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ORIGINAL ARTICLE

Management of Adults With Congenital Heart Disease and Pulmonary Arterial Hypertension in the UK: Survey of Current Practice, Unmet Needs and Expert Commentary

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Background Pulmonary arterial hypertension (PAH) is a well-recognised complication of adult congenital heart disease (CHD). However, management is not currently standardised between centres and specific guidelines are lacking. In order to identify and understand the unmet needs related to PAH associated with CHD (PAH-CHD), a survey of physicians was performed.

Methods An electronic survey was sent to two physician groups: (1) cardiologists registered in a UK cardiology directory; (2) specialist pulmonary hypertension (PH) physicians known to manage patients with adult PAH-CHD. The questions related to referral pathways, screening, therapy and palliative care.

Results 821 surveys were distributed and 106 were returned. Respondents included a broad mix of specialist physicians with many patients along with general cardiologists managing only a small number of PAH-CHD patients. Although 97% of respondents have access to a specialist PH centre, patients are still being managed in non-specialist settings. Shared care arrangements are widespread but only 41% have formal shared care protocols. Palliative care services are limited and general cardiologists rarely perform 6-minute walk tests (6MWT) or quality of life assessments. People with PAH-CHD are often undertreated, with 39% of respondents reporting that fewer than 25% of these patients were receiving PAH-specific therapies.

Conclusions The survey revealed gaps and inconsistencies in the management of patients with PAH-CHD therefore patient-specific guidance is needed for many of these aspects.

Keywords Pulmonary arterial hypertension • Congenital heart disease • Survey • Unmet needs

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Introduction

Pulmonary arterial hypertension (PAH) is defined as a mean pulmonary arterial pressure of ≥25 mmHg at rest, with a pulmonary artery wedge pressure or left atrial pressure ≤15 mmHg and pulmonary vascular resistance (PVR) >3 Wood units [1–3]. Congenital heart disease (CHD) that causes unrestricted pressure and volume overload of the pulmonary circulation can lead to PAH [4]. Recent paediatric medical and surgical advances have enabled people with CHD to live longer, leading to an increase in the number of patients who go on to develop PAH associated with CHD (PAH-CHD) [3]. The 2015 UK audit of specialised PH centres reported a rate of 17 people with PAH due to CHD per million of the population [5]. Five to ten per cent of adults with CHD also have PAH [1,2].

The European Society of Cardiology/European Respiratory Society (ESC/ERS) guidelines provide recommendations for the management of PAH-CHD [1,2]. However, evidence to support the use of PAH-specific therapies in CHD is limited and the management is not standardised, with variation between treating centres and clinicians.

The Congenital Heart disease And pulMonary arterial hyPertension: Improving Outcomes through education and research Networks (CHAMPION) program was set up to improve the care of patients with PAH-CHD in the UK and to help inform clinician decision-making. Objectives include identification of evidence gaps, unmet needs, challenges associated with the management, and development of educational resources and initiatives to ensure sharing of best practice. Within this remit, a survey was conducted to gain an understanding of the challenges faced by physicians in the day-to-day management of PAH-CHD in the UK. This publication covers various aspects of patient management, presenting the survey results and providing expert commentary on each aspect.

Methods

Respondents and Inclusion/Exclusion Criteria

The survey targeted both general cardiologists and experts in PAH-CHD and therefore included two groups of clinicians: those proposed by the CHAMPION Steering Committee as currently managing these patients, and all adult cardiologists registered with the UK Directory of Cardiology (http://cardiodirectory.co.uk) with an email address. This second source was used to ensure that physicians who manage PAH-CHD outside specialist centres were also identified and included. At the start of the survey, consenting respondents were asked to confirm whether they worked in a general cardiology department, specialist adult CHD centre or specialist PH centre, and that they managed PAH-CHD. Other respondents were excluded from the survey. An introductory email, followed by two reminder emails, was sent to all participants together with a hyperlink to the survey. All responses were anonymised, but some demographic information was requested and analysed.

