

Executive Summary of Key Findings

1. Pulmonary hypertension is a rare disease which occurs at any age, has many causes, and often shortens life expectancy.
2. In the UK, Channel Islands and Isle of Man seven hospitals have been designated to diagnose and treat pulmonary hypertension in adults and one hospital for children.
3. The NAPH is a prospective audit of processes and outcomes and has the participation of all eight designated centres.
4. This first annual report covers the period 1 April 2009 to 31 March 2010 during which database development was taking place.
5. The data provides a responsible overview of clinical activity and prescribing for patients with pulmonary hypertension in the UK, Channel Islands and Isle of Man.
6. In the first year of this audit the data in this report should be considered as indicative of the national picture subject to the limitations of the quality of the data at the time of the audit. The accuracy of the data cannot be verified without an audit of source data and this has not been undertaken.
7. During the audit UK pulmonary hypertension services saw 5478 patients. Since 60 patients were seen in more than one centre during the year, the workload of the centres is represented by 5538 patients. The mean age was 56 years and the median age was 60 years. There were 1.9 females to every male.
8. Of the total number of patients, 12 per cent were discharged and 10 per cent died during the one year audit. Ten patients were transplanted.
9. Investigations at designated centres totalled 1659 cardiac catheterization procedures, 3823 echocardiograms and 6562 exercise tests.
10. Disease targeted drug therapy was prescribed for 2980 patients, mean age 56 years and median age 59 years. The ratio of females to males was 1.9:1.
11. The most commonly prescribed drugs were phosphodiesterase 5 inhibitors followed by endothelin receptor antagonists.
12. There has been progressive growth in the number of patients managed by designated centres in the UK since 2004.
13. It is recommended that the audit should continue to collect and report data in the future.

Quality of the Data Used for This Report

The new NAPH database described in the introduction was work-in-progress during the period of this audit. During this period the database required changes and additions to data already being collected by designated centres before the audit began. This in turn required designated centres to carry out a large amount of work retrospectively on existing patient data as well as to enter new data prospectively. For users this was further complicated by the addition and adjustment of database fields during the audit period. Technical issues with data uploads resulted in missing or corrupted data when it finally populated the central database. These difficulties made complete data collection unattainable. Further important technical issues arose within the database which were highlighted after the data for the annual report had been extracted. During this period communication between the lead clinician, users and the NHS IC was maintained by monthly newsletters and six monthly meetings of the Users Group in London.

Data from selected key fields was used for this report. The dataset itself is more extensive than this report belies. Data entry on existing patients was not unexpectedly incomplete when this audit data was extracted from the database in May 2010.

Specifically, some important key data items are incomplete in this report and this is its major limitation. These are highlighted in individual sections. The quality of the data entered by designated centres was not audited because the major focus of the project during the audit period was on database adjustment and data entry.

There were technical issues with the system whereby duplicate records were created or allowed. A data cleaning process was implemented after data extraction to remove duplicates.

Transcription errors in NHS numbers were identified where the same NHS number was applied to different patients. These records were not excluded from the report as the patients were manually identified as distinct.

The difficulties with developing this national audit were not unexpected. The problems described above were identified and remedied as they arose. They should not detract from most of the data which is reported in subsequent sections which gives at the least an indicative contemporary description of pulmonary hypertension in the UK.

Simple statistics were used to describe data and an example of Kaplan Meier survival analysis was used to describe survival in selected patients.

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 2009 and 31 March 2010 inclusive. The findings of this report are based on those cases entered into the database.

Sixty patients were seen in more than one centre (duplicate patients) and hence the total number of individual patients seen was less than the total number of patients managed by designated centres.

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 2010. It therefore ignores patients who were seen during the audit period and who were discharged, transplanted or who died before the 31 March 2010.

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 seven years.

