E13/S(HSS)/g

2013/14 NHS STANDARD CONTRACT
FOR PRIMARY CILIARY DYSKINESIA (PCD) DIAGNOSIS AND MANAGEMENT SERVICE (CHILDREN)

SECTION B PART 1 - SERVICE SPECIFICATIONS

Service Specification No.	E13/S(HSS)/g
Service	Primary Ciliary Dyskinesia (PCD)Diagnosis and Management Service(Children)
Commissioner Lead	
Provider Lead	
Period	12 months
Date of Review	

1. Population Needs

1.1 National/local context and evidence base

The expertise required to detect and manage early signs of lung disease in Primary Ciliary Dyskinesia (PCD) is specific to this disease, and without this specialist input, PCD patients can develop severe progressive lung disease and bronchiectasis (Noone 2004). Bronchiectasis can occur in infancy demonstrating the need for early instigation of appropriate treatments. Once irreversible lung damage is established, significant medical input is required with regular hospitalisation for antibiotics, ventilatory support and sometimes, surgical intervention including lobectomy (Pedison 1983, Noon 2004, Kennedy 2007) and lung transplantation (Noon 2004, Pifairy 2010).

A number of longitudinal studies have reported that lung function deteriorates before diagnosis of PCD but stabilises following diagnosis and appropriate treatment (Corki 1981, Elerman 1997 and Hellinckx 1998) demonstrating that lung damage results from under-treatment and can be prevented by early treatment and intensive therapy (Ellerman 1997). A more recent longitudinal study has shown that lung function may continue to deteriorate in a minority of cases despite therapy (Marthin 2010) but the decline in lung function is reduced.

Importantly, poor lung function improves in some patients once aggressive treatment is instigated (Marthin 2010).

The early detection and management of infections in children with PCD is difficult, in comparison to patients with, for example, cystic fibrosis (CF). Unlike CF patients, children with PCD have a persistent wet cough even when well. Infection is generally

accompanied by a normal chest x-ray, the chest may sound within normal limits for that individual and infection is rarely accompanied by pyrexia.

Bronchoscopy is sometimes the only way to obtain samples for microbiological analysis, particularly in children <6 years. The indications for bronchoscopy are different in PCD patients. Additionally, the bronchoscopy need to be undertaken by someone who regularly scopes children with PCD as the gross findings are different to patients with other diseases such as CF. Because of the problems with secretions in PCD, patients are at high risk of atelectasis and collapse if a specialist physiotherapist is not present during the procedure to clear secretions. An expert knowledge of this patient group is therefore essential to detect early changes and instigate vigorous management to prevent the long term damage that may lead to early respiratory failure in adulthood.

Chronic otitis media management differs from those with non-PCD related ear disease. In particular, grommet insertion leads to chronic otorrhea, persistent perforation and often is not accompanied by improved hearing (Campbell 2009). Constant nasal symptoms are a major complaint of PCD children and are associated with sinusitis. Audiology and Ear, Nose & Throat (ENT) assessment by people experienced in PCD is therefore required to ensure appropriate management required for normal hearing, prevention of speech delay and ultimately, normal educational achievement.

A PCD service is markedly different in structure to a CF or general respiratory clinic. In addition to the specialist skills required of the respiratory clinician and hysiotherapist, the service needs staff not available in CF clinics, in particular ENT specialists and audiologists with an interest in PCD. Patients with PCD do not have pancreatic insufficiency or malabsorbtion, and the dietetic input available in CF clinics is inappropriate for this patient group. There is evidence that patients managed in CF clinics or non-specialist clinics have inappropriate management due to inappropriate clinic personnel e.g. missed hearing impairment and unsuitable vitamin and dietary supplements. The patient literature and educational media provided by the service will be disease appropriate and specific, coming from the PCD Support Group rather than the CF Trust.