Survey

The survey was electronically administered using a professional survey website (SurveyMonkey, Palo Alto, CA, USA) (see Supplementary Figure S1 for survey questions). Questions were either multiple choice or answered by five-point Likert scales or free text comment boxes. Prior to the survey, a pragmatic literature review of PAH-CHD was performed and used by the CHAMPION Steering Committee to identify unmet needs related to their care. As part of the survey, respondents were asked to rank these unmet needs in order of importance. For each unmet need, weighted averages were calculated by assigning five points for each first place

Table 1 Respondent characteristics.

Characteristic[a]	n/N	%
Geographic region of practice		
North	5/32	16
Midlands and East	6/32	19
South, excluding London	12/32	38
London	3/32	9
Scotland	4/32	13
Northern Ireland	0/32	0
Wales	2/32	6
Speciality		
General physician	0/32	0
Rheumatologist	0/32	0
Respiratory physician	2/32	6
General cardiologist	18/32	56
ACHD cardiologist	12/32	38
Paediatric cardiologist	0/32	0
Years in practice		
<5 years	0/32	0
5 to 15 years	4/32	13
>15 years	28/32	88
Years in practice as a consultant		
<5 years	8/31	25
5 to 15 years	13/31	41
>15 years	10/31	31
Approximate number of PAH-CHD patients seen annually[b]		
<10	23/55	42
10 to 49	14/55	26
50 to 100	13/55	24
>100	5/55	9

n/N: Number of respondents with each characteristic/Total number of respondents who completed the question.

Abbreviations: ACHD, adult congenital heart disease; PAH-CHD, pulmonary arterial hypertension associated with congenital heart disease.

[a]Not all respondents provided demographic information.

[b]Respondents were asked to specify for the year 2015.

Table 2 **Unmet needs identified by the Steering Committee: Ranking by survey respondents.**

Unmet needs and rank (n = 30)	Rank[a]	%[b]	Weighted average[c]
Development of a standardised approach to the care of PAH-CHD patients across centres	1	60	3.3
Establishment of a screening protocol for PAH in CHD patients	2	53	3.7
Definition of treatment targets for different PAH-CHD subgroups	3	53	2.6
National guidance on when to initiate and escalate PAH-specific therapies in PAH-CHD patients	4	50	3.3
Guidance on risk stratification for PAH-CHD patients	5	47	3.4
Guidance on how to manage non-Eisenmenger's syndrome patients	6	40	3.1
Guidance on non-pharmacological therapy for PAH-CHD patients	7	40	2.8
Establishment of a registry to determine the size and characteristics of the PAH-CHD population	8	33	3.1
Palliative care strategy	9	33	2.8
Identification of biomarkers and other non-invasive strategies for screening CHD patients for PAH	10	30	2.4
Management of Down's syndrome patients	11	27	2.4
Creation of a biobank of CHD samples for genomic analysis	12	20	2.0

n: Number of respondents who completed the question.

Abbreviations: PAH-CHD, pulmonary arterial hypertension associated with congenital heart disease; PAH, pulmonary arterial hypertension; PH, pulmonary hypertension; CHD, congenital heart disease.

[a]Ranking was assigned in descending order according to % of respondents who indicated the unmet need within their top 5, followed by weighted average in cases where % was identical.

[b]% of respondents who indicated that this was one of their top 5 unmet needs.

[c]5 points were assigned for each 1st place ranking, 4 points for each 2nd place ranking, 3 points for each 3rd place ranking, 2 points for each 4th place ranking and 1 point for each 5th place ranking. The average was calculated by dividing by the total number of points assigned to each unmet need by the total number of respondents who provided a ranking for that need.

ranking, four points for each second place, etc. The total was divided by the number of respondents in order to determine an average ranking for each unmet need. Categorical values were presented as numbers (percentage).

