

# National Audit of Pulmonary Hypertension 2011



Electronic copies of this report can be found at: [www.ic.nhs.uk](http://www.ic.nhs.uk).  
If you have any queries or comments on this publication,  
please contact The NHS Information Centre for health and  
social care 0845 300 6016 or email: [enquiries@ic.nhs.uk](mailto:enquiries@ic.nhs.uk)  
quoting document reference IC22090311.

For further information about this report, email:  
[enquiries@ic.nhs.uk](mailto:enquiries@ic.nhs.uk) or contact:  
Clinical Audit Support Unit (CASU)  
The NHS Information Centre for health and social care  
1 Trevelyan Square  
Boar Lane  
Leeds  
LS1 6AE

**Prepared in partnership with:**



**The NHS Information Centre for Health and Social Care (The NHS IC)** is England's central, authoritative source of essential data and statistical information for frontline decision makers in health and social care. The NHS IC managed the publication of the 2010/11 annual report.



**The Pulmonary Hypertension Association (PHA UK)** is the only charity in the UK especially for people with pulmonary hypertension (PH). They are committed to helping improve the lives of people with PH, and supporting the PH community in the UK. They do this by funding research into PH, raising awareness of the condition, and helping people in their day-to-day lives.



**National Pulmonary Hypertension Centres of the UK and Ireland Physicians Committee** The Committee comprises all physicians from the designated centres and coordinates the national centres. These centres diagnose and treat all the Pulmonary Hypertension in the UK using a set of National Guidelines. This structure ensures that all patients have a firm diagnosis and receive evidence based treatment to ensure morbidity and survival are amongst the best in the world.



**The Specialised Commissioning Groups** commission the services from the designated centres for Pulmonary Hypertension.



**National Services Division (NSD)** is a division within NHS National Services Scotland (NSS). NSD receives funding from the Scottish Government Health Directorates (SGHD) to commission and performance manage nationally designated specialist services and screening programmes.



**Welsh Health Specialised Services Committee (WHSSC)** is responsible for the commissioning and planning of all specialised services for the population of Wales.

# National Audit of Pulmonary Hypertension 2011

Second annual report: Key findings  
from the National Audit of  
Pulmonary Hypertension for the  
United Kingdom, Channel Islands,  
Gibraltar and Isle of Man

Report for the Audit Period  
April 2010 to March 2011

# Contents

<b>Acknowledgements</b>	<b>5</b>
<b>List of Abbreviations</b>	<b>5</b>
<b>Foreword by Professor Dame Carol Black</b>	<b>6</b>
<b>Introduction</b>	<b>7</b>
<b>Executive Summary of Key Findings</b>	<b>8</b>
<b>Participation of Designated Centres and Number of Patients Seen</b>	<b>9</b>
<b>Patient Demographics</b>	<b>11</b>
Age and Sex of All Patients	11
<b>Diagnostic Classification of Pulmonary Hypertension</b>	<b>12</b>
Distribution of Patients in Great Britain According to Pulmonary Hypertension Diagnosis	14
New Referrals	25
Diagnosis in Inpatients	28
<b>Patients treated with disease-targeted drug therapies</b>	<b>29</b>
Disease-Targeted Drug Therapies During the Audit	30
Disease-Targeted Therapy Census Data	34
<b>Clinical Activity</b>	<b>37</b>
Disposition of patients	37
Deaths	38
Clinical Investigations: Cardiac Catheterization	39
Clinical Investigations: Echocardiography	39
Clinical Investigations: Exercise Testing	40
<b>Survival Outcome Analysis</b>	<b>41</b>
<b>Audit of Pulmonary Endarterectomy</b>	<b>42</b>
Assessment	42
Referrals to Papworth Multidisciplinary Team Meeting with Suspicion of CTEPH	42
Outpatient activity (surgical assessment )	44
Surgery	44
Follow up 3 months after PEA	44
Audit of Pulmonary Endarterectomy: Acknowledgements	45
<b>Key findings</b>	<b>46</b>
<b>Recommendations</b>	<b>47</b>
<b>References</b>	<b>47</b>
Appendix 1: Dana Point clinical classification of pulmonary hypertension	48
Appendix 2: Specialised Commissioning Drug Policy for England	49
Appendix 3: Members of Staff who participated in the Audit at the Designated Pulmonary Hypertension Centres	50
Appendix 4: Members of the NAPH Project Board	51

## Acknowledgements

The National Pulmonary Hypertension Audit was funded in its first year by the Pulmonary Hypertension Patients Association (PHA-UK) to whom we are most grateful for ensuring the start of this important project. It has subsequently been funded by Specialised Commissioners in England and hosted by the NHS Information Centre for health and social care

The project wishes to acknowledge the following who have provided unwavering support for the audit and provided guidance during the data collection period, analysis and writing of this report:

- PHA-UK
- Clinicians and audit database users at each designated pulmonary hypertension centre (Appendix 3)
- Specialised commissioners
- National Commissioning Group
- National Service Division, NHS Scotland
- Welsh Health Specialised Services
- National Audit of Pulmonary Hypertension Project Board (Appendix 4) chaired by Simon Gibbs who designed and drafted this report
- The NHS IC who provided project management, data analysis and database development

## List of Abbreviations

NAPH	National Audit of Pulmonary Hypertension
NHS	National Health Service
NHS IC	National Health Service Information Centre for health and social care
PHA-UK	Pulmonary Hypertension Association (UK)
WHO	World Health Organisation

## Foreword by Professor Dame Carol Black

National audit in the UK plays a major role in influencing healthcare delivery. This annual report of the second year of the National Audit of Pulmonary Hypertension represents the tremendous progress and collaborative effort of all eight of the hospitals designated to manage pulmonary hypertension in the UK. It builds on the previous report to describe new and current epidemiological data for different types of pulmonary hypertension nationwide and the pattern of referral of new patients to designated centres. This type of information plays a crucial role in improving and planning health services for patients, now and in the future. Outcomes have started to be assessed and it is important that these feature more prominently as the audit matures.

As research continues to shed new light on this serious disease and develop promising new therapies and treatment strategies, this audit will be able to assess their impact on clinical outcomes. In doing so, it will build on the evidence derived from clinical research studies and describe long-term outcomes.

# Introduction

Pulmonary hypertension is a rare disease which occurs at any age, has many causes, and often shortens life expectancy. In the UK, Channel Islands, Gibraltar and Isle of Man seven hospitals have been designated to diagnose and treat pulmonary hypertension in adults and one hospital for children.

The National Audit of Pulmonary Hypertension (NAPH) is a prospective audit of processes and outcomes and has the participation of all eight designated centres. This report of the second year brings together the audit data collected from the nationally designated centres (Table 1). It builds on the first report one year ago<sup>1</sup> while expanding new areas of information. Since the last report the lack of completeness of some key data fields has been resolved. Database processes have been kept under constant review and improved. New information has been added to this year's report about diagnosis, new referrals and survival of selected diagnostic groups of patients.

The data presented in this report is for the year from 1 April 2010 to 31 March 2011 and covers all patients seen at the designated centres. This audit includes England, Scotland, Wales, Northern Ireland, the Channel Islands, Gibraltar and the Isle of Man.

**Table 1**  
Designated Pulmonary Hypertension Centres in the United Kingdom. For hospitals in England their Strategic Health Authority is shown in parentheses

Freeman Hospital, Newcastle-upon-Tyne (North East SCG)
Golden Jubilee Hospital, Glasgow, Scotland
Great Ormond Street Hospital for Children, London (London SCG)
Hammersmith Hospital, London (London SCG)
Papworth Hospital, Papworth Everard (East of England SCG)
Royal Brompton Hospital, London (London SCG)
Royal Free Hospital, London (London SCG)
Royal Hallamshire Hospital, Sheffield (Yorkshire and Humber SCG)

The designated centres follow current UK and European guidelines<sup>2, 3</sup> and are responsible for making the diagnosis (Appendix 1), initiating disease-targeted drug therapy and where appropriate, undertaking the long-term management of patients. Patients are treated with disease targeted drug therapies according to the prescribing policy drawn up by Specialised Commissioners (Appendix 2).

Pulmonary endarterectomy surgery for chronic thromboembolic pulmonary hypertension is carried out at Papworth Hospital under the separately designated Pulmonary Endarterectomy service. Data from a separate audit of the pulmonary endarterectomy service are reported here for the first time. Although this service is distinct from the pulmonary hypertension service, it provides surgical treatment for patients with chronic thromboembolic pulmonary hypertension.

The audit holds quarterly meetings of its User Group and twice yearly meetings of the Project Board. The Lead Clinician of the audit provides direct links with the National Pulmonary Hypertension Centres of the UK and Ireland Physicians Committee and the National Joint Commissioners Forum. PHA-UK is represented on the Project Board. Since the beginning of 2011, designated centre reimbursement for drug therapy has been linked to information entry into the database. The funding for the work carried out at the NHS Information Centre for this audit year was provided by Specialised Commissioning.

The strategy for the next year is to continue the audit, introduce an assessment of the accuracy of data entry at each centre and develop a comparison of outcomes between designated centres. This latter work has already commenced with the formation of a group of physicians, the Pulmonary Hypertension Outcomes Group, who are reviewing the information required to achieve this.

# Executive Summary of Key Findings

1. This report describes the clinical activity and prescribing for patients with pulmonary hypertension in the UK, Channel Islands, Gibraltar and Isle of Man.
2. There is significant under reporting of patients in Northern Ireland which is a matter of concern.
3. There has been progressive growth in the number of patients managed by designated centres in the UK since 2004.
4. During the audit UK pulmonary hypertension services saw 6196 patients. Since 36 patients were seen in more than one centre during the year, the workload of the centres is represented by 6232 patients. 86 per cent of patients came from England. The median age was 62 years which is 2 years older than the previous year. There were 1.8 females to every male.
5. In Great Britain designated centres saw 97 patients per million population. In England the prevalence of patients with pulmonary hypertension seen at designated centres was highest down the east side of the country where the centres are located.
6. Of the total number of patients, 12 per cent were discharged and 11 patients were transplanted.
7. There were 444 deaths (7 per cent of the population) at a median age of 67 years.
8. Pulmonary arterial hypertension was the commonest diagnosis in 46 per cent of patients followed by chronic thromboembolic hypertension in 18 per cent. 18 per cent did not have pulmonary hypertension.
9. There were 2083 patients admitted to designated centres as inpatients for 12093 bed days. Investigations at designated centres totalled 1895 cardiac catheterization procedures, 4231 echocardiograms and 6621 exercise tests.
10. There were 2089 new patients referred to designated centres. The commonest diagnoses were pulmonary arterial hypertension (31 per cent) and no pulmonary hypertension (31 per cent). There was wide variation in referral rates in different geographical regions.
11. Disease targeted drug therapy was prescribed for 3163 patients, mean age 56 years and median age 59 years. The ratio of females to males was 1.9:1.
12. The standardised rate of treatment of patients with disease-targeted drug therapies was similar in England, Scotland and Wales but there was wide variation in treatment rates across England.
13. The most commonly prescribed drugs were phosphodiesterase 5 inhibitors followed by endothelin receptor antagonists.
14. One year survival for new cases of idiopathic, heritable and anorexogen-induced pulmonary arterial hypertension was 87 per cent and for connective tissue disease associated pulmonary arterial hypertension was 87 per cent. New patients seen with congenital heart disease associated pulmonary arterial hypertension had a one year survival of 92 per cent.
15. In a separate audit of pulmonary endarterectomy, 129 operations were performed with an in hospital mortality of 5.4 per cent and with improvement in WHO functional class 3 months post operatively.

# Participation of Designated Centres and Number of Patients Seen

All designated centres participated in the audit. [Table 2](#) shows the number of patients seen at each designated centre. The numbers of patients seen at each hospital include any patient seen in the Pulmonary Hypertension Service between 1 April 2010 and 31 March 2011 inclusive. Duplicate patients are those who are seen in more than one pulmonary hypertension centre during the audit year. A comparison is made with data from the previous year.

Designated Pulmonary Hypertension Centre	Number of Patients 2010	Number of Patients 2011
Freeman Hospital	375	377
Golden Jubilee Hospital	409	475
Great Ormond Street Hospital for Children	376	376
Hammersmith Hospital	851	938
Royal Brompton Hospital	567	736
Papworth Hospital	857	859
Royal Free Hospital	811	1005
Royal Hallamshire Hospital	1292	1466
<b>Total Patients Seen at Designated Centres</b>	<b>5538</b>	<b>6232</b>
Duplicate Patients	60	36
<b>Total Patients Seen in the UK, Channel Islands, Gibraltar and Isle of Man</b>	<b>5478</b>	<b>6196</b>

A snapshot of the clinical service on one day is shown in [Table 3](#) and [Figure 1](#). This data was collected as a census of all patients alive and not discharged on 31 March. It therefore ignores patients who were seen during the audit period and who were discharged, transplanted or who died before the 31 March of that year.

