SCHEDULE 2 – THE SERVICES

A. Service Specifications

Service Specification No.	A14/S(HSS)/c
Service	Primary Ciliary Dyskinesia management service (adults)
Commissioner Lead	Sarah Watson, Commissioning Manager
Period	
Date of Review	

1. Population Needs

1.1 National/local context and evidence base

Primary Ciliary Dyskinesia (PCD) is a rare (estimated between 1:26000 - 1:40000) [http://erj.ersjournals.com/content/36/6/1248.long] hereditary disorder of ciliary dysfunction leading to multisystem abnormalities which include chronic respiratory, sinus and ear infections, cardiac abnormalities, infertility, and ectopic pregnancies. Chronic infection of the upper and lower airway is the most common. Without appropriate specialised treatment, progressive chronic lung disease and bronchiectasis develop. If diagnosis is delayed or managed sub optimally permanent bronchiectasis and deterioration of lung function will occur. (http://www.atsjournals.org/doi/pdf/10.1164/rccm.200303-365OC).

Highly specialised management is needed to be provided for this condition by a multidisciplinary team of professionals trained in the multisystem manifestations of PCD. The importance of this is highlighted by a number of longitudinal studies demonstrating an improved patient course with diagnosis and the institution of appropriate management. (http://erj.ersjournals.com/content/10/10/2376.long) (http://www.atsjournals.org/doi/pdf/10.1164/rccm.200811-1731OC).
There is evidence that the lack of intervention results in up to 25% of adult patients developing respiratory failure, all of whom will die early unless they have a lung transplant. (http://www.atsjournals.org/doi/pdf/10.1164/rccm.200303-365OC). Poor management leads to a

cycle of damage, increased risk of infection, further damage, leading to accelerated decline.

Specialised care is very important for this group of patients and will have a direct impact on disease progression, morbidity and mortality, satisfying all of the NHS Framework domains. In addition to the substantial patient benefits, it will also be cost effective, reducing the pressures placed on resources by poorly managed patients with reduced hospitalisations, need for advanced respiratory support and socioeconomic costs of chronic ill health.

This service will collaborate closely with the PCD paediatric service commissioned by NHS England. The service will ensure excellence in transition to adulthood, a vulnerable period for patients. Adult patients will continue to receive specialist input and the significant gains that have been made in the paediatric specialist centres will be maintained. The adult specialist service is crucial to build on the successes in the paediatric population and care for a group of patients with more severe disease. In addition to the improvement in care and outcome that would be provided by a specialised adult service, the service will also ensure an improved patient experience.

2. Outcomes

2.1 NHS Outcomes Framework Domains & Indicators

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

The purpose of this service is in line with the domains of the NHS Framework. Appropriate management of this patient group has been demonstrated to reduce disease progression and should therefore reduce premature death and enhance the quality of life for patients with PCD. Ensuring that all patients have access to the optimal standards of care should provide improved recovery from ill health and also ensure that services are delivered so that patients are treated in appropriate environments and protected from harm (e.g. inpatient or adequately monitored home intravenous antibiotics as needed). Patient groups indicate that patients want a nationally commissioned adult service to ensure an equitable and positive patient experience.

For recording outcomes, data from annual review measurements will be collected in a national database which allows documentation of variables including number of patients in each centre, referrals to each centre, annual reviews performed, outpatient episodes per patient, inpatient episodes per patient.

Specific outcome measures that will be recorded to assess the performance of the specialist centre and how they map to the NHS outcome framework domains are detailed below.

% of adult patients diagnosed with PCD managed in adult PCD specialist centres (Domain 1)
% of patients in the PCD management service offered an annual review appointment (Domain 2
% of patients seen by a physiotherapist at annual review (Domain 3)
% of patients seen by a nurse specialist at annual review (Domain 4)
% PCD inpatients at or discussed with the specialist centre (Domain 5)

3. Scope

3.1 Aims and objectives of service

This service should provide a specialised management service for adult patients with PCD. The core objective of this service is to ensure that all patients with PCD have access to the optimal standards of care to enable a reduction in the morbidity and mortality associated with the condition.