Results and Expert Commentary

Respondent Characteristics

A total of 821 physicians were invited to participate in the survey and 106 responded. Demographic information is shown in Table 1. The proportion of respondents managing more than 50 people with PAH-CHD per year was 33%, hence many worked in a specialised setting. However, 56% of respondents described themselves as general cardiologists and 42% of respondents managed less than 10 people with PAH and CHD annually. Four of the respondents were from a PAH specialist centre. We have shown that 47% of respondents were from the South and London, where the majority of specialist PH centres are located. Eighteen of 55 (33%) of respondents saw >50% PAH-CHD patients annually, of these 5/18 (28%) stated that they worked in a general cardiology department (so therefore were NOT either PH specialists or ACHD specialists).

Unmet Needs in the Management of Pulmonary Arterial Hypertension Associated with CHD

The unmet needs in the management of people with PAH-CHD were previously identified by the Steering Committee. When asked to rank these, respondents were in broad agreement (Table 2). The five unmet needs considered to be most important by the Steering Committee are discussed below.

Standardised Approach to Care

Seven adult and one paediatric centre were designated in 2001 to provide specialist PH services in the UK (see Supplementary Table S1). Each centre has to meet standards including the use of specialist nurses and expert consultants providing 24-hour care, prescription and monitoring of PAH therapies in addition to collection of data for the annual UK National PH audit. Furthermore, the National Health Service, England recently conducted a review of CHD services in order to standardise patient care [6]. Current UK adult CHD services comprise regional networks of specialist adult CHD (including surgical) and local adult CHD centres. The proportion of respondents who referred people with PAH-CHD to a specialist PH centre was 97% (57% of whom were general cardiologists and respiratory physicians) (see Table 3). The majority of respondents (81%) share the management of patients with a specialist PH centre, with 56% of respondents reporting having a shared care clinic within their own unit. Shared care is driven by physicians with an interest in PH, enabling the diagnosis, prescribing and monitoring of people with PAH-CHD to be provided at the patient's adult CHD centre. Formal shared care protocols were in place in only 41% of shared care clinics (see Table 3), revealing a lack of adherence to the recommendations outlined in the 2015 New CHD review, which states that all Level one and Level two adult CHD centres are required to have evidence of joint protocols of care with a national PH centre [6].

Table 3 Referral/care pathways for PAH-CHD patients.

Elements of care	Respondents who answered yes	
	n/N	%
Do your PAH-CHD patients attend the following types of clinic?[a]		
General cardiology clinic	15/44	34
Specialist ACHD clinic	27/44	61
Specialist PAH-CHD clinic	23/44	52
General PH clinic	6/44	14
In your centre do you refer patients to a specialist PH service?	35/36	97
Do you share management of patients with a specialist PH service?	29/36	81
Do you have a shared care clinic within your own unit?	18/32	56
Do you have a written, formal shared care protocol with a specialist PH centre?	13/32	41
Do your PAH-CHD patients have access to the following specialist staff?		
Nominated PAH clinician	27/44	61
PAH specialist nurse[b]	14/42	33
PAH specialist pharmacist	8/42	19

n/N: Number of respondents who answered yes/Total number of respondents who completed the question.
Abbreviations: PAH-CHD, pulmonary arterial hypertension associated with congenital heart disease; PAH, pulmonary arterial hypertension; PH, pulmonary hypertension; ACHD, adult congenital heart disease.
[a]Respondents could specify more than one type of clinic.
[b]Included only responses that stated ≥ 1 full time equivalent nurse.

The proportion of respondents that ranked the need for a standardised approach to the care of PAH-CHD as one of their top five unmet needs was 60% (Table 2) and highlighted the need for clearer referral pathways and easier access to specialist PH centres (see Supplementary material survey data slides). Furthermore, although 61% of respondents reported that patients had a nominated PH physician, specialist PH nurses and specialist pharmacists were reported by only 33% and 19% of respondents respectively as being available, indicating that certain aspects of specialist care are lacking (Table 3). Current ESC/ERS guidelines recommend that specialist PH centres should follow at least 50 patients on PAH treatment with at least two new referrals per month [1,2]. They should be able to offer full diagnostic services including computerised tomography, magnetic resonance imaging and right heart catheterisation and should have specialist nurse support [1,2]. Furthermore, they should be able to offer all PAH therapies available in that country and should also enrol patients in clinical trials [1,2]. Although specialist adult CHD centres may be able to fulfil many of these requirements, they may not be able to offer more complex therapies such as therapies targeting the prostacyclin pathway and, therefore, a clear referral link with a designated PH centre is vital.

