SCHEDULE 2 – THE SERVICES

A. Service Specifications

Service Specification No.	17009/S
Service	Interstitial Lung Disease Service Adult
Commissioner Lead	
Provider Lead	

1. Scope

1.1 **Prescribed Specialised Service**

This service specification covers the provision of **Interstitial Lung disease (ILD).**

1.2 **Description**

Interstitial lung diseases comprise a broad spectrum of conditions, all of which are characterised by inflammation or fibrosis of the alveolar wall with impairment of gas exchange. The commonest of these conditions are idiopathic pulmonary fibrosis (IPF), sarcoidosis and extrinsic allergic alveolitis (EAA). Together these conditions affect between 1,500 and 3,000 individuals in England each year. In addition up to 30% of patients with connective tissue diseases (also known as collagen vascular diseases) such as rheumatoid arthritis and scleroderma develop ILD. There are also a myriad of less common ILDs (non-specific interstitial pneumonitis, desquamative interstitial pneumonitis, respiratory bronchiolitis interstitial lung disease, acute interstitial pneumonia, lymphocytic interstitial pneumonitis, pulmonary alveolar proteinosis, histiocytosis X and lymphangioleiomyomatosis (LAM)) each with an estimated incidence of between 0.1-5 per 100,000 individuals per year

Between 2,000 and 4,000 new patients are diagnosed with ILD in England each year with the majority having either sarcoidosis or IPF. Disease-specific management plans are drawn up following Multi-Disciplinary Team (MDT) assessment at regional specialist units. Networks of care need developing so that the majority of subsequent follow-up is provided in local secondary care units.

1.3 **How the Service is Differentiated from Services Falling within the Responsibilities of Other Commissioners**

NHS England commissions interstitial lung disease services from Specialist Respiratory Centres. CCGs commission existing secondary and primary care services supporting the local care of patients with ILD as determined by the Specialised centres. This service includes specified activity at specified centres.

2. Care Pathway and Clinical Dependencies

Currently management and diagnosis of the ILDs is guided by a number of guidelines:

- NICE Idiopathic Pulmonary Fibrosis Guideline (CG163) June 2013 http://guidance.nice.org.uk/CG163
- The 2008 British Thoracic Society SIGN guidelines on interstitial lung disease
- The diagnosis of IPF has been formalised in a consensus guideline developed by the American Thoracic Society (ATS) and European Respiratory Society (ERS) (published in 2011)
- NICE IPF Pirfenidone Technology Appraisal (TA 282) Published April 2013
- The Map of Medicine http://eng.mapofmedicine.com/evidence/map/interstitial_lung_disease1.html
- Sarcoidosis is covered by the 1999 ATS/World Association of Sarcoidosis and other Granulomatous Disorders (WASOG) guidelines
- LAM is addressed by 2010 ERS management and diagnosis guidelines
- The Gold Standards Framework for palliative care (Gold Standards Framework)
- NICE IPF Quality Standard QS79 IPF in adults published January 2015
- NICE IPF Nintedanib Technology Appraisal (TA 379) published January 2016

The ICD 10 codes to be used for interstitial lung disease are given below:

D76.0	Langerhan's cell histiocytosis
J67	Hypersensitivity pneumonitis
D86.0	Sarcoidosis
D86.2	Sarcoidosis
J84.0 - J84.9	Interstitial Lung Disease (including IPF and CTD-associated)

Therapies associated with the diagnostic codes will also be identified for pirfenidone, nintedanib and cyclophosphamide.

Sarcoidosis will only be considered for specialist centre assessment in cases where there is extensive multi-organ involvement (to include lungs, heart or brain and other organ

involvement) or in individuals who develop progressive pulmonary fibrosis despite standard therapy.

The flows and pathways required at specialist centres will be those centred around

1) diagnosis, 2) treatment planning, 3) in some cases, treatment administration and 4) in cases requiring the above named therapies, disease monitoring.