The service aims to:

1. Ensure that all PCD adult patients, wherever they live, have access to and are managed according to optimal standards of care, by ensuring:

a) all PCD adult patients have an annual review by a specialist PCD MDT consisting of a PCD specialist consultant, physiotherapist, nurse specialist and ENT specialist with access to a dietician and psychologist as needed.

b) all PCD adult patients have access to appropriate home and inpatient antibiotic services

c) care and management of patients with PCD is coordinated so that patients receive appropriate respiratory, ENT, cardiac, obstetric and physiotherapy care, fertility advice, palliative care and care for other conditions associated with PCD.

d) patients/families and other health professionals are educated on the implications and management of PCD.

2. Raise awareness in the Adult Respiratory community about the diagnosis of PCD to ensure that these patients are transferred from bronchiectasis clinics to the PCD specialist service to monitor their condition and limit disease progression.

3. Reduce the morbidity and mortality related to PCD, as well as the economic burden, associated with late diagnosis and poorly managed disease.

4. Enter patients into a national database.

3.2 Service description/care pathway

The national adult PCD service will work closely with the existing paediatric services and will provide a geographically accessible service for patients offering seamless transition for patients from the paediatric PCD centres. Each centre will additionally provide an outreach service to ensure equality of access.

As detailed below, the centrally commissioned specialised service will provide all patients with an annual review, access to the multidisciplinary team, and a detailed management plan. Further outpatient appointments at the specialist centre will be dictated by clinical need and stability. Some PCD patients will require intravenous antibiotics either at home or as inpatients. Inpatient care, with appropriate specialised physiotherapy and MDT input, will be provided either at the specialist centre or at local centres with supervision by the specialist centre.

1.2.1 Entry to the Service

Patients entering the service are likely to be:

1) paediatric transition patients with a confirmed diagnosis of PCD
2) adult patients with a confirmed diagnosis of PCD made by the PCD Diagnostic Service
3) adult patients with clinically suspected PCD but indeterminate results of investigations by the PCD Diagnostic services.

1.2.2 Routine Care (see Figure 1)

i. Annual review

All patients will have an annual review at the specialist centre. This will involve a day in the specialist centre and include the following in all patients:

1) Formal lung function testing
2) physiotherapist review
3) ENT specialist review
4) PCD nurse specialist review
5) PCD consultant review
6) Sputum culture sent for bacterial, mycobacterial and fungal culture
7) Blood tests - FBC, U&Es, LFTs, CRP, ESR, ABPA markers
8) Chest X-Ray
9) Audiology assessment
10) Database entry
11) Written annual plan for patient, primary and secondary care.

Additionally the following will be available for all annual review patients as needed:

1) Dietician review
2) Psychology review
3) Further blood tests including immunoglobulins, pneumococcal antibodies, aspergillus ICAP IgG, drug monitoring
4) HRCT scan
5) Echocardiogram
6) Fertility advice (limited dates / year)

ii. Additional clinic review

It is anticipated that for some patients, only an annual review will be necessary in the specialist centre. For other patients 6 monthly, or for the most severe or progressive patients, 3 monthly appointments in the specialist centre will be needed. The following will be performed at each additional routine clinic visit for all patients:

1) Spirometry
2) Physiotherapist review
3) PCD nurse specialist review
4) PCD doctor review
5) Sputum culture sent for bacterial, mycobacterial and fungal culture.