The data for 2004 to 2009 in [Figure 1](#) was collected before the audit began by the National Pulmonary Hypertension Centres of the UK and Ireland Physicians Committee. This figure shows the progressive growth of the service over eight years.

Hospital	Total 2010	Total 2011
Freeman Hospital	294	320
Golden Jubilee Hospital	291	329
Great Ormond Street Hospital for Children	300	319
Hammersmith Hospital	664	764
Royal Brompton Hospital	492	640
Papworth Hospital	591	625
Royal Free Hospital	581	774
Royal Hallamshire Hospital	1074	1291
<b>Total</b>	<b>4287</b>	<b>5062</b>

**Figure 1**  
Number of patients active in the service and alive on 31 March in eight successive years



# Patient Demographics

## Age and Sex of All Patients

Table 4 shows the age of all patients who were seen during the audit year. Patients were assigned to a country according to their postcode. Since the previous year the median age of the UK cohort has increased by 2 years. The low number of patients in Northern Ireland and their age difference from the rest of the country suggests that there is either under reporting of patients in the database or under diagnosis. For this reason a separate number is shown for Great Britain (England, Scotland and Wales) to avoid biasing the data for the UK.

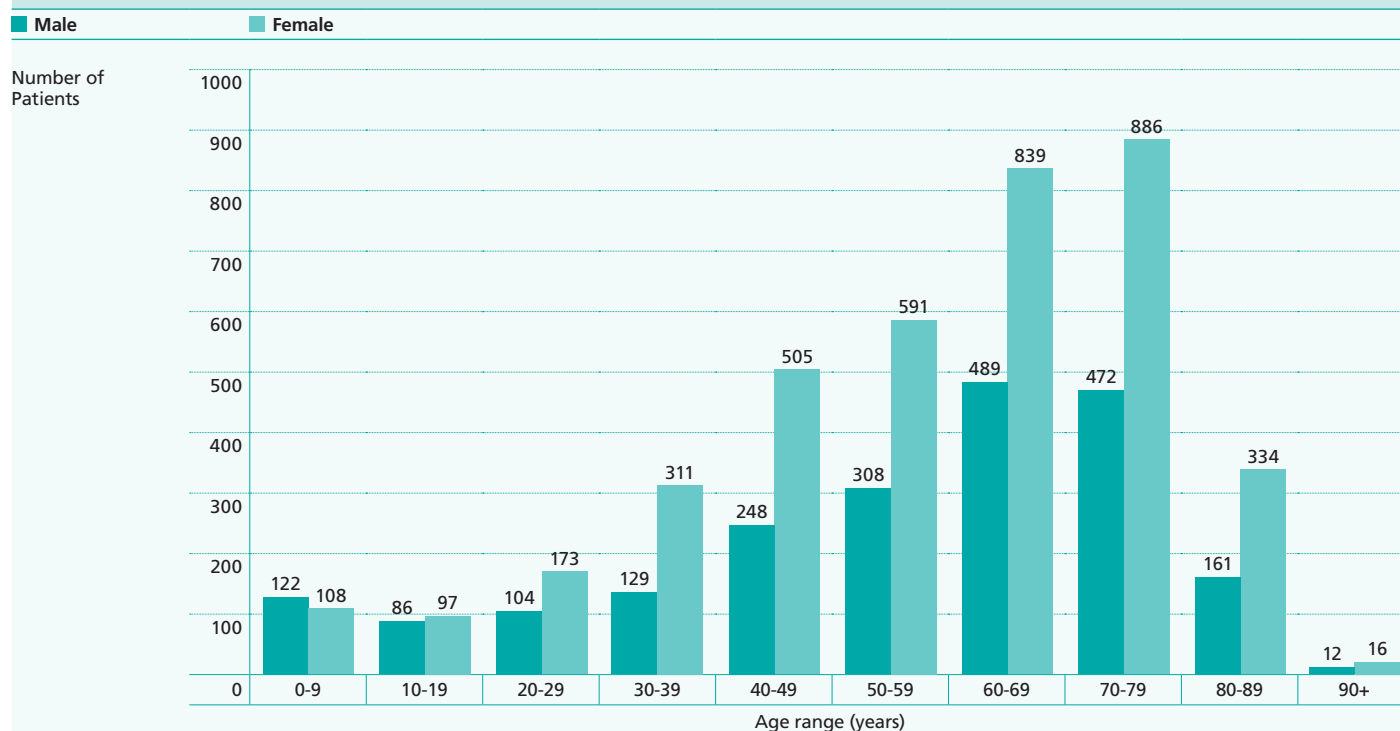
Figure 2 shows the number of males and females by age. The distribution of patients is skewed towards older age. The ratio of females : males was 1.8.

**Table 4**  
Age of all patients by country. Date of birth was recorded in 6001 patients. Ages have been rounded to the nearest whole number

Region	Number of patients 2010-11	Number of patients per million population*	Mean age (years) 2010-11	Median age (years) 2010-11	Median age (years) 2009-10
UK (all patients)	6001	-	57	62	60
Great Britain	5869	97	57	62	-
England	5140	98	57	62	61
Scotland	492	94	59	63	60
Wales	237	79	54	60	58
Northern Ireland	36	20	31	34	37
Other/Unknown	96	-	55	58.5	58

\* Population data is based on mid year 2010 estimate (Office of National Statistics). Other includes Channel Islands, Gibraltar, and the Isle of Man. Unknown indicates that the country of the patients has not been identified.

**Figure 2**  
Distribution of age and sex of all patients. Patients were excluded if they had no date of birth or sex recorded



# Diagnostic Classification of Pulmonary Hypertension

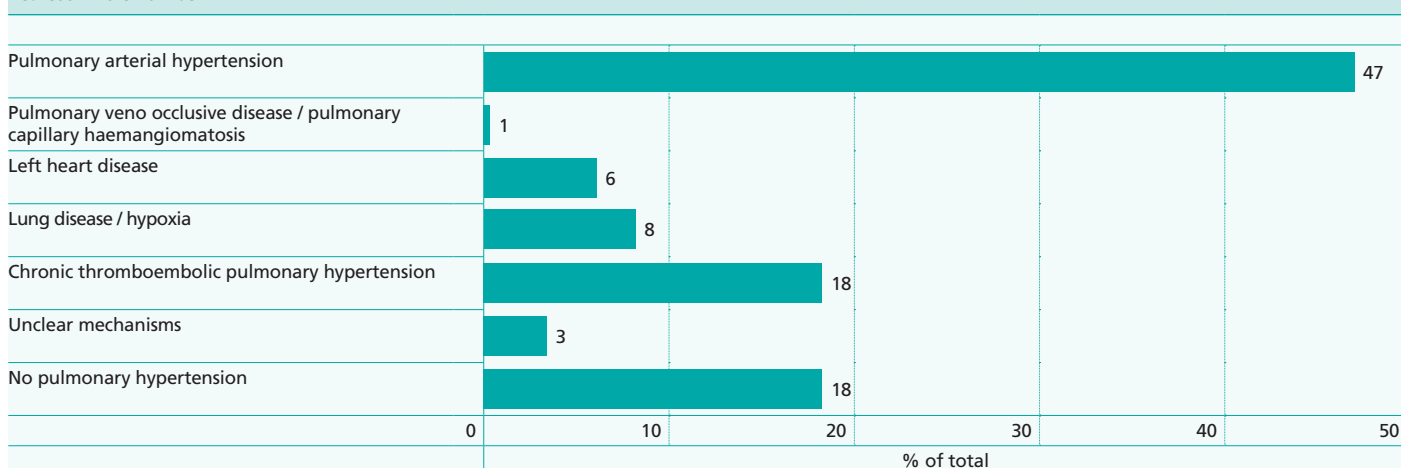
The diagnosis of pulmonary hypertension is made according to the Dana Point clinical classification<sup>4</sup> (Appendix 1). Diagnosis is key to the treatment.

The number of patients in whom a diagnosis had not been recorded in the patient's record at the time of the audit was 302 (5 per cent) of 6196 patients. Those patients without a diagnosis were in the process of being seen for the first time and being investigated and so would not have had a diagnosis.

Figure 3 shows the diagnostic classification of patients with a recorded diagnosis. The most common diagnoses are pulmonary arterial hypertension and chronic thromboembolic pulmonary hypertension. It is well recognised that pulmonary

veno-occlusive disease and pulmonary haemangiomatosis are rare. Patients who were investigated and found not to meet the criterion of a mean pulmonary arterial pressure of greater than or equal to 25 mm Hg were recorded as "not pulmonary hypertension". These patients included those in whom echocardiography had suggested a diagnosis of pulmonary hypertension, patients with breathlessness at risk of pulmonary arterial hypertension or first degree relatives of patients with heritable pulmonary arterial hypertension who underwent screening for pulmonary hypertension. Figure 4 show the distribution of different subcategories of pulmonary arterial hypertension.

**Figure 3**  
Dana Point diagnostic classification of all patients with a final diagnosis showing the proportion in each diagnostic group. (See Appendix 1 for details of the classification). "No pulmonary hypertension" has been added to the classification for the purposes of this figure. Percentages have been rounded to the nearest whole number



**Figure 4**  
Proportion of patients in different subcategories of pulmonary arterial hypertension

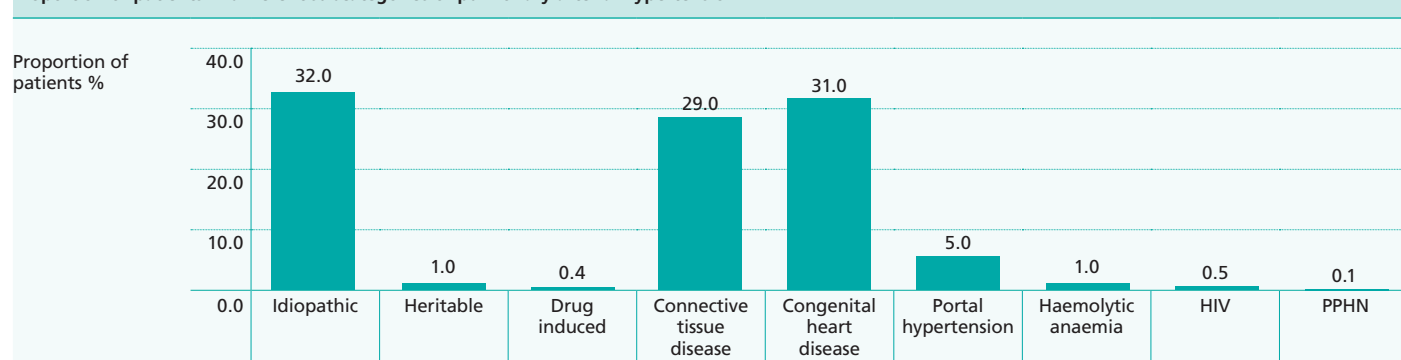
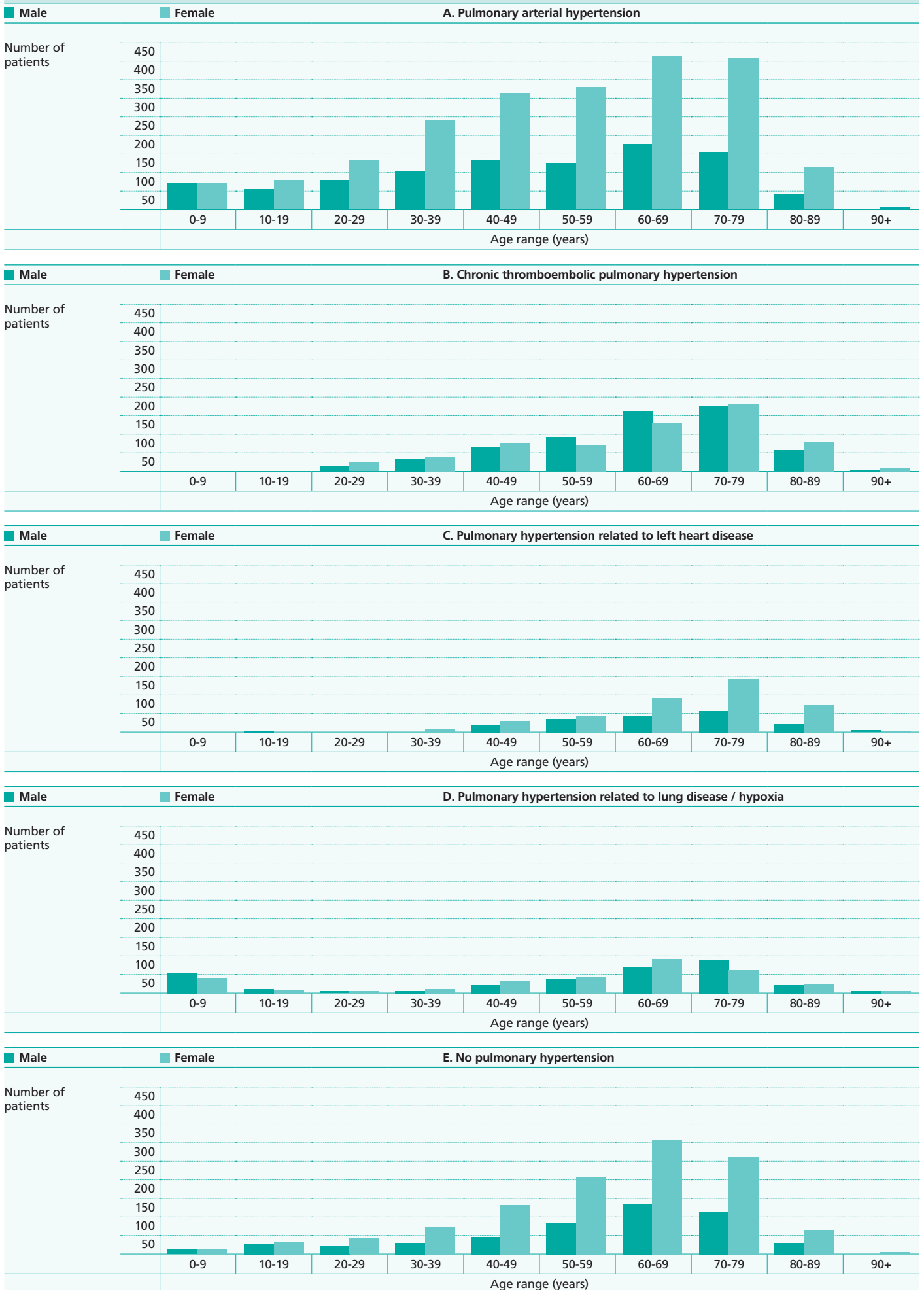