Table 4 Screening for PAH in CHD patients.

Aspects of Screening	Respondents who answered yes	
	n/N	%
Do you consider PH when evaluating each of the following groups of patients?[a]		
ASD	34/36	94
VSD	34/36	94
Systemic to pulmonary shunts	35/36	97
Operated left to right shunts	30/36	83
Complex defects	33/36	92
Other	3/36	8
When do you screen CHD patients for PAH?		
First evaluation only	3/36	8
Every time I review them	22/36	61
Periodically	11/36	31

n/N: Number of respondents who selected this option/Total number of respondents who completed the question.
Abbreviations: ASD: atrial septal defect; CHD: congenital heart disease; PAH: pulmonary arterial hypertension; VSD: ventricular septal defect.
[a]Respondents could specify more than one group of patients.

Table 5 Therapy recommendations for PAH-CHD patients.

PAH-CHD classification	Recommended treatments	ESC/ERS recommendation (class/level of evidence) [1,2]	Evidence to date
Eisenmenger's syndrome	Bosentan [1,2]	IB	BREATHE 5 [21] + BREATHE 5 OLE [22]
Eisenmenger's syndrome	Other ERAs, PDE5is and prostanoids should be considered [1,2]	IIaC	Data from retrospective, observational studies and small RCTs [23–27]
PAH associated with prevalent systemic to pulmonary shunts	Selected cases may benefit from closure. As data are lacking, no recommendations can be made for or against the use of PAH-specific therapies [4]	NA	Data from observational study [23]
PAH with small/coincidental defects[a]	Avoid defect closure, treat with PAH-specific therapies [4]	NA	Data from RCTs and observational study [23,28]
PAH after defect correction	Do not close residual defects. Treat with PAH-specific therapies [4]	NA	Data from RCTs with PAH-CHD subgroup [29–34]. Data from observational study [23]

NA: No specific recommendations are provided in the ESC/ERS Guidelines.
Abbreviations: ERA, endothelin receptor antagonist; ERS, European Respiratory Society; ERS, European Society for Cardiology; OLE, open label extension; PAH, pulmonary arterial hypertension; PAH-CHD, pulmonary arterial hypertension associated with congenital heart disease; PDE5i, phosphodiesterase type 5 inhibitor; RCT, randomised controlled trial; BREATHE 5, The Bosentan Randomized Trial of Endothelin Antagonist Therapy-5.
[a]Many patients with small defects are considered to be idiopathic PAH patients with a coexistent defect and hence are eligible for treatment.

Screening

The importance of identifying CHD patients with a high PVR has long been accepted [7]. This is achieved by estimating pulmonary artery pressure on echocardiography and referring for cardiac catheterisation. It, therefore, comes as no surprise that physician awareness of the potential for pulmonary vascular disease in CHD is high, with more than 90% of respondents reporting that they screen all patients with atrial septal defects, ventricular septal defects, systemic-to-pulmonary shunts and complex defects and 83% screen patients with repaired left-to-right shunts for late development of PH (Table 4). Furthermore, 92% of respondents screen patients either periodically or at every clinic visit, with just 8% only screening for PAH at the initial clinic visit (Table 4). These data confirm that clinicians managing CHD routinely assess pulmonary haemodynamics; such screening should remain part of the long-term management of these patients.

