

E05/S(HSS)/a

**2013/14 NHS STANDARD CONTRACT
FOR PULMONARY HYPERTENSION SERVICE (CHILDREN)**

PARTICULARS, SCHEDULE 2 – THE SERVICES, A – SERVICE SPECIFICATION

Service Specification No.	E05/S(HSS)/a
Service	Pulmonary hypertension service (Children)
Commissioner Lead	
Provider Lead	
Period	12 months
Date of Review	

1. Population Needs

1.1 National/local context and evidence base

Description

‘Pulmonary hypertension’ means a high pressure in the circulation of blood through the lungs. The term describes a condition in which the millions of small blood vessels in the lungs gradually become obstructed by a growth of tissue from the vessel wall (pulmonary vascular disease). It is a progressive, incurable disease leading to right heart failure and death. In idiopathic pulmonary hypertension (IPAH) the mean survival time from diagnosis to death in the untreated patient is 2.8 years in adults but only 10 months in children. The causes of pulmonary hypertension associated with other diseases or congenital abnormalities (APH) are more varied and complex in children than adults.

By 2001 evidence of drug efficacy led to the establishment of the UK pulmonary hypertension service for children (a managed clinical network).

Evidence base

The service has considerably improved the historical 50% mortality from 0.8 years to about 10.7 years (average for idiopathic pulmonary hypertension).

[historical 50% mortality rate of 0.8 years vs cohort of 2002 to 2005 of 7.2 years, vs cohort of 2005-2008 of 10.7 years (published in Moledina et al, HEART 2010)]. The figures below give a graphical representation of the survival analysis described by disease type.

Figure 52: Survival rates for patients treated at the Pulmonary Hypertension clinics between 2002 and 2007, with either idiopathic pulmonary hypertension (iPAH) or acquired pulmonary hypertension (aPAH).

Cummulative Survival: iPAH vs aPAH

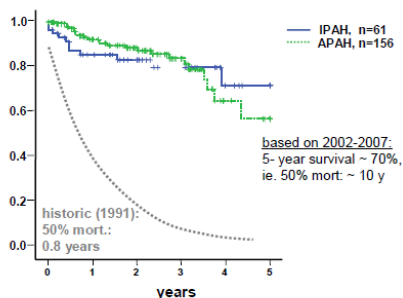
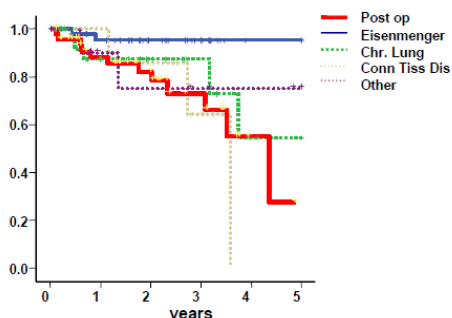


Figure 53: Survival rates for children treated at the pulmonary hypertension clinics between 2002 and 2007, by type of disease.

Cummulative Survival: ass. PAH



2. Scope

2.1 Aims and objectives of service

The service for pulmonary hypertension (PH) in children provides care for children with all forms of pulmonary hypertension, the newborn form (PPHN) being excluded.

Great Ormond Street Hospital for Children NHS Foundation Trust (GOSH) is the only UK centre allowed to design treatment plans and to authorise the prescription of the drugs.

The structure of the service is unique and without parallel in Europe and worldwide, internationally leading the practice and science of paediatric pulmonary

hypertension.

Its aims are to establish a managed clinical network in order to:

- ensure **equity of care throughout the UK**
- improve **survival**
- improve **quality of life**
- continue to **research** the clinical management and pathobiology of pulmonary hypertension in childhood;
- **educate** health professionals to improve disease recognition and prompt referral

The diagnosis of pulmonary hypertension has a devastating impact on both child and family and ensuring rapid diagnosis prompt treatment and support in the home, school and community is vital.

The service works closely with the patients' organisation Pulmonary Hypertension UK.

