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Adults' experiences of living with pulmonary hypertension: a thematic synthesis of qualitative studies

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ABSTRACT

Objectives Pulmonary hypertension is a life-shortening disease that has a considerable impact on quality of life. Improving our understanding of how individuals are affected and cope with the disease will help to improve services and outcomes. This review synthesises the published qualitative research that has listened to adults discuss their experiences of living with the disease.

Design A comprehensive systematic search of four databases was conducted in May 2020: Web of Science, PubMed, PsycINFO and Cochrane Library. Suitable studies were evaluated using the Critical Appraisal Skills programme. Findings from the studies were extracted and subjected to a thematic synthesis.

Results Nineteen articles were identified reflecting the experiences of over 1900 individuals impacted by pulmonary hypertension from Europe, North and South America and Asia. Ten studies did not report participant's WHO functional class of pulmonary hypertension, which resulted in comparing experiences between different severity difficult. All studies met the majority of the quality assessment items. Six descriptive themes emerged discussing participant's experiences of diagnosis, treatment, prognosis, healthcare professionals, impact and coping with pulmonary hypertension. Four higher order analytical themes were developed from the descriptive themes, reflecting: (i) uncertainties and anxiety that participants encountered related to pulmonary hypertension; (ii) lack of recognition of the impact of the condition; (iii) frustration at the paucity of awareness of pulmonary hypertension in society and healthcare settings and (iv) participant's accounts of transitioning through different stages of living with the disease.

Conclusions These findings form the first synthesis of experiences of life in individuals impacted by pulmonary hypertension and illustrate the multifaceted impact of the condition. The voices of numerous groups are missing from the literature highlighting the need for additional research. The results have implications for clinical practice emphasising the role of educational and psychological therapies to support those with the disease.

Strengths and limitations of this study

- This is the first systematic review of the impact of pulmonary hypertension on adults' experiences of living.
- A thematic synthesis approach was used, a key aim of which is to help inform future interventions, policy and clinical practice.
- A comprehensive search strategy was undertaken screening 9362 articles from four electronic databases.

INTRODUCTION

Pulmonary hypertension (PH) describes a group of conditions characterised by elevated pulmonary artery pressure, which untreated results in right heart failure and premature death. Five groups each sharing clinical and pathophysiological features can be described: group 1: pulmonary arterial hypertension (PAH); group 2: PH due to left heart disease; group 3: PH due to lung disease or hypoxia; group 4: chronic thromboembolic PH and group 5: PH due to unclear or miscellaneous disorders.[1 2] The WHO functional classification system in PH is an important tool used to reflect disease severity, and monitor progression of the disease and response to treatment. This index ranges from I, suggestive of patients experiencing no or minimal symptoms during physical activity, to IV indicative of patients experiencing severe symptoms at rest.[2]

Despite improved outcomes in PH-related mortality associated with advancements in medical therapies,[3–5] it remains, a life-shortening condition with significant physical and psychological morbidity and a profound impact on quality of life (QoL).[4] Recent research has explored patient's experiences of living with the disease,[6 7] demonstrating the importance of psychosocial factors in PH on QoL[8–10] and engagement in care.[11] A greater understanding of how individuals are affected and cope with PH should help to improve treatment pathways and patient-reported outcomes.[7]

Table 1 Search terms

Concept 1	AND	Concept 2
Pulmonary hypertension OR Pulmonary arterial hypertension		Qualitative OR thematic OR mixed*methods OR experience* OR perspective* OR semi*structured OR interview OR phenomenolog*

This review systemically synthesises the published qualitative evidence examining adults' experiences of PH. The majority of researchers in this area have employed quantitative methodologies.[7] These studies have investigated a specific difficulty asking individuals to answer a series of structured questions by endorsing predefined responses.[12–14] While this has produced large generalisable datasets, participants may not have been able to discuss experiences most important to them. Qualitative methods, however, allow participants to describe their experiences, in their own words, meaning rich and fine-grained data can be collected. The objective of this review was to provide a more in-depth and nuanced understanding of experiences, highlight unmet needs and identify new areas of investigation.[15] For instance, the current findings were used to help guide the development of a self-help intervention for anxiety in PH.

METHODS

Search strategy

This systematic review has been conducted in accordance with Preferred Reporting Items for Systematic Reviews and Meta-Analyses and Enhancing Transparency in Reporting the Synthesis of Qualitative Research (ENTREQ)[16] (online supplemental 1 and 2). A comprehensive search of four databases was performed in January 2020, which was repeated in May 2020 prior to submission: Web of Science, PubMed, PsycINFO and Cochrane Library. The references of suitable articles were searched for relevant studies: this helped to identify one additional article[17] (tables 1 and 2). Search terms were informed by author's experience of previously published articles on PH. We used the term 'pulmonary hypertension' to remain inclusive as this is likely to also identify other groups of PH. Keywords of several suitable articles were checked to confirm salient words had not been omitted.

Quality assessment

Relevant studies were evaluated using the 10-item Critical Appraisal Skills Programme for qualitative research.[18] An overall score was given to each article demonstrating that the study had achieved the majority or all (++), most (+) or few (−) of the criteria. The lead author (GHR) assessed all articles. A second researcher (CG) independently re-assessed approximately one-quarter of the papers (n=5) chosen as random. Following discussion, a 92% level of agreement per study was observed for all 11 items (including the overall score) per study, compared with an 87% level of agreement prior to discussion. All findings, regardless of quality, were included in the synthesis to remain inclusive.

Data analysis

A thematic synthesis was adopted using four stages[19]:

1. GHR read each paper before extracting relevant information.

Table 2 Inclusion and exclusion criteria—based on the spider tool[61]

	Inclusion	Exclusion
Sample	Adults (18 years or older) diagnosed with PH; participants who self-reported being impacted by a diagnosis of PH. We assumed participants were adults unless otherwise stated.	Children (aged <18 years), individuals without a diagnosis of PH.
Phenomenon of interest	Exploring individual's experiences of PH, that is, diagnosis, treatment, psychosocial impact.	Exploring other cardiovascular conditions. Using qualitative methodology to address a quantitative research goal, that is, development of a quantitative clinical measure.
Design	Any study that reported utilising a qualitative research methodology that is, open-ended question (sometimes known as Big 'Q') was considered. If participants with PH were grouped with individuals with a different diagnosis, their unique contribution must be made explicit.	Solely quantitative research methodologies or converted experiences to numbers, that is, standardised psychometric measures. Participants with PH grouped with other individuals without PH and it is not clear from whom data were collected from.
Evaluation	Any form of qualitative analysis that aim to report on subjective experiences, that is, thematic analysis, interpretative phenomenological analysis.	Solely quantitative analysis.
Research type	Published in English in a scientific journal and peer reviewed. No date restrictions were applied.	Grey literature, that is, books, abstracts, blogs, unpublished studies, reviews.

PH, pulmonary hypertension.