Figure 5 provides a graphical representation of the distribution of patients by age and sex according to the main diagnostic categories. This allows the reader to see the different age and sex distributions of these diagnoses. Patients with chronic thromboembolic pulmonary hypertension and left heart disease are older than patients with pulmonary arterial hypertension. Note that the age and sex distribution of patients without pulmonary hypertension is similar to those with the disease.

**Figure 5**

The age and sex distribution of patients according to the Dana Point clinical classification diagnosis



## Distribution of Patients in Great Britain According to Pulmonary Hypertension Diagnosis

In order to understand the current epidemiology of some common causes of pulmonary hypertension, a map of patients was created for each major diagnosis. The distribution of each patient diagnostic group is described both by country (England, Scotland and Wales) and for England only by its ten Strategic Health Authorities. Designated pulmonary hypertension centres in England are located in Strategic Health Authorities in the North East, Yorkshire and Humber, East of England and London. Northern Ireland has been excluded because of the lack of data. Prevalence has been standardised, age and sex according to the Office of National Statistics mid-year population estimates in 2010. Prevalence would not be expected to differ geographically.

The patient distribution for pulmonary arterial hypertension of all causes is shown in [Figures 6 and 7](#); of idiopathic, heritable and anorexogen-induced pulmonary arterial hypertension in [Figures 8 and 9](#); for connective tissue associated pulmonary arterial hypertension in [Figures 10 and 11](#); for congenital heart disease associated pulmonary arterial hypertension in [Figures 12 and 13](#); and for chronic thromboembolic pulmonary hypertension in [Figures 14 and 15](#).

The prevalence of pulmonary arterial hypertension is broadly similar across England, Scotland and Wales ([Figure 6](#)) although there is wider variation in the prevalence of the subcategories ([Figures 8, 10, 12](#)). While idiopathic, heritable and anorexogen-induced pulmonary arterial hypertension is most prevalent in Scotland, connective tissue disease associated pulmonary arterial hypertension is most prevalent in England and congenital heart disease associated pulmonary arterial hypertension in Wales.

In England in general the greatest numbers of patients diagnosed with pulmonary hypertension are down the east of the country in proximity to designated centres. For example, chronic thromboembolic disease is most prevalent in England and the highest prevalence is in the East of England where the UK pulmonary endarterectomy surgical centre is located. There is also variation in the local prevalence of various subcategories of pulmonary arterial hypertension between centres.

Overall the picture is of wide variation in prevalence of the described types of pulmonary hypertension across the country. Although 5 per cent of the patients were not included in this analysis because of a missing diagnosis this is not enough to explain the differences being observed. Rather it suggests different patterns of referral and types of pulmonary hypertension seen at different designated centres across the country.

**Figure 6**

Distribution of patients with pulmonary arterial hypertension managed by designated pulmonary hypertension centres in Great Britain during 2010-11. The prevalence per million population per annum has been standardised for age and sex and is shown in the legend

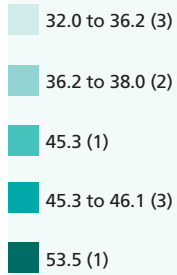
Per million population



© Crown Copyright and database rights 2011 Ordnance Survey 0100044406

**Figure 7**  
 Distribution of pulmonary arterial hypertension managed by designated pulmonary hypertension centres in England during 2010-11. The prevalence per million population per annum has been standardised for age and sex and is shown in the legend. The numbers in parentheses show the number of SHAs which fall within the range

Per million population



© Crown Copyright and database rights 2011 Ordnance Survey 0100044406

**Figure 8**

Distribution of idiopathic, heritable and anorexogen-induced pulmonary arterial hypertension managed by designated pulmonary hypertension centres in Great Britain during 2010-11. The prevalence per million population per annum has been standardised for age and sex and is shown in the legend

Per million population



© Crown Copyright and database rights 2011 Ordnance Survey 0100044406

**Figure 9**  
 Distribution of idiopathic, heritable and anorexogen-induced pulmonary arterial hypertension managed by designated pulmonary hypertension centres in England during 2010-11. The prevalence per million population per annum has been standardised for age and sex and is shown in the legend. The numbers in parentheses show the number of SHAs which fall within the range

Per million population

- 9.2 to 10.4 (2)
- 10.4 to 12.5 (2)
- 12.5 to 13.3 (2)
- 13.3 to 13.7 (3)
- 15.2 (1)



© Crown Copyright and database rights 2011 Ordnance Survey 0100044406

**Figure 10**

Distribution of connective tissue disease associated pulmonary arterial hypertension to designated pulmonary hypertension centres in Great Britain during 2010-11. The prevalence per million population per annum has been standardised for age and sex and is shown in the legend

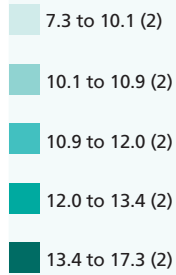
Per million population



© Crown Copyright and database rights 2011 Ordnance Survey 0100044406

**Figure 11**  
 Distribution of connective tissue disease associated pulmonary arterial hypertension to designated pulmonary hypertension centres in England during 2010-11. The prevalence per million population per annum has been standardised for age and sex and is shown in the legend. The numbers in parentheses show the number of SHAs which fall within the range

Per million population



© Crown Copyright and database rights 2011 Ordnance Survey 0100044406

**Figure 12**

Distribution of congenital heart disease associated pulmonary arterial hypertension to designated pulmonary hypertension centres in Great Britain during 2010-11. The prevalence per million population per annum has been standardised for age and sex and is shown in the legend

Per million population

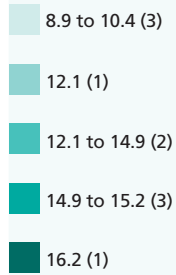


© Crown Copyright and database rights 2011 Ordnance Survey 0100044406

**Figure 13**

Distribution of congenital heart disease associated pulmonary arterial hypertension to designated pulmonary hypertension centres in England during 2010-11. The prevalence per million population per annum has been standardised for age and sex and is shown in the legend. The numbers in parentheses show the number of SHAs which fall within the range

Per million population



© Crown Copyright and database rights 2011 Ordnance Survey 0100044406

**Figure 14**

Distribution of chronic thromboembolic pulmonary hypertension to designated pulmonary hypertension centres in Great Britain during 2010-11. The prevalence per million population per annum has been standardised for age and sex and is shown in the legend

Per million population

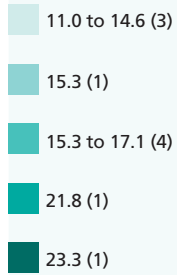


© Crown Copyright and database rights 2011 Ordnance Survey 0100044406

**Figure 15**

Distribution of chronic thromboembolic pulmonary hypertension to designated pulmonary hypertension centres in England during 2010-11. The prevalence per million population per annum has been standardised for age and sex and is shown in the legend. The numbers in parentheses show the number of SHAs which fall within the range

Per million population



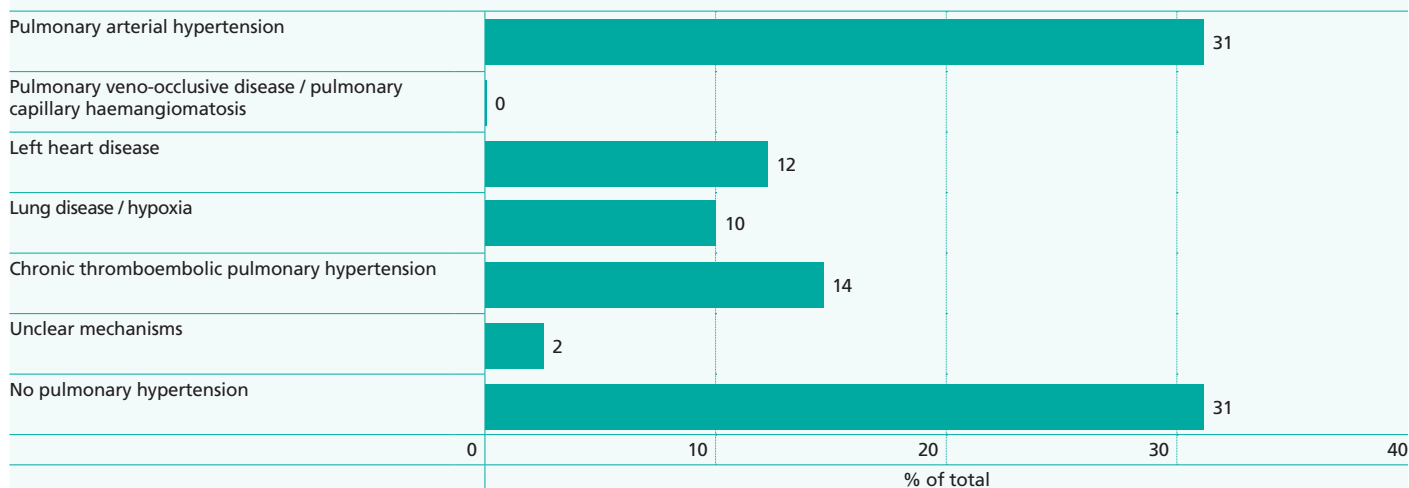
© Crown Copyright and database rights 2011 Ordnance Survey 0100044406

## New Referrals

During the audit year, 2089 patients were referred to pulmonary hypertension centres. Of these 1089 underwent cardiac catheterization and 626 received disease-targeted drug therapies. Figure 16 shows the final diagnosis of this group of patients in those in whom a final diagnosis was recorded. Almost one third of patients did not have pulmonary hypertension. The commonest diagnosis was pulmonary arterial hypertension.

Figures 17 and 18 describe the geographical source of new referrals. Referral rates were similar in England and Scotland but lower by 31 per cent in Wales. There was wide variation in referral rates in England between 21.5 and 71 per million population per annum.

