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# Examining the impact of pulmonary hypertension on nonprofessional caregivers: A mixed-methods systematic review

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## Abstract

Pulmonary hypertension (PH) is a heterogeneous condition, associated with a high symptom burden and high rates of disability. While nonprofessional caregivers are essential in helping patients live better, little is known about the impact on caregivers and support that is currently available. This review has synthesised evidence examining experiences of caregivers of adults with PH. Web of Science, PubMed, PsycINFO, and Cochrane Library were searched for all types of study design. Articles were evaluated and analysed using a Joanna Briggs Institute approach. Eight articles primarily focussed on pulmonary arterial hypertension and chronic thromboembolic PH were identified investigating 456 caregivers from at least 10 countries. Four categories were identified describing caregiver demographics, responsibilities, impact, and support. Four integrated themes emerged identifying possible unmet needs and therapeutic targets: (1) Change, reflecting the various demands caring had on people as they attempted to balance the needs of the patient and their own; (2) Preparedness, discussing how caregivers could feel uncertain and unskilled for their duties and unsupported; (3) Isolation, with caregivers often encountering challenges to gaining information on PH turning to PH organisations and others affected for support and connection; and (4) Physical and mental demands, reflecting the multifaceted impact of caring. Findings add to the evidence demonstrating that PH can have a considerable impact on patients' support network. Providing support for caregivers of people with PH is an unmet need and may have a positive impact on patients and is an area that requires further research.

## KEYWORDS

caregiver burden, carers, pulmonary arterial hypertension, qualitative, quality of life

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## INTRODUCTION

Pulmonary hypertension (PH) is a heterogeneous condition and is characterised by an elevation in pulmonary artery pressure.<sup>1</sup> There are five forms of PH: pulmonary arterial hypertension (PAH), PH due to left heart disease, PH due to lung disease or hypoxia, chronic thromboembolic PH (CTEPH) and PH due to unclear causes. Approximately, 1% of the global population are estimated to have PH, with an even higher prevalence in more elderly patients.<sup>2</sup>

Achieving the correct diagnosis is essential in terms of determining the most effective and appropriate form of treatment, and for understanding prognosis. However, it is not uncommon for people to experience a delay of several years before being diagnosed.<sup>3</sup> Treatments attempt to reduce right ventricular afterload using pulmonary vasodilators in patients with PAH or removing obstructions in the vasculature with interventions such as surgery or balloon pulmonary angioplasty in patients with CTEPH. The aim of these therapies is to improve symptoms, survival and health-related quality of life (HRQoL). Despite initial improvements with drug therapies, the majority of patients with PAH have a progressive condition, whereas for selected patients with CTEPH, there is the prospect of cure following pulmonary endarterectomy.<sup>4</sup> PH referral centres have been advocated as a model for providing care for this patient group, reflecting the complex nature of the disease.

The commonest symptoms of PH are exertional breathlessness, fatigue, leg swelling and chest pain.<sup>5</sup> Severity can be assessed using the World Health Organisation (WHO) functional classification system, ranging from Class I symptoms with patients with near-normal exercise capacity, to Class IV, where individuals will have symptoms at rest or on minimal activity.<sup>6</sup> Most experience Classes II and III symptoms. Patients with severe disease may be breathless at rest or during minimal exercise and may require oxygen. A recent thematic synthesis exploring patients' experience of living with PH demonstrated the multifaceted impact of the disease, the effect of which can go beyond the cardiorespiratory symptoms.<sup>7</sup> This is supported by quantitative findings as individuals with PH commonly report low HRQoL and high rates of morbidity.<sup>8</sup>

The chronic and debilitating nature of PH means that nonprofessional caregivers have an important role to play in providing support to patients.<sup>9</sup> Caregivers of people with a long-term condition often include immediate family members or those belonging to the patient's social network. Given the differences in responsibility, professional training and potential impact, we have made the distinction between professional (i.e., paid professionals

working within health settings) and nonprofessional caregivers (i.e., family members or friends who are typically unpaid for their duties).<sup>10</sup>

The responsibility of caregiving may have a significant effect on caregivers—as well as on the patient.<sup>10</sup> Research has shown caregivers of people with a chronic illness are likely to experience, anxiety and depression in addition to finding it burdensome.<sup>11</sup> The perspective and needs of caregivers are frequently overlooked and without their input, it is recognised that current healthcare services would struggle to meet the needs of the patient.<sup>12</sup>