The medical leads of the service have published national and international evidence based guidelines for the management of PCD (Barbato 2009; Bush 2007)

2. Scope

2.1 Aims and objectives of service

PCD is a relatively rare (estimated between 1:26000 and 1:40000) hereditary disorder characterised by chronic infection of the upper and lower airway. In addition, approximately half the patients have situs inversus and male infertility is common.

The airway symptoms are caused by impaired mucociliary clearance which results in accumulation of airway secretions often containing bacteria and allergens, leading to inflammation and chronic infection. The impaired mucociliary clearance is a consequence of abnormal ciliary beat function. Without appropriate early treatment progressive chronic lung disease and bronchiectasis develop, and mismanagement of hearing impairment is common.

The service will provide access to specialised clinical care for all children in England with PCD.

The multi-system problems related to PCD are highly disease-specific and children with PCD require access to a multi-disciplinary team (MDT) uniquely specialised in these systems to optimise their care.

Aims

The service aims are:

- to ensure that all patients, wherever they live, have access to and are managed according to current PCD standards of care
- to ensure children are seen at least annually by the specialist PCD MDT. Their care throughout the year will be provided by local healthcare professionals as recommended by the specialist team. to
- to diagnose early, and then monitor and limit disease progression
- to coordinate care and management of patients with PCD ensuring that all receive appropriate respiratory, ENT, cardiac and physiotherapy care and care for other conditions associated with PCD e.g. learning difficulties, hydrocephalus, biliary atresia etc
- to provide education to health care professionals, patients/families and educators on the implications and management of PCD making it a priority to emphasise the potential severity of the condition and prevent the poor outcomes in under treating this disease
- to reduce the morbidity and mortality related to PCD, as well as the economic burden, associated with late diagnosis and poorly managed disease.

The objectives of the service are to:

- provide a specialist multidisciplinary service for children diagnosed with PCD throughout England
- to limit the long-term complications of PCD by ensuring that all patients receive specialist care to enable them to enter adult services with optimal lung function and hearing, and the health-education to maintain good PCD management in adulthood.

The purpose of the service will be to:

- develop an equitable national PCD service in which children with PCD have access to a specialist PCD MDT at least annually

- provide personal management plans for each patient annually, the provision of which will be delivered by local services
- provide specialist advice and support to local providers of care, and in difficult cases to review the patient between annual visits
- develop and share national PCD protocols and guidelines, ensuring that 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
- provide a specialist PCD bronchoscopy service
- improve awareness and management of PCD within the UK by education and provision of an excellent service
- offer patient-centred assessment and management regarding the many organ specific problems associated with PCD
- minimise impact on the patient and their family life, education and work practice.

Outcome measures

The service must collect the following data annually to monitor clinical outcomes throughout childhood:

- lung function (% predicted forced expiratory volume (FEV_1) and forced vital capacity (FVC))
- audiometry (decibels of hearing loss) relative to at diagnosis
- health-related quality of life, disease-specific and generic, for children and their families.

At transition to adult services must report:

- percentage of patients with normal lung function
- improvement in hearing (decibels) since diagnosis.

2.2 Service description/care pathway

Care and referral pathways

The attached care pathway figure is set out below. At the outset of the service, all children who have previously been identified with PCD will enter the service. From that point, newly diagnosed patients must be referred to the service from the PCD diagnostic service. Occasionally patients diagnosed at PCD specialist centres in other countries will be referred.