**Figure 16**  
Diagnosis in 1636 new patient referrals with a final diagnosis recorded



**Figure 17**  
Distribution of the source of new referrals to designated pulmonary hypertension centres in Great Britain during 2010-11. The prevalence per million population per annum has been standardised for age and sex and is shown in the legend

Per million population

23.4

34.0

35.1



© Crown Copyright and database rights 2011 Ordnance Survey 0100044406

**Figure 18**

Distribution of source of new referrals to designated pulmonary hypertension centres in England during 2010-11. The prevalence per million population per annum has been standardised for age and sex and is shown in the legend. The numbers in parentheses show the number of SHAs which fall within the range

Per million population

- 21.5 to 22.5 (2)
- 22.5 to 28.1 (2)
- 28.1 to 34.0 (2)
- 34.0 to 36.9 (3)
- 70.8 (1)

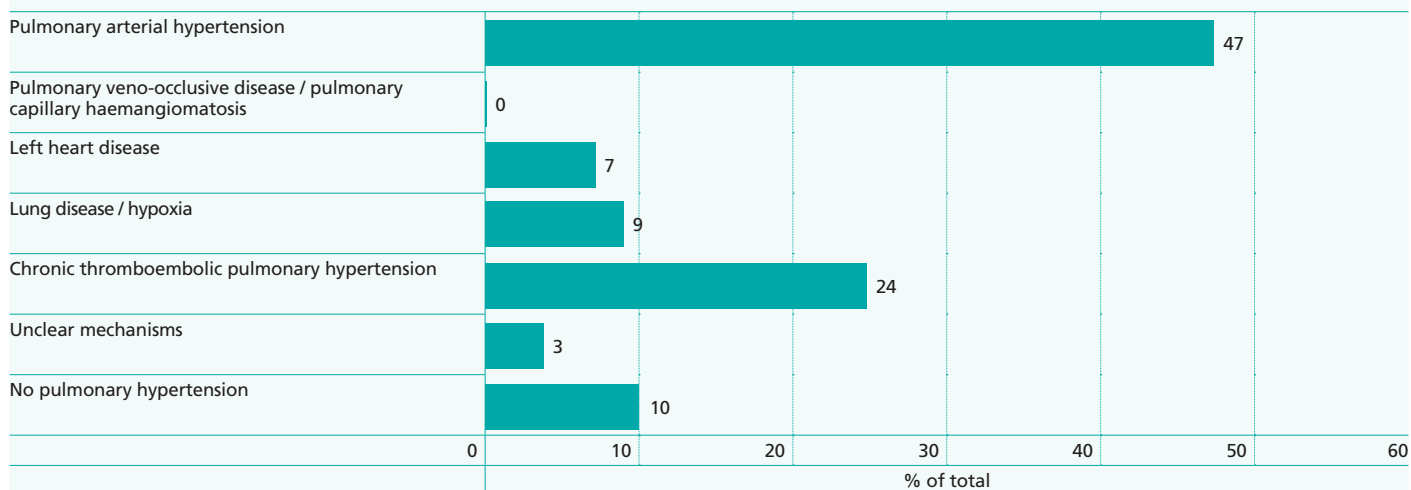


© Crown Copyright and database rights 2011 Ordnance Survey 0100044406

## Diagnosis in Inpatients

A total of 2083 patients were admitted to hospital for 12093 bed days. Figure 19 shows that the commonest diagnosis was pulmonary arterial hypertension followed by chronic thromboembolic pulmonary hypertension. Significantly fewer patients have no pulmonary hypertension compared to the entire patient cohort.

**Figure 19**  
Diagnosis of patients admitted to hospital under the pulmonary hypertension team at a designated centre for any reason. A final diagnosis was missing in 75 inpatients



# Patients Treated With Disease-Targeted Drug Therapies

This section describes the patient population who were prescribed disease-targeted drug therapies by the designated centres.

The disease-targeted drug therapies fall into four groups:

- Endothelin receptor antagonists which are administered orally
- Phosphodiesterase 5 inhibitors which are administered orally
- Prostanoids which are administered by intravenous infusion, by subcutaneous infusion or intermittently by nebuliser
- Calcium channel blockers for vasoreactive pulmonary arterial hypertension which are administered orally

The NHS policy for prescribing drug therapies is shown in Appendix 2 and was unchanged from the last report. Unlike guidelines, NHS policy dictates which treatments are available for use and in what circumstances. It also requires all prescriptions to be issued only by designated centres for the duration of treatment. Note that the policy in force during this audit did not permit the treatment of patients in WHO functional II (WHO functional class is used to describe symptom severity and is akin to the New York Heart Association functional class). Prescription of drugs outside this policy was only permitted when a case for treatment on grounds of exceptionality was made to the Primary Care Trust of the general practitioner of the patient and approved by their Exceptional Circumstances Panel.

Two types of data are presented. First an analysis of all patients treated during the audit. Second data collected on a single day (census data). The number of patients differs between these two datasets as would be expected.

## Disease-Targeted Drug Therapies During the Audit

Table 5 shows that 3163 patients received disease targeted drug therapy at some time during the audit period 2010 - 2011. This represents a 6 per cent increase in patients treated over the previous year. The number of patients who received drug therapies is underestimated since at least some patients in Northern Ireland were not recorded on the database as described above.

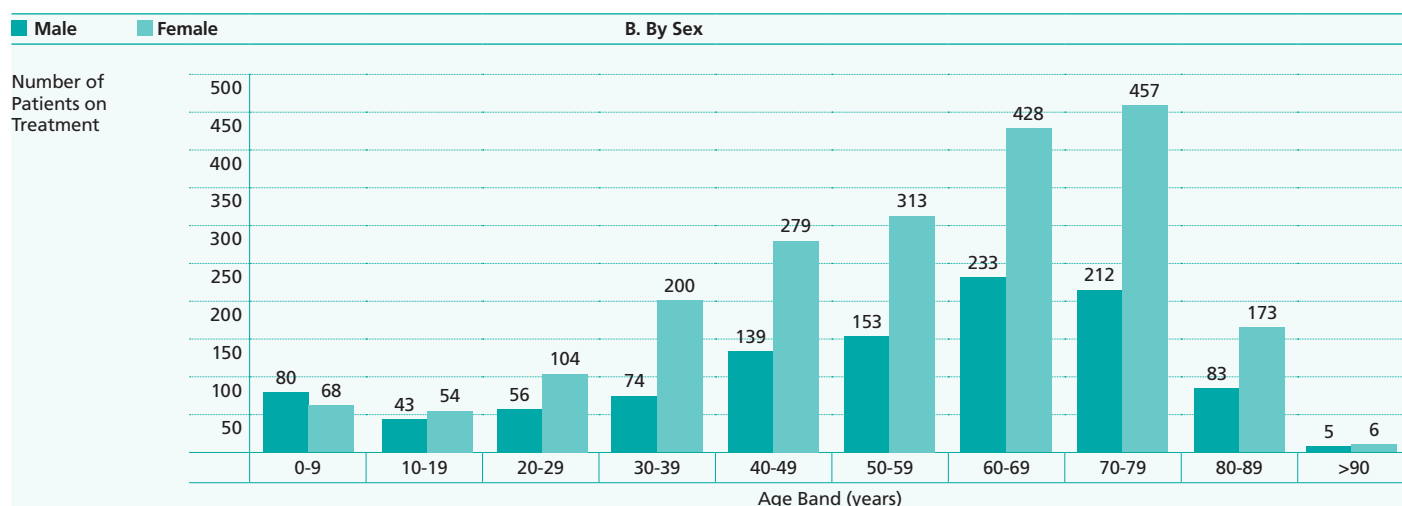
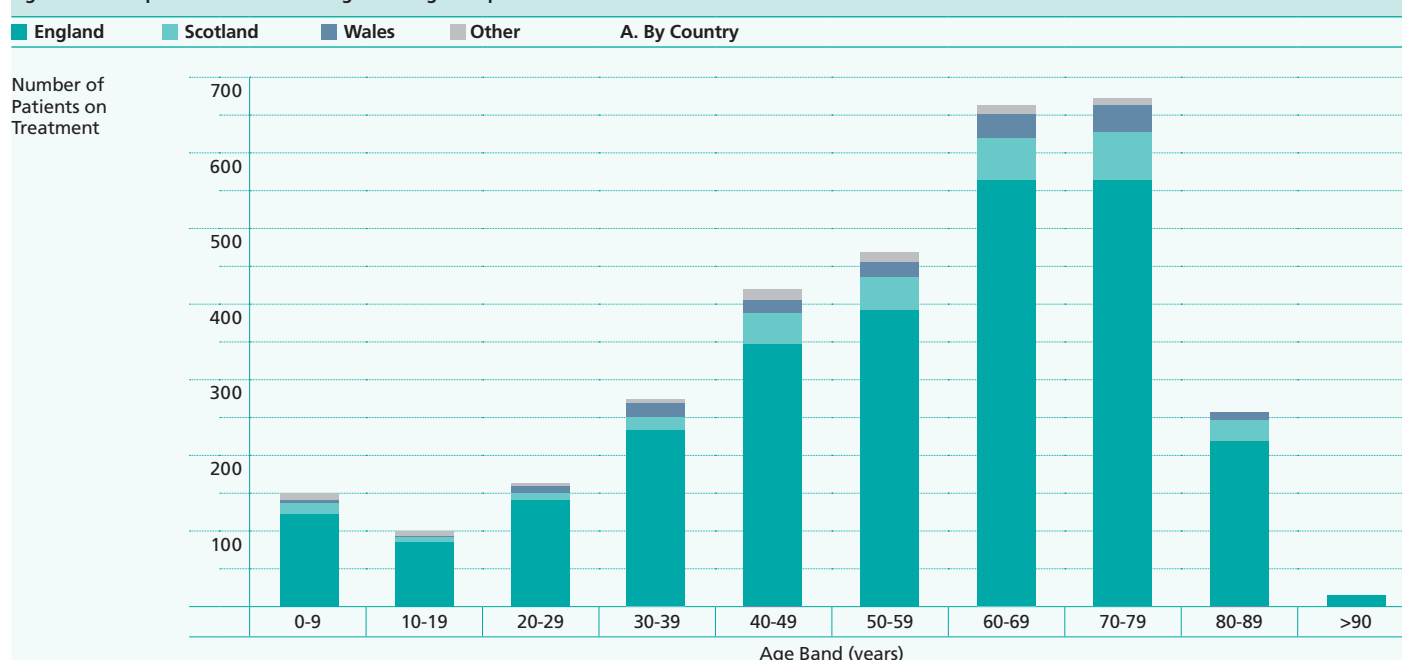
**Table 5**  
Patients treated with disease-targeted therapy by country at any time during the audit year

Region	2009-10		2010-11			
	Patient Count	Mean Age (years)	Median Age (years)	Patient Count	Mean Age (years)	Median Age (years)
UK (all patients)	2980	56	59	3163	55.3	60
England	2527	57	59	2666	55.3	60
Scotland	252	55	60	272	57.6	62
Wales	151	54	59	155	55.7	60
Northern Ireland*	29	45	33	26	25	20.5
Other/Unknown	21	31	32	44	52.8	55

\* Data for Northern Ireland was under reported as noted in the text. Other includes the Channel Islands, Gibraltar and the Isle of Man. Unknown indicates that the country of the patients has not been identified.

Figure 20 displays the number of patients on disease-targeted therapy, by country and by sex according to age. Northern Ireland data has been excluded to avoid bias. The geographical distribution of patients treated with disease targeted therapies is described in Figures 20 and 21. There are similar treatment rates seen between England, Scotland and Wales. Within England there is marked variation with the highest treatments rates seen in the Strategic Health Authorities which host designated centres.

**Figure 20**  
Age and sex of patients on disease-targeted drug therapies 2010-11.



**Figure 21**  
Distribution of patients treated with disease targeted therapies by designated pulmonary hypertension centres in Great Britain during 2010-11. The prevalence per million population per annum has been standardised for age and sex and is shown in the legend

Per million population

49.9

50.8

51.3

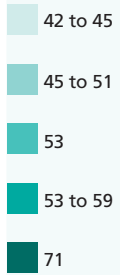


© Crown Copyright and database rights 2011 Ordnance Survey 0100044406

**Figure 22**

Distribution of patients treated with disease targeted therapies by designated pulmonary hypertension centres in Great Britain during 2010-11. The prevalence per million population per annum has been standardised for age and sex and is shown in the legend. The numbers in parentheses show the number of SHAs which fall within the range

Per million population



© Crown Copyright and database rights 2011 Ordnance Survey 0100044406

## Disease-Targeted Therapy Census Data

Census data was collected on 31 March 2011. The census collects data on patients who are alive and under the care of designated centre pulmonary hypertension services on that day. Treatment data reflects the number of patients on treatment that day and the number of drugs they were taking, monotherapy indicates one drug and combination therapy more than one drug. The increase in numbers since the last audit period is a consequence of more patients receiving treatment but under reporting of drug therapies cannot be excluded.

Table 6 shows monotherapy data and Table 7 shows combination therapy data. A total of 2844 patients were receiving therapy on 31 March 2011. Figure 24 shows the distribution of monotherapy and combination therapy treatment by age. Up to the age of 19 years 47.5 per cent of patients are receiving combination therapy. This falls with age: between 20 and 59 years 30 to 33 per cent are treated with combination therapy, 27.5 per cent between 60 and 69 years, 24 per cent between 70 and 79 years and 16 per cent in the range 80 and 89 years. Finally in this section, Table 8 lists the number of prescriptions for the individual drugs themselves irrespective as to whether they were prescribed as monotherapy or in combination.