Initiation and Escalation of Therapy/Risk Stratification

Table 5 summarises therapy recommendations for PAH-CHD patients. The survey revealed that many people with PAH-CHD are not receiving treatment, with almost 40% of respondents reporting a low usage of PAH-specific therapies (defined as less than 25% receiving a PAH-specific therapy) (Table 6). The proportion of respondents reporting low usage of PAH-specific therapies was greater among those working in general cardiology departments than among those working in specialist adult CHD centres (49% versus 18%, respectively) (Figure 1). These data suggest that patients may benefit from further assessment in a specialist setting. Over half (56%) of respondents with a high case load of Eisenmenger's syndrome (defined as more than 50% of their PAH patients) reported high or very high usage of PAH-specific therapies (defined as 50–74% or 75–100%), as compared with 30% (17/56) for all respondents combined. Conversely, among respondents with prevalent systemic-to-pulmonary shunts accounting for more than 50% of all people with PAH-CHD, only 11% (1/9) reported high or very high usage of PAH-specific therapies. This reflects differences in the guidance for PAH-specific therapies for each of the CHD subtypes.

To further explore how therapy is initiated and managed in people with PAH-CHD, the survey respondents were asked whether they initiate and/or manage therapy, or whether they refer all patients who need PAH-specific therapy to a PH specialist. Over half (54%) of respondents stated that they initiate and/or manage specific therapies (Table 6). Of these, 35% reported both initiating and managing specific therapies, while the remainder only manage therapies after initiation by another clinician. The survey indicates that therapy is more likely to be initiated by physicians working in specialist centres than in general cardiology departments; 65% of respondents from specialist adult CHD centres both initiate and manage PAH-specific therapies, whereas this was only true for 15% of respondents working in general cardiology departments, possibly leading to under-treatment.

Table 6 Management of PAH-specific therapy in PAH-CHD patients.

Aspects of PAH-specific therapy management	Respondents	
	n/N	%
Approximately what percentage of your PAH-CHD patients are currently receiving a PAH-specific therapy?		
0 to 24%	22/56	39
25 to 49%	17/56	30
50 to 74%	14/56	25
75 to 100%	3/56	5
Do you initiate and manage PAH-specific therapies for patients with PAH-CHD?		
Yes, both initiate and manage	22/63	35
Yes, manage but not initiate	12/63	19
No, I refer all patients who need PAH specific therapies to a PH specialist	29/63	46
Which therapies do you initiate/manage?[a]		
Sildenafil	30/30	100
Bosentan	25/30	83
Ambrisentan	20/30	67
Macitentan	16/30	53
Tadalafil	15/30	50
Riociguat	7/30	23
Iloprost i.v.	7/30	23
Epoprostenol i.v.	6/30	20
Iloprost inhaled	5/30	17
Treprostinil s.c.	1/30	3
Treprostinil i.v.	1/30	3
Vardenafil	0/30	0
Do you perform walk tests for your PAH-CHD patients?		
Yes	25/41	61
No	16/41	39
Do you measure quality of life for your PAH-CHD patients?		
Yes	19/40	50
No	20/40	50

n/N: Number of respondents who selected this option/Total number of respondents who completed the question.

Abbreviations: i.v., intravenous; PAH-CHD, pulmonary arterial hypertension associated with congenital heart disease; PH, pulmonary hypertension; s.c., subcutaneous.

[a]Respondents could specify more than one therapy.

Bosentan is widely used in PAH-CHD patients (83% of respondents who initiate/manage therapy) (see Table 6). This frequent use is likely due to the fact that bosentan is the only therapy with a class I recommendation in Eisenmenger's syndrome patients in the ESC/ERS guidelines [1,2]. A current trial is being undertaken with macitentan (MAESTRO, Clinical study to assess the efficacy, safety, and tolerability of macitentan in subjects with Eisenmenger Syndrome; ClinicalTrials.gov Identifier: NCT01743001) and should report in 2017. All respondents reported managing patients using phosphodiesterase type −5 inhibitors, which is likely the result of the wide availability of sildenafil, low cost and clear commissioning guidance supporting sildenafil as first-line treatment for all PAH patients in the UK; 67% and 53% of respondents reported that they also managed patients with the endothelin receptor antagonists ambrisentan and macitentan, respectively. In total, 43% of respondents reported that they managed their patients using prostanoids (Table 6). A greater use of prostanoids was reported among specialist PH and adult CHD centres (61%) versus general cardiology clinics (15%).