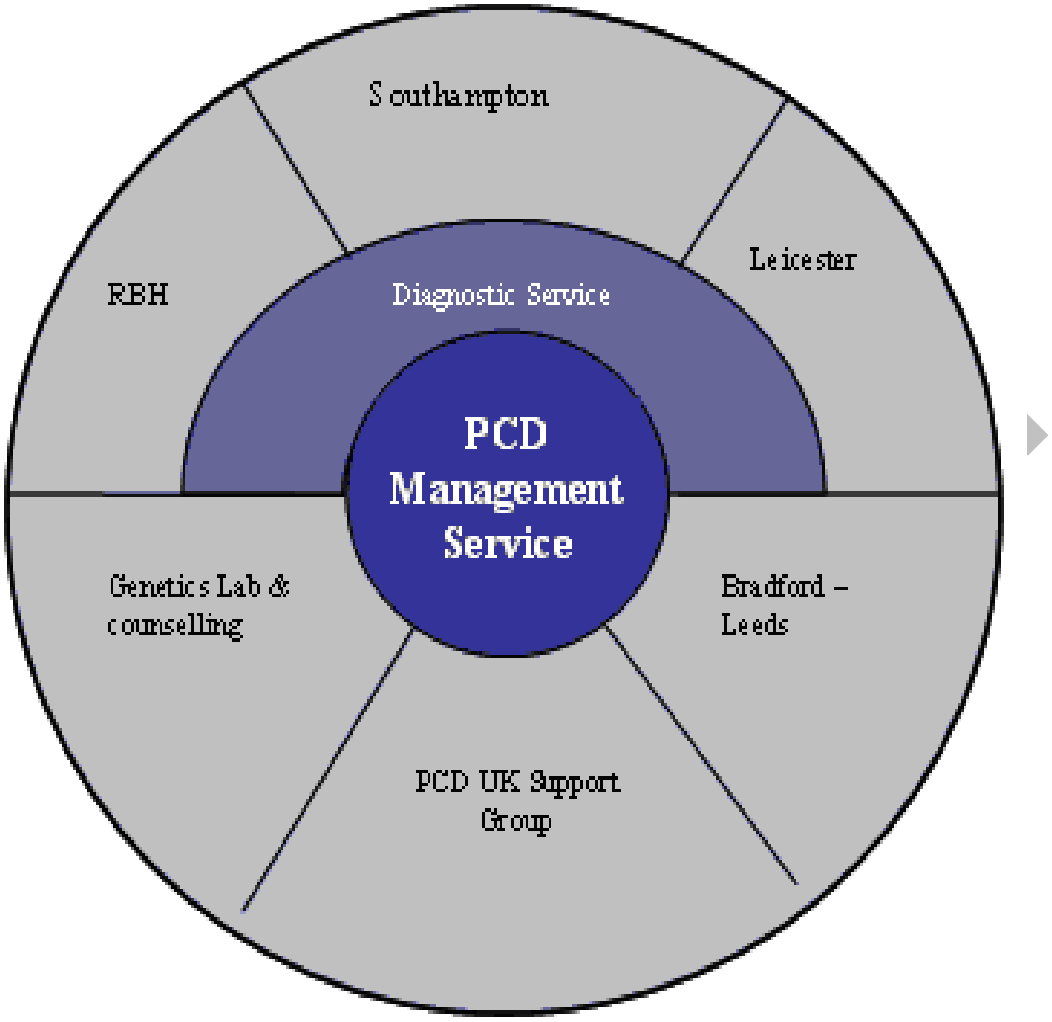
The patients must be seen at least annually by the PCD MDT as an outpatient. Care throughout the year will be recommended by the specialist PCD team, for delivery by local teams.

Patients must transition to adult services with recommendations for on-going care.

Structure of proposed service

- Out-patient based service for patients referred to diagnostic centres
- Four PCD management centres in England. Patients will be shared between centres based on geographical location.
- Based at the three existing PCD Diagnostic Centres (Southampton, Leicester and the Royal Brompton Hospital), with an additional centre (Bradford/Leeds) to reflect the high prevalence of PCD in their local population and to provide a geographically accessible centre for patients in the North of England.
- MDT clinic structure to provide patient access to all the specialists involved in the patient's care. The required combination of specialists is unique to the specific problems encountered by PCD patients and cannot be met by combining with other respiratory specialist clinics catering for patients with other forms of chronic suppurative lung disease.
- A disease monitoring and management programme with a remit to provide advice, oversee and coordinate management.
- A service tailored to key life stage events to maximise the benefits of the programme for the changing needs of patients with transitional care into adult practice.
- In-patient admissions at local NHS Trust. Supported by this service where necessary but in-patient care is not part of the proposed service.