While the effect of caregiving can be wide and varied, we know that some types of conditions can be associated with a greater burden than others highlighting the need to investigate the impact in certain groups. Such themes are explored further in the review, caregivers of people with PH may have to travel long distances to see specialist clinicians and services, manage the physical demands of their role and navigate the barriers associated with a rare condition. A clinical review examining caregivers' burden of PH was conducted by Verma et al.<sup>9</sup> recognising that while the needs of caregivers have been extensively examined in other conditions, there is a lack of research in PH. The authors synthesised the data from three reports before making recommendations for clinical practice. This included highlighting the need to screen for the burden of care required, educate caregivers, and signpost caregivers for support. However, articles were not identified using a systematic search strategy and therapies for PH have since evolved, potentially changing the landscape. The aim of this review was to systematically identify and assess the quality of the literature that has investigated the impact on caregivers of adults with PH and propose areas for future research and clinical practice.

## METHODS

### Search strategy

A systematic search was performed in May 2021 of Web of Science, PubMed, PsycINFO, and Cochrane Library. Search terms and criteria for eligibility are reported in Tables 1 and 2. The title and abstract of papers were screened by the lead author assessing eligibility. If relevant, articles were then subject to a full-text review.

### Quality assessment

The Mixed Methods Appraisal Tool (MMAT) was used to evaluate relevant articles.<sup>13</sup> First, articles were screened

**TABLE 1** Search terms

Concept 1	AND	Concept 2
"pulmonary hypertension" OR "pulmonary arterial hypertension"		"caregiver*" OR "carer*" OR "partner*" OR "spouse*" OR "dyad"

**TABLE 2** Inclusion and exclusion criteria

	Inclusion	Exclusion
Sample	Individuals providing unpaid care for an adult (18 years or older) with PH. Participants did not need to be referred to as or accept the label of caregiver or carer.	Paid healthcare professionals or researchers.
Phenomenon of interest	Investigating individuals' experience of supporting an adult that they know with PH.	Investigating individuals' experience of supporting a child with PH or an adult with a condition other than PH.
Design and analysis	Any methodology.	-
Type	Published in English language and peer reviewed.	Grey literature.

Abbreviation: PH, pulmonary hypertension.

by the lead author using two questions which examined whether a clear research question(s) was asked and whether it was addressed by the data. If both were met, studies were assessed using five items that were specific to the methodology used. Scores varied from 1 to 5. The second author second-rated 50% of the articles at random. An interrater reliability score of 95% was observed (100% after discussion).

## Data analysis

Joanna Briggs Institute approach to mixed-method systematic review was used,<sup>14</sup> adopting a convergent integrated approach as equal weight was given to quantitative and qualitative data. A four staged approach was used:

1. The lead author read each article before key findings were extracted. Quantitative data were transformed using a process of "qualitising," in which results were translated into textual descriptions.
2. Data were coded with the aim of creating textual categories. Categories emerged by iteratively pooling data across studies based on the similarity and difference of findings. This was a data-driven approach.
3. Categories were then aggregated to produce integrated findings of the review. This was guided by the purpose of mixed methods reviews, which is to synthesise evidence to guide practice and policy.<sup>15</sup> Therefore, themes had relevance to unmet needs and possible therapeutic targets.
4. Themes and proposed clinical implications were discussed with PH clinicians and caregivers exploring

meaningful translation of the findings into useful recommendations.

## RESULTS

### Search results

The search identified 609 unique articles, of which eight articles were relevant (Figure 1). Samples ranged from 7 to 129 caregivers. Data were collected from 456 caregivers from Canada, China, France, Germany, Italy, Spain, Sweden, Turkey, the United Kingdom, and the United States (Table 3).