Once a patient with PAH has been initiated on therapy, the current ESC/ERS guidelines recommend regular assessment in specialist PH centres to collect prognostic information, including exercise testing, which allows stratification of patients in a low (estimated 1-year mortality <5%), intermediate (estimated 1-year mortality 5 to 10%) or high risk (estimated 1-year mortality >10%) group. If patients do not meet their treatment goals, and their response to therapy is deemed inadequate, the treatment regimen should be escalated [1,2]. In the current study, 39% of respondents (the majority working in general cardiology units) reported

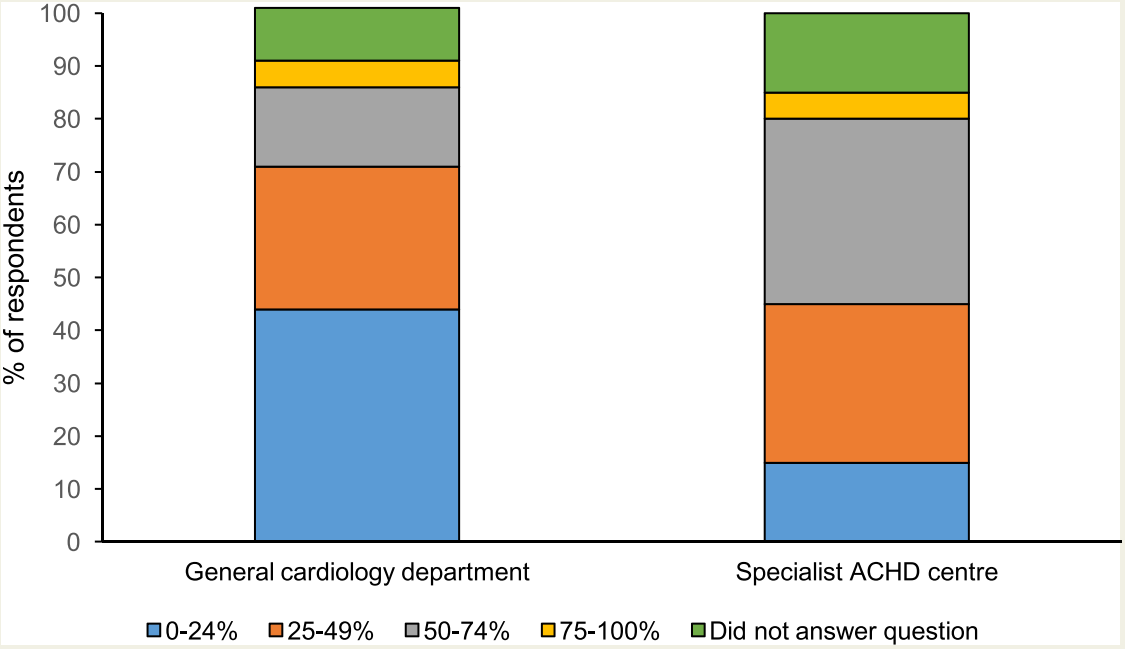

Figure 1 Proportion of PAH-CHD patients treated with PAH-specific therapies, classified according to the type of practice. Abbreviations: ACHD, adult congenital heart disease; PAH-CHD, pulmonary arterial hypertension associated with congenital heart disease.

that they do not perform walk tests, while these are routinely performed in specialist PH centres (Table 6). In this survey, 50% of respondents also stated that they do not perform quality of life assessments (90% of these worked in general cardiology units). Quality of life is an important treatment goal in PAH [1,2] and a decreasing quality of life has been shown to be a determinant of mortality in people with PAH-CHD [8], highlighting the need for regular assessment.