**Table 6**  
Number of patients on monotherapy on 31 March 2010 and 2011

Type of drug	Total 2010	Total 2011
Endothelin receptor antagonist	775	800
Phosphodiesterase 5 inhibitor	1038	1111
Prostanoid therapy	111	82
Calcium Channel Blockers for Vasoreactive PAH	29	6
Unknown	32	36
<b>Total</b>	<b>1985</b>	<b>2035</b>

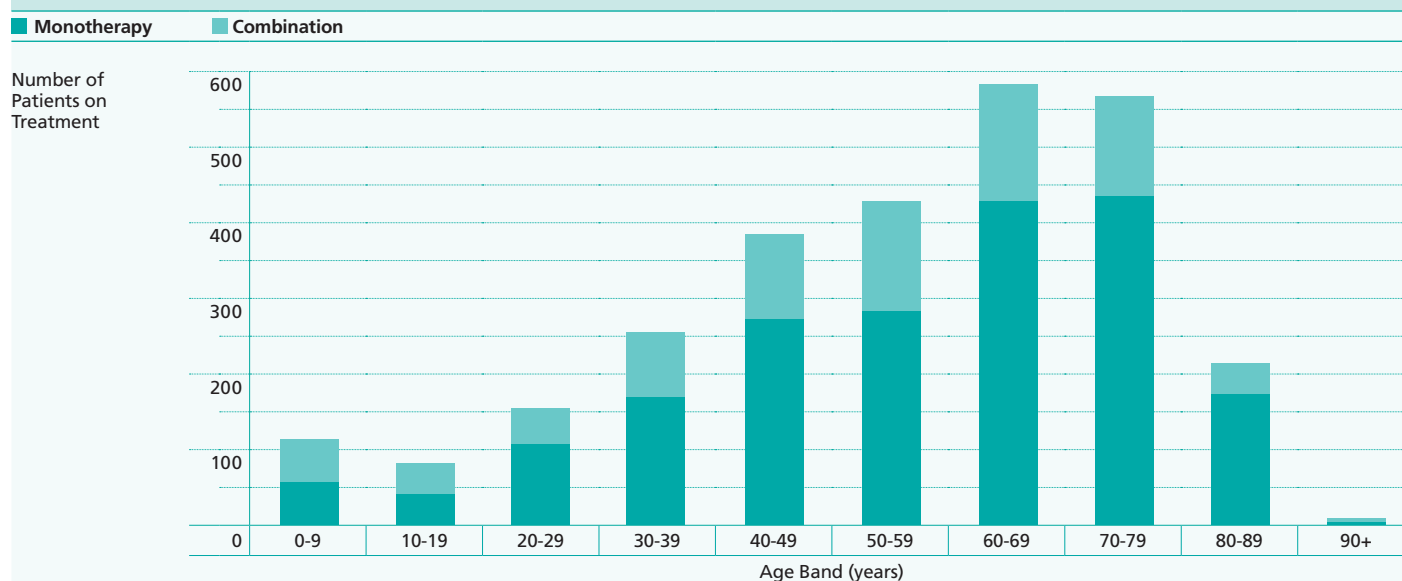
PAH - pulmonary arterial hypertension. Unknown indicates clinical trial medication or the drug therapy type was unknown.

**Table 7**  
Number of patients on combination therapy on 31 March

Combination Therapy	Total 2010	Total 2011
Endothelin receptor antagonist & phosphodiesterase 5 inhibitor	282	578
Endothelin receptor antagonist & phosphodiesterase 5 inhibitor & prostanoid therapy	25	60
Endothelin receptor antagonist & phosphodiesterase 5 inhibitor & calcium channel blocker for vasoreactive PAH	2	8
Endothelin receptor antagonist & prostanoid therapy	26	36
Phosphodiesterase 5 inhibitor & prostanoid therapy	65	104
Phosphodiesterase 5 inhibitor & prostanoid therapy & calcium channel blockers for vasoreactive PAH	1	0
Unknown combination	22	23
<b>Grand Total</b>	<b>423</b>	<b>809</b>

PAH - pulmonary arterial hypertension. Unknown indicates clinical trial medication or which combination drug therapy was not recorded. One drug may be known but a second unknown, or two known and a third unknown, or both/all unknown.

**Figure 23**  
Age of patients treated with monotherapy and combination therapy on 31 March 2011



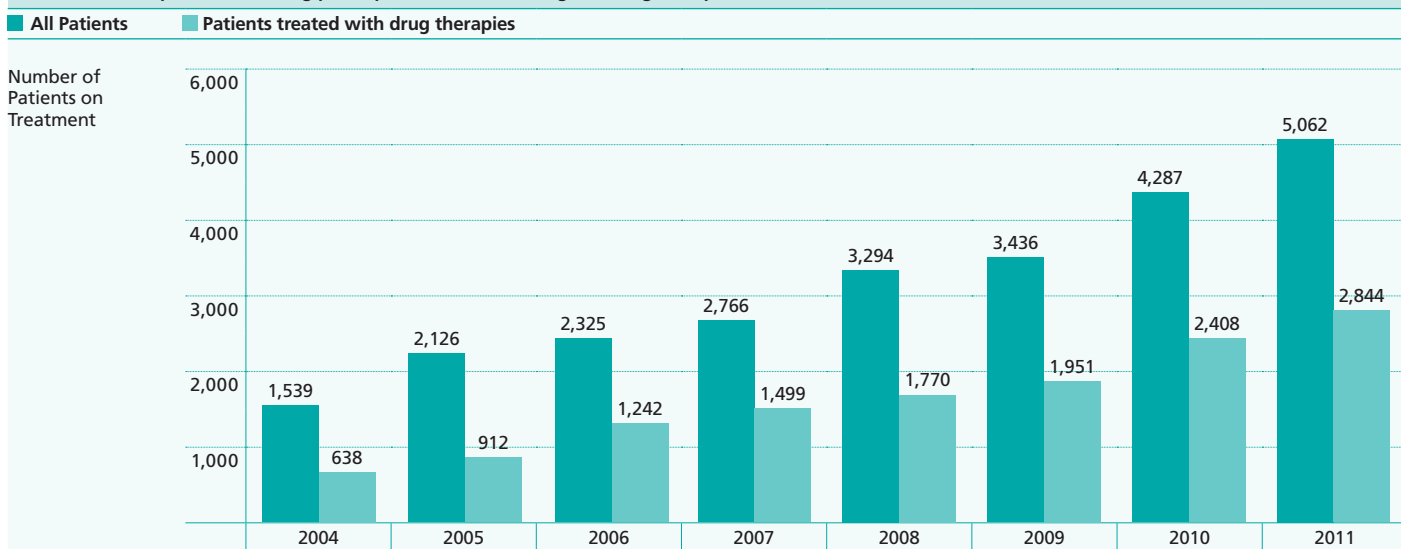
**Table 8**  
Number of drug prescriptions on 31 March including monotherapy and combination therapy. Drugs administered by different routes appear more than once in the table

Drug	Total Prescriptions 2010	Total Prescriptions 2011
Sildenafil	1414	1869
Bosentan	909	1208
Ambrisentan	120	277
Sitaxsentan	88	0
Iloprost iv	49	54
Iloprost nebulised	47	66
Treprostinil sc	44	47
Treprostinil iv	42	36
Treprostinil nebulised	2	6
Epoprostenol iv	39	69
Amlodipine	12	18
Nifedipine	8	6
Tadalafil	5	6
Diltiazem	1	0
Nicardipine	1	1
Unknown	91	78
<b>Total</b>	<b>2872</b>	<b>3741</b>

Key:  
iv - intravenous infusion; sc - subcutaneous infusion.  
Unknown indicates clinical trial therapy or that the class of drug therapy was recorded but not the actual drug by name.

Census data describing the number of patients prescribed disease-targeted therapy compared to previous years is shown in [Figure 24](#). The number of patients seen is also included in this histogram for comparison and is the same data as shown in [Figure 1](#).

**Figure 24**  
**Number of patients active under the service and number of patients on disease- targeted therapies for pulmonary hypertension on 31 March. Drug therapy refers to the number of patients receiving prescriptions for disease-targeted drug therapies**



# Clinical Activity

## Disposition of Patients

Table 9 shows the number of patients who had been seen by the designated centres during the audit and identifies the number who remain under the service at the end of the year. A total is given for the number of patients who have been discharged, undergone lung or heart- lung transplantation or died during the year. In the audit database, death is normally entered by the treating hospital, but if this is missed then it is identified by automatic cross-checking of the audit database with the Office of National Statistics database.

Note that patients who moved between centres, and hence were seen in more than one centre during the year, have been intentionally double counted as they represent clinical work for each centre at which they are seen. The total number of these patients is 36.

The most common reasons for patients being seen at more than one centre were:

- Patients who were transitioned from the UK Children's Pulmonary Hypertension Service
- Patients who moved their place of residence within the UK, Channel Islands and Isle of Man
- Patients who wished to transfer to another hospital

**Table 9**  
Clinical activity for the whole population showing number and percentage of patients per clinical outcome per annum. Percentages have been rounded to the nearest whole number

A. For all patients		
Clinical Outcome	Total 2010	Total 2011
Alive & Not Discharged	4327	5035
	78%	81%
Died	560	444
	10%	7%
Discharged	651	727
	12%	12%
Transplanted	10	11
	0%	0%
Total	5538	6196
	100%	100%

PAH - pulmonary arterial hypertension. Unknown indicates clinical trial medication or the drug therapy type was unknown.

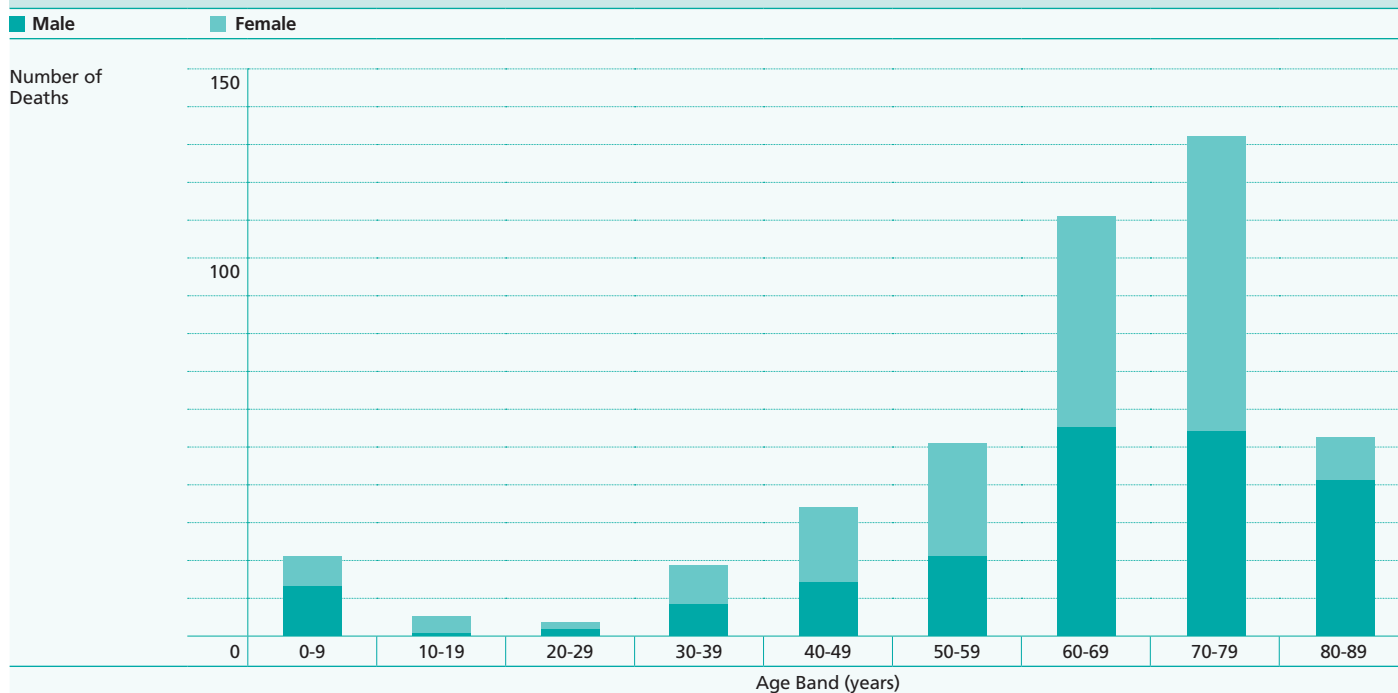
B. By Country					
Clinical Outcome	England	Scotland	Wales	Ireland	Other / Unknown
Alive & Not Discharged	4378	351	207	29	70
	82%	70%	86%	78%	67%
Died	354	44	21	4	21
	7%	9%	9%	11%	20%
Discharged	582	104	13	4	14
	11%	21%	5%	11%	12%
Transplanted	10	0	0	0	1
	0%	0%	0%	0%	1%
Total	5314	499	241	37	105
	100%	100%	100%	100%	100%

Other includes the Channel Islands, Gibraltar and the Isle of Man. Unknown indicates that the country of the patients has not been identified.