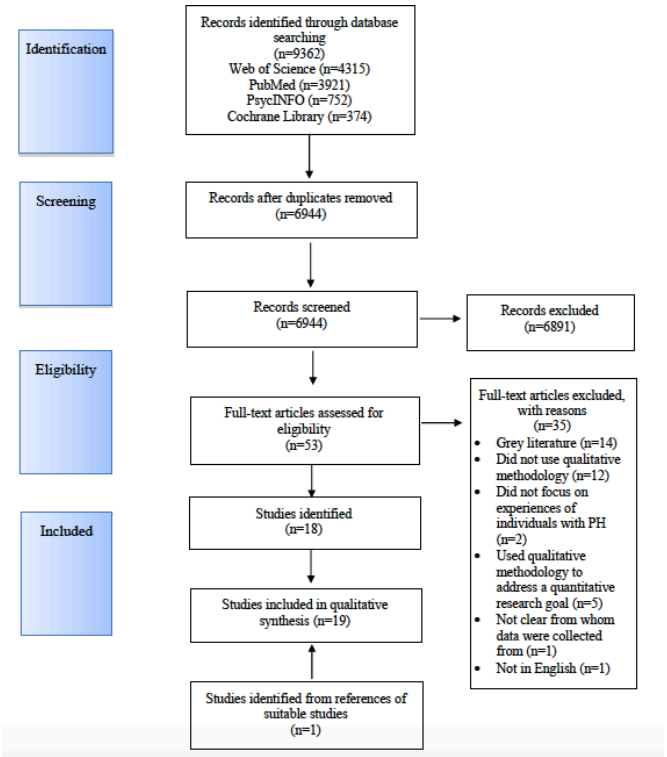

Figure 1 Preferred Reporting Items for Systematic Reviews and Meta-Analyses diagram. PH, pulmonary hypertension.

2. Findings were iteratively coded by GHR with the aim of developing descriptive themes. Themes were informed by frequency and saliency of codes, as opposed to study characteristics (ie, sample size).
3. Analytical themes were developed, which aimed to go beyond the original data and descriptive themes in context of the review's objective. Themes were discussed among other members of the research team.
4. Final report was written, and suitable quotations were selected. " " have been used when reporting participants' words and ' ' for authors.

Patient public involvement

There was no patient and public involvement in this systematic review.

RESULTS

Findings of search

Overall, 19 studies were included in the synthesis (figure 1). Studies were published between 2005 and 2020; however, 18/19 were published in the last 10 years.[20] Data reflect over 1900 individuals across Europe, North and South America and Asia. Ages ranged from 19 to 91 years. In all but one study, the samples were predominantly female[21]—likely reflecting the gender bias observed in PH.[22] Only three studies provided sufficient detail regarding sample ethnicity.[17 23 24] Participants had been diagnosed with PH ranging from <1 year to 24 years. Ten studies failed to report medical information concerning participant's functional class—this is important given that experiences seemed to differ depending on disease-specific issues.[17 25] The majority (n=14) of studies used research interviews for data collection, whereas greater variation of analytical methods was used (table 3).

Quality assessment

All studies met the majority of the 10 quality assessment items (table 4). Seven studies failed to justify their decision for using a qualitative approach and one study provided a limited description of their data analysis method. Surprisingly, 15 articles did not report their method of reflexivity.

Descriptive themes

Six descriptive themes emerged:

Diagnosis

Participants reported experiencing PH-related symptoms a "*long*" time [[27], p. E19] prior to obtaining a diagnosis.[21 26 27] This period was characterised by a decline in physical functioning, QoL and emotional and social difficulties.[20 25 28 29] During this time, and thereafter, individuals expressed confusion over the cause of their experiences[28]—worries that were not necessarily reduced by the diagnosis, which was described as having posed "*more questions than it answers*"[22, p. 3]. Participants tended to avoid, adapt to or explain away their symptoms: "[I] *just assumed that everything was down to smoking*" [27, 28, p. 4].

Participants described undergoing a series of examinations, referrals and misdiagnoses to find an explanation for their experiences.[26–29] This process left participants feeling '*frustrated*' [21, p. 103], '*uncertain*' [26, p. 611], angry and "*disillusioned*" with the perceived meaning of their symptoms [28, p. 6]. Receiving the diagnosis was described as '*life-changing*' [37, p. 4]. While people responded differently, for instance, expressing feelings of '*relief*'—as they can now receive treatment [22, p. 3]—'*shock*' [30, p. 38], helplessness[30] and confusion,[26] a strong emotional response was evident.[31] Following the diagnosis, individuals ruminated over the cause of their disease.[27]

Treatment

Participants discussed their reliance on specialist centres,[6 21] which helped reduce feelings of uncertainty by answering questions and dispelling false narratives about PH: '*it* (the diagnosis) *wasn't as devastatingly unhopeful as first appeared*' [28, p. 7]. A disparity in care between services was described,[6] specifically, between specialist centres and community services in terms of knowledge, treatments and approach to care.[21 28 31] Many participants expressed the need for greater collaboration between services.[6 21 26 27]

Individuals often discussed how they felt their care could be improved, which included, regular checkups,[24] receiving more information,[27 32] promoting shared decision making[26] and holistic care[33] and involving family members.[27]

Participants did not view medication as a "*cure*" [29, p. 2], but rather to alleviate symptoms, improve health and

Table 3 Summary of studies

Study	Country	Aim	N	% female	Age (mean unless stated) and range	Ethnicity	Years since diagnosis (mean unless stated) and range	WHO classification of PH described by authors	WHO functional class	Data collection and analysis
Alami *et al*[26]	France	To explore adult's experiences of PH and identify potential improvements in care	16	63	49 24–75	NR	NR	75%=idiopathic PAH 25%=heritable PAH	I=0% II=38% III=50% IV=12%	Semi-structured interviews; grounded theory
Armstrong *et al*[28]*	UK	To investigate participant's experiences of being diagnosed with PH	30	60	56.3 26–80	NR	<1to >10	Group 1 PAH (86%) 37%=idiopathic PAH 23%=connective tissue disease-associated PAH 17%=congenital heart disease-associated PAH 3%=portopulmonary hypertension 3%=heritable PAH 3%=drugs/toxins-associated PAH Group 3 PH-lung disease (3%) Group 4 CTEPH (4%) NR (3%)	I=0% II=30% III=63% IV=7%	Semi-structured interviews; thematic analysis
Carroll *et al*[24]	USA	To understand the motivations of why patients with PH partake in randomised controlled trials	26	85	Median 56 25th percentile 41 75th percentile 61	65% white, 19% black, 15% Hispanic	Median 5.5 25th percentile 4 75th percentile 11	Group 1 PAH (85%) 50%=idiopathic PAH 23%=connective tissue-associated PAH 4%=heritable PAH 4%=congenital heart disease-associated PAH 4%=portopulmonary Other/unknown 15%	I–III=100%	Randomised participants to review a hypothetical randomised controlled trial that did or did not allow continuation of pre-existing PAH therapies followed by semi-structured interviews; thematic analysis
Chiang *et al*[30]	Taiwan	To explore treatment experiences in individuals with PH	17	76	53.6 27–84	NR	6.4 2–15	Form of PH not specified	NR	Semi-structured interviews; thematic analysis