Additionally the following will be available as needed:

1) ENT specialist review
2) Dietician review
3) Psychology review
4) Blood tests - FBC, U&Es, LFTs, CRP, ESR, ABPA markers
4) Chest X-Ray
5) HRCT scan.

iii. Transition clinic

Transition to the adult service will be prepared by the paediatric management service involving an integrated care pathway. It will culminate in the transition clinic which will be a joint clinic performed with the associated paediatric centre with the members of the paediatric MDT and attended by the adult PCD consultant and the adult PCD specialist nurse. At this clinic appointment, in addition to the standard clinic review, all aspects of the patient care will be handed over and the adult team will describe the practicalities of the adult service and answer questions from the patient (and family). The same adult PCD consultant (where possible) and PCD nurse will see the patient at the first adult clinic following on from transition.

iv. Relationship with non-specialist centres / Outreach clinics

All PCD patients will be reviewed in the specialist centre at least annually with the provision for more regular follow-up appointments as above. Each specialist centre will provide outreach clinics with a PCD consultant, nurse specialist and physiotherapist to cater for those patients who require more than an annual review in the specialist centre, but find the specialist centre harder to access.

v. Urgent clinic review

Weekly urgent appointments /assessments will be available at the specialist centres where patients would be reviewed and the following provided:
1) spirometry
2) physiotherapist review
3) PCD nurse specialist review
4) PCD doctor review
5) sputum culture sent for bacterial, mycobacterial and fungal culture.

Additionally the following will be available as needed
1) Blood tests - FBC, U&Es, LFTs, CRP, ESR, ABPA markers
2) Chest X-Ray
3) HRCT scan.

vi. Phone advice

Each centre will provide a specialist nurse staffed phone line during working hours with direct consultant input which is available for both patients and healthcare workers.

vii. Other available services

There will be availability for specialist bronchoscopy for adult PCD patients as needed at the specialist centres. The service will deliver evidence-based management developed through the best use of educational resources that are easily accessible by patients and professionals.