## Deaths

The distribution of all cause mortality is shown in [Figure 25](#). The median age at death was 67 years (interquartile range 55 – 76 years) and 55 per cent were in females. A diagnosis of pulmonary arterial hypertension had been made in 48 per cent of patients who died.

**Figure 25**  
Age and sex distribution of deaths of patients under the pulmonary hypertension centres in 2010-11



## Clinical Investigations: Cardiac Catheterization

This section reports one of a number of selected investigations commonly carried out in patients with pulmonary hypertension as a measure of clinical workload.

Cardiac catheterization is required to make a definitive and accurate diagnosis. Table 10 shows the number of procedures undertaken on patients according to their country of origin and the proportion of the total. Some cardiac catheterizations are undertaken at another hospital before referral to a designated centre. Such procedures are not counted here.

**Table 10**  
Number of cardiac catheterization procedures carried out at designated centres by country. Percentages have been rounded to the nearest whole number

Country	Number of procedures 2010	Number of procedures 2011	Number of patients 2011	Percentage of patients 2011
England	1451	1662	1447	87
Northern Ireland*	8	5	5	0
Scotland	126	124	122	7
Wales	62	53	50	3
Other	4	7	4	0
Unknown	8	44	39	3
<b>UK Total</b>	<b>1659</b>	<b>1895</b>	<b>1667</b>	<b>100%</b>

\* Very limited data was obtained from Northern Ireland. Other includes the Channel Islands, Gibraltar and the Isle of Man. Unknown indicates that the country of the patients has not been identified.

The number of procedures performed per patient was 1.1. This number only reflects repeat procedures if two or more procedures were carried out during the audit and thus may underestimate the number of patients who have repeat procedures if these were carried out either side of the audit period.

## Clinical Investigations: Echocardiography

This section reports one of a number of selected investigations commonly carried out in patients with pulmonary hypertension as a measure of clinical workload.

Echocardiography is used to assess whether a patient is likely to have pulmonary hypertension, to diagnose causes such as left ventricular disease, aortic or mitral valve disease and congenital heart disease, and to monitor progress during serial follow-up. Echocardiography is in addition to other investigations. Table 11 shows the number of procedures undertaken on patients according to their country of origin and the proportion of the total. Echocardiograms performed at other hospitals have not been counted here.

**Table 11**  
Number of echocardiograms carried out at designated centres by country. Percentages have been rounded to the nearest whole number

Country	Number of procedures 2010	Number of procedures 2011	Number of patients 2011	Percentage of patients 2011
England	5489	5349	3323	80
Northern Ireland*	54	91	21	0
Scotland	539	569	344	8
Wales	209	150	98	2
Other	10	10	7	0
Unknown	17	81	59	1
<b>UK Total</b>	<b>3823</b>	<b>4231</b>	<b>3285</b>	<b>100%</b>

\* Very limited data was obtainable for Northern Ireland. Other includes the Channel Islands, Gibraltar and the Isle of Man. Unknown indicates that the country of the patients has not been identified.

The number of procedures performed per patient was 1.2. This number only reflects repeat procedures if two or more procedures were carried out during the audit and thus may underestimate the number of patients who have repeat procedures if these were carried out either side of the audit period.

### Clinical Investigations: Exercise Testing

This section reports one of a number of selected investigations commonly carried out in patients with pulmonary hypertension as a measure of clinical workload.

Exercise testing is used to undertake a functional assessment of exercise capacity. This is important because most patients with pulmonary hypertension initially present with exercise-induced symptoms including breathlessness, angina and / or syncope. These symptoms, and hence exercise capacity, worsen as disease progresses.

The most commonly used exercise test is the six minute walk test. In this submaximal test patients walk up and down a thirty metre long corridor, unencouraged, for six minutes at their own pace. The total distance walked is measured, as well as oxygen saturation and Borg dyspnoea score before and after exercise. One designated centre (Royal Hallamshire Hospital) uses the shuttle test instead of the six minute walk test.

Cardiopulmonary exercise testing comprises a maximal exercise test with measurement of exhaled gases. Like submaximal tests it is used to assess patients at diagnosis and follow-up. The database field for cardiopulmonary exercise testing was new in 2009 and may not have been filled in consistently during the audit by all designated centres. It is likely underestimated.

The number of procedures undertaken on patients according to their country of origin and the proportion of the total is shown in Table 12. Exercise tests performed at other hospitals have not been counted here.

**Table 12**  
Number of echocardiograms carried out at designated centres by country. Percentages have been rounded to the nearest whole number

Exercise Type	Country	Number of procedures 2010	Number of procedures 2011	Number of patients 2011	Percentage of patients 2011
Six-minute walk test or Shuttle test	England	5489	5349	3323	80
	Northern Ireland	54	91	21	0
	Scotland	539	569	344	8
	Wales	209	150	98	2
	Other	10	10	7	0
	Unknown	17	81	59	1
<b>Six-minute walk test or Shuttle test Total</b>		<b>6318</b>	<b>6250</b>	<b>3852</b>	
Cardiopulmonary	England	193	292	243	6
	Scotland	1	30	29	1
	Wales	19	27	22	1
	Unknown	1	1	6	0
<b>Cardiopulmonary Total</b>		<b>214</b>	<b>350</b>	<b>300</b>	
Type of exercise test not recorded	England	28	21	18	1
	Wales	2	0	2	0
<b>Unknown Total</b>		<b>30</b>	<b>21</b>	<b>20</b>	
<b>UK Total</b>		<b>6562</b>	<b>6621</b>	<b>4172</b>	<b>100%</b>

Other includes the Channel Islands, Gibraltar and the Isle of Man. Unknown indicates that the country of the patients has not been identified.

The number of six minute walk or shuttle tests performed per patient was 1.6 and any exercise tests was 1.6.

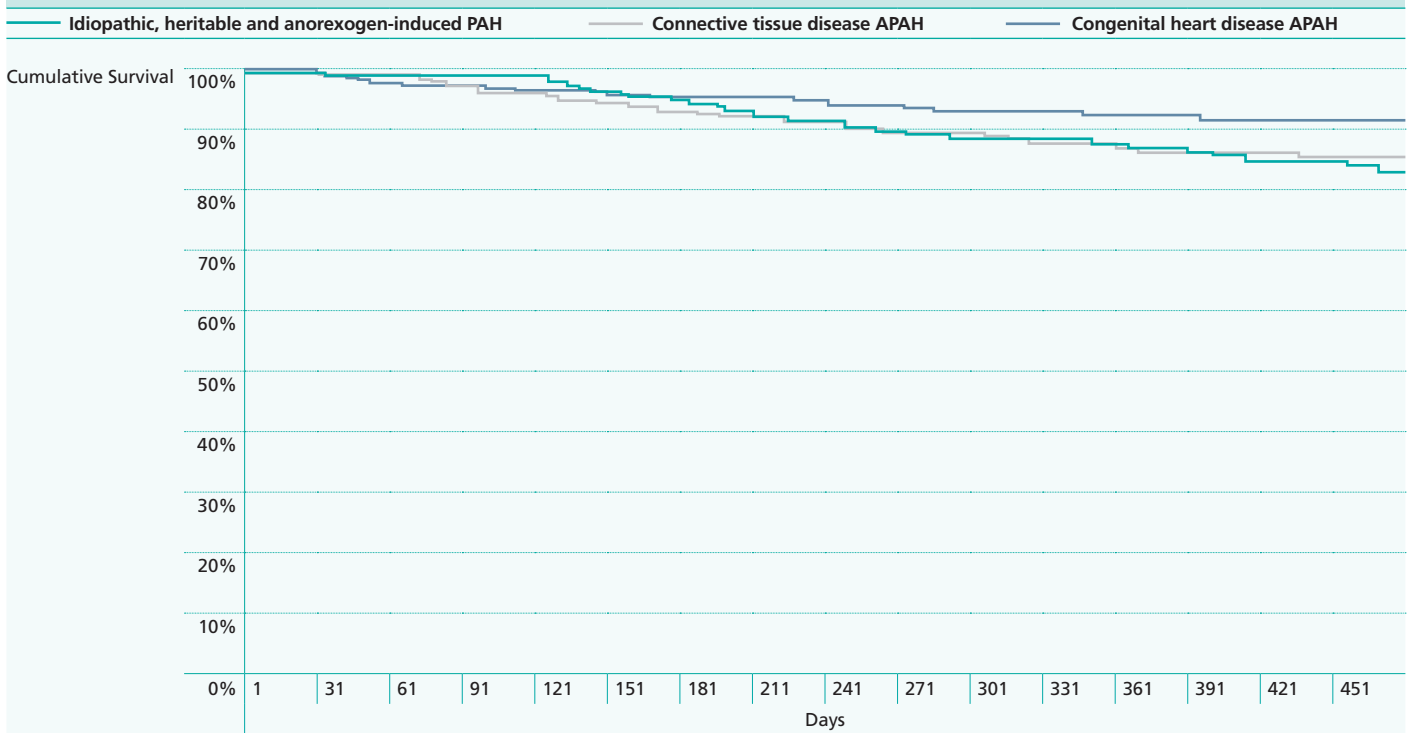
# Survival Outcome Analysis

In any chronic disease with a significant mortality survival is an important outcome measure. Disease-targeted drug therapies used in pulmonary arterial hypertension and chronic thromboembolic pulmonary hypertension slow disease progression but are not curative. This audit provides the opportunity to follow up a large cohort of patients over a long period of time. It also offers the opportunity to compare survival for the same condition between designated centres to identify outliers, hence correct less satisfactory outcomes should they arise. This will follow in the future.

In its second year, a survival curve of patients who were diagnosed on or after 1 April 2009 has limited value but it is shown to demonstrate the valuable information that will become available in future years.

Figure 26 shows data for patients who were seen in designated centres for the first time on or after 1 April 2009 with three types of pulmonary arterial hypertension. Patients with idiopathic, heritable or anorexigen-induced pulmonary arterial hypertension had a median age of 64 years and 60 per cent were female. One year survival was 87 per cent. Patients with connective tissue disease associated pulmonary arterial hypertension had a median age of 64 years and 80 per cent were female. One year survival was 87 per cent. Patients with congenital heart disease associated pulmonary arterial hypertension had a median age of 33 years and 57 per cent were female. One year survival was 92 per cent.

**Figure 26**  
Kaplan Meier survival curve of patients newly diagnosed with idiopathic, heritable or anorexigen-induced pulmonary arterial hypertension, connective tissue disease associated pulmonary arterial hypertension and congenital heart disease associated pulmonary arterial hypertension



Idiopathic PAH	337	284	229	187	143	101
Connective tissue disease	263	224	181	145	105	82
Congenital heart disease	261	231	199	169	124	96

# Audit of Pulmonary Endarterectomy

This section reports data from the Pulmonary Endarterectomy (PEA) Audit. Although this is not part of the National Audit of Pulmonary Hypertension, pulmonary endarterectomy is an important treatment for many patients with chronic thromboembolic pulmonary hypertension (CTEPH). It is now included in this section to provide a more complete picture of the pulmonary hypertension workload and activity in the UK.

Patients with chronic thromboembolic disease are investigated and managed by designated pulmonary hypertension centres and referred to the PEA service for surgical management. The PEA service is designated and funded separately from pulmonary hypertension centres.

As the single national centre commissioned to provide PEA, Papworth Hospital is responsible for the provision of the surgical treatment for patients with CTEPH. This includes assessment of all patients with possible CTEPH referred directly to Papworth Hospital, review of patients diagnosed with CTEPH and referred to Papworth by the designated pulmonary hypertension centres, performing PEA surgery and assessment of surgical outcome by follow up of all patients three months and one year following PEA. Long-term follow up post operatively is carried out at designated pulmonary hypertension centres.

The data presented has been collected prospectively and collated by Papworth Hospital. The data records activity between 1 April 2010 and 31 March 2011. Where follow up is concerned, only patients having follow up from surgery within the year are considered and therefore their full 3 and 12 month follow up data is pending. This will be updated in the subsequent year's report.

