take place at the appropriate stage of progression of the disease.	Many patients are not referred within 3 to 6 months of diagnosis as NICE guidance suggests. This can result in the assessment being too late	Recommendations in NICE Guideline 163.
4.3 & 4.5	British Lung Foundation	**Prompt access to treatment**	IPF frequently has a rapidly debilitating effect on patients. Immediately following diagnosis patients must be given the appropriate treatment to best reduce disease progression. This may include access to drugs, assessment for transplants, and clinical trial options.	It is important that patients have the full range of treatment options explained and offered to them at the earliest possible opportunity. While there is no cure for IPF, effective and quick treatment can have a considerable impact on clinical outcomes. Patients also worry that they are not fully informed of trials and transplants.	"…people with IPF and their families have the right to… full details of all treatment, clinical trials, transplant, support and service provision options available to them" Quote from the 'IPF Patient Charter', September 2013, published by the BLF and developed alongside clinicians and patients.
4.3	SCM4	Early transplant opinion	Limited efficacy of conventional treatments		
4.3	SCM1	Key area for quality improvement 4	Oxygen assessment	Too many patients are not being given oxygen for exercise as they don't desaturate on a 6 minute walk test but they encounter problems as soon as they exert themselves.	
4.3	Action for Pulmonary Fibrosis	Appropriate and timely access to Ambulatory Oxygen	The patient's quality of life and level of independence is significantly improved by access to the appropriate type and levels of ambulatory oxygen.	The quality of assessment for ambulatory oxygen is variable resulting in many patients receiving inappropriate or delayed assessment, This is often conducted by a non-IPF medical specialist. For the patient this results in greater dependence on others and limits their ability to take part in day-to-day	NICE Guideline 163 identifies ambulatory oxygen as a key priority for implementation. Again it is recognised as an important aspect of Best Supportive Care. In addition, reports from patients highlight the need for timely assessment and indicate that this is variable across the country

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				activities.	
4.3	Association Respiratory Nurse Specialists /Royal Brompton Hospital & Harefield NHS Trust	Key area for quality improvement 3 Oxygen therapy	Oxygen is a treatment that is prescribed and should be used according to instructions within that prescription. It is recognised that variations in oxygen therapy provision increases the risk for poor quality care and increases waste.	Many patients diagnosed with IPF require ambulatory oxygen, many require overnight oxygen and some patients progress to needing continuous oxygen therapy. NHS Improvement – lung recognise that it is critical that patients are formally assessed in respect of their individual clinical need prior to the oxygen supply being issued.	Please see Improving the quality and safety of home oxygen services: the case for spread issued as part of the NHS improvement programme. The scoping exercise undertaken subsequent to the DH good practice guideline (2011) identifies good practice standards to ensure quality of oxygen service provision. Further research is needed in respect of determining the impact that the community respiratory nurse has on the safe initiation delivery and monitoring of patients on home oxygen therapy.
4.4	Sheffield Teaching Hospitals Foundation Trust	Key area for quality improvement 5	Adequate medical resource with clear follow up regimens	Growing population – limited previous investment in this area	
4.5	Sheffield Teaching Hospitals Foundation Trust	Key area for quality improvement 1	Detailed and accurate data collection with submission of data to central databases	Understand populations, effects of treatment, variation in presentation and clinical needs. May lead to increased research study recruitment and benchmarking of practice. Possibility of early case finding exercises.	ATS/ERS Consensus Statement for ILD
4.5	Association Respiratory Nurse Specialists /Royal	Key area for quality improvement 4 IPF Registry	IPF is a rare disease in the UK and its incidence is rising Epidemiological evidence is limited.	Accurate knowledge is needed regarding the true incidence and prevalence of IPF in the UK. A national registry will enable the setting up of national Public Health	BTS established a pilot Sarcoid registry - the data obtained from this enabled the characterisation of demography of sarcoid disease. This has become a valuable

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	Brompton Hospital & Harefield NHS Trust			and epidemiological studies – such studies will increase our understanding of IPF trends and inform public Health Policy and national guidelines	resource for both clinicians and research. The BTS ILD registry project will establish the burden of IPF and reduce diagnostic delay through the collection of accurate information.
4.5	SCM4	Optimise registry entry for consideration of clinical trials			
NA	NHS England	Thank you for the opportunity to comment the scope consultation for the above Quality Standard I wish to confirm that NHS England has no substantive comments to make regarding this consultation			
NA	Royal College of Nursing	This is to inform you that the Royal College of Nursing have no comments to submit to inform on the above topic engagement at this present time. Thank you for the opportunity to participate, we look forward to the next stage of the			

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		development process.			