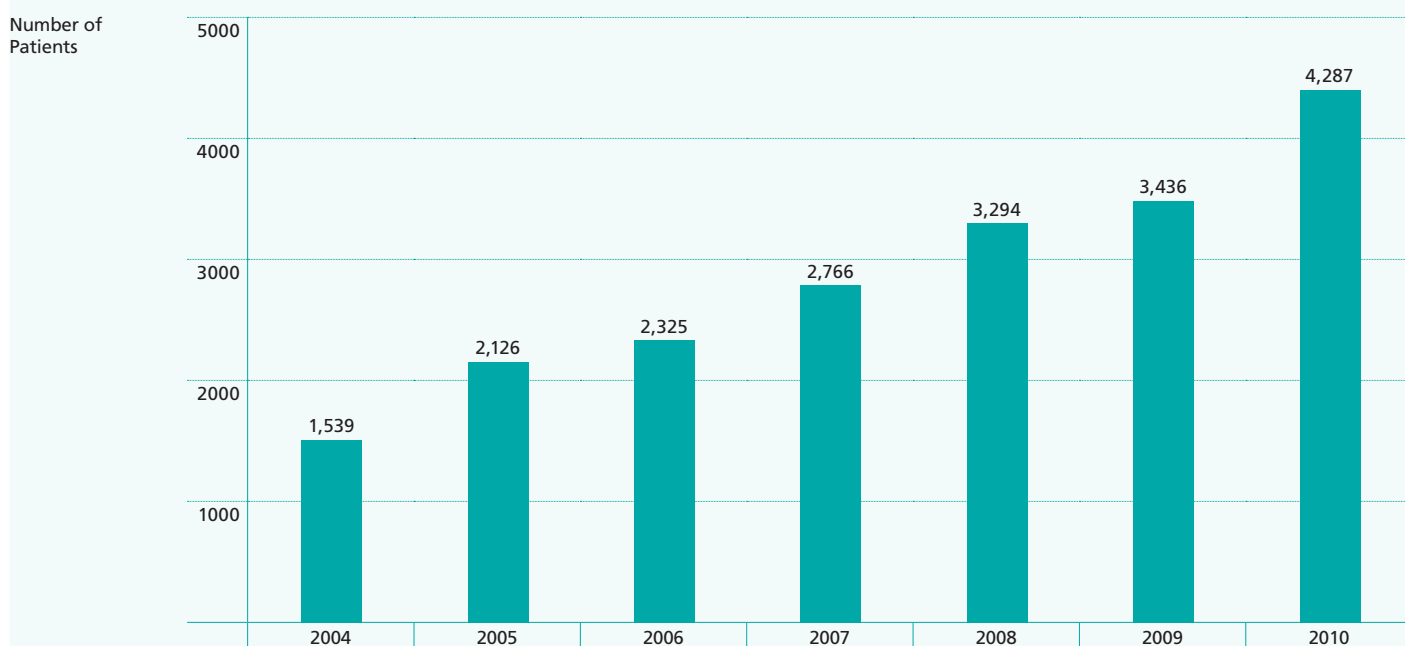
Table 2
Number of patients seen during the audit by each designated centre during the audit.

Designated Pulmonary Hypertension Centre	Number of Patients
Freeman Hospital	375
Golden Jubilee Hospital	409
Great Ormond Street Hospital for Children	376
Hammersmith Hospital	851
Royal Brompton Hospital	567
Papworth Hospital	857
Royal Free Hospital	811
Royal Hallamshire Hospital	1292
Total Patients Seen at Designated Centres	5538
Duplicate Patients	60
Total Patients Seen in the UK, Channel Islands and Isle of Man	5478

Table 3
Number of patients active in the service and alive on 31 March 2010 by designated centre.

Hospital	Total
Freeman Hospital	294
Golden Jubilee Hospital	291
Great Ormond Street Hospital for Children	300
Hammersmith Hospital	664
Royal Brompton Hospital	492
Papworth Hospital	591
Royal Free Hospital	581
Royal Hallamshire Hospital	1074
Total	4287

Figure 1
Number of patients active in the service and alive on 31 March 2010 in seven successive years. In 2009 no data was available from the Royal Brompton Hospital which explains the stalling of the upward trend in numbers in this year.



Patient Demographics

Age and Sex of All Patients

Table 4 shows the age of all patients who were seen during the audit. The majority of patients came from England. Some patients in Northern Ireland were treated locally and did not become part of the audit in year. This will hopefully be rectified for future reports. The small number of patients from Northern Ireland may have skewed the age in this population.