2.2 Service description/care pathway

The service comprises of the following:

Patient Referral

Patients who are diagnosed or have a high level of suspicion of, having pulmonary hypertension, can either be seen directly at the outpatient clinic at Great Ormond Street Hospital for Children NHS Foundation Trust being referred from another hospital, or are seen at an outreach clinics, being referred by that hospital and a PH clinical network member. In general, direct referrals by a general practitioner (GP) are rare

Patient Assessment

The patient is assessed by clinical examination which includes:

- electrocardiogram (ECG)
- echocardiogram
- vital signs
- six minute walking test, where appropriate, when presenting in the outpatient clinics

When presenting as an inpatient, a number of other tests may be added, such as:

- magnetic resonance imaging (MRI)
- computerised tomography (**CT**) scan,
- cardio-pulmonary exercise **testing** (CPEX)
- cardiac catheterisation

All tests are checked and seen by the lead consultant or in uncomplicated cases by the registrars of the pulmonary hypertension (PH) Team

Patient Treatment

Patients are treated under packages of care using a treatment algorithm modified from the adult equivalent. Almost all patients undergo cardiac catheterisation to determine pulmonary vascular resistance before and after vasodilator testing to identify the best treatment option – unless they are too ill, in which case treatment is instituted immediately. For the majority (95%) this normally entails intravenous epoprostenol, various prostacyclin analogues, the dual endothelin receptor antagonist bosentan, and the phosphodiesterase III inhibitor sildenafil, in different combinations depending on severity

Children with severe pulmonary hypertension who have syncopal attacks (fainting) when they experience a sudden, further increase in pressure, need a 'balloon atrial septostomy' (BAS), as an attack can be fatal. This involves making a small hole between two fore-chambers in the heart, to enable blood flow to the brain in cases of prohibitively high pulmonary vascular resistance.

Cardiac catheterisation may need 3 days of hospital stay, or when connected with iv Epoprostenol start, 10 days including parent education. The 'balloon atrial septostomy' as such does not add to the length of hospital stay.

Ongoing Support

Established pulmonary vascular disease is incurable. Therefore long-term support in the community is essential. This is secured by a clinical nurse specialist who has established widespread links with paediatric community nurses throughout the UK and with several children's hospices.

Long Term treatment

Many children eventually fail medical treatment, need lung or heart lung transplantation and are assessed for cardiothoracic transplantation by the service at GOSH.

Discharge planning

There are four pathways for patients to leave this service:

- PH resolved
- patient transplanted
- patient died
- patient transitioned to adult PH service

PH resolved

Some forms of pulmonary hypertension, in the 'associated PH' category, begin with

an event causing the PH (infection, cardiac surgery) and resolve when the original cause has disappeared and the pulmonary vascular system has recovered; such recovery can be highly variable across individual patients. Further, while this process is observed in individual patients, there are few published data.

Patient transplanted

The availability of thoracic organs (lung or heart/lung block) is very scarce. Only a few patients have been successfully transplanted with good/intermediate result so far. Patients with idiopathic pulmonary hypertension, and patients with PH due to associated disease with severe changes, are seen by the transplant team and are assessed, having been formally referred. Following the first initial assessment (and acceptance) they continue to be seen on a long term routine follow up basis by the transplant team, as well as (and coordinated with the cardiothoracic transplant service appointments) by the PH team as indicated. During that time, the primary care responsibility remains with the PH team, in cooperation with the transplant team. Once transplanted, the primary care responsibility shifts to the transplant team.

Patient deceased

Most deaths occurs outside GOSH, and most outside of their local hospitals. The PH service aims to obtain all information around the death and tries to establish the precise cause (lung bleeding, arrhythmia, right ventricular (RV) failure, pulmonary artery (PA) rupture). When appropriate, a limited (small thoracic) autopsy is suggested, with appropriate family consent; this is actually done in a minority of patients, in whom the biopsy could answer a question important to that patient (pulmonary/myocardial changes, confirmation of diagnosis and severity).

Patient transitioned to adult PH service

Most patients are transferred to the appropriate adult PH services (idiopathic, congenital heart disease, connective tissue disease) based on diagnosis and place of residence, when they are 16 years old. In cases where 'paediatric issues' remain, their time with GOSH may be extended to their 18th birthday, and in exceptional cases even longer if clinically necessary. Typical patients for this are those with trisomy 21. The transition process is formal, including an outpatient visit where the paediatric PH consultant joins the patient and attends together the first outpatient clinic of the adult PH service. On these occasions, a small number of PH patients may have been transferred at the same time.