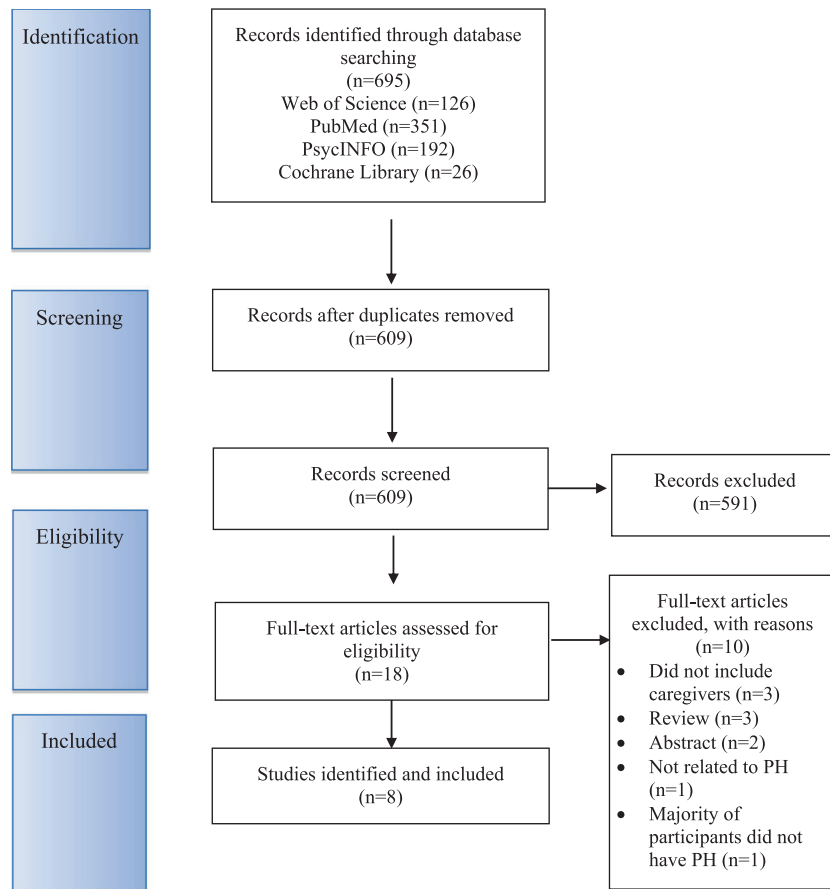
### Quality assessment

There was agreement that all studies met the two screening questions. Risk of bias varied from 2 to 5 (median 3.75/5). Participants were more likely to be recruited from hospital settings ( $n = 5$ ) than in the community ( $n = 3$ ). Studies rarely reported their sampling method or how many people were invited to take part and as such the level of bias is unknown. The use of participant's quotations and figures to represent findings in qualitative studies were limited.

### Categorical themes

Four categories were identified:

FIGURE 1 PRISMA diagram



## Caregivers' demographics

Spouses or partners living with the patient represented the greatest proportion of caregivers. Identified caregivers were typically in the fourth-sixth decade of life, with an even split in the sex of caregivers across studies. An association between carer demographics and difficulties encountered was observed. In one study involving 72 caregivers in Turkey, female caregivers reported a higher degree of burden than males ( $p = 0.01$ ) and people who were unemployed scored greater than those employed ( $p = 0.005$ ). Caregiver burden did not differ significantly ( $p = 0.81$ ) between spousal ( $n = 30$ ), children ( $n = 33$ ), or other caregivers ( $n = 9$ ). Greater caregiver burden was associated with worse HRQoL in patients ( $r = 0.39$ ,  $p < 0.003$ ); however, other factors such as the patient's age, PH class, 6-min walking distance, NT pro brain natriuretic peptide and systolic pulmonary arterial pressure were not significantly related ( $p > 0.05$ ).<sup>24</sup> Demographic and clinical factors of patients with PH who participants cared for are reported in Table 3.

## Caregivers' responsibilities

Most caregivers had provided care for several years. The amount of care ranged from 1 h per week to 24 h a day, with an average of 60.9/168 h per week.<sup>21</sup> Responsibilities differed greatly between interviewees; some provided *around the clock, hands-on care*, while others reported that their loved one retained a high degree of independence [19, pg. 40].

One study asked 35 caregivers in the United States to complete a non-PH specific questionnaire which investigated the type of support they provided in patients in WHO functional Classes I to III. On average, participants gave emotional support to patients most often, followed by treatment or practical care, motivational change, and finally personal care. Most caregivers felt that it was their role to help patients manage their anxiety and depression,<sup>21</sup> or were the recipients of patient's emotions.<sup>18</sup> Caring for their emotional needs could be challenging, as patients would often deny or avoid discussing their difficulties.<sup>18</sup> Caregivers also supported others with the impact of PH, with some spouses explaining that they were the one who told people about the patient's diagnosis.<sup>18</sup>