Figure: Structure of the proposed service



Clinic provision:

Total PCD clinics – based on 304 patients nationally per annum for annual review, transition and follow up, will require 100 clinics per year nationally:

- annual review - six patients per consultant clinic requiring the equivalent of 60 annual review clinics per year (15 clinics per centre)
- follow up clinics for patients who need more intense levels of specialist care. It is estimated that 40 clinics nationally (10 per centre) per annum will cater for this.
- transition clinics – each centre will lead the adolescent patient towards transition into their local adult clinic. These will be incorporated into the proposed 25 clinics.
- outreach clinic capacity will be available within the proposed staffing levels in this service. A visiting PCD consultant, physiotherapist, clinical nurse specialist [CNS] and respiratory technician visit the centre to join the local team in seeing patients. It is proposed that local centers hosting outreach clinics will also invite local audiology and ENT specialists to attend.
- pre- and post-clinic MDT meetings.

- all patients are discussed prior to each visit, so that all diagnostic and management needs can be met and dealt with on the patient visit.
- post-clinic meetings will ensure that clear guidelines for on-going management are discussed and disseminated to the shared care team. Sputum cultures will be checked and acted upon where required.

Each PCD clinic will therefore require an additional four hours of time for meetings, administration processes and follow up of results.

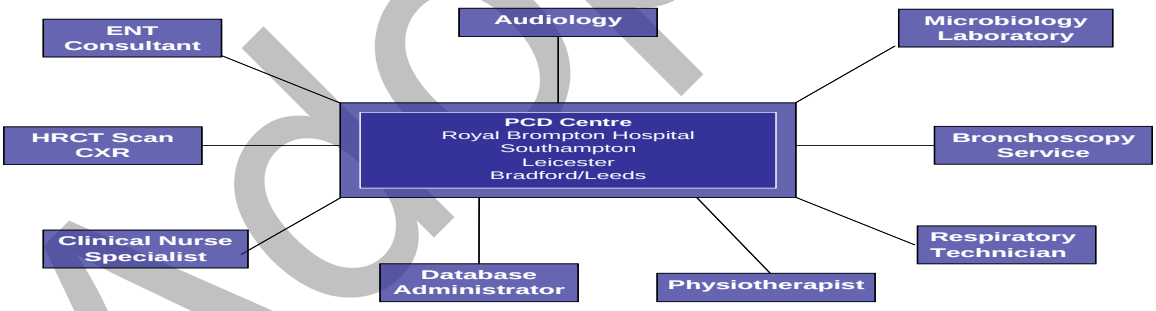
Advisory phone service

A direct line must be available to families and shared care professionals within working hours. It is anticipated that phone advice will contribute to a half consultant session, and two nursing sessions per week. The Clinical Nurse Specialist (CNS) and physiotherapist will also carry mobile phones to extend this advisory role when they are away from the office.

Clinic structure

Each patient will have access to the following specialists at each visit; a respiratory PCD consultant, an ENT consultant, an audiologist, a respiratory technician (lung function and any outstanding diagnostics), PCD clinical nurse specialist, PCD physiotherapist, audiology technician.

Figure: clinic structure



Additional services

- lung function. Annual review patients must have formal lung function testing in a specialist laboratory. Longitudinal lung function trends will be recorded and acted upon
- sputum culture for micro-organisms
- bronchoscopy - run by the PCD consultant for:
    - patients not responding to treatments as expected, or where there is a difficulty obtaining sputum samples for microbiology. It is expected that this

will involve about 10-12 patients/year
- newly diagnosed patients [aged <5years] will be offered bronchoscopy to determine their infection/colonization status. This is impossible to ascertain in this age group and will provide essential guidance to the future management of each patient.