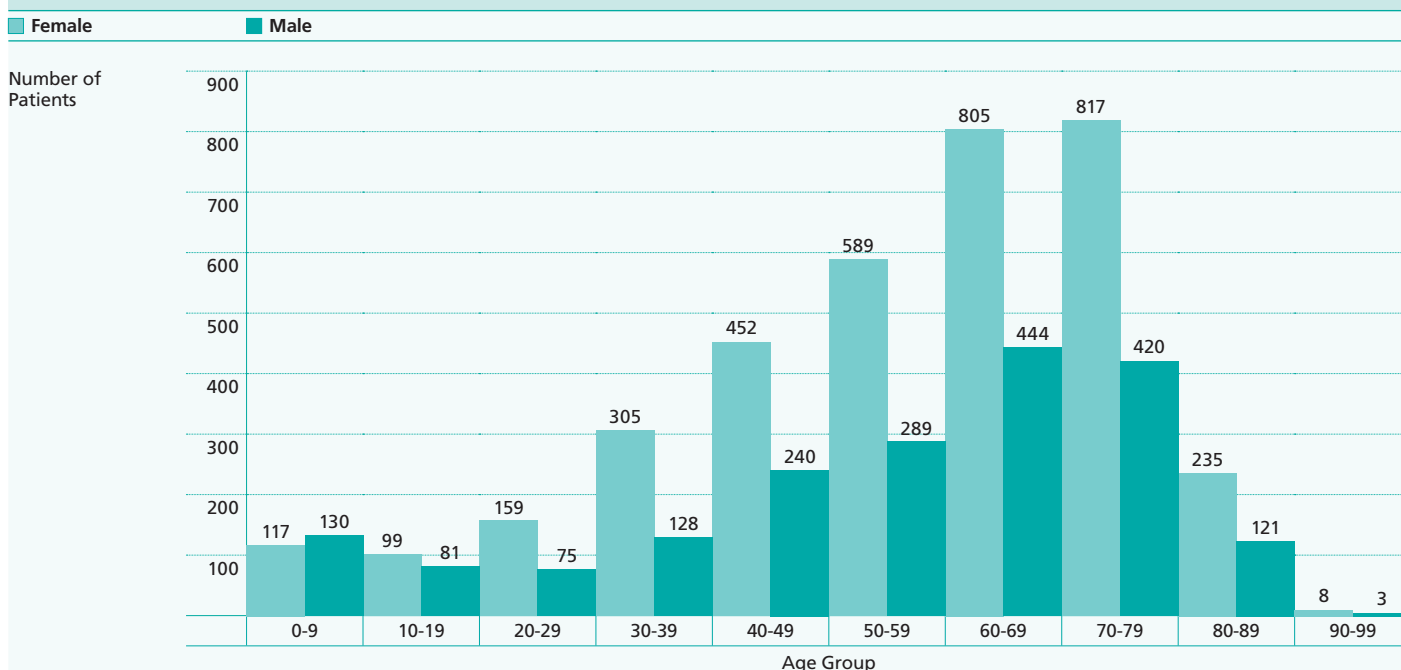
Figure 2 shows the number of males and females by age and includes all patients. The ratio of females: males was 1.9:1.

Table 4
Age of all patients by country.

Region	Patient Count	Mean (years)	Median (years)
UK (all patients)	5538	56	60
England	4819	56	61
Scotland	434	57	60
Wales	209	55	58
Northern Ireland*	40	34	37
Other	36	50	58

* Very limited data was obtainable for Northern Ireland. Other includes Channel Islands, Isle of Man and country not identified.

Figure 2
Distribution of age and sex of all patients. Twenty one patients whose sex was not recorded have been excluded from the chart.



Total patient counts	Female	3586
	Male	1931
	Unknown	21
	Total	5538

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. 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 2980 patients received disease targeted drug therapy at some time during the audit period. 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.

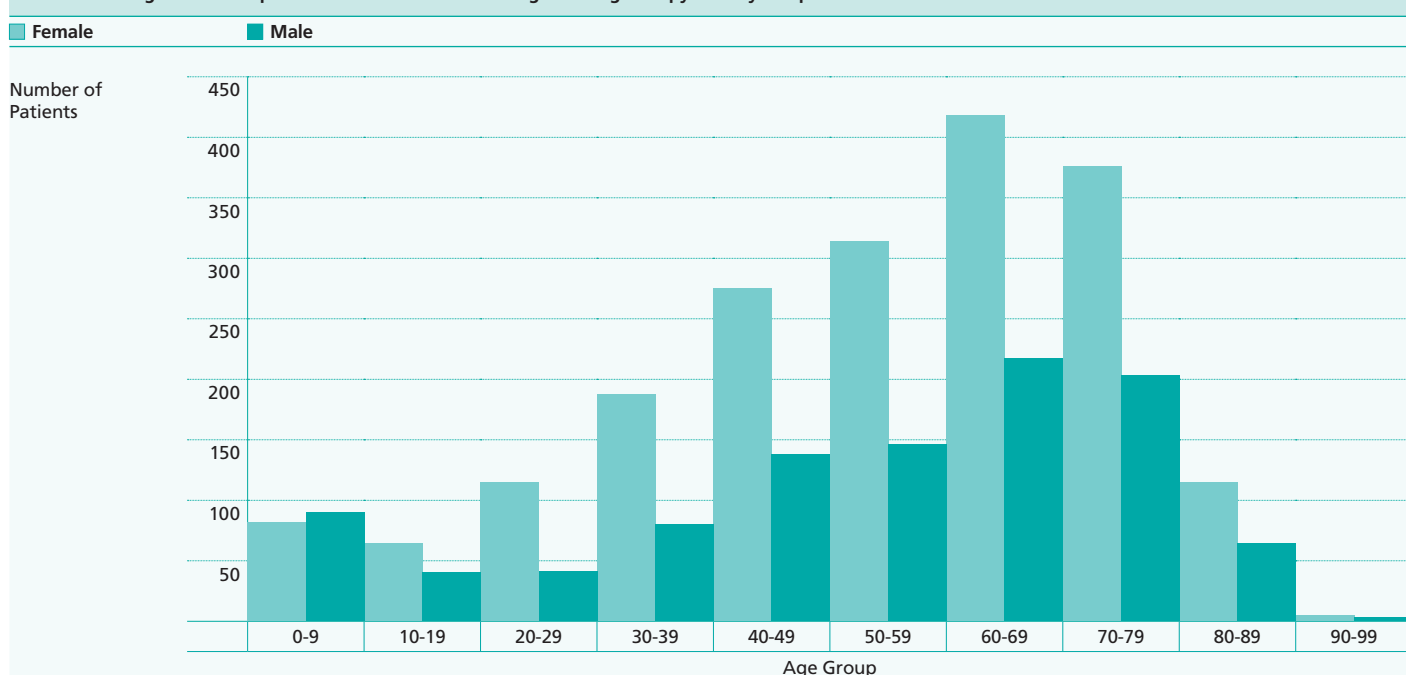
Figure 3 displays the number of patients on disease-targeted therapy, by age band and sex. The number patients who received treatment with disease-targeted drug therapies peaked in the seventh decade.