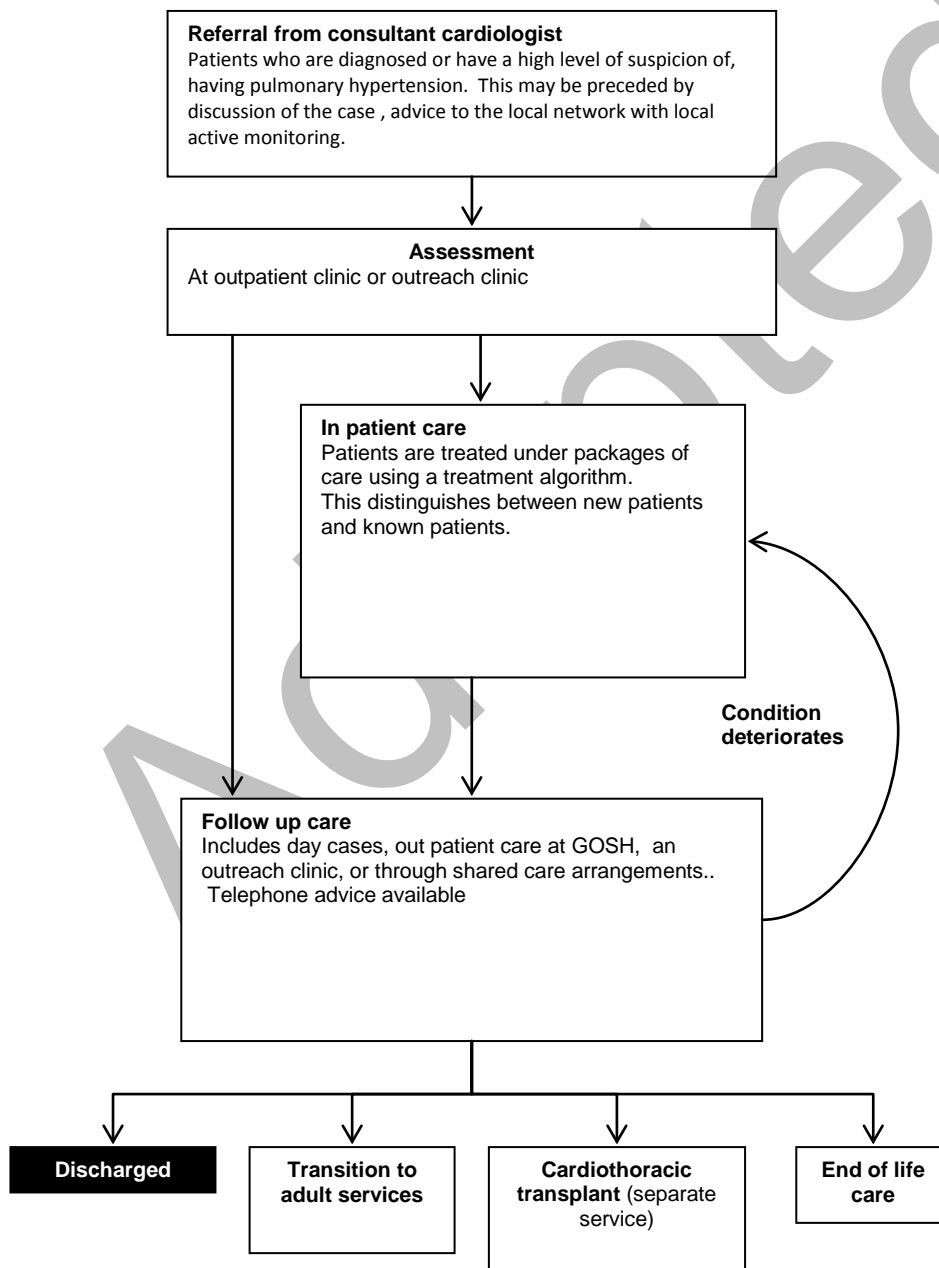
Patient and carer information

The correct information about meaning and prognosis of the disease, as well as ensuing procedures and plan, is a priority when meeting any new patient. This is crucial in establishing trust and a good patient relationship. Ensuring this good communication between all the professionals involved essential. Thus, the first appointment is usually in the day care ward to avoid time pressure and physical

crowdedness. Letters are copied to the parents/patients and all others involved.

The parents are given general information about pulmonary hypertension and the treatment together with specific information about their own child and his/her treatment and emergency telephone numbers. General practitioners, satellite centres, local paediatricians, community nurses, school and 'Healthcare at Home', (the company contracted to deliver drugs to the home; epoprostenol, plus infusion pumps etc. and bosentan) are kept informed.