Continued

Table 3 Continued

Study	Country	Aim	N	% female	Age (mean unless stated) and range	Ethnicity	Years since diagnosis (mean unless stated) and range	WHO classification of PH described by authors	WHO functional class	Data collection and analysis
Flattery *et al*[20]	USA	To describe patients experiences of PAH and explore the impact	11	73	58.5 40–72	'Most were white' (p. 101)	3.5 0.5–14	Group 1 PAH 55%=idiopathic PAH 18%=systemic lupus erythematosis-associated PAH 9%=mixed connective tissue disease 9%=portopulmonary hypertension; 9%=other PAH Not specified (45%)	NR	Semi-structured interviews; Colaizzi's approach to phenomenological analysis
Goddard *et al*[21]	UK	To use narrative analysis to explore the impact of PH	12	50	20–79	NR	NR	Group 1 PAH (100%)	NR	Unstructured narrative interview; narrative analysis
Hall *et al*[31]	Canada	To describe patient's experiences of adjusting to PH and continuous intravenous epoprostenol	7	71	55 43–70	NR	3 0.5–7	Group 1 PAH (100%)	NR	Semi-structured interviews; constant comparison method
Ivarsson, Ekmehag B[27]+	Sweden	To describe patient's experience of information provision in PH	17	76	56 28–73	NR	5 1–12	Group 1 PAH (71%) 35%=idiopathic PAH 24%=systemic sclerosis-associated PAH 12%=PAH Group 4 CTEPH (29%) 11% treated by pulmonary endarterectomy	NR	Semi-structured interviews; content analysis
Ivarsson, Ekmehag B[34]+	Sweden	To describe patient's experiences of support in PH	17	76	56 28–73	NR	5 1–12	Group 1 PAH (71%) 35%=idiopathic PAH 24%=systemic sclerosis-associated PAH 12%=PAH Group 4 CTEPH (29%) 11% treated by pulmonary endarterectomy	NR	Semi-structured interviews; content analysis
Keen *et al*[6]	UK	To explore the nature of physiotherapy for PH in patients	3	67	44–66	NR	8–10	Pulmonary hypertension Breakdown of classification not specified	NR	Semi-structured interviews; framework analysis

Continued

Table 3 Continued

Study	Country	Aim	N	% female	Age (mean unless stated) and range	Ethnicity	Years since diagnosis (mean unless stated) and range	WHO classification of PH described by authors	WHO functional class	Data collection and analysis
Kingman *et al*[29]	International study—Europe, Asia, North America, South America	To examine patient's perspectives of PH, including impact of living with PH, management and treatment	39	75	19–91	NR	NR	Group 1 PAH (87%) Group 4 CTEPH (13%)	I=5% II=36% III=44% IV=15%	Video of participants in their home, field notes and participant's diaries; ethnography
Lee *et al*[33]	International study	To investigate different data sources to better understand perspectives of those with PH	1087 from online discussion board; 27 from archival data	NR	NR	NR	NR	Group 1 PAH (100%)	NR	Collected from online discussion boards and Food and Drug Administration archival data; NR
Lo *et al*[17^]	Canada	To examine the presentation of existential distress in those with PAH	30	77	52 24–77	57% white, 20% South Asian, 3% black, 3% Arab, 3% Indigenous, 13% other	6.3 0.5–24	Group 1 PAH (93%) 40% PAH associated with CTD Group 2 PH left heart disease (7%)	I=10% II=60% III=23% IV=7%	Semi-structured interview; thematic analysis
Matura *et al*[35]	International study	To examine how people with PH are using online discussion boards	549 data available for n=155	92	45.6 19–78	NR	NR	Form of PH not specified but n=95 used prostanoid therapy, which is approved for group 1 PAH	Participants who self-identified with PH group I–V were included	Data were collected from online discussion board posted by individuals with PH; qualitative descriptive methodology
Martin *et al*[37^]	Canada	To identify avoidant techniques in patients with PH when discussing difficult topics associated with the disease	30	77	52 24–77	57% white	6.3 0.5–24	Group 1 PAH (100%) 40% PAH associated with connective tissue disease	II=60%	Semi-structured interview; qualitative content analysis
McDonough, Matura LA[23]	USA	To explore symptoms and their impact on participants with PH experience	10	70	65 38–81	80% white 10% Asian-American 10% other	5.3 1–10	Group 1 PAH (100%)	I=10% II=10% III=70% IV=10%	Semi-structured interviews; qualitative descriptive methodology

Continued

Table 3 Continued

Study	Country	Aim	N	% female	Age (mean unless stated) and range	Ethnicity	Years since diagnosis (mean unless stated) and range	WHO classification of PH described by authors	WHO functional class	Data collection and analysis
Muntingh *et al*[25]	The Netherlands	To explore mental health needs in patients with PH	24	83	26–69	NR	0–16	Group 1 PAH (100%)	NR	Semi-structured interviews; mixed methods—constant comparison method
Uhlenbusch, Löwe B (2019)[32]	Germany	To explore burden of living with a rare disease, including PH	4	NR	NR	NR	NR	Group 1 PAH (100%)	NR	Focus groups; content analysis
Yorke J, Armstrong I[36]*	UK	To explore participant's experiences of daily life with PH	30	60	56.3 26–80	NR	<1to >10	Group 1 PAH (89%) 37%=idiopathic PAH 23%=connective tissue disease-associated PAH 17%=congenital heart disease-associated PAH 3%=heritable PAH 3%=portopulmoary hypertension 3%=drugs/toxins-associated PAH Group 3 PH-lung (3%) Group 4 CTEPH (7%) NR 3%	I=0% II=30% III=63% IV=7%	Semi-structured interviews; thematic analysis

*, +, ^ same sample investigated across studies.
CTEPH, Chronic thromboembolic pulmonary hypertension; n, number; NR, not reported; PAH, pulmonary arterial hypertension; PH, pulmonary hypertension.;

Table 4 Quality assessment using critical appraisal skills programme[18]

Study	Criteria										Rating
	1	2	3	4	5	6	7	8	9	10	
Alami *et al*[26]	+	+	+	+	+	+	+	+	+	+	++
Armstrong *et al*[28]*	+	+	+	+	+	−	+	+	+	+	++
Carroll *et al*[24]	+	+	−	+	+	−	+	+	+	+	++
Chiang *et al*[30]	+	+	−	+	+	−	+	+	+	+	+
Flattery *et al*[20]	+	+	+	+	+	−	+	+	+	+	+
Goddard *et al*[21]	+	+	+	+	+	−	+	+	+	+	++
Hall *et al*[31]	+	+	+	+	+	+	+	+	+	+	++
Ivarsson B, Ekmehag B[27]+	+	+	−	+	+	−	+	+	+	+	+
Ivarsson B, Ekmehag B[34]+	+	+	−	+	+	−	+	+	+	+	+
Keen *et al*[6]	+	+	−	−	−	−	+	+	+	+	−
Kingman *et al*[29]	+	+	+	+	+	−	+	+	+	+	++
Lee *et al*[33]	+	+	+	+	+	−	+	−	+	+	+
Lo *et al*[17]^	+	+	−	+	+	−	+	+	+	+	+
Matura *et al*[35]	+	+	+	+	+	+	+	+	+	+	++
Martin *et al*[37]^	+	+	−	+	−	−	+	+	+	+	+
McDonough A, Matura LA[23]	+	+	+	+	+	+	+	+	+	+	+
Muntingh *et al*[25]	+	+	+	+	+	−	+	+	+	+	++
Uhlenbusch N, Löwe B[32]	+	+	+	+	+	−	+	+	+	+	++
Yorke J, Armstrong I[36]*	+	+	+	+	+	−	+	+	+	+	++

*, +, ^ same sample investigated across studies.
Was the research aim(s) (1) clearly stated and (2, 3) addressed using a qualitative approach. Evaluation of the: (4) recruitment strategy, (5) data collection method, (6) issues concerning reflexivity, (7) ethical implications, (8) data analysis, (9) clarity of findings and (10) overall value of the research.

ultimately, survival: "[if medication was stopped] *Well, all of us would die. It's as simple as that"* [25, 27, 29, 31–33, 38, p. 458]. Many reported an almost immediate reduction in symptoms attributed to treatment,[27 34] with a '*strong emotional attachment*' to medication being noted [25, 33, p. 5].