PCD Routine Care

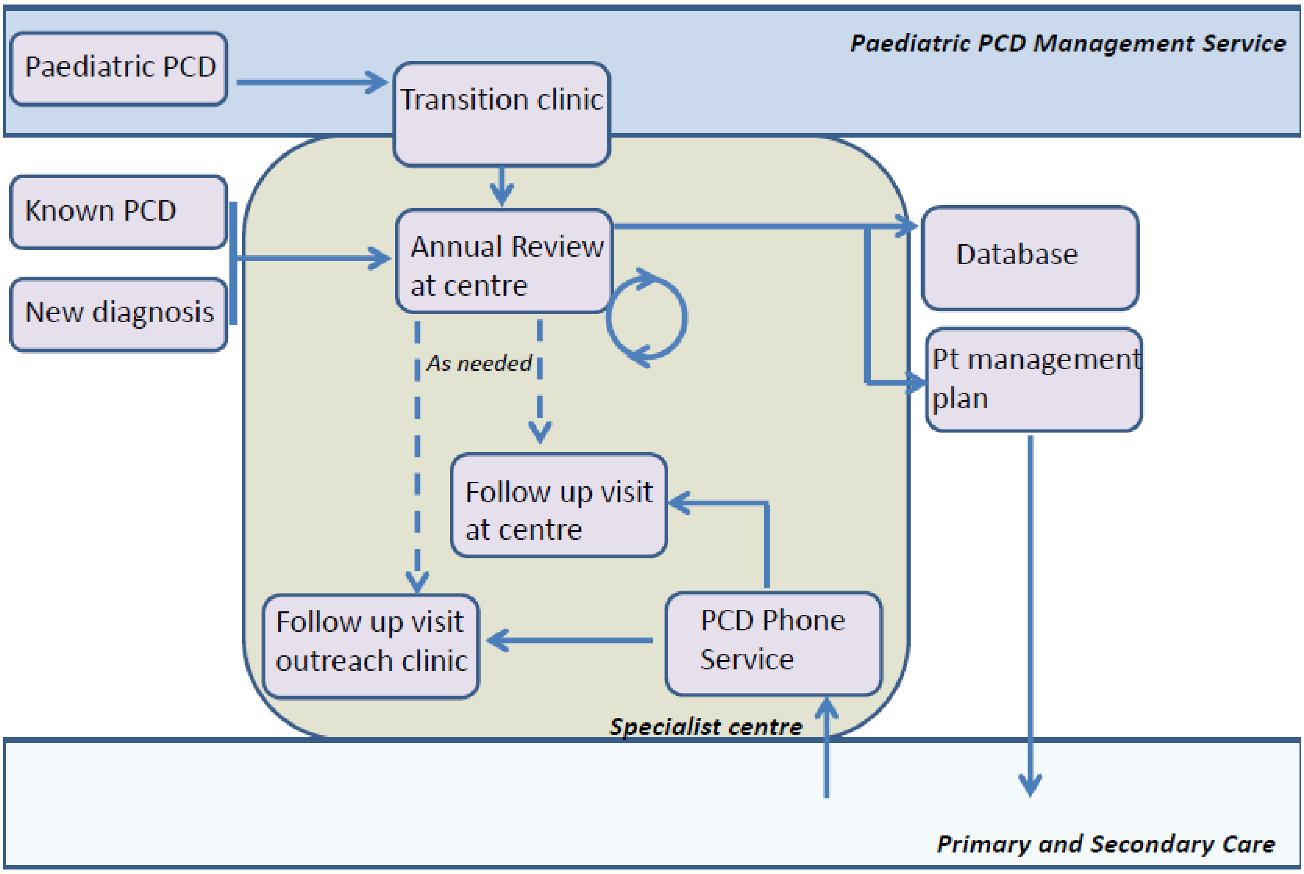

Figure 1

3.0 Acute care (See Figure 2)

All patients will have a management plan for when they are unwell. Within working hours further advice will be available from the specialist nurse staffed phone line with direct consultant input. For acute problems outside working hours, patients will seek advice from their local hospital. There will be provision for weekly urgent appointments /assessments at the specialist centres as above. Local hospitals that perform acute patient assessments will be able to liaise with specialist centres as needed.

3.1 Hospital admissions

The specialist centres will have some involvement in all inpatient admissions for intravenous antibiotics. The flow chart below demonstrates the various entry points to inpatient admission. Assessments and inpatient admissions can be arranged directly in the specialist centres. Acute presentations will also be to local hospitals based on severity and practicalities. Specialist centres will then liaise with the local centres daily to provide advice and if necessary transfer.

3.2 Home IV antibiotics

All PCD patients will have access to a suitable home IV antibiotic service, if appropriate. If patients are unwell and need IV antibiotics, they will be assessed in the specialist centre or local hospital. This will be in conjunction with their management plan. Specialist centres will liaise with the local centres and arrange for the delivery of a suitable course of home IV antibiotics and monitoring as needed.