The relevant trusts must provide the facilities and equipment for the PCD service:
- imaging – Chest X-ray (CXR) and high resolution computed tomography (CT) [HRCT] scanning are required to assess and monitor extent of disease. Around 10% of patients may require HRCT per annum, with most patients having at least one scan in their period of paediatric care.
- database – all patient data must be entered following annual review. Each centre will require the same database and technical support for data entry and maintenance support.

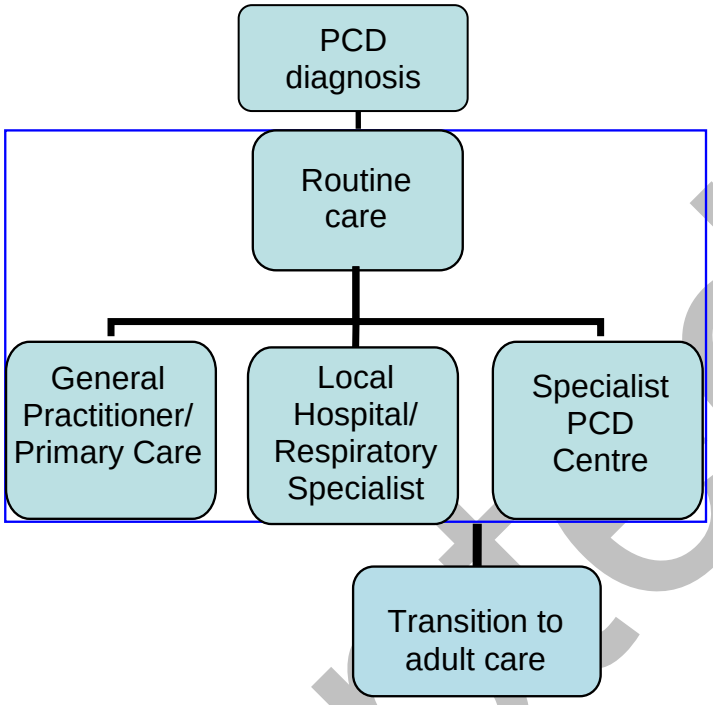
Risk management

Care delivered by the PCD management service providers must be of a nature and quality to meet the care standards, specification and agreement for the service. It is the trust's responsibility to notify the commissioner on an exceptional basis should there be any breaches of the care standards. Where there are breaches any consequences will be deemed as being the trust's responsibility.

Patients must be managed in line with the specification and care standards. Any deviation from these which has not been approved by NHS England is at the trust's risk both clinically and financially. It is the trust's responsibility to inform the commissioners of any such non-approved deviations on an exceptional basis.

Where a patient's presentation challenges the assumptions that underpin the specification, service standards and contractual arrangements, it is the trust's responsibility to inform the commissioners on an exceptional basis, prior to any treatment (except for emergency treatment) so that the implications of the patient's requirements can be considered. This does not affect situations where the Individual Funding Application process applies.

Service model and care pathways:
PCD pathway
Routine care



Routine care

Entry to pathway: PCD Diagnosis

- diagnosis of PCD made by English PCD Diagnostic Service
- diagnosis of PCD made by recognised international diagnostic centre
- occasionally, patients with a strong likelihood of PCD but in whom a definitive diagnosis remains difficult, will be reviewed in the service.

GP

- The specialist centres will provide paper and internet-based guidelines for the management of PCD
- The GP will remain responsible for non-PCD related health care issues
- Immunisations remain the responsibility of primary care. Specific guidelines (e.g. for epidemic influenza) will be provided by the centres
- The GP will inform the specialist centre of significant treatments or patient issues.

Local hospital

- The specialist centres will provide paper and internet based guidelines for the management of PCD
- Patients should be offered appointments at least 3 monthly at either their local

hospital or specialist centre.