Table 5
Age of patients treated with disease-targeted therapy by country.

Region	Patient Count	Mean Age (years)	Median Age (years)
UK (all patients)	2980	56	59
England	2527	57	59
Northern Ireland*	29	45	33
Scotland	252	55	60
Wales	151	54	59
Other/Unknown	21	31	32

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

Figure 3
Distribution of age and sex of patients treated with disease-targeted drug therapy. Twenty one patients whose sex was not recorded have been excluded from the chart.



Disease-Targeted Therapy Census Data

Census data was collected on 31 March 2010. The census collects data on patients who are alive and under 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.

Table 6 shows monotherapy data and Table 7 shows combination therapy data. A total of 2408 patients were receiving therapy on 31 March 2010. Finally, in this section, Table 8 lists the number of prescriptions for the individual drugs themselves. Note that few patients receive calcium channel blockers for vasoreactive pulmonary hypertension indicating how uncommon this condition is in clinical practice.

Table 6
Number of patients on monotherapy on 31 March 2010.

Type of Drug	Total
Endothelin receptor antagonist	775
Phosphodiesterase 5 inhibitor	1038
Prostanoid therapy	111
Calcium Channel Blockers for Vasoreactive PAH	29
Unknown	32
Total	1985

Unknown indicates clinical trial medication or the drug therapy type was unknown.

Table 7
Number of patients on combination therapy on 31st March 2010.

Combination Therapy	Total
Endothelin receptor antagonist & phosphodiesterase 5 inhibitor	282
Endothelin receptor antagonist & phosphodiesterase 5 inhibitor & prostanoid therapy	25
Endothelin receptor antagonist & phosphodiesterase 5 inhibitor & calcium channel blocker for vasoreactive PAH	2
Endothelin receptor antagonist & prostanoid therapy	26
Phosphodiesterase 5 inhibitor & prostanoid therapy	65
Phosphodiesterase 5 inhibitor & prostanoid therapy & calcium channel blockers for vasoreactive PAH	1
Unknown combination	22
Grand Total	423

Key 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.

Census data describing the number of patients prescribed disease-targeted therapy compared to previous years is shown in Figure 4. The number of patients seen is also included in this histogram for comparison and is the same data as shown in Figure 1.