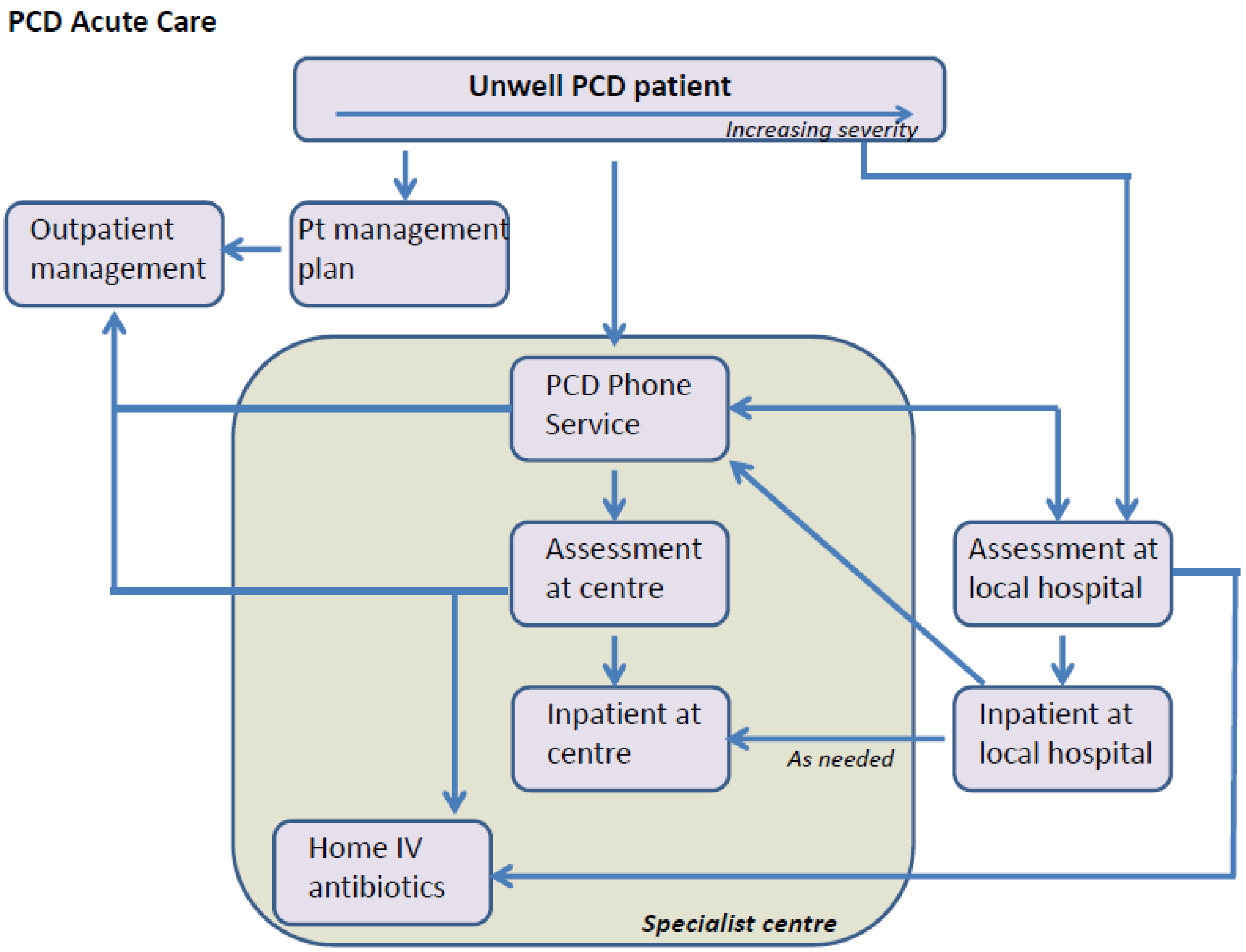

Figure 2

3.3 Population covered

This service is for all adult patients with a diagnosis of PCD. This can be patients with a diagnosis made by the PCD Diagnostic Service, patients with a diagnosis of PCD made by a diagnostic service outside the UK, with diagnostic criteria compatible with those in England, or in rare cases, patients in whom a definitive diagnosis has not been made, but in whom the diagnosis is extremely likely (in the consideration of the PCD Specialist). It will be a lifelong service for these patients. The service can be accessed by any eligible patient with a diagnosis of PCD irrespective of gender, age, sex, disability, religious belief. Interpreters or use of a language line will be provided for families for whom English is not their first language.

This service covers patients registered with an English General Practitioner, resident in resident in the European Union and eligible for treatment in the NHS under reciprocal arrangements. Patients from Wales, Scotland and Northern Ireland are not part of this commissioned service and the NHS Hospital Trusts must have separate arrangements in place in order to accept patients from these countries.

3.4 Any acceptance and exclusion criteria and thresholds

This service is a lifelong service for **all** adult patients with a diagnosis of PCD. Children (<16) and patients found not to have PCD are not covered by this service.

3.5 Interdependencies with other services/providers

Figures 1 and 2 illustrate the interdependencies between the Highly Specialised Centre, secondary and primary care. It is proposed that the service is wholly commissioned by NHS England. There will be significant liaison with local hospitals that will provide some acute assessments and inpatient care in liaison with the specialist centres.