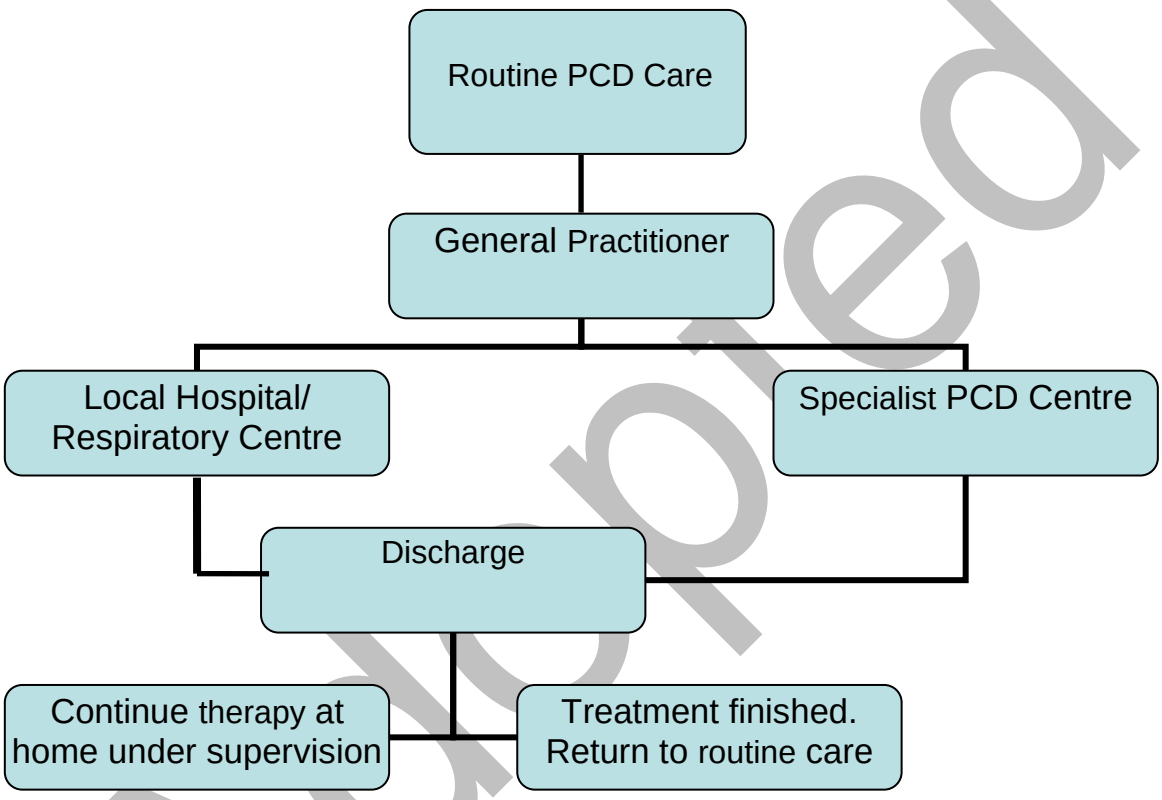
- The clinician reviewing the patient should have a specialist interest in paediatric respiratory care.
- The patient should be seen by a multi-disciplinary team including a physiotherapist.
- The patient should have lung function measured (if age appropriate) and sputum collected for microscopy, culture and sensitivity (MC&S) at each visit.
- All communication should be copied to the specialist centre, GP and parents (or patient if >14 years).
- Open access for advice from local team. The local team will also have access to the specialist team to refer on questions and receive advice that can be passed back to the families.

Specialist centre

- The specialist centre is responsible for providing local hospitals and GP's with paper based and/or internet based guidelines.
- The specialist centre will see every patient at least once a year.
- Patients may be bought to additional clinics for evaluation where they continue to deteriorate despite discussion and phone advice.
- Every patient shall have an annual review conducted by a specialist centre.
- At the annual review the patient and family shall be seen by a multi-disciplinary team to include:
    - clinicians – paediatric respiratory and ENT
    - specialist nurse
    - physiotherapist
    - respiratory technician (lung function)
    - audiologist
- At every annual review the patient shall have the following monitored if age appropriate:
    - lung function
    - pulse oxymetry
    - sputum (MC&S)
    - physio techniques – monitor and update
    - exercise plan – monitor and update
    - lung clearance index
    - any bloods etc.
- In addition the centre shall provide the following as required:
    - additional clinic review and assessment for patients not doing well
    - support for Disability Living Allowance (DLA) forms
    - bronchoscopy
    - assessment for planned general anaesthetic
    - fitness to fly assessments
    - chest x-rays and High Resolution Computed Tomography (HRCT)
    - advice regarding hearing/provide hearing aids.