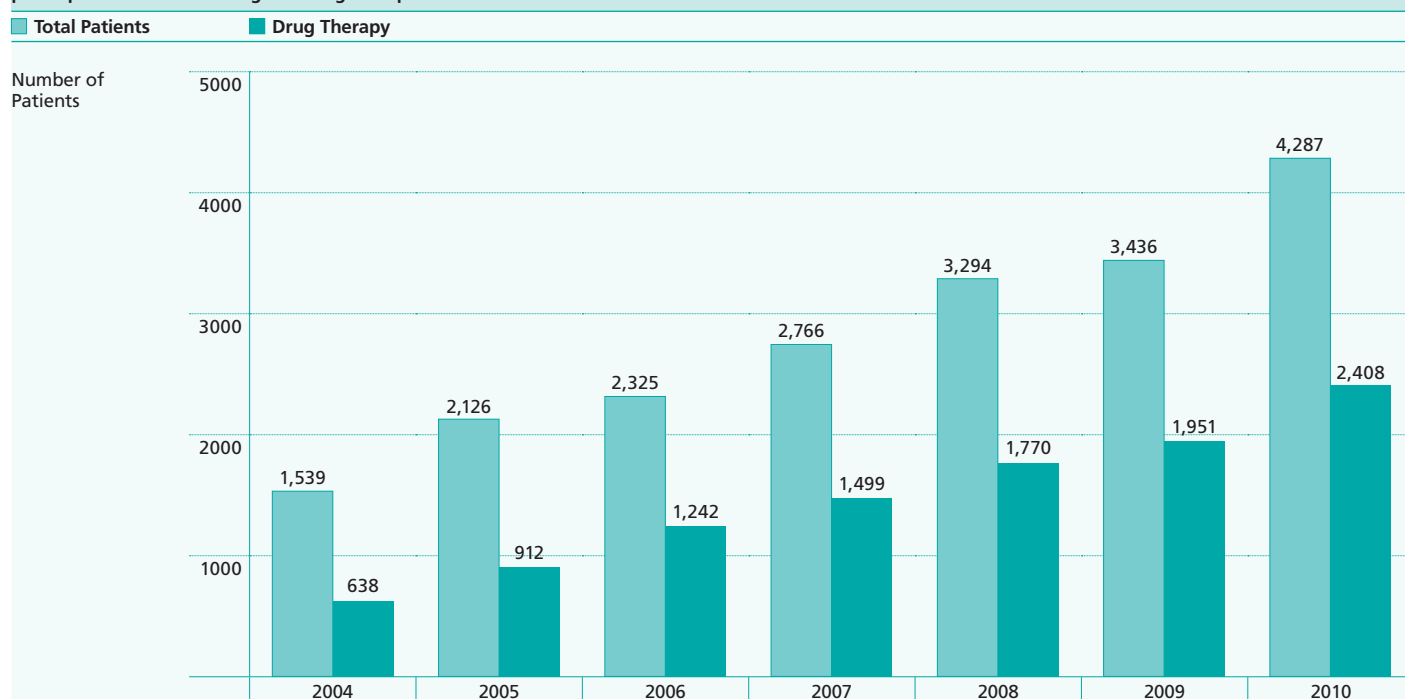
Table 8
Number of drug prescriptions on 31 March 2010 including monotherapy and combination therapy. Drugs administered by different routes appear more than once in the table.

Drug	Total Prescriptions
Sildenafil	1414
Bosentan	909
Ambrisentan	120
Sitaxsentan	88
Iloprost iv	49
Iloprost nebulised	47
Treprostinil sc	44
Treprostinil iv	42
Epoprostenol iv	39
Amlodipine	12
Nifedipine	8
Tadalafil	5
Treprostinil nebulised	2
Diltiazem	1
Nicardipine	1
Unknown	91
Total	2872

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.

Figure 4
Number of patients active under the service and number of patients on disease-targeted therapies for pulmonary hypertension on 31 March. In 2009 no data was available from the Royal Brompton Hospital which explains the stalling of the upward trend in numbers in this year. Drug therapy refers to 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 60.

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

- Patients who underwent pulmonary endarterectomy at Papworth Hospital and were under the care of another designated centre were sometimes double counted
- 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 showing number and percentage of patients per clinical outcome per annum. Percentages have been rounded to the nearest whole number.

Clinical Outcome	Total
Alive & Not Discharged	4327 78%
Died	560 10%
Discharged	651 12%
Transplanted	10 0%
Total	5538 100%

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

	Number of catheters	Number of patients	Percentage of patients
England	1451	1317	87
Northern Ireland*	8	6	0
Scotland	126	122	8
Wales	62	58	4
Other	4	4	0
Unknown	8	8	1
UK Total	1659	1515	100

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

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.

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.

	Number of procedures	Number of patients	Percentage of patients
England	3430	2794	90
Northern Ireland*	42	26	1
Scotland	165	146	5
Wales	161	134	4
Other	5	5	0
Unknown	20	16	0
Grand Total	3823	3121	100

* Very limited data was obtainable for Northern Ireland. Other includes the Channel Islands 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.

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

The number of six minute walk or shuttle tests performed per patient was 1.5 and cardiopulmonary exercise tests was 1.1.

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

Exercise Type	Country	Number of procedures	Number of patients	Percentage of Patients
Six-minute walk test or Shuttle test	England	5489	3638	83
	Northern Ireland*	54	32	1
	Scotland	539	306	7
	Wales	209	153	3
	Other	10	8	0
	Unknown	17	12	0
Six-minute walk test or Shuttle test Total		6318	4149	95
Cardiopulmonary	England	193	181	4
	Scotland	1	1	0
	Wales	19	15	0
	Unknown	1	1	0
Cardiopulmonary Total		214	198	4
Type of exercise test not recorded	England	28	28	1
	Wales	2	2	0
Unknown Total		30	30	1
UK Total		6562	4377	100

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

Diagnostic Classification of Pulmonary Hypertension

The diagnosis of pulmonary hypertension is made according to the Dana Point clinical classification (Appendix 1). This section shows completeness of diagnosis fields in the database and then describes the diagnosis in patients in whom the diagnosis has been recorded.