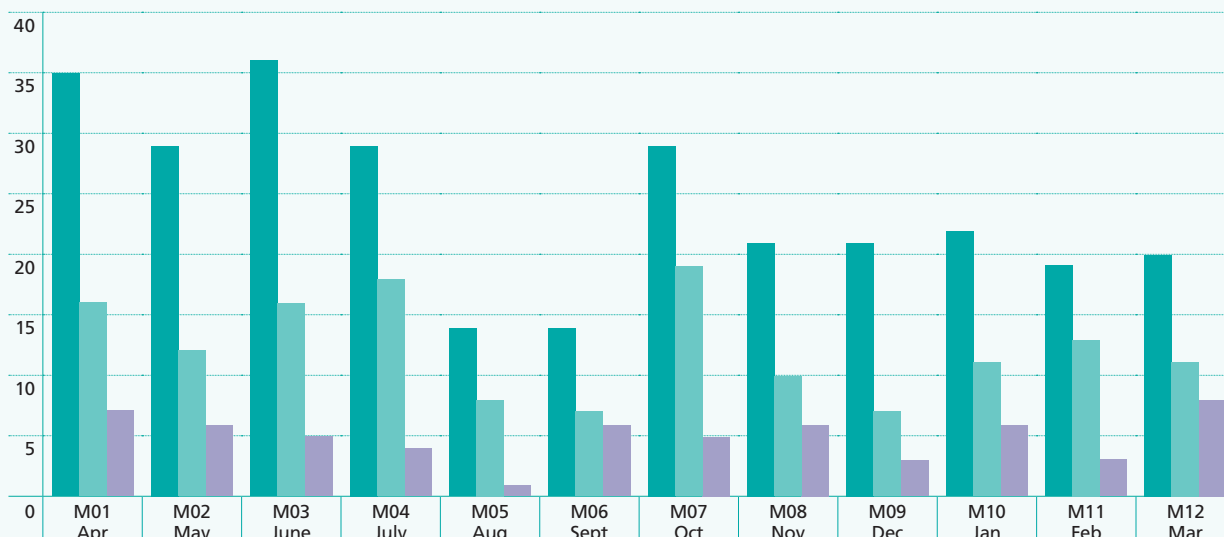
## Assessment

### Referrals to Papworth Multidisciplinary Team Meeting with Suspicion of CTEPH

Figure 28 shows the number of new referrals received (from the designated National Pulmonary Hypertension centres and direct to Papworth) for consideration of PEA. A proportion of referrals are immediately accepted for further surgical work up, and a proportion require further information before a decision can be made. Figure 29 shows the geographical distribution of referrals.

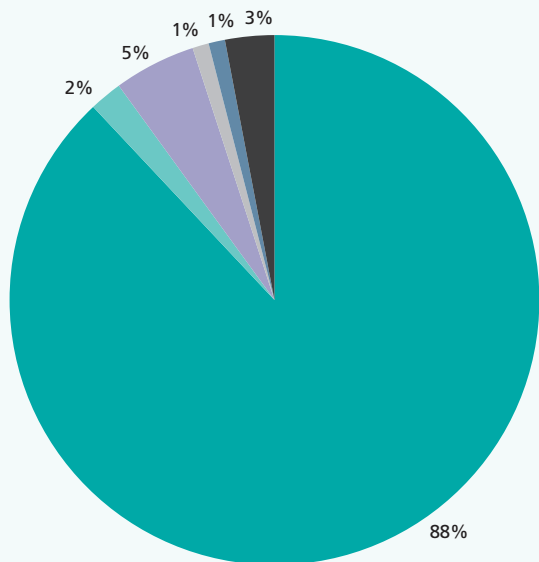
**Figure 27**  
New referrals received for PEA

Number of patients



	M01 Apr	M02 May	M03 June	M04 July	M05 Aug	M06 Sept	M07 Oct	M08 Nov	M09 Dec	M10 Jan	M11 Feb	M12 Mar
Referrals assessed	35	29	36	29	14	14	29	21	21	22	19	20
Immediately accepted for further work up	16	12	16	18	8	7	19	10	7	11	13	11
Further information required before decision	7	6	5	4	1	6	5	6	3	6	3	8

**Figure 28**  
Source of referral for PEA by country



- England – 88%
- Wales – 2%
- Scotland – 5%
- Northern Ireland – 1%
- Republic of Ireland – 1%
- Other – 3%

## Outpatient activity (surgical assessment)

All patients considered suitable for PEA following discussion at the multidisciplinary team meeting are invited to an outpatient appointment for assessment and discussion with the surgeon and specialist nursing team.

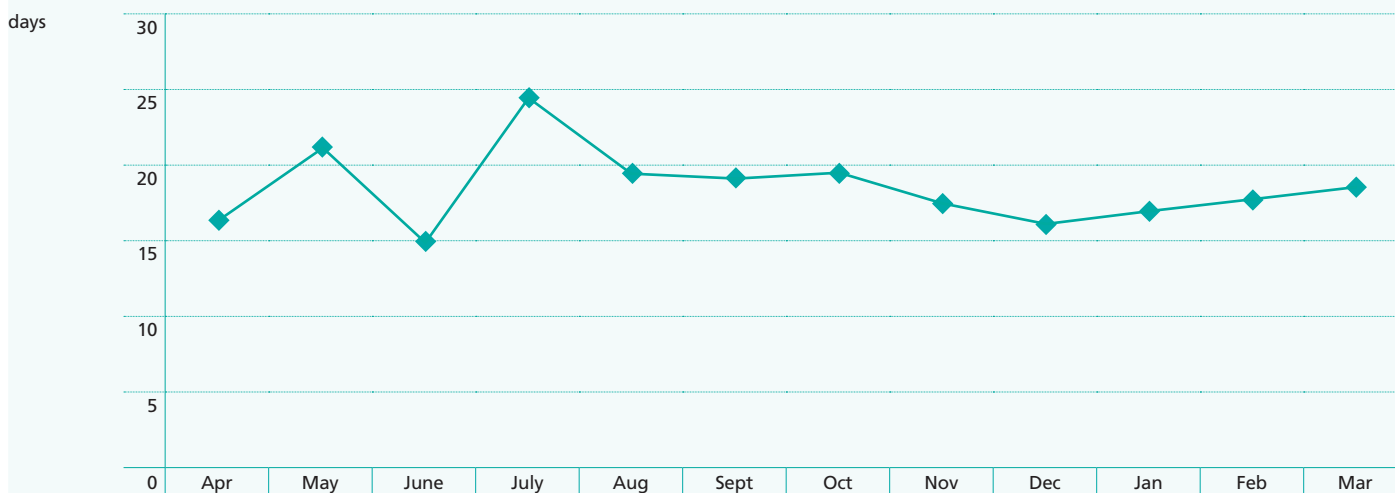
	2010-2011
Number of pre-operative outpatient appointments	133

## Surgery

Table 13 Patients undergoing surgery	
	2010-2011
Age (mean)	58
Number of patients on targeted therapy – pre surgery	86 (67%)
Number of operations	129
Number of major concomitant operative procedures	20
ECMO* usage peri-operative	8
Redo PEA surgery	3
Total in-hospital mortality	5.43%

\*ECMO, extracorporeal membrane oxygenation

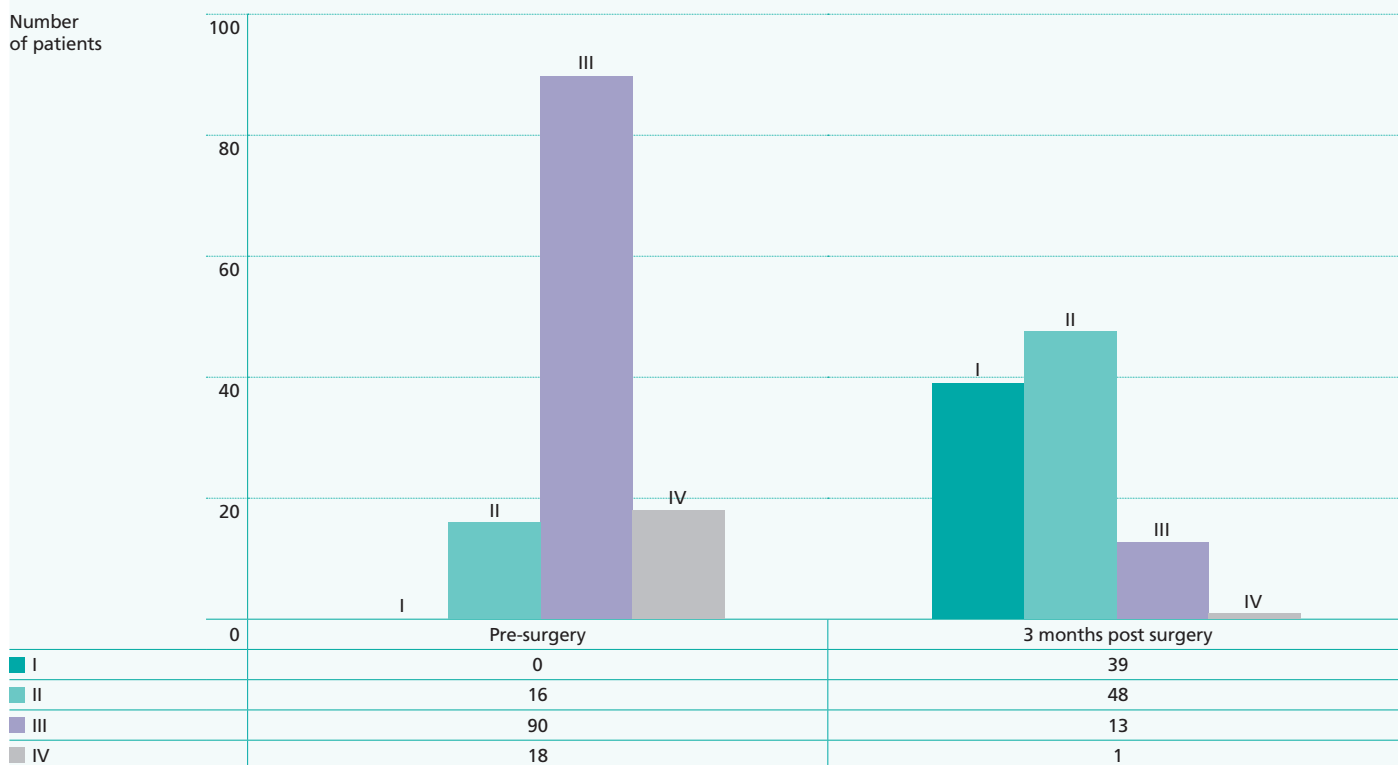
**Figure 29**  
Length of hospital stay for patients undergoing pulmonary endarterectomy



## Follow up 3 months after PEA

	2010-2011
Number of 3 month follow ups	108
No. patients on targeted therapy – post op	7 (6.5%)

**Figure 30**  
**WHO functional class of patients undergoing PEA, pre-operatively and 3 months after surgery**



### Audit of Pulmonary Endarterectomy: Acknowledgements

The project wishes to acknowledge the following who have provided support for the audit and provided guidance during the data collection period, analysis and writing of this report (in alphabetical order): Mr. John Dunning, Alison Gibson, Dr Deepa Gopalan, Mr. David Jenkins, Dr Robert MacKenzieRoss, Dr Joanna Pepke-Zaba, Anie Ponnaberanam, Maureen Rootes, Dr. Nicholas Screatton, Dr. Karen Sheares, Carrie Symington, Carmen Treacy, Mr Steven Tsui.

## Key Findings

1. All designated pulmonary hypertension centres in the UK participated in this report of the second year of the National Audit of Pulmonary Hypertension.
2. There was significant under reporting of patients in Northern Ireland.
3. There has been progressive growth in the number of patients managed by designated centres in the UK since 2004.
4. During the audit UK pulmonary hypertension services saw 6196 patients. Since 36 patients were seen in more than one centre during the year, the workload of the centres is represented by 6232 patients. 86 per cent of patients came from England. The median age was 62 years which is 2 years older than the previous year. There were 1.8 females to every male.
5. In Great Britain designated centres saw 97 patients per million population. In England the prevalence of patients with pulmonary hypertension seen at designated centres was highest down the east side of the country where the centres are located.
6. Of the total number of patients, 12 per cent were discharged and 11 patients were transplanted.
7. There were 444 deaths (7 per cent of the population) at a median age of 67 years.
8. Pulmonary arterial hypertension was the commonest diagnosis in 46 per cent of patients followed by chronic thromboembolic hypertension in 18 per cent. 18 per cent did not have pulmonary hypertension.
9. There were 2083 patients admitted to designated centres as inpatients for 12093 bed days. Investigations at designated centres totalled 1895 cardiac catheterization procedures, 4231 echocardiograms and 6621 exercise tests.
10. There were 2089 new patients referred to designated centres. The commonest diagnoses were pulmonary arterial hypertension (31 per cent) and no pulmonary hypertension (31 per cent). There was wide variation in referral rates in different geographical regions.
11. Disease targeted drug therapy was prescribed for 2980 patients, mean age 59 years and median age 60 years. The ratio of females to males was 1.9:1.
12. The standardised rate of treatment of patients with disease-targeted drug therapies was similar in England, Scotland and Wales but there was wide variation in treatment rates across England.
13. The most commonly prescribed drugs were phosphodiesterase 5 inhibitors followed by endothelin receptor antagonists.
14. One year survival for new cases of idiopathic, heritable and anorexigen-induced pulmonary arterial hypertension was 87 per cent and for connective tissue disease associated pulmonary arterial hypertension was 87 per cent. New patients seen with congenital heart disease associated pulmonary arterial hypertension had a one year survival of 92 per cent.
15. In a separate audit of pulmonary endarterectomy, 129 operations were performed with an in hospital mortality of 5.4 per cent and with improvement in WHO functional class 3 months post operatively.