- provide adjuncts for physiotherapy.
- peer specialist advice to local hospital (physio to physio, nurse to nurse, etc)
- home assessment
- school reviews.

PCD pathway
Acute care



PAEDIATRIC PCD CLINICAL CARE PATHWAY

ACUTE

Entry to pathway:

- Patient under the routine care of PCD Management service. Acutely unwell.

GP

- The specialist centres will provide Standard Operating Procedures (SOPs) for common management problems including indications for discussion with the centre.
- Specific advice may be given by a local hospital or specialist centre to go to the GP or A&E etc. This will depend on the nature of the symptoms.

- Guidelines will be available to all shared care professionals in bound form or online based via the PCD Family Support Group website.
- It is not unusual for advice to start oral antibiotics prior to a hospital visit. The hospital may fax the GP to provide a prescription.
- The GP may be contacted about non-PCD related illness. The centre should be informed of significant events or treatments. 'Normal childhood illnesses', e.g. chickenpox, gastroenteritis, should be managed in the usual way.
- The centre should be contacted if there are any concerns.

Local hospital

Unplanned hospital admissions may be required for a number of reasons, most usually respiratory exacerbation.

The service will encourage an open admissions policy for PCD patients, possibly on a named patient basis, to facilitate these admissions.

- The PCD centre should be informed within working hours
- Whilst in hospital adherence to cross-infection recommendations is essential Treatment for chest exacerbations should follow Standard Operating Procedures provided by the centre, or individualised plans provided by the centre. In particular, mono-therapy is not advised. Treatment is usually 10-14 days
- Full multi-disciplinary care including nursing and physiotherapy.

Specialist centre

In complex cases not responding to standard treatment, review in a specialist centre may be required for assessment of condition and treatment regimes. It is the aim that alterations in therapy can be continued in the local centre, but on occasion, these changes and assessments may require a short admission to the specialist hospital.

This is most likely where specialist tests and procedures, such as bronchoscopy are required. Completion of treatment can then be completed at the local hospital closer to home.

Discharge

After discharge early follow-up is advised. If appropriate, this may be by phone. The patient is then returned to usual care.

Days/hours of operation

The PCD service operates non-emergency NHS hours - Monday to Friday, 9-5pm. It does not operate bank holidays.

Discharge criteria & planning including any transition arrangements

A process of transition must commence at least two years prior to discharge to adult services

2.3 Population covered

Geographic coverage/boundaries

The service covers patients registered with an English General Practitioner, resident in the European Union and eligible for treatment in the NHS under reciprocal arrangements. Patients from Scotland, Wales and Northern Ireland are not part of this commissioned service and the trust must have separate arrangements.

2.4 Any acceptance and exclusion criteria

Accessibility/acceptability

The service is commissioned by NHS England for all eligible patients from England and Scotland. The service can be accessed by any eligible child 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.

The service will monitor equitable geographical access across the country and take actions to address gaps in access. The provider will provide information to patients on public transport access and accommodation for patients and relatives as needed.

Close family members are encouraged to attend the clinic in order to follow a family-orientated approach.

The provider will co-operate with the commissioner in undertaking Equality Impact Assessments.

Referral criteria, sources and routes

- Patients with a diagnosis of PCD made by the PCD diagnostic service.
- Patients with a diagnosis of PCD made by a diagnostic service outside the UK. The service will be expected to have diagnostic criteria compatible with those in England; otherwise the patient should be seen in an English diagnostic centre prior to entry to this service.
- 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 diagnostic service specialist).