The number of patients in whom a diagnosis had not been recorded in the patient's record at the time of the audit was 1077 (19 per cent) of 5538. It is expected that patients who were in the process of being seen for the first time and being investigated would not have had a diagnosis. The number of missing diagnoses is a limitation of the current audit.

The diagnosis of many patients had to be re-entered into the database when the audit commenced in 2009. Individual hospital databases had recorded diagnosis until then according to the Venice clinical classification. Although the new Dana Point clinical classification used in the NAPH has some minor changes compared to Venice, some of these do not map easily to the previous Venice classification. These have required manual input. Checking and updating diagnoses was work-in-progress in all hospitals at the time of the audit.

Figure 5 shows the diagnostic classification of patients with pulmonary hypertension. It obviously excludes the 19 per cent of patients without a 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 haemangiomas are rare.

Figure 6 provides a graphical representation of the distribution of patients by age and sex according to the largest diagnostic categories (see Appendix 1 for more details). This allows the reader to see the different age and sex distributions of these diagnoses. Pulmonary arterial hypertension affected a younger population than chronic thromboembolic pulmonary hypertension and left heart disease. The sexes were equally affected in chronic thromboembolic disease while females predominated in pulmonary arterial hypertension and left heart disease.

Figure 5
Dana Point diagnostic classification of all patients showing the proportion in each diagnostic group. (See Appendix 1 for details of the classification). Percentages have been rounded to the nearest whole number.

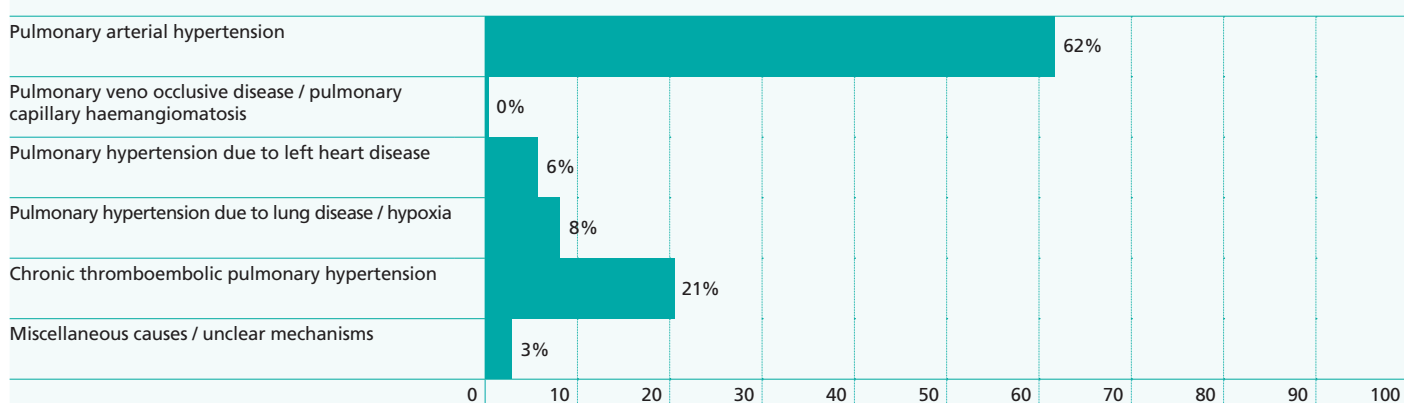


Figure 6

The age and sex distribution of patients according to the Dana Point clinical classification diagnosis. Six patients with missing sex were omitted from pulmonary arterial hypertension, 3 from chronic thromboembolic pulmonary hypertension, and 1 from left heart disease. Note that the vertical axis scales are different in each graph since the purpose of the figure is to compare age and sex distributions.