Diagnostic assessment

Patients will be referred to the centre that is geographically most convenient for them and will then be systematically assessed. Where possible, this will require a single visit to the centre, but in cases of diagnostic uncertainty assessment may require two to three day case visits (including for bronchoscopy). In approximately 10-15% of cases, diagnostic video assisted surgical lung biopsy will be necessary. It is expected that specialist centres will have the facilities to undertake, as required by individual cases, the following investigations:

- Full pulmonary function tests (PFT) including carbon monoxide diffusing capacity (DLco) and estimation of total lung capacity (either by plethysmography or helium dilution)
- 6 minute walk test (or equivalent)
- High Resolution Computed Tomography of the thorax
- Bone densitometry (dual energy X-ray absorptiometry (DEXA))
- Immunology including extended autoimmune panel and serum precipitins
- Echocardiogram with right heart assessment
- Overnight oximetry
- Bronchoscopy with bronchoalveolar lavage (BAL) and availability of a pathology service able to provide formal BAL differential cell counts
- Surgical lung biopsy (pre-operative planning to include a case conference between physician, surgeons and, when necessary, thoracic radiologist)
- Genotyping for relevant ILDs (e.g. tuberous sclerosis, alveolar microlithiasis, Burt-Hogg-Dubae syndrome etc.)
- Cardiac magnetic resonance imaging (MRI), for cases of suspected cardiac sarcoid
- Anti granulocyte/macrophage colony stimulating factor (anti-GM CSF) antibody testing for suspected cases of alveolar proteinosis.

Following the completion of the assessments the results of test will be discussed at a diagnostic multi-disciplinary team (MDT) meeting. A fully constituted MDT will consist of a respiratory physician with specialist training in ILD, a thoracic radiologist with expertise in ILD, a thoracic pathologist, an ILD Specialist Nurse and an MDT co-ordinator. The necessary level of clinical expertise and training required by MDT members is described in detail in the NICE IPF Guidelines 2013.

Treatment planning

Following diagnosis, treatment will be planned in accordance with national and international guidelines. This will involve input from respiratory physicians with training in ILD, an ILD specialist nurse and, where necessary, respiratory physiotherapists, occupational therapists and physicians from other disciplines (e.g. rheumatologists, ear, nose and throat (ENT), cardiology, transplant, pulmonary hypertension, palliative care etc.). Provision should be made to ensure patient access to pulmonary rehabilitation. In most cases, it is envisaged that treatment will be delivered locally and reviewed annually at specialist centres.

Treatment Administration

In certain cases, it will be necessary for specialist centres to administer therapy to patients rather than rely on local services. Such treatments include:

- parenteral cytotoxic agents (intravenous cyclophosphamide, intra muscular methotrexate)
- biological agents
- chemotherapeutic agents
- plasmapheresis
- intra venous immunoglobulins
- whole lung lavage for pulmonary alveolar proteinosis (this will only be available in only two centres to ensure adequate case load and expertise)
- monitoring of immunosuppressant serum levels

Disease monitoring

For patients receiving treatment at specialist centres, follow-up and monitoring of disease (in most cases through clinical assessment and full lung function testing) will be dictated by disease severity and treatment regimen, but is likely to be once every 3–4 months. For the majority of individuals for whom care is being delivered locally, specialist centre review will occur annually. These reviews will comprise clinical assessment, full lung function and thoracic imaging. Patients will also, where necessary, be seen at the same visit by other members of the multi- disciplinary team and in advanced disease should undergo appropriate assessment of oxygen requirements.

Discharge

Individuals will be discharged from specialist centre care in the following circumstances:

- if diagnostic assessment fails to confirm a diagnosis of an ILD
- patients with disease that remains stable at two consecutive visits following withdrawal of treatment
- individuals with end-stage disease transitioning to palliative care services
- individuals with end-stage disease who have undergone lung transplantation.

Additional roles of specialist centres

- Patient education
- Patient support groups
- Outreach support to local centres, electronically or by teleconference, this should include annual audit
- Inpatient care and transfer of individuals with treatment responsive ILD or patients requiring emergency, inpatient assessment following first presentation of acute onset ILD
- Intensive care support for ILD inpatients
- Education of health care professionals.

Please note that access to treatment will be guided by any applicable NHS England national clinical commissioning policies.