Side effects of treatments were common, which seemed to mostly manifest physically.[20 23 32] There was uncertainty surrounding side effects as participants reflected that it was not always clear whether their symptoms were associated with the disease itself or psychosocial burden of PH.[33 35] Side effects posed as a barrier to adherence, as while some viewed the negatives of treatment as a tradeoff: "*it's added years to my life…but it's bittersweet"* [21, p. 102], others stopped despite the risk.[36]

Other barriers were discussed such as, accessibility and cost of treatment, emotional distress and problems with self-administration.[23 24 31–34 36] Lack of perceived need also affected uptake as while many experienced emotional difficulties, some felt they did not need psychological support,[34] perceiving: "*the disease itself is the problem*"[29, p. 4]. However, some expressed the need for help with managing anxiety and low mood.[25]

Participants described a number of worries associated with their treatment, such as, whether it was effective[20] and if so for how long,[36] potential risks,[31] what other options are available[29 31 36] and the wait to receive treatment.[17] Visible treatments could be a source of '*social stigma'* [27, 38, p. 456], leaving participants feeling *'self-conscious'* [33, p. 4].

Prognosis

Participants were saddened,[30] "t*errified*"[35, p. 162] and anxious over the prospect of their death.[17 25] Individuals described many uncertainties associated with their prognosis including, how their disease would progress,[23 35] what their future would hold,[34] the impact of their death on loved ones[17 21 25] and how long they had remaining: "*I am clueless as to how long my heart will hold out"* [29, p. 3]. One women wondered if her death would make it "*easier*" due to the impact of PH on her family [17, p. 4]. The pervasive fear over when or how their symptoms would worsen '*plagued'* participants [24, p. 129], resulting in emotional and psychological difficulties.[23 34]

Healthcare professionals

Developing a therapeutic relationship with a PH "*expert"* was important [33, p. 5]. Professionals were described as a source of support,[34] advice,[6] knowledge,[31] reassurance,[28] motivation and '*hope*' [29, 31, p. 4]. That said, participants

reported some difficulties accessing or discussing their worries with professionals,[26 27] as well as previous encounters in which professionals lacked competency,[32] empathy or sensitivity: one participant, for example, reported being told "*It's [PH] worse than cancer, we can't treat it*" [28, p. 3].

Participants expressed their frustration over healthcare professionals' poor understanding of PH.[28 31 32] Individuals spoke of pleading or taking it on themselves to explain the disease to staff.[21 27]

Impact

A range of symptoms were experienced by participants including, dyspnoea, pain, dizziness, fatigue, palpitations and cognitive difficulties.[20 21 23 26 32 35] Breathing problems, however, were described as the "*main symptom*" [27, p. E16], which was associated with feelings of breathlessness, '*suffocating*', '*choking*' and '*pain*'—this was closely interlinked with anxiety [24, 27, p. E16]. Breathing could become a '*conscious act*' [27, p. E16], as participants provided precise details over which activities would cause them shortness of breath [27, p. E17]. That said, some symptoms were '*unpredictable*' and likened to being on a '*rollercoaster*' [30, 38, p. 456].

Decline in physical ability, mobility and energy levels were prevalent,[25 30] which could be caused by any form of activity.[23 26 31] Participated expressed '*fear*' of engaging in activities [33, p. 4] due to the onset of symptoms, pain[23] or it requiring too much energy.[34] There seemed to be a discrepancy between participant's ideal and actual self, in terms of ability,[26] which manifested in feelings of '*anger*' [24, p. 129], "*failing*" [22, p. 4], '*disappointment*' [32, p. 38] and shame.[34]

Individuals had to reduce or cease many activities,[17 26 37] leading them to relate their situation to a "*shackle*" [33, p. 4]. Reflected in this, was the feeling of restriction, the nature of which had a considerable impact on profession, family and social roles.[17 21 25 26 30 32 33] This appeared to be, somewhat, moderated by age, gender and stage of the disease.[17 25] For example, younger participants had to limit themselves to activities that matched their '*activity tolerance*' opposed to interests [33, p. 4]. Participants reported sexual difficulties largely due to fatigue and shortness of breath, which negatively affected their relationship.[17 26 34] Financial problems associated with the cost of care and loss of earnings.[25 30 32] Practical and emotional problems caused by travelling.[31 34] Male and female participants discussed difficulties associated with having or caring for their children.[17 21 25 27 34]

There was a general sense that the disease could be invisible and "*hidden*" from others [38, p. 456]. Family, friends, the public, local authorities, insurance companies and even healthcare professionals were all described, at times, as failing to understand the disease and participant's physical limitations.[17 20 26 27 32 34 36] Participants found themselves having to '*battle*' for their illness to be recognised [38, p. 458], and justify their difficulties.[27 34]

Life with PH could have a negative effect on participant's mental health, with anxiety, low mood, isolation[23 25 29 32 35] and suicidal ideation being discussed.[30 37]

Coping

Participants required a period of time to adjust to life with PH.[20 23 30 36] Individuals coped differently, for example, an international study observed two approaches: '*disease dominated*' whereby participants harboured a '*passive attitude towards PH*', were dependent on others, and likely to experience depression; whereas '*solution seekers*' developed strategies to manage, were less dependent and maintained a social life [33, p. 4].

A common approach to coping was preparation,[21] '*extensive planning*' and developing '*daily routines*' [30, 33, p. 5]. Individuals discussed weighing up how activities would impact them, rather than "*act on impulse*" [24, p. 127]. Participants reported feeling like they had to '*hold back or take it* [daily activities] *slow*' [21, 24, p. 127]. Personal limits were learnt through '*testing*' their own ability [30, p. 39].