**TABLE 3** Summary of articles included

Primary author	Country	Aim	n	Caregiver	Patient	Research design	Standardised measures	Quality assessment
Giri <sup>16</sup>	US	To assess the effect of participating in support groups in those with PH and their caregiver.	41	Could not be ascertained	Could not be ascertained	Quantitative—online questionnaire	Patients HRQoL measured using emPHasis-10	2
Guillevin <sup>17</sup>	France (22%), Germany (9%), Italy (39%), Spain (24%) and the UK(6%)	To improve the understanding of patients' and carers' experience of living with PAH	129	Mean age 52, Female 56% 55% cared for a spouse or partner with PH, 21% a child, 18% parent, 6% family or friends.	Fifty-four percent cared for someone with idiopathic PAH, 14% PAH associated with CHD, 12% PAH associated with another disease, 10% PAH associated with CTD, 4% family history of PAH, 6% not sure. Class III 43%, II 31%, IV 23%, I 3%	Qualitative study on 1:1 basis and quantitative survey	-	2
Hall <sup>18</sup>	Canada	To describe the experiences of patients and their support persons adjusting to PAH and continuous intravenous epoprostenol	7	Age range 23–71 Female, 43% English Canadian, 57% French Canadian	N = 6 Female 83% Age range 43–70 French Canadian 83%, black Canadian 17% Time since diagnosis of PAH range 0.5–7 years Time since epoprostenol therapy 0.5–2 years	Qualitative—semi-structured interview with both patient and caregiver—constant comparison method	-	5
Hwang <sup>19</sup>	US	To examine depression, care tasks, support and impact of caregiving on family caregivers or patients with PAH.	35	Mean age 51.7 (range 18–72) 60% female 66% white, 20% Hispanic/Latino, 8% Black, 6% Asian 51% high school educated or less education Income = 26% <20k, 34% 20–75k, 29% >75k 63% currently married 68% spouse/partner to patient, 9% adult child, 9%	N = 35 Female 63% mean age 50.5 (range 20–72) 60% white, 20% Hispanic/Latino, 8% African American, 12% Asians/Pacific Islanders, 66% high school educated or less Income = 37% <20k, 32% 20–75k, 20% >75k 46% married 23% idiopathic PAH, 3% heritable PAH, 74% associated PAH 46% Class	Quantitative—structured questionnaires	Health Survey Short Form-36 v2, Patient Health Questionnaire-8, Caregiver Reaction Assessment, Dutch Objective Burden Inventory, Medical Outcome Study Social Support Survey	3

(Continues)

TABLE 3 (Continued)

Primary author	Country	Aim	n	Caregiver	Patient	Research design	Standardised measures	Quality assessment
				parent, 9% other 57% quit or reduced hours to provide care 14% lived with the patient months of providing care 51.7 (range 2–240) Caregiving hours per week 60.9 (1–168)	II, 54% Class III 31% continuous intravenous PAH therapy Epoprostenol or Treprostinil, 23% continuous oxygen therapy, Charlson comorbidity index = 57% 1–2, 29% 3–4, 14% <5, number of hospital admission in last 12 months = 1.7 (range 0–5), number of emergency department visits 1.5 (range 0–10). Days since last hospital discharge = 40% <3 months, 31% 3–12 months, 29% >12 months			
Ivarsson <sup>20</sup>	Sweden	To investigate spouse's experience of living with a partner with PAH or CTEPH.	14	All spouses 36% were female Median age 68 (range = 40–87) 50% retired/employed 29% had families with children 50% college/university education, 50% high school or elementary school	Patients lived with PH for 1–14 years.	Qualitative content analysis—interviews		5
Kocakaya <sup>21</sup>	Turkey	To evaluate the relationship between patients' QoL and caregiver burden	72 patient-care-giver dyad	Mean age 43.7 (min–max 20–70) 55.6% female 72.2% married, 11.1% never married, 11.1% widowed, 5.6% divorced 66.7% unemployed Primary school 30.6%, middle	N = 72 mean age 60.8 (min–max 19–84) 73.6% female, 3.2 years mean time of diagnosis (min–max 0.5–34 years), 45.8% Class II, 33.3% III, 20.8% IV, Mean NT-proBNP = 1644.5 Mean sPAP = 61.9 Mean 6-MWD = 277.6	Quantitative	Zarit Caregiver Burden Scale (patients' HRQoL measured using emPHasis-10)	3



TABLE 3 (Continued)

Primary author	Country	Aim	n	Caregiver	Patient	Research design	Standardised measures	Quality assessment
				school 19.4%, high school 26.4%, university 23.6%, 27.8% housewife/ husband, 22.2% retired, 18.1% freelance, 12.5% worder, 8.3% government worker, 2.8% student 41.7% cared for a spouse, 45.8% were children of patients, 12.5% other relation	CTEPH = 76.4%, CHD = 6.9%, IPAH = 5.6%, CTD = 5.6%, other = 5.6%			
Lichenstein <sup>22</sup>	PH Association	To gain an understanding of how caregivers of people with PH are using an online discussion board.	98	Forty-six percent mother, 3% father, 11% wife, 14% husband, 2% daughter, 1% son, 12% parent, 2% partner, 1% stepfather, 2% sibling, 5% offspring	N = 98 Female 60%, male 18%, 22% missing Mean age 15.8 years (range = 3–74) Medications: endothelin receptor antagonist 34%, phosphodiesterase type 5 inhibitors 31%, diuretics 10%	Data collected from PH discussion board—thematic analysis		5
Zhai <sup>23</sup>	China	To describe the impact of PAH on patients and their caregivers.	60	Mean age 41.8 Female 40.4%	Class 1 (2.4%) II (41.5%), III (51.2%), IC (4.9%)	Quantitative survey via semi-structured interview—descriptive statistics		3