Exclusion criteria

- Patients who are too old for paediatric services (16-18 years)

2.5 Interdependencies with other services

Patients must be seen in the PCD specialist clinic annually. The clinic will provide individualised and standard guidelines for the management of the patient throughout the year to the patient's local MDT and GP.
Telephone support will be available during normal NHS working hours (9 to 5) for local services.
Non-specialist management e.g. IV antibiotics will be recommended by the specialist service, to be delivered locally.
Specialist investigations (bronchoscopy, sputum induction) will be provided by the PCD centre. For this reason, patients may require to be seen between annual review visits, following discussion with the specialist centre.
The patient pathway (attached) reflects the interdependency for the service.

Relevant networks and screening programmes

None

3. Applicable Service Standards

3.1 Applicable national standards e.g. NICE, Royal College

The nationally designated PCD Management service must be fully integrated into their trust's corporate and clinical governance arrangements.

See also NHS England Service Standards (to be developed)

Continual Service Improvement Plan

Commissioner and provider have a commitment to working together to continually improve the service and react to innovative and dynamic ideas. The provider and commissioner have a responsibility to continually review and redesign services and consider and act upon requests of the other party.

Service improvement may be stimulated through areas such as:

- complaints
- monitoring information
- provider feedback
- learning from other services
- needs assessments
- service user feedback/patient and public involvement
- research
- policy/ guidance on best practice e.g. NHS Institute for Innovation and Improvement
- other communication with stakeholders

This must be an on-going and dynamic process.

4. Key Service Outcomes

Lung function (% predicted FEV_1 and FVC).
Audiometry (decibels of hearing loss) relative to at diagnosis

- Health related quality of life, disease-specific and generic, for children and their families

Transition Measures

- % with normal lung function
- Improvement in hearing (decibels) since diagnosis

Quality & Performance Standards

All centres must work within the clinical governance structure of their acute medical trust. Where outreach clinics are to be held agreements must be made regarding clinical governance with the host trust.

The lead consultant for this service must be on the General Medical Council (GMC) specialist register for paediatric respiratory medicine.

The centres must develop protocols for all clinical investigations and management which will be auditable.

In keeping with the diagnostic service, regular visits will be made between the three centres. It is has been established that this is an excellent method of developing best practice, particularly in areas where the evidence base is weak It ensures a vibrant and moving national service with all centres adhering to the same standards.

Protocols for audit will be developed and shared amongst the centres and where appropriate with health professionals who share the care of our patients.

Critical incidents must be reported within the acute medical trusts. All serious adverse events must be discussed and reported to NHS England. In addition, it will be shared with the other centres if this seems appropriate.

Summaries of audits must be presented to NHS England.

Formal meetings between the centres must be held at least annually:

- to review audit results
- review all publications relating to the management of PCD
- in light of the above, to amend the management guidelines if required.
- to discuss management of ‘difficult cases’.

5. Location of Provider Premises

Four PCD management centres in England. Patients will be shared between centres based on geographical location:

Royal Brompton & Harefield
Sydney St
London SW3 6NP

University Hospitals Leicester
Gwendolen Rd
Leicester LE5 4PW

Southampton University Hospital Trust
Southampton General Hospital
Tremona Road
Southampton
Hampshire SO16 6YD

Leeds General Infirmary
Consultant Respiratory Paediatrician.
Great George Street
Leeds LS1 3EX

In addition, outreach clinic capacity must be available within the proposed staffing levels in this service. By prior agreement with local providers, a visiting PCD consultant, physiotherapist, CNS and respiratory tech must visit outreach clinics to join the local team in seeing patients. It is proposed that local centres hosting outreach clinics must also invite local audiology and ENT specialists to attend.

Sub-contractors

Bradford Hospital NHS Trust
Duckworth Lane
Bradford BD9 6RJ