Many acknowledged the importance of developing a helpful or positive *"mindset*" [26, 29, p. 4], for example, focusing on activities that were still within their ability[23]: "*I tell myself that I have to look at what I can still do…*" [30, p. 39]. Self-talk,[31] focusing on having overcome previous adversity,[37] faith[20] and self- humour were also used to cope.[20 21]

Altering expectations of ability, recovery and future[25–27 36] allowed some to develop a new sense of purpose[31] or "*redefine*"their lives [24, p. 128, 27, 35]. Accepting their limits gave participants the perception that they were not letting their disease "*dominate*" [25, 26, 33, p. 4] and provided a sense of control[23]: *"…it's just my life, it's just what I do"* [21, p. 102]. Keeping active was described as helping to shift their focus away from their symptoms,[35] control disease progression and have psychological benefits.[6]

Family, friends and employers were described as offering practical and emotional support.[25 29 30 34 35] However, participants appeared conflicted, describing their own position as "*enforced dependency*" [17, 33, p. 3]. Social support could also be a barrier or counterproductive to coping, as how others dealt with the disease was not always helpful.[32 34]

Participants spoke about concealing, avoiding, diminishing and making excuses for their difficulties.[17 21 29 30 36 37] This helped reduce some of the emotional impact of PH, as well as feeling less of a burden or protecting others.[31 34 37] However, this could lead to social withdrawal and isolation, further straining relationships and causing psychological difficulties.[25 29 30]

Peer supports groups and organisations were common sources of reassurance, support and advice.[32 34] Individuals sought validation and normalisation of their symptoms from people with shared experience[35] and also looked to help others.[24]

Analytical themes

Four key analytical themes emerged:

Managing uncertainty

Participants acknowledged many uncertainties caused by internal, such as bodily sensations, or external events linked to PH, including the reaction of others[26] or what the future may hold.[17] Uncertainty was often associated with a perception of risk or threat, which precipitated and perpetuated psychological, emotional and physiological distress. Many of the approaches that participants employed to manage their difficulties related to uncertainty seemed to only provide brief relief, until the next unknown situation occurred, or their strategy fed back into their level of distress, for example:

Participants sought information and reassurance from a range of sources,[20] in the hope of gaining knowledge, understanding and some sense of certainty.[28] Unfortunately, given the complexity and lack of understanding of PH, this could result in even greater uncertainty.[28]

Individuals seemed to engage in worry as a helpful means to problem solve, plan and minimise negative outcomes. However, planning could be impractical due to the unpredictable nature of PH[36] or it removed *'spontaneity in their life'* [24, p. 127]. Worrying was also closely linked with hypervigilance to the impact of everyday activities[23] and somatic changes as an '*indicator*' to alter behaviour [22, 27, p. E17]. This resulted in anticipatory anxiety and panic.[23]

Some described avoiding[25 37] or concealing their disease, however, this often prolonged distress and resulted in emotional difficulties, such as anxiety, guilt and loneliness.[17]

Physical nature of PH

PH was described as a physically limiting disease,[32] as was there a tendency for individuals to report somatic experiences.[21 23 26] Furthermore, despite experiencing emotional difficulties, participants often resisted the idea or need for psychological support.[25 32 37] A notion further reflected in the relief on hearing the disease was not psychosomatic in nature.[28]

Conversely, participant's social environment did not always recognise the physical manifestations of the disease—participants assumed because "*you can't see anything*" and most of the time they "*look fine*" as there was a lack of visible signs [26, 38, p. 455]. Discordance between the meaning and impact participants and others attached to the disease seemed most salient on occasions when physical adaptations were required due to fatigue or breathlessness—symptoms of which could be invisible to others.[25 34] It seemed the support participant's received was, at times, influenced by other's representation of the disease.[21] Likewise, participant's own perceptions of their circumstance may have affected their experiences of coping, which could help, in part, to explain resistance to psychological support.

Living with the rarity of PH

The rarity of PH was highlighted in discussions,[25 26] no more so than when individuals expressed their frustration at the lack of PH awareness in society and healthcare.[27 32] Lack of understanding and misconceptions over PH resulted in participants feeling stigmatised and discriminated against.[21 25 32] Participants found themselves having to justify and legitimise their sick role, despite having an incurable disease[32 36] or withdrawing to avoid difficulties.[17] Individuals felt the need to become experts in PH, which made it all the more frustrating when others assumed that they knew better or when participant's voices were unheard in care.[21 27] Living with a rare condition left individuals feeling lonely and isolated: "*forced to carry a burden no one wants, or understands fully*" [33, p. 4]. Participants found understanding from experts in PH and PH communities.[20]

Transitional nature of PH

Participants progressed through different transitions living with PH.[25] At first, experiencing a decline in general functioning, which appeared to improve following the diagnosis and administration of treatment.[28] Participants next faced the process of adapting to life with PH and treatment,[20 31] the goal of which was to stabilise the disease,[6] maintain a good QoL and survive.[25] Participants had to find a balance between feeling underactive, in which they perceived their identity as changing[17] and themselves "*failing*" [22, p. 4], and overactive, whereby participants attempted to resume normal life despite the risk and increase in PH symptoms, or dissonance feeling torn between societal norms and their own ability.[23] This balance was often achieved through '*trial and error*' [30, p. 39] and becoming more aware of physical, psychological, family and society cues.

DISCUSSION

This review presents the first systematic synthesis of qualitative data exploring adults' experiences of living with PH. Four analytical themes emerged reflecting: how individuals navigate uncertainty associated with PH; the physical and somatic nature of PH; implications of living with a rare disease and the transitional nature of adapting to life with PH.

Feelings of uncertainty are commonly experienced by individuals with a rare medical condition.[38] For participants in the current review, uncertain events were often perceived as threatening resulting in distress. Intolerance to uncertainty is one of the main theories aimed at understanding generalised anxiety disorder (GAD).[39] In GAD, individuals find the feeling of uncertainty difficult. This distress is often managed by engaging in worry as a method to prepare for possible eventualities. The associated anxiety however influences individuals to appraise and overestimate events as negative and threatening. As observed here, this can generate greater uncertainty and anxiety.[39 40] A study investigating GAD in PH found 93%

reported some symptoms.[41] Symptoms of GAD in PH has been found to be associated with depression,[41] which may impact patient's ability to adapt to the challenges of PH.[42] Although anxiety and depression in PH is common, just over one-third of patients receive treatment.[43] Strategies aimed at helping individuals to better manage distress associated with uncertainty are likely to be useful. A range of psychological treatments have been shown to be effective for GAD,[44] however, there is paucity of evidence regarding psychological treatments in PH.[45]

Participants focused primarily on the physical difficulties of PH. While this may indeed be how the disease presents, individuals described becoming more aware of somatic feedback to help gauge their own body. Individuals investigated elsewhere who experienced chest pain have also reported hypervigilance to cardiopulmonary sensations. The degree of vigilance was positively related to chest pain, the association of which was mediated by fear of bodily sensations.[46] A similar relationship can be observed in Clark's model of panic, whereby perceptions of threat and anxiety over somatic symptoms increase the likelihood of experiencing, and also misinterpreting, a bodily sensation as negative.[47] Individuals with PH should be supported to better understand and differentiate the shared overlap between cardiovascular symptoms and anxiety.[48] Interventions, such as mindfulness, that help individuals disengage their attention from sensations may also be beneficial.[49]

Disparity in illness perceptions between participants and others was a common source of distress. This has also been reported in other diagnoses that are associated with poor understanding and uncertainty.[50] Such incongruences have been suggested to influence how individuals present to services with their symptoms and adherence to treatment.[51] Healthcare professionals may benefit from additional training on PH, as well as developing skills to help explore individual's understanding of the disease, and perceived psychosocial impact.[52] While greater consideration of individual's social support in PH care will likely minimise the impact and degree of conflicting perceptions, helping patients to develop skills to manage conversations when other people are confused about PH is also important.[53]

PH care should reflect the transitional nature involved in adapting to life with PH. This may help to minimise or prevent worsening of psychosocial symptoms following their PH diagnosis.[54] Initially, individuals are likely to value from additional support in understanding PH, managing the emotional impact of the diagnosis and integrating the disease into their identities exploring existential worries. For example, caregivers should help individuals to explore their difficulties and construct new meaning.[55] Experiences of fatigue appeared a prominent theme thereafter as individuals aimed to find a balance between their energy and engaging in activities. Fatigue is the second most common symptom patients with PH present with.[52] Supporting individuals to manage fatigue through education, pacing and prioritising valued actions is likely to helpful—this may also reduce mood disorders.[56]

Limitations

Most studies did not discuss reflexivity. This involves the researcher attending to the wider content of knowledge acquisition, discussing the potential role of relationships between participants and themselves in their investigation.[57] This is an important stage in some qualitative research approaches and also contributes to rigour and trustworthiness.[58]

Due to lack of reporting, the analysis was unable to distinguish between experiences based on PH-related factors, which has been shown to be associated with psychological distress.[59 60] Grouping individuals based solely on their diagnosis of PH may have resulted in idiosyncratic findings being missed. Moreover, experiences relating to palliative care, pregnancy, surgery and psychological interventions were largely unrepresented despite being commonly encountered in practice.