Abbreviations: CHD, congenital heart disease; CTD, connective tissue disease; CTEPH, pulmonary arterial hypertension Congenital Heart Disease; HRQoL, Health-Related Quality of Life; N, number of participants; PAH, pulmonary arterial hypertension; QOL, Quality of Life.



Adhering to PH treatment regimens could be particularly challenging for caregivers with some explaining that they felt a *pressure to perform* [19, pg. 40], which increased their sense of burden, responsibility, and anxiety. One difficulty spoken about specifically, was the preparation and administration of medication as participants were concerned that if they did it incorrectly, it could endanger the patient's life.<sup>19</sup> Other duties included helping patients adhere to their diet and fluid intake, and exercise.<sup>21</sup>

Most participants described taking on more responsibility of household tasks, errands<sup>18,20,21</sup> and other physically demanding tasks. This was also reflected by the fact 85% of caregivers from China reported feeling exhausted more often.<sup>20</sup>

## Impact on caregivers

As in patients with PH,<sup>7</sup> caregivers were also affected by the condition long before the diagnosis was made.<sup>18,23</sup> Caregivers expressed a sense of fear, frustration and *uncertainty* [pg. 320] at the delay and need for countless investigations.<sup>23</sup> This was perpetuated by witnessing the patient suffer and deteriorate, often feeling powerless to help.<sup>18</sup>

Feelings of confusion, shock, fear and helplessness seemed common reactions to the diagnosis.<sup>18,23</sup> However, some also felt a sense of relief when patients were diagnosed, which appeared in response to having answers, and ending the ordeal of attending many investigations and the legitimacy of their needs not always being taken seriously.<sup>18</sup> Once diagnosed, caregivers expressed anxiety, fear and a sense of unpredictability when thinking about the future.<sup>18,22,22</sup> Some struggled to come to terms with the diagnosis or hoped that there was another possibility.<sup>23</sup>

Partners of patients were concerned about the impact on their children and worried how much they understood the diagnosis; as one parent explained: *Children so understand what we say...and probably understood that NN [patient] might die* [20, p. 4.]. Following the diagnosis, caregivers' concern soon shifted to treatment; for instance, whether the patient was receiving the most suitable treatment, possible side effects and adherence.<sup>18,23</sup>

One study in the United States asked 35 family caregivers to complete a standardised measure of depression, reporting 14% experienced moderate to severe levels.<sup>21</sup> The emotional impact was also reflected across cultures as: over one-fifth of caregivers from five European countries felt often or very often stressed;<sup>22</sup> 87.7% of caregivers in China reported feeling stressed and 58.9% that their health had suffered;<sup>20</sup> and in 72

caregivers from Turkey, 43% reported mild-moderate, 22% moderate-severe, and 1.4% severe levels of caregiver burden.<sup>24</sup>

Nearly one-fifth (23%) of spousal caregivers in Europe felt less close to the patient, and caregivers in Europe and China described feeling more of a caregiver than a lover.<sup>20,22</sup> Sexual intimacy had also declined for most caregivers.<sup>22</sup> This was linked to the patient's progressive symptoms, lack of interest and self-esteem, and caregiver's fears about making the patient more ill.<sup>20,22</sup>

Caregivers across cultures did report positive aspects of their role.<sup>18</sup> Over 90% of caregivers in Europe believed that they were contributing to the patient's HRQoL and nearly two-thirds felt their family had become closer.<sup>22</sup> Similarly, caregivers in the United States viewed their role as rewarding and a source of self-esteem.<sup>21</sup>

## Support for caregivers

In a study from the United States, the more support caregivers perceived themselves as having, the less symptoms of depression they reported ( $r = -0.5$ ,  $p = 0.002$ ). More specifically, people who had less emotional and informational support or positive social interactions, were more likely to experience depression. Overall, caregivers described social and affectionate support as being the most available, followed by tangible support and lastly emotional/informational support.<sup>21</sup>