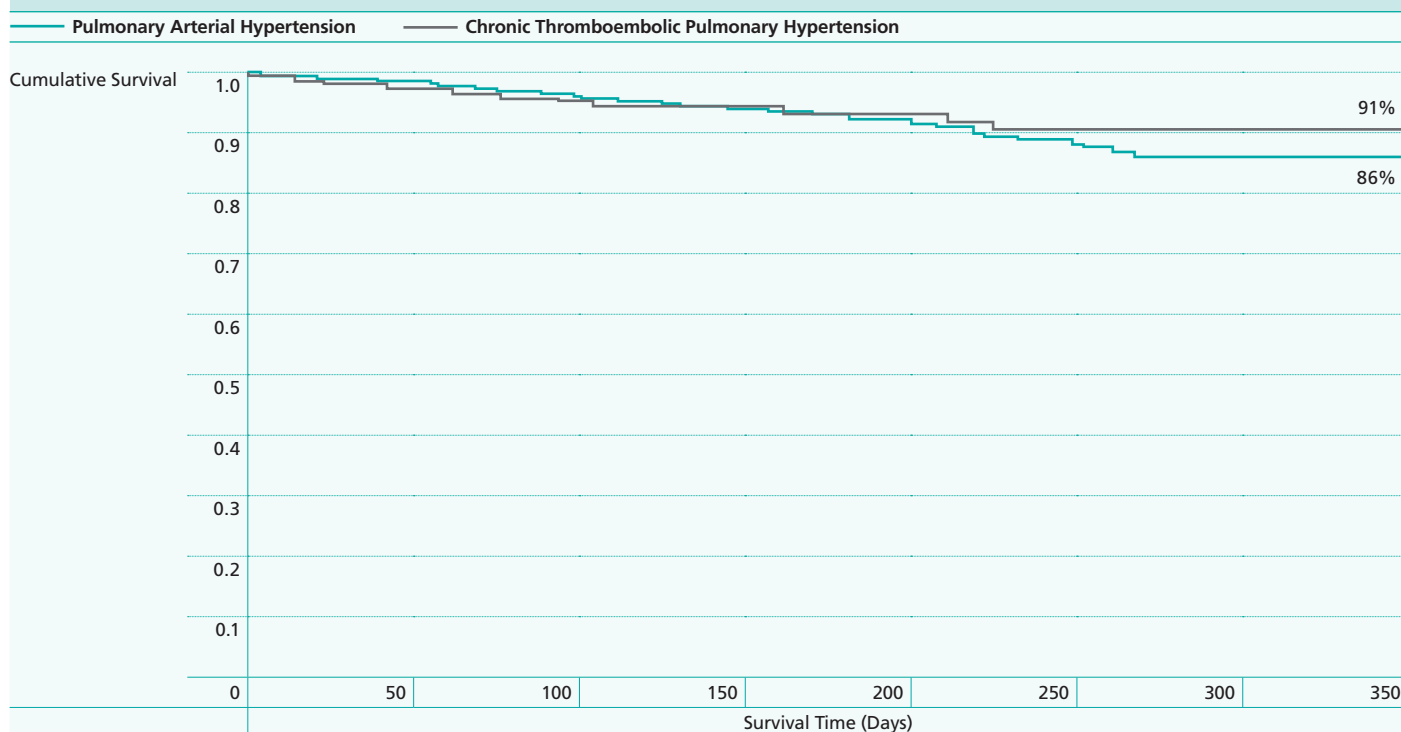
Example of Survival Outcome Analysis

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 and 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 and hence correct less satisfactory outcomes should they arise.

In its first year, a survival curve of patients who were diagnosed on or after 1 April 2009 has very limited value but it is shown to demonstrate the valuable information that will become available in future years. Figure 7 shows data for patients with a new diagnosis of pulmonary arterial hypertension and chronic thromboembolic disease. Since the follow-up times (shown below) are only 167 – 181 days the one year mortality is unreliable. There were too few patients to draw separate survival curves for subgroups of these diagnoses on this occasion, but this will become possible in future years as the cohort size expands and follow-up times become much longer.

Figure 7

Kaplan Meier survival curve of patients newly diagnosed with pulmonary arterial hypertension or chronic thromboembolic pulmonary hypertension. Note that pulmonary arterial hypertension includes all causes of pulmonary arterial hypertension as listed in Appendix 1 and not only idiopathic.



	Patients with survival time > 0 days	Mean follow-up (days)	Median follow-up (days)
Pulmonary Arterial Hypertension	459	181	181
Chronic Thromboembolic Pulmonary Hypertension	172	175	167.5

Key Findings

1. This report describes the first audit of its kind worldwide. The data provides a responsible overview of clinical activity and prescribing for patients with pulmonary hypertension in the UK, Channel Islands and Isle of Man.
2. All designated centres for pulmonary hypertension in the UK agreed to participate and submitted data for the year 1 April 2009 to 31 March 2010.
3. In the first year of this audit the data in this report should be considered as indicative of the national picture subject to the limitations of the quality of the data at the time of the audit. The accuracy of the data cannot be verified without an audit of source data and has not been undertaken.
4. During the audit UK pulmonary hypertension services saw 5478 patients. Since 60 patients were seen in more than one centre during the year, the workload of the centres is represented by 5538 patients. The mean age was 56 years and the median age was 60 years. There were 1.9 females to every male.
5. Of the total number of patients, 12 per cent were discharged and 10 per cent died during the one year audit. Ten patients were transplanted.
6. Investigations at designated centres totalled 1659 cardiac catheterization procedures, 3823 echocardiograms and 6562 exercise tests.
7. Disease targeted drug therapy was prescribed for 2980 patients, mean age 56 years and median age 59 years. The ratio of females to males was 1.9:1.
8. The most commonly prescribed drugs were phosphodiesterase 5 inhibitors followed by endothelin receptor antagonists.