Finally, the review protocol has been registered on Open Science Framework registries; while it was not registered prior to data extraction, authors confirm that the protocol had not been altered since it was initially devised and prior to data extraction.

CONCLUSION

This review uniquely thematically synthesises qualitative data from over 1900 individuals with PH across four continents, with the majority of participants recruited in two international studies.[33 35] The review finds numerous voices are missing, which needs addressing, including those from palliative care, non-white background and individuals who have experienced pregnancy in PH. The results have implications for clinical practice highlighting the potential role of education and psychological therapies to support those with the disease.

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Contributors GHR was responsible for the conception of the review, data collection and analysis, and writing the report for publication. He approved the final version for publication. NB made substantial contributions to research design and provided feedback on data analysis and final report. He approved the final version for publication. IA made contributions to data analysis and provided feedback on the final report. He approved the final version for publication. RC made contributions to data collection, analysis and provided feedback on the final report. He approved the final version for publication. DGK made contributions to data analysis and provided feedback on the final report. He approved the final version for publication. IS made contributions to data collection, analysis and provided feedback on the final report. He approved the final version for publication. ART was responsible for the conception of the review, made substantial contributions to the research design and provided feedback on data analysis and final report. He approved the final version for publication.

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receives an unrestricted educational grant from GSK for an annual clinical education meeting.

Patient consent for publication Not required.

Provenance and peer review Not commissioned; externally peer reviewed.

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REFERENCES

1 Hoeper MM, Bogaard HJ, Condliffe R, *et al*. Definitions and diagnosis of pulmonary hypertension. *J Am Coll Cardiol* 2013;62:D42–50.
2 McGoon M, Gutterman D, Steen V, *et al*. Screening, early detection, and diagnosis of pulmonary arterial hypertension. *Chest* 2004;126:14S–34.
3 Hoeper MM, Ghofrani H-A, Grünig E, *et al*. Pulmonary hypertension. *Dtsch Arztebl Int* 2017;114:73–84.
4 Kiely DG, Elliot CA, Sabroe I, *et al*. Pulmonary hypertension: diagnosis and management. *BMJ* 2013;346:f2028.
5 Galiè N, Humbert M, Vachiery J-L, *et al*. ESC/ERS guidelines for the diagnosis and treatment of pulmonary hypertension. *European Heart Journal* 2016;37:67–119.
6 Keen C, Fowler-Davis S, McLean S, *et al*. Physiotherapy practice in pulmonary hypertension: physiotherapist and patient perspectives. *Pulm Circ* 2018;8:2045894018783738.
7 McGoon MD, Ferrari P, Armstrong I, *et al*. The importance of patient perspectives in pulmonary hypertension. *Eur Respir J* 2019;53:1801919.
8 Halimi L, Marin G, Molinari N, *et al*. Impact of psychological factors on the health-related quality of life of patients treated for pulmonary arterial hypertension. *J Psychosom Res* 2018;105:45–51.
9 Cole E, Armstrong I, Cutts K. Links between psychological factors and health- related quality of life in pulmonary hypertension. *The European Respiratory Journal* 2016;48.
10 Delcroix M, Howard L. Pulmonary arterial hypertension: the burden of disease and impact on quality of life. *Eur Respir Rev* 2015;24:621–9.
11 Graarup J, Ferrari P, Howard LS. Patient engagement and self-management in pulmonary arterial hypertension. *Eur Respir Rev* 2016;25:399–407.
12 McKenna SP, Doughty N, Meads DM, *et al*. The Cambridge pulmonary hypertension outcome review (camphor): a measure of health-related quality of life and quality of life for patients with pulmonary hypertension. *Qual Life Res* 2006;15:103–15.
13 Bonner N, Abetz L, Meunier J, *et al*. Development and validation of the living with pulmonary hypertension questionnaire in pulmonary arterial hypertension patients. *Health Qual Life Outcomes* 2013;11:161.
14 Yorke J, Corris P, Gaine S, *et al*. emPHasis-10: development of a health-related quality of life measure in pulmonary hypertension. *Eur Respir J* 2014;43:1106–13.
15 Harper D, Thompson AR. *Qualitative research methods in mental health and psychotherapy: a guide for students and practitioners*. Oxford: Wiley-Blackwell, 2012.
16 Tong A, Flemming K, McInnes E, *et al*. Enhancing transparency in reporting the synthesis of qualitative research: ENTREQ. *BMC Med Res Methodol* 2012;12:181.
17 Lo C, Sarker T, Canning O, *et al*. Clinical presentation of existential distress in pulmonary arterial hypertension. *Canadian Journal of Respiratory, Critical Care, and Sleep Medicine* 2019;3:56–62.
18 Critical appraisal skills programme. CASP qualitative checklist, 2018. Available: https://casp-uk.net/wp-content/uploads/2018/03/CASP-Qualitative-Checklist-2018_fillable_form.pdf;
19 Thomas J, Harden A. Methods for the thematic synthesis of qualitative research in systematic reviews. *BMC Med Res Methodol* 2008;8:45.
20 Flattery MP, Pinson JM, Savage L, *et al*. Living with pulmonary artery hypertension: patients' experiences. *Heart Lung* 2005;34:99–107.
21 Goddard JC, Armstrong IJ, Kiely DG, *et al*. Combining creative writing and narrative analysis to deliver new insights into the impact of pulmonary hypertension. *BMJ Open Respir Res* 2017;4:e000184.
22 Hoeper MM, Simon R Gibbs J, Gibbs SR. The changing landscape of pulmonary arterial hypertension and implications for patient care. *Eur Respir Rev* 2014;23:450–7.
23 McDonough A, Matura LA, Carroll DL. Symptom experience of pulmonary arterial hypertension patients. *Clin Nurs Res* 2011;20:120–34.
24 Carroll R, Antigua J, Taichman D, *et al*. Motivations of patients with pulmonary arterial hypertension to participate in randomized clinical trials. *Clin Trials* 2012;9:348–57.
25 Muntingh ADT, Gerritsen S, Batelaan NM, *et al*. Pathways to experiencing mental problems and need for mental support in patients with pulmonary arterial hypertension. *Pulm Circ* 2017;7:609–16.
26 Alami S, Cottin V, Mouthon L, *et al*. Patients', relatives', and practitioners' views of pulmonary arterial hypertension: a qualitative study. *Presse Med* 2016;45:e11–27.
27 Ivarsson B, Ekmehag B, Sjöberg T. Information experiences and needs in patients with pulmonary arterial hypertension or chronic thromboembolic pulmonary hypertension. *Nurs Res Pract* 2014;2014:1–8.
28 Armstrong I, Rochnia N, Harries C, *et al*. The trajectory to diagnosis with pulmonary arterial hypertension: a qualitative study. *BMJ Open* 2012;2:e000806.
29 Kingman M, Hinzmann B, Sweet O, *et al*. Living with pulmonary hypertension: unique insights from an international ethnographic study. *BMJ Open* 2014;4:e004735.
30 Chiang Y-C, Hu L-Y, Couper J, *et al*. Exploring the experiences and psychosocial stresses of Taiwanese patients with pulmonary hypertension: a qualitative interview study. *Pulm Circ* 2018;8:2045894018787479.
31 Hall H, Côté J, McBean A, *et al*. The experiences of patients with pulmonary arterial hypertension receiving continuous intravenous infusion of epoprostenol (Flolan) and their support persons. *Heart Lung* 2012;41:35–43.
32 Uhlenbusch N, Löwe B, Depping MK. Perceived burden in dealing with different rare diseases: a qualitative focus group study. *BMJ Open* 2019;9:e033353.
33 Lee C, St Clair C, Merenda CC, *et al*. Assessment of public and patient online comments in social media and food and drug administration archival data. A pilot qualitative analysis Research in social & administrative pharmacy. *Research in Social and Administravei Pharmacy* 2019;10:S1551.
34 Ivarsson B, Ekmehag B, Sjöberg T. Support experienced by patients living with pulmonary arterial hypertension and chronic thromboembolic pulmonary hypertension. *Heart Lung Circ* 2016;25:35–40.
35 Matura LA, McDonough A, Aglietti LM, *et al*. A virtual community: concerns of patients with pulmonary hypertension. *Clin Nurs Res* 2013;22:155–71.
36 Yorke J, Armstrong I, Bundock S. Impact of living with pulmonary hypertension: a qualitative exploration. *Nurs Health Sci* 2014;16:454–60.
37 Martin V, Sarker T, Slusarek E, *et al*. Conversational avoidance during existential interviews with patients with progressive illness. *Psychol Health Med* 2020;25:1073–82.
38 von der Lippe C, Diesen PS, Feragen KB. Living with a rare disorder: a systematic review of the qualitative literature. *Mol Genet Genomic Med* 2017;5:758–73.
39 Dugas MJ, Robichaud M. *Practical clinical guidebooks. Cognitive-behavioral treatment for generalized anxiety disorder: From science to practice*. New York: Routledge, 2007.