This lack of information on PH and its impact was observed across samples.<sup>22,23</sup> For example, in China most caregivers described needing more information on a range of topics including patient organisation contacts (98.2%), the disease (98.2%), financial impact (94.2%), psychological consequences (90.5%) and treatment (83%).<sup>20</sup> Participants described wanting to be involved in patient's care, however, felt restricted over confidentiality and were concerned how their involvement may affect the patient's relationship with services.<sup>18</sup> Caregivers described feeling exasperated and had to find advice and support themselves. This was sought from disease-specific brochures produced by healthcare services, the internet, and other caregivers.<sup>18,23</sup>

PH organisation and discussion boards were considered to be of value to caregivers with one person writing in a group: *...I talk and think about...this website and how you all got me through (one of) the roughest times of our lives* [22, p. 586.]. Other benefits from engaging with organisations included gaining a better understanding of the disease and how to cope with it; support with managing the emotional impact; and discussing experiences which felt cathartic and less isolating.<sup>17,23</sup> Caregivers ( $n = 41$ ) recruited in the United States were asked

about their use of PH support groups with the majority (73%) having used them. Some caregivers were more likely to use PH support groups than others;<sup>17</sup> for example, in another study, nearly 50% of caregivers using an online discussion board were mothers caring for a child with PH.<sup>23</sup>

## Integrated themes

Four themes emerged:

### Change

PH had a considerable disruption to daily life,<sup>20,21</sup> with 57% of European caregivers feeling caring had a profound effect.<sup>22</sup> Among the changes commonly investigated, differences in interpersonal dynamics between caregivers and the patient (and others) were often described. Caregivers noted having to prioritise the needs of others above their own;<sup>22</sup> for example, planning, scheduling, changing, and adapting to the needs of the patient.<sup>20</sup> This was further reflected by the fact over two-thirds of participants in China felt they had lost control of their life.<sup>20</sup> Some participants found it difficult to achieve a balance between supporting patients to be independent and being overprotective<sup>18</sup>—or recognise that their own needs were not being met. It was common for caregiver to feel that they spent as much time caring for the patient as they did on themselves,<sup>22</sup> and most (57%) of caregivers experienced a lack of privacy.<sup>20</sup> Some described adjusting to their duties over time sometimes seeing their increasing list of responsibilities as a continuation of their role.<sup>19</sup>

### Preparedness

Caregivers described feeling underprepared and unskilled for some of their caregiving responsibilities and was not always related to how long they had been a caregiver, but associated with roles, for example, the challenges in administering medication. This theme was also related to a fear of failure and inadequacy, which was most notably reflected in participants' descriptions of feeling uncertain of how to help the patient;<sup>20,22</sup> as one caregiver explained: *I need some advice or maybe just an ear. I just don't know what to do to help* [22, p. 586,]. Caregivers questioned whether they could be doing more or be more helpful,<sup>18</sup> or felt they were not doing well enough,<sup>22</sup> with over two-thirds of caregivers in China feeling guilty.<sup>20</sup> Many participants were open about such

emotions and their difficulties. Some were proactive in seeking opportunities for further clarity and accessing information from various sources acknowledging that this made them more knowledgeable, with the hope of feeling better skilled or prepared.<sup>23</sup>

### Isolation

Caregivers voiced feeling isolated and lonely,<sup>17,22</sup> which appeared to have a considerable impact on their social and emotional wellbeing. Nearly two-thirds (63.8%) of participants felt caregiving affected how much time they spent with others.<sup>20</sup> Participants described feeling that their social systems reduced overtime, which for some was influenced by the nature of their caring responsibilities and deterioration in the patients' ability.<sup>18</sup> Caregivers for whom the patient was their main source of support<sup>19</sup> or those who were under more financial pressure (possibly making social resources limited) were more vulnerable to isolation.<sup>18</sup>

Isolation was linked by caregivers to the rarity and invisibility of PH.<sup>20,22</sup> Caregivers found it difficult to relate to others noting that they did not understand what they were experiencing<sup>18</sup> or lacked any knowledge of the disease.<sup>20</sup> Caregivers looked for other people with similar stories describing PH organisations as important sources of support, to create a sense of camaraderie,<sup>23</sup> and feeling less isolated.<sup>17</sup> One carer explained: *I found myself on the internet, in this PAH forum...there is a need to connect with others with the same family situation as us* [2, p. 4].