2.2 **Interdependence with other Services**

Co-located services:

- Immediate onsite access to critical care.
- Advanced diagnostics including lung physiology, and bronchoscopy
- Day unit available for patient assessment and high cost novel biological agent administration.
- In-patient beds available for management of acute exacerbations of ILD
- Specialist thoracic radiology services
- Dedicated thoracic pathology
- Palliative care services
- Thoracic surgery

Interdependent services:

- Rheumatology
- Clinical Immunology
- ENT
- Physiotherapy for exercise and pulmonary rehabilitation
- Pulmonary hypertension
- Lung transplant
- Cardiology with expertise in cardiac sarcoid
- Dermatology with expertise in cutaneous sarcoid
- Pharmacy
- Neurology with expertise in neurosarcoid
- Opthalmology with expertise in occular sarcoid

Related services:

- Occupational lung disease

3. Population Covered and Population Needs

3.1 Population Covered By This Specification

The service outlined in this specification is for adult patients ordinarily resident in England; or otherwise the commissioning responsibility of the NHS in England (as defined in Who Pays?: Establishing the responsible commissioner, and other Department of Health guidance relating to patients entitled to NHS care or exempt from charges).

3.2 Population Needs

Estimates of the incidence of the different ILDs vary. The commonest, IPF and sarcoidosis, have an incidence of between 2000–4000 in the England per annum. Whilst the prognosis for individuals with sarcoidosis is reasonably good, the median survival for those with IPF is only 3–3.5 years and the disease now accounts for more than 3000 deaths in England each year. Rarer conditions such as LAM and histiocystosis X have an incidence of 2-6 per million per annum whilst a recent epidemiological study utilising the UK GPR database identified only 563 new cases of CTD-ILD diagnosed in the last decade. Importantly however, mortality for those with CTD-ILD was 40% higher than for those with CTD alone. Clinical trial data are emerging to support the value of a number of therapies in the various ILDs. The novel anti-fibrotic drugs slow disease progression in IPF. Intravenous cyclophosphamide improves outcomes in individuals with CTD associated ILD. Appropriate use and monitoring of all these therapies requires the integration of several disciplines and is therefore best suited to regionally delivered specialist centres.

3.3 Expected Significant Future Demographic Changes

Epidemiological studies suggest that the incidence of ILD (in particular IPF) has been increasing over the last three decades. It might be anticipated that if this trend continues there will be between 300–800 new cases per annum over the next 10 years.

3.4 Evidence Base

The specification has been developed by expert consensus taking in to account BTS ILD Guidelines, NICE IPF guidelines (CG163), NICE Pirfenidone TA (ID334), NICE Nintedanib TA (ID752) and NICE IPF Quality standards (QS79) as well as relevant international consensus statements and guidelines cited in section 2.

4. Outcomes and Applicable Quality Standards

4.1 Quality Statement – Aim of Service

The overall aim of the specialist service is to ensure equality of patient access to multi-disciplinary team diagnosis, to guarantee that patients with ILD have equal access to current treatment modalities and that their disease-specific management plans are drawn up following MDT assessment at regional specialist units. Networks of care need developing so that the majority of subsequent follow-up and, where necessary, end-of-life care, is provided in local secondary care units.

The objectives of the service are to:

- provide a specialist multi-disciplinary service for diagnosis (thus improving diagnostic accuracy for individuals with ILD)
- initiate appropriate pharmacological and non-pharmacological treatment for individuals with ILD
- reduce morbidity and mortality due to ILD, including reducing hospitalisation
- ensure equity of access to specialised therapies for all patients with ILD in England
- identify individuals requiring referral to lung transplant centres
- oversee those aspects of care that fall out with the expertise of local units.

The purpose of the service will be to:

- develop an equitable national ILD service whereby individuals with ILD will have access to specialist ILD-MDT diagnosis.
- where appropriate, provide personal management plans for each patient annually, the provision of which will, in most cases, be delivered locally.
- provide specialist advice and support to local providers of care, and in difficult cases to review patients between annual visits.
- provide equitable access to specialist therapies and to provide appropriate support and ancillary services (e.g. drug level monitoring) to ensure the safe local management of individuals requiring cytotoxic or immunosuppressant therapy.
- develop and share national ILD protocols and guidelines. Ensure local clinical teams are provided with management guidelines, and have access to specialist advice when needed.
- provide a national forum to discuss difficult management decisions.
- improve awareness and management of ILD within England by education and provision of an excellent service.
- raise standards of care for patients with ILD in England so as to improve prognosis and reduce disease and treatment related morbidity.
- offer patient centred assessment and management regarding the disease complications and organ specific problems associated with certain ILDs (e.g. sarcoidosis, CTD-associated ILD).
- minimise the disease impact of ILDs on the patient and their family life and work practice.
- enable integration of clinical services with clinical trials and translational research to

ensure on going developments in the care of individuals with these rare diseases.

