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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, California, 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

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.

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