### Physical and mental demands

Caregivers noted being asked about or more commonly provided information on the physical and practical impact of PH—rather than the wider and emotional effects.<sup>22</sup> Despite caregivers in the United States who completed a health survey demonstrating their physical and mental wellbeing were impacted equally.<sup>21</sup> Caring for someone with PH seemed to be associated with the patient always being on their carer's mind.<sup>19,21</sup> Caregivers frequently found it difficult to switch off or have a break from their duties as they always needed to be available.<sup>23</sup> Some caregivers' descriptions reflected the feeling that they had no choice to be a caregiver whilst others struggled to find fulfilment<sup>22</sup> and voiced concerns about being unsure how long they could continue.<sup>20</sup>

Some caregivers described the importance of having time to engage in self-care and other meaningful activities, with one participant explaining: *I have a large need to take care of myself* [20, p.5]. Caregivers described

talking to others as an important source of support, in particular close friends, family, and those familiar with PH—as well as using their own spirituality to cope.<sup>17,23</sup> However, not everyone had someone to turn to<sup>21</sup> or they found it difficult to express themselves.<sup>20</sup> Patients with PH reported a greater number of benefits from engaging in PH organisations than caregivers, which may suggest that services are more focused on patients' needs.<sup>17</sup>

Figure 2 shows the relationship between categorical and integrated themes and how they contribute to caregiver burden.

## DISCUSSION

This is the first systematic review investigating the impact of caring for someone with PH. Caregivers described a range of psychological, emotional, physical, relational, financial, and social difficulties associated with their role, which affected their day-to-day life in addition to having a long-term impact. They experienced high levels of caregiver burden, which may also reflect the duration and intensity of their roles, in addition to the nature and perceived pressure of their responsibilities. Caregivers described a lack of support, information and advice, which contributed to thoughts and feelings of depression, anxiety, uncertainty and under-preparedness.

Finding ways to best support caregivers is important. Despite several authors making recommendations for clinical interventions based on their findings, no evidence has been gathered assessing the effectiveness

of specific interventions for this group. This is particularly concerning given the high rates of emotional and psychological difficulties reported by caregivers. The various challenges associated with caring for someone with PH means a range of interventions targeting different needs would most likely be beneficial. Recommendations for supporting caregivers of patients with other chronic conditions include education, helping to develop problems solving skills and coping strategies, support from healthcare workers, developing social goals, and medication, such as anxiolytics and antidepressants. Moreover, interventions aimed at caregivers have been most effective when tailored to the specific needs of individuals.<sup>16</sup> As such, it is crucial for support to reflect PH-specific issues specifically those that are not routinely seen in other conditions.

In terms of providing support for caregivers, several considerations can be made based on the current results. There seems to be a need for care to be provided by different services, including health and social care and PH organisations. This may help to overcome numerous barriers associated with caregivers accessing some services. Given the intensity of caring for someone with PH with some patients requiring care 24 h a day, any intervention needs to be easily accessible and flexible towards carer's availability. Self-help interventions could serve this role in addition to helping to overcome challenges to engagement, such as caregivers finding it difficult to express themselves. However, it is important to recognise other approaches are also needed; for example, opportunities for social support providing a