40 Bottesi G, Ghisi M, Carraro E, *et al*. Revising the intolerance of uncertainty model of generalized anxiety disorder: evidence from UK and Italian undergraduate samples. *Front Psychol* 2016;7:7.
41 Harzheim D, Klose H, Pinado FP, *et al*. Anxiety and depression disorders in patients with pulmonary arterial hypertension and chronic thromboembolic pulmonary hypertension. *Respir Res* 2013;14:104.
42 Wryobeck JM, Lippo G, McLaughlin V, *et al*. Psychosocial aspects of pulmonary hypertension: a review. *Psychosomatics* 2007;48:467–75.
43 Somaini G, Hasler E, Muller-Mottet S, *et al*. Anxiety and depression are highly prevalent in pulmonary hypertension and might improve with target therapy. *European Respiratory Journal* 2015;46:PA2112.
44 Hunot V, Churchill R, Teixeira V, Silva de Lima M, *et al*. Psychological therapies for generalised anxiety disorder. *Cochrane Database Systematic Reviews* 2007;7:CD001848.
45 Bussotti M, Sommaruga M. Anxiety and depression in patients with pulmonary hypertension: impact and management challenges. *Vasc Health Risk Manag* 2018;14:349–60.
46 White KS, Craft JM, Gervino EV. Anxiety and hypervigilance to cardiopulmonary sensations in non-cardiac chest pain patients with and without psychiatric disorders. *Behav Res Ther* 2010;48:394–401.
47 Clark DM. A cognitive approach to panic. *Behav Res Ther* 1986;24:461–70.
48 Fleet RP, Beitman BD. Cardiovascular death from panic disorder and panic-like anxiety: a critical review of the literature. *J Psychosom Res* 1998;44:71–80.
49 Tulloh RMR, Garratt V, Tagney J, *et al*. A pilot randomised controlled trial investigating a mindfulness-based stress reduction (MBSR) intervention in individuals with pulmonary arterial hypertension (PAH): the pathways study. *Pilot Feasibility Stud* 2018;4:78.
50 Whitehead K, Kandler R, Reuber M. Patients' and neurologists' perception of epilepsy and psychogenic nonepileptic seizures. *Epilepsia* 2013;54:708–17.
51 Monzoni CM, Duncan R, Grünewald R, *et al*. Are there interactional reasons why doctors may find it hard to tell patients that their physical symptoms may have emotional causes? A conversation analytic study in neurology outpatients. *Patient Educ Couns* 2011;85:e189–200.
52 Fenstad ER, Shanafelt TD, Sloan JA, *et al*. Physician attitudes toward palliative care for patients with pulmonary arterial hypertension: results of a cross-sectional survey. *Pulm Circ* 2014;4:504–10.
53 Clarke A, Thompson AR, Jenkinson E, *et al*. *CBT for appearance anxiety : psychosocial interventions for anxiety due to visible difference*. Chichester: Wiley Blackwell, 2014.
54 Halimi L, Molinari N, Marin G, *et al*. Quality of life in worsening and non-worsening PAH patient's in a 1 year follow-up: What's going on? *European Respiratory Journal* 2015;46:PA705.
55 Egnew TR. A narrative approach to healing chronic illness. *Ann Fam Med* 2018;16:160–5.
56 Connolly D, O'Toole L, Redmond P, *et al*. Managing fatigue in patients with chronic conditions in primary care. *Fam Pract* 2013;30:123–4.
57 Dodgson JE. Reflexivity in qualitative research. *J Hum Lact* 2019;35:220–2.
58 Larkin M, Thompson A. Interpretative Phenomenological Analysis in Mental Health and Psychotherapy Research. In: Harper D, Thompson AR, eds. *Qualitative research methods in mental health and psychotherapy*. Wiley-Blackwell, 2012: 101–16.
59 Yorke J, Deaton C, Campbell M, *et al*. Symptom severity and its effect on health-related quality of life over time in patients with pulmonary hypertension: a multisite longitudinal cohort study. *BMJ Open Respir Res* 2018;5:e000263.
60 Yorke J, Armstrong I, Deaton C, *et al*. Patient rated who functional class and psychological distress are strongly associated with dyspnoea in pulmonary hypertension. *European Respiratory Journal* 2015;46:PA331.
61 Methley AM, Campbell S, Chew-Graham C, *et al*. PICO, PICOS and spider: a comparison study of specificity and sensitivity in three search tools for qualitative systematic reviews. *BMC Health Serv Res* 2014;14:579.

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Version 1

Title
Adult's experiences and perspectives of living with pulmonary hypertension: a systematic review and thematic synthesis of qualitative studies

Start date
10/1/20

Completion date
30/4/20

Lead researcher
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Review question
The object of the present review was to undertake the first systematic synthesis, that the authors are aware of, of published qualitative evidence examining lived experiences of PH in individuals diagnosed with the condition.