There will be a specialist nurse staffed phone line during working hours with direct consultant input to help provide this link. Home IV antibiotics will be organised through the specialised centres with an external provider and with the collaboration of local hospitals.

Outreach clinics will be provided jointly with local and specialised centres. Transition clinic will be held jointly by the adult and paediatric PCD management services.

The PCD database will span both the adult and paediatric services. For patients with severe disease there will be interdependencies with ventilation services for patients that need assisted ventilation and interdependencies with transplant services for appropriate patients.

The service will link closely with the Diagnostic service for Primary Ciliary Dyskinesia.

4. Applicable Service Standards

4.1 Applicable national standards e.g. NICE

Best practice guidelines from specialist centres were collated to produce a European consensus statement in children with PCD (Barbato et al Eur Respir J 2009; 34:1264-76) which recommends regular follow up in specialist centres with an MDT approach as described in this service specification. The British Thoracic Society guidelines on bronchiectasis were produced in 2010 (Pasteur et al Thorax 2010 65S). These provide general bronchiectasis advice but do not provide comprehensive PCD specific guidance.

- There is a requirement to hold national audit meetings involving all designated centres on an annual basis.
- Each centre must assure that:

1. All practitioners participate in continuous professional development and networking
2. Patient outcome data is recorded and audited across the service
3. They participate in the national audit commissioned by NHS England. Audit meetings should address:

- Clinical performance and outcome
- Process-related indicators e.g. efficiency of the assessment process, prescribing policy, bed provision and occupancy, outpatient follow-up etc.
- Stakeholder satisfaction, including feedback from patients, their families, referring clinician and General Practitioners doctors and GPs.

4.2 Applicable standards set out in Guidance and/or issued by a competent body (e.g. Royal Colleges)

Nothing additional.

5. Applicable quality requirements and CQUIN goals

5.1 Applicable quality requirements (See Schedule 4 Parts A-D)

There is a requirement to hold national audit meetings involving all designated centres on an annual basis.

Each centre must assure that:-

1. All practitioners participate in continuous professional development and networking
2. Patient outcome data is recorded and audited across the service
3. They participate in the national audit commissioned by NHS England. Audit meetings should address:
    - Clinical performance and outcome
    - Process-related indicators e.g. efficiency of the assessment process, prescribing policy, bed provision and occupancy, outpatient follow-up etc.
    - Stakeholder satisfaction, including feedback from patients, their families, referring clinician and General Practitioners doctors and GPs.

5.2 Applicable CQUIN goals (See Schedule 4 Part E)

To be agreed with the Commissioner.

6. Location of Provider Premises

The Provider's Premises are located at:

No more than 4 centres are proposed, linking into the 4 paediatric PCD management services.

7. Individual Service User Placement

Not applicable.

Appendix Two

Quality standards specific to the service using the following template:

Quality Requirement	Threshold	Method of Measurement	Consequence of breach
Domain 1: Preventing people dying prematurely			
% of adult patients diagnosed with PCD managed in adult PCD specialist centres	90%	Comparison of the PCD diagnostic records and the PCD management records	To be addressed at the annual audit meeting
Domain 2: Enhancing the quality of life of people with long-term conditions			
% of patients in the PCD management service offered an annual review appointment	90%	Annual review records	As per Standard NHS Contract General Conditions Clause 9 (GC9) Remedial Action Plan
Domain 3: Helping people to recover from episodes of ill-health or following injury			
% of patients seen by a physiotherapist at annual review	90%	Annual review records	As per Standard NHS Contract General Conditions Clause 9 (GC9) Remedial Action Plan
Domain 4: Ensuring that people have a positive experience of care			
% of patients seen by a nurse specialist at annual review	90%	Annual review records	As per Standard NHS Contract General Conditions Clause 9 (GC9) Remedial Action Plan
Domain 5: Treating and caring for people in a safe environment and protecting them from avoidable harm			
% PCD inpatients at or discussed with the specialist centre	70%	Record of inpatient stays collected from patient at inpatient review and compared	To be addressed at the annual audit meeting