## Recommendations

1. To maintain this audit in the future it is imperative that trusts ensure that staff with appropriate skills and access to the necessary resources are in place to complete data entry.
2. Resourcing of data collection needs to be built into pulmonary hypertension service tariff arrangements in a sustainable way.
3. An audit of source data should be carried out to assess data accuracy of key data fields.
4. Continued review and improvement of database systems is required to ensure high quality data.
5. The inequality of access of patients in different diagnostic groups to designated centres needs to be addressed by all stakeholders.
6. There is a need to describe the experience of patients who attend designated centres by the selection and implementation of patient reported outcomes.
7. Over the next year the audit should start to make comparisons between designated centres of survival of selected patient diagnostic groups.
8. The lack of adequate data from Northern Ireland needs to be investigated and addressed by the NHS in Northern Ireland.
9. The clinical findings in this report should continue to be reported annually.
10. The NAPH should continue to be supported and funded since it has demonstrated that important information can be collected and analysed for the benefit of patients and future service planning.

## References

1. The NHS Information Centre. National Audit of Pulmonary Hypertension, 1st Annual Report. 2010. <http://www.ic.nhs.uk/services/national-clinical-audit-support-programme-ncasp/heart-disease/pulmonary-hypertension>.
2. National Pulmonary Hypertension Centres of the UK and Ireland. Consensus statement on the management of pulmonary hypertension in clinical practice. *Heart* 2008; 94 (Suppl I):i1-i41. *Thorax* 2008; 63 (Suppl II):ii1-ii41.
3. Galiè N, Hoeper MM, Humbert M, Torbicki A, Vachiery JL, Barbera JA, Beghetti M, Corris P, Gaine S, Gibbs JSR, Gomez-Sanchez MA, Jondeau G, Klepetko W, Opitz C, Peacock A, Rubin L, Zellweger M, Simonneau G. Guidelines for the diagnosis and treatment of pulmonary hypertension: The Task Force for the Diagnosis and Treatment of Pulmonary Hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS), endorsed by the International Society of Heart and Lung Transplantation (ISHLT). *Eur Heart J.* 2009;30:2493-2537 and *Eur Respir J.* 2009;34:1219-1263.
4. Simonneau G, Robbins IM, Beghetti M, et al. Updated clinical classification of pulmonary hypertension. *J Am Coll Cardiol* 2009 Jun 30;54(1 Suppl):S43-S54.

# Appendix 1: Dana Point clinical classification of pulmonary hypertension

## 1 – Pulmonary arterial hypertension (PAH)

- 1.1 Idiopathic PAH
- 1.2 Heritable
  - 1.2.1 BMPR2
  - 1.2.2 ALK1, endoglin (with or without hereditary haemorrhagic telangiectasia)
  - 1.2.3 Unknown.
- 1.3 Drugs and toxins induced
- 1.4 Associated with (APAH):
  - 1.4.1 Connective tissue diseases
  - 1.4.2 HIV infection
  - 1.4.3 Portal hypertension
  - 1.4.4 Congenital heart disease
  - 1.4.5 Schistosomiasis
  - 1.4.6 Chronic haemolytic anaemia
- 1.5 Persistent pulmonary hypertension of the newborn

## 1' Pulmonary veno-occlusive disease and/or pulmonary capillary haemangiomatosis

## 2 – Pulmonary hypertension due to left heart disease

- 2.1 Systolic dysfunction
- 2.2 Diastolic dysfunction
- 2.3 Valvular disease

## 3 – Pulmonary hypertension due to lung diseases and/or hypoxia

- 3.1 Chronic obstructive pulmonary disease
- 3.2 Interstitial lung disease
- 3.3 Other pulmonary diseases with mixed restrictive and obstructive pattern
- 3.4 Sleep-disordered breathing
- 3.5 Alveolar hypoventilation disorders
- 3.6 Chronic exposure to high altitude
- 3.7 Developmental abnormalities

## 4 – Chronic thromboembolic pulmonary hypertension

## 5 – Pulmonary Hypertension with unclear and/or multifactorial mechanisms

- 5.1 Haematological disorders: myeloproliferative disorders, splenectomy
- 5.2 Systemic disorders, sarcoidosis, pulmonary Langerhans cell histiocytosis, lymphangioleiomyomatosis, neurofibromatosis, vasculitis
- 5.3 Metabolic disorders: glycogen storage disease, Gaucher disease, thyroid disorders
- 5.4 Others: tumoral obstruction, fibrosing mediastinitis, chronic renal failure on dialysis

ALK-1: Activin receptor-like kinase 1 gene

APAH: associated pulmonary arterial hypertension

BMPR2: Bone morphogenetic protein receptor, type II

HIV: human immunodeficiency virus

PAH: pulmonary arterial hypertension

## Appendix 2: Specialised Commissioning Drug Policy for England

This table was extracted from National Specialised Commissioning Group, Interim Commissioning Policy, Target therapies for the treatment of pulmonary arterial hypertension in Adults, 1st July 2008

	Pulmonary Arterial Hypertension (IPAH, FPAH, Anorexogen-induced, and where associated with Portal hypertension or HIV infection)	PAH associated with significant venous or capillary involvement	PAH associated with connective tissue disease	PAH associated with congenital heart disease	PH due to chronic thrombotic and/or embolic disease
<b>First line</b>	WHO III: S WHO IV: P	PVOD only: WHO III/IV, S, consider surgery PCH: no disease targeted treatment supported consider surgery	WHO III/IV: B	WHO III: B WHO IV: B as a bridge to transplant	WHO III/IV: S Consider surgery
<b>Second line/ Alternative*</b>	WHO III: B,P or X WHO IV: B		WHO III/IV: S, P or X		WHO III/IV: B
<b>Combination</b>	B+S or X+S Patients should be entered into a clinical trial where possible		B+S or X+S Patients should be entered into a clinical trial where possible	B+S will be considered as a bridge to surgery	B+S will be considered as a bridge to surgery
<b>Alternate combination</b>	P+S Patients must be entered into a clinical trial (NB trial participation previously agreed by commissioners)				

B= Bosentan, P= Prostanoids, S= Sildenafil, X= Sitaxsentan OR Ambrisentan

WHO= Functional classification of PH modified after the New York Heart Association functional classification according to the World Health Organisation 1998

IPAH= Idiopathic PAH; FPAH= Familial PAH; PVOD= Pulmonary veno-occlusive disease; PCH= Pulmonary capillary haemangiomas

\* If first line is contraindicated, ineffective in controlling symptoms or poorly tolerated

## Appendix 3: Members of Staff who participated in the Audit at the Designated Pulmonary Hypertension Centres

\* indicates staff who entered data

	Freeman Hospital	Golden Jubilee Hospital	Great Ormond Street Hospital	Hammersmith Hospital	Papworth Hospital	Royal Brompton Hospital	Royal Free Hospital	Royal Hallamshire Hospital
<b>Lead Clinician</b>	Paul Corris	Andrew Peacock	Ingram Schulze-Neick	Simon Gibbs	Joanna Pepke-Zaba	John Wort	Gerry Coghlan*	David Kiely
<b>Supporting Physicians</b>	James Lordan Andrew Fisher Guy MacGowan	Martin Johnson		Luke Howard Rachel Davies	Nick Morrell Karen Sheares	Michael Gatzoulis Kostas Dimopoulos Phil Marino Gerhard Diller	Clive Handler* Benji Schreiber*	Charlie Elliot Robin Condliffe Ian Sabroe
<b>Surgeons (PEA)</b>					David Jenkins John Dunning Steven Tsui			
<b>Clinical Nurse Specialists</b>	Margaret Day Rachel Crackett* Julia de Soyza	Agnes Crozier Alison Curran Val Pollock	Yvette Flynn Harriet Foster	Wendy Gin-Sing Chantal Torpy Eilish Lawlee	Natalie Doughty* Sam Clare* Anie Ponnaberanam* Maureen Rootes* Nicola Speed* Kathy Page*	Carl Harries Lisa Parfitt	Sally Redcliffe* Adele Gallimore* Katie Mullard* Clare Das Joanna Lansell	Iain Armstrong (nurse consultant) Paul Sephton Lisa Martin Jane Wilkinson
<b>Data Managers</b>	Paul McAlinden* Anne Ramsey*	Simon Kerridge*	Barbara Margetts*	Lucia Heath*	Carmen Treacy*	Sweeta Dhakan*	Anthony Pivowarski	Sheila Forshaw
<b>Pharmacist</b>	Maria Allen	Allan Smith	Lynne Cochrane	Lynn Humphrey	Duncan Grady	Beejal Shah	Jay Pang	Neil Hamilton
<b>Radiologists</b>	Leslie Mitchell Michelle Muller	Michael Sproule	Cathy Owens Andrew Taylor	Mary Roddie Christoph Juli James Jackson	Nick Screaton Deepa Gopalan	Michael Rubens Simon Padley	Charlotte Cash Jamanda Haddock	Christine Davies Catherine Hill
<b>Service Manager</b>	Liz Bailey			Jo Nicholson Michelle Andrews	Alison Gibson Carrie Symington	Kelly Goulding	Olivia Ellah	Lisa Needham
<b>IT Staff</b>					Mike Moore Jim Butler	Colin Gordon George Lampadariou		Stephen Stewart Theresa Dodd H Jon Wrend
<b>Administrative Staff</b>	Kim Clay	Veronica Ferry Lorraine James	Barbara Margetts*	Jemaine Charles Claire Gin-Sing Byron Wong	Karin Johnson Tanya Vonseld Jackie Richmond Lesley Humphrys	Lilly Morgans Sylvia Dedman	Inness Hammel-Crowe* Alison Narcisse*	Denise Stephenson Natalie Johnson

PEA, pulmonary endarterectomy

## Appendix 4: Members of the NAPH Project Board

Name	Role	Organisation
Roger Boyle	Policy Lead	Department of Health
Geoffrey Carroll (Wales)	Medical Director	Welsh Health Specialised Services
Natalie Doughty	Senior User (Nursing)	Papworth NHS Foundation Trusts
Cathy Edwards	Senior User (Commissioners)	Director Yorkshire & Humber SCG, England
Simon Gibbs (Chair)	Clinical Lead	Imperial College London & Imperial College Healthcare NHS Trust
Julie Henderson	Acting Head for Clinical Analysis	The NHS Information Centre
Martin Johnson	Senior User (Physician)	NHS Greater Glasgow & Clyde
Lesley King	Senior User (Commissioners)	The Eastern Health and Social Services Board (Northern Ireland)
Julie Michalowski	Project Manager	The NHS Information Centre
Lisa Needham	Senior User (Manager)	Sheffield Hospitals NHS Trust
Andrew Peacock	Senior User (Physician)	Chair of Physicians Committee from 2009
Ingram Schulze-Neick	Senior User (Physician)	Paediatric Pulmonary Hypertension Service
Dr Carol Wilson	Senior User (Commissioners)	Consultant Cardiologist Royal Victoria Hospital Site, Belfast Health & Social Care Trust
Mike Winter	Senior User (Commissioners)	Medical Director, National Services Division, Scotland
Kay Yeowart	Senior User (Patients)	Pulmonary Hypertension Association UK

The NHS Information Centre for health and social care (The IC) is working to make information more relevant and accessible to the public, regulators, health and social care professionals and policy makers, leading to improvements in knowledge and efficiency.

The IC is a special NHS health authority that collects, analyses and distributes data to reduce the burden on frontline staff, releasing more time for direct care.

This work remains the sole and exclusive property of The IC and may only be reproduced where there is explicit reference to the ownership of The IC. This work may be re-used by NHS and government organisations without permission. Commercial re-use of this work must be granted by The IC.

Copyright © 2011. The NHS Information Centre, National Pulmonary Hypertension Audit. All rights reserved.

Ref: IC22090311

**Need to know more?**

T. 0845 300 6016

E. [enquiries@ic.nhs.uk](mailto:enquiries@ic.nhs.uk)

[www.ic.nhs.uk](http://www.ic.nhs.uk)

The NHS Information Centre  
for health and social care  
1 Trevelyan Square  
Boar Lane  
Leeds  
LS1 6AE