Paediatric pulmonary hypertension service care pathway



2.3 Population covered

Patients are referred to a hospital in the PH clinical network from all parts of the UK. Also, children are referred directly to GOSH from other European Union (EU) countries.

In addition, the service provides a widely used telephone advisory service to the medical profession, such as other paediatric cardiologists, anaesthetists, intensivists, and respirologists, providing advice regarding patients who do require this special PH support.

Homecare delivery and drugs are commissioned by NHS England for England only. Local arrangements need to be in place for patients from other devolved administrations.

NHS England commissions the service for the population of England. Commissioning on behalf of other devolved administrations is reviewed annually, and a current list is available from NHS England commissioners.

At the moment, NHS England contract includes provision for the service to treat eligible overseas patients under S2 [Under EU regulations, patients can be referred for state funded treatment to another European Economic Area (EEA) member state or Switzerland, under the form S2 (for EU member states) or the form E112 (for Iceland, Norway, Liechtenstein and Switzerland)] referral arrangements. Providers are reimbursed for appropriately referred and recorded activity as part of NHS England contract.

Trusts performing procedures on EU-based patients outside of S2 arrangements will need to continue to make the financial arrangements directly with the governments involved, separately from their contract with NHS England.

With regard to S2, the mechanism for recovery of costs has been via the Department for Work and Pensions Overseas Healthcare Team. They are responsible for agreeing reconciliation and recovery of costs with European administrations. These arrangements were implemented in October 2009, though a similar process existed previously. The financial flows are therefore back into the Treasury rather than back to trusts.

2.4 Any acceptance and exclusion criteria

Acceptance criteria

Urgent referrals are accepted at any time via the on-call cardiology registrar.
The PH Team at GOSH operates 5 days a week.

From 0800 to 1800 hours Monday to Friday, telephone advice is available. If advice

is needed outside these times, the on-call cardiology registrar at GOSH can be contacted or, in specific cases and for individual patients during certain times in their

care, the lead clinician.

The service gives advice on children with all forms of PH even on suspicion of the diagnosis of pulmonary hypertension. The service accepts referrals for this patient group. This clinical group includes such forms of PH as idiopathic and familial PH, pulmonary veno-occlusive disease, post-operative PH, inoperable congenital heart disease with PH, parenchymal lung disorders/disease with PH, miscellaneous causes of PH, and PH of other or uncertain cause. Persistent neonatal PH is specifically excluded due to its different pathophysiology (difficult birth, immaturity and thus belongs to the neonatal speciality.

Referrals are triaged by the consultant. Most referrals come from cardiology, respiratory or intensive care departments at other hospitals.

This service is delivered in accordance with the Great Ormond Street Hospital Single Equality Scheme.

Exclusion criteria

The service is designated for children 16 years and under.

Cases can be excluded where there is clear evidence of absent pulmonary hypertension. General practitioners (GPs) can refer children for screening at GOSH or one of the outreach clinics when pulmonary hypertension has been diagnosed in another family member. In most cases, we need some imaging of cardiac function in order to accept into the service, but exceptions need to be made in cases of urgency.

2.5 Interdependencies with other services

The Great Ormond Street Hospital for Children NHS Foundation Trust PH service is the heart of a national clinical network for the support of PH.

The network is organised on a hub and spoke principle, the hub being Great Ormond Street Hospital for Children NHS Foundation Trust and the spokes being, as per 2009, eight centres of paediatric cardiology, which are:

- Belfast
The Royal Belfast Hospital for Children (Northern Ireland) Belfast Health and Social Care Trust
- Birmingham
Birmingham Children's Hospital NHS Foundation Trust
- Bristol
Bristol Royal Hospital for Children (Wales), University Hospitals Bristol NHS Foundation Trust

- Glasgow
Royal Hospital for Sick Children (Yorkhill)
- Leeds
Leeds General Infirmary, Leeds Teaching Hospitals NHS Trust
- Manchester
Royal Manchester Children's Hospital, Central Manchester University Hospitals NHS Trust
- Newcastle,
Freeman Hospital, The Newcastle Hospitals NHS Foundation Trust
- Southampton
Southampton General Hospital, University Hospital Southampton NHS Foundation Trust

Each of the satellite centres has at least one paediatric cardiologist with a special interest in pulmonary hypertension. The Great Ormond Street Hospital for Children NHS Foundation Trust team holds joint outreach clinics with the local team 4-6 times per year depending on number of patients. The busiest clinics are the mid-England ones (Birmingham, Leeds, Manchester), corresponding with the distribution of population. Teleclinics are offered as an additional option. The care of Scottish children is supported by Scottish National Services Division. Welsh children are seen in the Bristol clinic, with a paediatric cardiologist from the Heath Hospital, Cardiff.