Recommendations

1. The problem of lack of completeness in the database of key data items required for commissioning reports needs to be addressed before the next report.
2. To ensure high quality data entry at designated centres dedicated staff is required. It remains the responsibility of trusts to ensure that appropriate skills and the necessary resources are in place.
3. Data quality should be verified by an annual site audit at each designated centre to ascertain the accuracy of database entries compared to source records.
4. A regular review of database system processes is required to prevent recurrence of the many technical bugs and issues which have been identified with the database.
5. The clinical findings in this report should continue to be reported annually. In addition, the next report should provide more details about new patients referred to the service, diagnosis and survival.
6. 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. 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.
2. 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.
3. 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. PH 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
First line	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
Second line/ Alternative*	WHO III: B,P or X WHO IV: B		WHO III/IV: S P or X		WHO III/IV: B
Combination	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
Alternate combination	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 haemangiomatosis

* 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
Lead Clinician	Paul Corris	Andrew Peacock	Ingram Schulze-Neick	Simon Gibbs	Joanna Pepke-Zaba	John Wort	Gerry Coghlan*	David Kiely
Supporting Physicians	James Lordan Andrew Fisher Guy MacGowan	Martin Johnson		Luke Howard	Nick Morrell Karen Sheares	Michael Gatzoulis Kostas Dimopoulos Phil Marino Gerhard Diller	Clive Handler* Benji Schrieber*	Charlie Elliot Robin Condliffe Ian Sabroe
Clinical Nurse Specialists	Margaret Day Rachel Crackett* Julia de Soya	Agnes Crozier Alison Curran	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	Iain Armstrong (nurse consultant) Paul Sephton Lisa Martin Jane Wilkinson
Data Managers	Paul McAlinden*	Simon Kerridge*	Barbara Margetts*	Lucia Heath*	Carmen Treacy*	Sweeta Dhakan*	Anthony Piwowarski	Sheila Forshaw
Pharmacist	Maria Allen	Allan Smith	Lynne Cochrane	Lynn Humphrey	Duncan Grady	Beejal Shah	Pang Jay	Neil Hamilton
Radiologists	Leslie Mitchell Michelle Muller	Michael Sproule	Cathy Owens Andrew Taylor	Mary Roddie Christoph Juli James Jackson	Nick Scream Deepa Gopalan	Michael Rubens Simon Padley	Charlotte Cash	Christine Davies Catherine Hill
Service Manager	Teresa Glennie			Jo Nicholson	Alison Gibson	Kelly Goulding	Olivia Ellah	Lisa Needham
IT Staff					Mike Moore Jim Butler	Colin Gordon George Lampadariou		Stephen Stewart Theresa Dodd Jon Wrend
Administrative Staff	Kim Clay	Veronica Ferry Lorraine James	Barbara Margetts*	Jermaine Charles Claire Gin-Sing Byron Wong	Karin Johnson Tanya Vonseld Jackie Richmond Lesley Humphrys	Lilly Morgans Sylvia Dedman	Innes Hammel* Alison Narcisse*	Denise Stephenson Natalie Johnson

Appendix 4: Members of the NAPH Project Board

Name	Role	Organisation
Brendan Barrett (retired 2010)	CCAD Developer	The NHS Information Centre
Roger Boyle	Policy Lead	Department of Health
Peter Bushell	CCAD Developer	The NHS Information Centre
Geoffrey Carroll	Senior User (Commissioners)	Medical Director, Health Commission, Wales
David Cunningham	Senior Supplier	The NHS Information Centre
Natalie Doughty	Senior User (Nursing)	Papworth NHS Foundation Trusts
Cathy Edwards	Senior User (Commissioners)	Director Yorkshire & Humber SCG, England
Nadeem Fazal	Service Manager	The NHS Information Centre
Stewart Fleming (retired 2009)	CCAD Developer	The NHS Information Centre
Simon Gibbs (Chair)	Clinical Lead & Chief Auditor (Project executive)	Imperial College London & Imperial College Healthcare NHS Trust
Martin Johnson	Senior User (Physician)	NHS Greater Glasgow & Clyde
Lesley King	Senior User (Commissioners)	The Eastern Health and Social Services Board (Northern Ireland)
Mark Lee (retired 2010)	Project Manager	The NHS Information Centre
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
Joanna Pepke-Zaba (retired 2009)	Senior User (Physician)	Chair of Physicians Committee until 2009
Thomas Pharaoh (retired 2009)	Senior User (Manager)	Imperial College Healthcare NHS Trust
Ingram Schulze-Neick	Senior User (Physician)	Paediatric Pulmonary Hypertension Service
Tracy Whittaker (retired 2010)	Project Manager	The NHS Information Centre
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

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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.

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