There were four responses from PH specialists. One was not managing PAH-CHD patients so did not proceed with the survey beyond q. 2 and another did not proceed beyond q. 4: Which therapies do you initiate/manage. Therapies initiated or managed were: Respondent 1: bosentan, macitentan, iloprost i.v., iloprost inhaled, treprostinil; respondent 2: bosentan, macitentan, ambrisentan, epoprostenol i.v., iloprost inhaled. Both respondents saw 50–100 patients annually and for respondent 1, 50–74% were on PAH specific therapy, and for respondent 2, 0–24% were. Both performed the 6MWT twice a year and both used the emPHasis-10 quality of life questionnaire.

In summary, the results of this survey indicate that many patients with PAH-CHD are not being treated with PAH-specific therapies. This may, in part, be because many patients are still managed in general cardiology clinics. The ESC/ERS guidelines recommend that PAH care should take place in specialist PH centres that routinely manage PAH-specific drug therapies and can prescribe all such therapies available in a particular country [1,2]. Patients who are managed in non-specialist centres may have less access to therapy, have lower awareness of available treatment options and do not routinely undergo exercise testing and quality of life assessment. Improving referral pathways and access to specialist centres, both raised as issues to be addressed by the survey respondents, may lead to an increase in the use of PAH-specific therapy for people with PAH-CHD. The lack of specific treatment recommendations for PAH-CHD subtypes other than Eisenmenger's syndrome needs to be addressed.

Non Pulmonary Arterial Hypertension-Specific Therapy

Non-pharmacological and supportive therapy are important components of the clinical management of PAH. This is reflected in the results of the current survey, with 40% of respondents stating that more information was needed on non-pharmacological management (Table 2). Furthermore, when asked to describe their top issues in management of PAH-CHD, respondents mentioned issues such as pregnancy and birth control, iron deficiency, exercise, anticoagulation, dental/skin care, arrhythmias and role of venesection.

There are currently no national protocols for the non-pharmacological management of PAH-CHD, however, recommendations are given in the ESC/ERS guidelines [1,2]. Pregnancy is associated with substantial mortality in PAH patients [9] and therefore should be avoided [1,2]. Most clinicians would recommend Mirena coil and progesterone-only preparations for contraception. As people with PAH are often iron-deficient, they should undergo regular monitoring of iron status and haemoglobin levels due to the potential impact of iron deficiency on exercise capacity and mortality [1,2]. If iron replacement therapy is required it may

need to be given intravenously due to poor oral absorption or intolerance [1,2,10]. When a patient is diagnosed, a degree of realistic expectation management is required for activities related to daily living. It is often necessary for patients to adapt their lifestyle to avoid strenuous exercise and sometimes to alter their working patterns in order to still have the maximum quality of life. The current ESC/ERS guidelines recommend that patients should undertake supervised physical activity within the limits of symptoms [1,2]. It is important for patients to avoid activity that causes syncope or severe breathlessness, but there is mounting evidence that 'keeping fit' is an important adjunct to pharmacological treatment [11,12]. Currently, it is not known which method of exercise rehabilitation should be used or the intensity and duration of exercise that should be applied [1,2]. No study has yet determined whether exercise-training programs improve prognosis. An exercise regime will therefore need to be tailored to the individual patient, with realistic expectations.

The most common arrhythmias seen in patients with PAH-CHD are atrial flutter (often relating to sites of previous incisions due to cardiac surgery) and ventricular tachycardia (resulting from postoperative chronic fibrosis, hypoxia or chronic volume loading) [13,14]. Both conditions require input from cardiac electrophysiologists and are poorly tolerated. The consensus view is that therapy involving high dose beta blockade should be avoided in patients with PAH, since it impairs their ability to generate an increased heart rate during exercise, impairs right ventricular function and can lead to symptoms of severe breathlessness [15]. Venesection is generally not recommended in PAH-CHD since it renders the patient iron deficient and may actually be associated with an increased risk of stroke. It should only be performed, with isovolumetric fluid replacement, when there are symptoms of hyperviscosity [1,2]. Unlike patients with idiopathic PAH, those with PAH-CHD are generally not anticoagulated, as they are prone to bleeding from dilated bronchial arteries and often have a low platelet count [1,2,16].