Searchers
Web of Science, PubMed, PsycINFO and Cochrane Library.

Condition being studied
Pulmonary hypertension (PH), also known as pulmonary arterial hypertension (PAH), is a progressive life limiting condition. It is characterised by a high mean blood pressure of ≥ 20 mm Hg, at rest, in the blood vessels that supply the lungs, otherwise known as the pulmonary arteries. The surrounding walls of the pulmonary arteries become thick and rigid, resulting in hypertension and impaired functioning, heart-failure and eventually death (Marius M. Hoeper et al., 2013; M. M. Hoeper & Humbert, 2019).

Participants
Inclusion - examining adults (18 years or older) diagnosed with PH.
Exclusion criteria - examining children (<18 years old) and/or individuals without a diagnosis of PH

Type of study to be included
Suitable studies must have utilised qualitative research methodologies i.e. research interviews, and used any form of qualitative analysis to interpretative the data. Qualitative research published in English in a scientific journal and peer reviewed.

Main outcomes
Main outcomes will emerge as descriptive and analytical themes from a thematic synthesis of the data.

Data extraction
Following the systematic search any duplicates will be deleted using Endnote Web. Articles will then be screen by title and abstract. Following this, articles will undergo a full-text screen. Included articles will be read several times by the lead researcher (Dr Rawlings) prior to data extraction. After this time the following data will be extracted: authors, year, country, aim, sample characteristics, qualitative methodology, key findings and discussion points relevant to the review's primary aim.

Risk of bias
All studies in the synthesis will be evaluated using the Critical Appraisal Skills Programme for qualitative research. This is a ten-item checklist used to appraise qualitative papers. The first three questions explore whether the research aim(s) are clearly stated and if it can be addressed using a qualitative approach. The following questions are then asked to help evaluate the: recruitment strategy, data collection method, issues concerning reflexivity, ethical implications, data analysis, clarity of findings, and overall value of the research. An overall score will be given to each article to demonstrate that the study had achieved all (++), most (+) or few (-) of the criteria. The lead author will assess each paper individually and 30% will be undergo another quality assessment by a second reviewer. The level of agreement will be reported. The quality of papers will be assessed to help evaluate the evidence and for any interpretations made to be in light of its source. All findings will be included in the analysis irrespective of quality.

Strategy for data synthesis
A thematic synthesis will be used to analyse the data. This approach aims to develop higher order themes in a transparent way as it explicitly reports and distinguishes between descriptive and analytical findings. Moreover, the new analytical themes can be used to inform future intervention and research. A four staged approach will be used, as outlined by Thomas & Harden (2008):

Dissemination plans
We plan to publish the review in an International and peer reviewed journal.

Key words
thematic synthesis, qualitative, pulmonary hypertension, pulmonary atrial hypertension

Appendix 2

PRISMA standards checklist

Section/topic	#	Checklist item	Reported on page #
TITLE			
Title	1	Identify the report as a systematic review, meta-analysis, or both.	1
ABSTRACT			
Structured summary	2	Provide a structured summary including, as applicable: background; objectives; data sources; study eligibility criteria, participants, and interventions; study appraisal and synthesis methods; results; limitations; conclusions and implications of key findings; systematic review registration number.	3
INTRODUCTION			
Rationale	3	Describe the rationale for the review in the context of what is already known.	4-5
Objectives	4	Provide an explicit statement of questions being addressed with reference to participants, interventions, comparisons, outcomes, and study design (PICOS).	5-6
METHODS			
Protocol and registration	5	Indicate if a review protocol exists, if and where it can be accessed (e.g., Web address), and, if available, provide registration information including registration number.	The review was conducted as part of a doctoral thesis. The protocol has been registered on Open Science Framework (OSF).
Eligibility criteria	6	Specify study characteristics (e.g., PICOS, length of follow-up) and report characteristics (e.g., years considered, language, publication status) used as criteria for eligibility, giving rationale.	6

Information sources	7	Describe all information sources (e.g., databases with dates of coverage, contact with study authors to identify additional studies) in the search and date last searched.	5
Search	8	Present full electronic search strategy for at least one database, including any limits used, such that it could be repeated.	6
Study selection	9	State the process for selecting studies (i.e., screening, eligibility, included in systematic review, and, if applicable, included in the meta-analysis).	6
Data collection process	10	Describe method of data extraction from reports (e.g., piloted forms, independently, in duplicate) and any processes for obtaining and confirming data from investigators.	7-8
Data items	11	List and define all variables for which data were sought (e.g., PICOS, funding sources) and any assumptions and simplifications made.	7-8
Risk of bias in individual studies	12	Describe methods used for assessing risk of bias of individual studies (including specification of whether this was done at the study or outcome level), and how this information is to be used in any data synthesis.	7
Summary measures	13	State the principal summary measures (e.g., risk ratio, difference in means).	7-8
Synthesis of results	14	Describe the methods of handling data and combining results of studies, if done, including measures of consistency (e.g., I^2) for each meta-analysis.	7-8

Section/topic	**#**	**Checklist item**	**Reported on page #**
Risk of bias across studies	15	Specify any assessment of risk of bias that may affect the cumulative evidence (e.g., publication bias, selective reporting within studies).	7-8
Additional analyses	16	Describe methods of additional analyses (e.g., sensitivity or subgroup analyses, meta-regression), if done, indicating which were pre-specified.	-
RESULTS			
Study selection	17	Give numbers of studies screened, assessed for eligibility, and included in the review, with reasons for exclusions at each stage, ideally with a flow diagram.	8-10

Study characteristics	18	For each study, present characteristics for which data were extracted (e.g., study size, PICOS, follow-up period) and provide the citations.	11-16
Risk of bias within studies	19	Present data on risk of bias of each study and, if available, any outcome level assessment (see item 12).	17-18
Results of individual studies	20	For all outcomes considered (benefits or harms), present, for each study: (a) simple summary data for each intervention group (b) effect estimates and confidence intervals, ideally with a forest plot.	11-16
Synthesis of results	21	Present results of each meta-analysis done, including confidence intervals and measures of consistency.	18-28
Risk of bias across studies	22	Present results of any assessment of risk of bias across studies (see Item 15).	18
Additional analysis	23	Give results of additional analyses, if done (e.g., sensitivity or subgroup analyses, meta-regression [see Item 16]).	18-28
DISCUSSION			
Summary of evidence	24	Summarize the main findings including the strength of evidence for each main outcome; consider their relevance to key groups (e.g., healthcare providers, users, and policy makers).	29-31
Limitations	25	Discuss limitations at study and outcome level (e.g., risk of bias), and at review-level (e.g., incomplete retrieval of identified research, reporting bias).	31-32
Conclusions	26	Provide a general interpretation of the results in the context of other evidence, and implications for future research.	29-33
FUNDING			
Funding	27	Describe sources of funding for the systematic review and other support (e.g., supply of data); role of funders for the systematic review.	33

Moher D., Liberati A., Tetzlaff J., & Altman D. G. (2009). Preferred reporting items for systematic reviews and meta-analyses: The PRISMA statement. *PLoS Med 6*: e1000097. doi:10.1371/journal.pmed1000097