Great Ormond Street Hospital for Children NHS Foundation Trust directly manages approximately 300 of the most severe PH patients using outpatient and inpatient services. If any PH patient needs a procedure involving hospital stay, then one of the 8 co-operating hospitals of the clinical network will either call for advice, or ask for transfer for that procedure. The support then consists of advice (written and/or telephonic), or accepting the patient for assessment and intervention.

Except from the above list of hospitals co-operating within our network and the shared care framework, there are no further programmes. Great Ormond Street Hospital for Children NHS Foundation Trust provide screening service when specifically asked for an individual patient, but do not have a screening programme as such.

3. Applicable Service Standards

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

The UK database of the service has been developed.

Great Ormond Street Hospital for Children NHS Foundation Trust multi-disciplinary meetings are held weekly. All members of the pulmonary hypertension team attend. Data on children requiring cardiac catheterisation are considered at the weekly meeting of all cardiologists.

Cases requiring transplantation are discussed at the weekly joint cardiac conference

with the transplantation team.

Any child with previously undiagnosed, potentially operable congenital heart disease is also presented at the same conference between cardiologists and cardiac surgeons.

Children with complex congenital heart disease seen at the clinical network centres and other units where either the diagnosis is uncertain or the operative risk is high are also discussed when the lead clinician believes that surgery may be feasible.

Quality of life

Every effort is made to ensure that the children have as fulfilled a life as possible and to keep the children out of hospital as much as possible. All those of nursery and school age go back to school, at least for some time, including all those receiving intravenous epoprostenol.

There is currently no quality of life quality assessment form which is specific for children with pulmonary hypertension. The internationally recognised quality of life assessment form designed for children; known as SF10 (Quality Metric Incorporated) was used for several years. It consists of the 10 questions found to have statistical significance out of the 50 questions on the original SF50 assessment. All the parents and the older children complete the form at each out-patient visit.

Great Ormond Street Hospital for Children NHS Foundation Trust works closely with the PHA UK to support the children and their families. The service contributes to the annual meeting and our clinical nurse specialist helps run an annual family weekend and this year has arranged a holiday weekend for teenagers on intravenous epoprostenol.

Summary of Progress to Date

A managed clinical network, the UK pulmonary hypertension service for children, has been established which covers the whole of the UK. We have fulfilled the original aims of setting up the network:

- quality of life has been improved, and the children are integrated rapidly back into society as soon as their condition permits
- survival has improved
- the children have ready access to all new therapies
- children deteriorating on maximal medical therapy have rapid access to the Paediatric Thoracic Transplantation Service
- there is equity of care throughout the UK, with no post-code prescribing
- education of health professionals is ongoing, in order to improve disease recognition and prompt referral
- research into the clinical management and pathobiology of pulmonary hypertension in childhood has continued

There is considerable user involvement through the Pulmonary Hypertension

Association UK.

4. Key Service Outcomes

<i>Quality Performance Indicator examples</i>	<i>Threshold</i>	<i>Method of measurement</i>	<i>Consequence of breach</i>	<i>Report Due</i>
Survival/Mortality	Statistical significance	Kaplan-Meier-Analysis	Review of Treatment Guidelines	q 6 months
Improvement in Clinical Class (WHO function)	Statistical significance	Statistical Analysis	Review of Treatment Guidelines	q 6 months
Quality of Life/PROM	Statistical significance	Statistical Analysis	Review of Treatment Guidelines	q 6 months
Length of Wait	>2 wks for ivEPO >6 wks for nl Pts	Date of Referral to Start of Therapy	Review of GOSH administrative pathways	q 6 months

5. Location of Provider Premises

Healthcare@Home provides the homecare delivery element of the service under the guidance of the clinical team and homecare manager. This arrangement is subject to the requirements to tender for services on a periodic basis.

Provider

Great Ormond Street Hospital for Children NHS Foundation Trust
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