General management measures are essential to improving both PAH-CHD outcomes and quality of life [4]. The ESC/ERS guidelines offer clear guidance on non-pharmacological management of PAH, but at the current time no specific guidance is given for people with PAH-CHD and as not all general measures are applicable to this patient subgroup, specific guidelines are warranted.

Palliative Care

The current ESC/ERS guidelines recommend proactive advanced care planning with timely referral of PAH patients to palliative care services [1,2]. A large proportion of physicians (82%) surveyed report having no formal palliative care service for PAH-CHD, but 81% of these feel that a service of this nature would be valuable to management (Table 7). To address this unmet need, it is recommended that each centre should have a palliative care service available, providing good quality end of life care. Of the 18% of physicians who do have a formal and appropriate palliative care service, all would consider referring a patient with a life expectancy of <6 months and 83% would also consider referral following repeated hospital admissions and/or deterioration to functional class IV. None of the respondents refer patients to palliative care at the point of diagnosis. There is, therefore,

Table 7 Palliative care in PAH-CHD patients.

Aspects of palliative care	Respondents who answered yes	
	n/N	%
Do you currently have a formal palliative care service for PAH-CHD patients at your centre?		
Yes	6/33	18
No	27/33	82
When do you refer PAH-CHD patients to a palliative care service?[a]		
On diagnosis	0/6	0
After repeated hospitalisations	5/6	83
Upon deterioration to FC IV	5/6	83
Upon listing for transplantation	0/6	0
Life expectancy <6months	6/6	100
Other	3/6	50
Do you think a palliative care service would be useful or valuable for the management of PAH-CHD patients in your practice?[b]		
Yes	22/27	81
No	5/27	19

n/N: Number of respondents who selected this option/Total number of respondents who completed the question.

Abbreviations: FC: functional class; PAH-CHD: pulmonary arterial hypertension associated with congenital heart disease.

[a]Respondents could specify more than one time point and only respondents who reported having a formal palliative care service were able to answer this question.

[b]Only respondents who reported not having a formal palliative care service for PAH-CHD patients were able to answer this question.

a gap in the provision of palliative care services and a need for guidance on the timing of referral to palliative care services [17–19]. Earlier referral to palliative care should be made when considering escalation of targeted vasodilator therapy.

Study Limitations

There are several limitations to this study, which are related to the electronic nature of the survey. The survey was optional and therefore the physicians, who responded (13%), may have been more likely to be those who considered themselves to be knowledgeable in the field or who held strong opinions on the subject. There might have been potential selection bias, if the survey was too lengthy for busy clinicians. The demographic data, however, indicated that responses were obtained from a broad range of physicians including both PH specialists and general cardiologists. Furthermore, the low response rate was not unexpected as PAH is a rare disease (only 1000 people with PAH associated with CHD were seen by the UK PH service in 2014) [20].

The survey results could have been affected by recall bias. However, over a quarter of respondents (26.3%) stated that they were using a database to answer the survey questions. This is likely to increase the reliability of the results.

Conclusions

Our survey managed to capture both general cardiologists and physicians working in close cooperation with PH centres. It demonstrated that physicians who were seeing a large number of patients with Eisenmenger's syndrome were more likely to initiate and manage patients with advanced PH therapy, and they were more likely to follow guidelines. In addition, the survey revealed a gap in the knowledge base for many therapies for patients with PAH-CHD. There might be other confounders, such as a patient or carer wish not to start therapy, but we did not see a reason why there might be a geographical variation in this but accept that there might be unrecognised alternative possibilities as to why the standard guidelines were not followed. This might even be more prevalent in a greater geographical area outside of this study, with the effect of cost containment, or other restrictions being in place.

General cardiologists report the need for improved guidelines and increased access to specialist care for their patients with PAH-CHD. Efforts are needed to increase referral of such patients into PH services by developing referral criteria and to identify a best practice protocol for screening patients with CHD for the presence of PAH.

Appendix A. Supplementary data

Supplementary data associated with this article can be found, in the online version, at https://doi.org/10.1016/j.hlc.2017.10.018.

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