- ensure equitable patient access to related services e.g. lung transplant assessment, pulmonary hypertension services, end-of life palliative care input etc.

NHS Outcomes Framework Domains

Domain 1	**Preventing people from dying prematurely**	**X**
Domain 2	**Enhancing quality of life for people with long-term conditions**	**X**
Domain 3	**Helping people to recover from episodes of ill-health or following injury**	**X**
Domain 4	**Ensuring people have a positive experience of care**	**X**
Domain 5	**Treating and caring for people in safe environment and protecting them from avoidable harm**	**X**

4.2 Indicators Include:

Number	Indicator	Data Source	Outcome Framework Domain	CQC Key question
Clinical Outcomes				
101	% patients discussed at ILD MDT	SSQD	1, 2, 3, 4, 5	Safe, effective, caring, responsive
102	Access to clinical trials	HES	1, 2, 3	Safe, effective, caring, responsive
103	Mortality rate.	SSQD	1	Safe, effective, caring, responsive
104	% patients having full lung function tests	SSQD	1, 2, 3, 4, 5	Safe, effective, caring, responsive
105	Proportion of emergency admissions for respiratory deterioration	SSQD	1, 2, 3, 4, 5	Safe, effective, caring, responsive
Patient Experience				
201	Are patients receiving a treatment plan and diagnosis at their first visit.	Self-declaration	1, 2, 3, 4, 5	Safe, effective, caring, responsive
202	Provision of patient information	Self-declaration	1, 2, 3, 4, 5	Safe, effective, caring,

				responsive
203	Quality of life measurement	Self-declaration	1, 2, 3, 4, 5	Safe, effective, caring, responsive
204	Patient experience survey	Self-declaration	1, 2, 3, 4, 5	Safe, effective, caring, responsive
Structure and Process				
301	Lead Clinician	Self-declaration	1, 2, 3, 4, 5	Safe, effective
302	MDT membership	Self-declaration	1, 2, 3, 4, 5	Safe, effective
303	Treatment Planning meetings	Self-declaration	1, 2, 3, 4, 5	Safe, effective
304	Facilities	Self-declaration	5	Safe
305	Annual Review	Self-declaration	1, 2, 3, 4	Safe, effective, caring, responsive
306	Outreach Services	Self-declaration	3, 4, 5	Safe, effective
307	Clinical guidelines	Self-declaration	1, 2, 3, 4, 5	Safe
308	Patient pathways	Self-declaration	1,2,3,4	Safe, effective, caring, responsive
309	Data Collection	Self-declaration	1, 2, 3, 4, 5	Safe
310	Audit	Self-declaration	1, 2, 3, 4, 5	Safe, effective, responsive, caring

Detailed definitions of indicators, setting out how they will be measured, are included in schedule 6.

4.3 Commissioned providers are required to participate in annual quality assurance and collect and submit data to support the assessment of compliance with the service specification as set out in Schedule 4A-C

4.4 Applicable CQUIN goals are set out in Schedule 4D

5. Applicable Service Standards

5.1 Applicable Obligatory National Standards

5.2 Other Applicable National Standards to be met by Commissioned Providers

- BTS ILD Guidelines 2008
- NICE IPF Guidelines (CG163, June 2013)
- NICE IPF Quality Standards (QS79 January 2015)

5.3 Other Applicable Local Standards

Not applicable

6. Designated Providers (if applicable)

Not applicable

7. Abbreviation and Acronyms Explained

The following abbreviations and acronyms have been used in this document:

Anti-GM CSF – anti-granulocyte/macrophage colony stimulating factor
BAL – bronchoalveolar lavage
CCG – clinical commissioning group
CTD – connective tissue disease
DEXA – dual energy X-ray absorptiometry
DLco – carbon monoxide diffusing capacity
EAA – extrinsic allergic alveolitis
ENT – ear, nose and throat
ILD – Interstitial Lung Disease
IPF – idiopathic pulmonary fibrosis
LAM - lymphangioleiomyomatosis
MDT – multi-disciplinary team
MRI – magnetic resonance imaging
NICE - National Institute for Health and Care Excellence
PFT – pulmonary function test