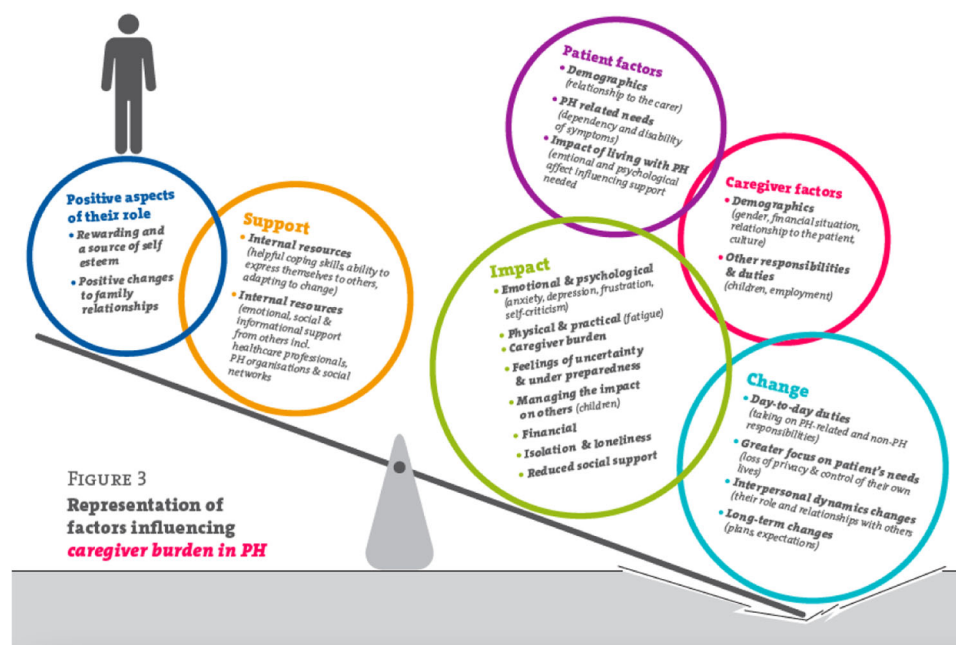


FIGURE 3  
Representation of  
factors influencing  
caregiver burden in PH

FIGURE 2 Representation of factors influencing caregiver burden in PH. PH, pulmonary hypertension

safe and compassionate space will be helpful for many participants—especially given the isolating nature of PH. Finally, it is crucial to recognise the heterogeneity of caregivers and provide tailored information and advice; for instance, caregivers differed in their roles and responsibilities, relationship to the patient or personal circumstance.

The lack of information on different aspects of living with PH was common across samples. This was shown to have a negative emotional impact on caregivers, but evidence elsewhere suggests it is likely to also affect patients. For example, coping strategies have shown to moderate the relationship between depression and health anxiety and HRQoL in patients with PH. More specifically, having access to information and emotional support helped to buffer the effects of psychological distress on HRQoL.<sup>25</sup> Further research is required to examine the relationship between caregiver burden on patients with PH, which will help to identify therapeutic targets for both groups.

All studies reviewed utilised a cross-sectional research design, therefore we do not know how caregiving for someone with PH changes over time. Moreover, results comparing factors (i.e., degree of burden) across subgroups of caregivers should be interpreted with caution as due to the small size of groups, any comparisons may be underpowered and covariate adjustments could not be performed. Finally, caregivers were not stratified based on the duration of how long they had been providing care or the nature of the patient's PH. This is particularly important given the progressive nature of PH. For example, in the qualitative synthesis of patients' accounts discussed previously,<sup>7</sup> a theme that emerged was the transitional nature of PH in which, initially individuals had to make sense of their diagnosis, after which it seemed participants worked to find a balance between being impacted by their symptoms and living their life. Indeed, it is likely the difficulties that caregivers experience—and possible therapeutic needs—will overlap with the stage of PH and time since diagnosis.<sup>26</sup>

Voices of some caregivers have not been heard in the published data as most interviewees were white, middle-aged, and living in Western countries. Indeed, we know from research examining ethnic differences in caregivers of people with other conditions that some groups are disproportionately affected, as caregivers from minority groups differ from white caregivers on the intrapersonal, interpersonal and environmental level.<sup>27</sup> In the current literature on PH examined here, any cross-cultural comparisons should be interpreted with caution given the variation in samples, how data was collected and differences in healthcare provision. However, it would

make sense for some to be more affected than others. For example, while the financial impact of PH was prevalent,<sup>21,22</sup> across eight major cities in China, low-income families were most impacted.<sup>20</sup> Financial pressures were associated with patients and caregivers having to reduce or leave work,<sup>21</sup> and costs of PH care—particularly for caregivers living in countries where costs are not covered by healthcare services.<sup>18</sup> This left some households below the average income,<sup>22</sup> further perpetuating or predisposing them to additional difficulties. For instance, over 50% of patients interviewed in China could not afford treatment.<sup>20</sup>

In conclusion, while there is growing evidence examining the impact of PH on patients, there is a paucity of research examining how it affects caregivers—with even less evidence examining interventions aimed at supporting this group. The findings demonstrate the multifaceted impact caregiving can have on individuals, thus suggesting multicomponent interventions are most likely to be helpful. More research is needed to investigate factors related to caregiver burden; this evidence may inform the development of treatments aimed at supporting caregivers and patients.

## AUTHOR CONTRIBUTIONS

Gregg H. Rawlings 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. Nigel Beail made contributions to data analysis including second rating articles, and provided feedback on the final report. He approved the final version for publication. Robin Condliffe made contributions to data analysis, analysis and provided feedback on the final report. He approved the final version for publication. David G. Kiely made contributions to data analysis and provided feedback on the final report. He approved the final version for publication. Andrew R. Thompson made contributions to data analysis and provided feedback on the final report. He approved the final version for publication. Ian Sabroe made contributions to data collection, analysis and provided feedback on the final report. He approved the final version for publication. Iain Armstrong made contributions to data analysis and provided feedback on the final report. He approved the final version for publication.

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## CONFLICTS OF INTEREST

GHR, NB, ART, RC, and IA have no conflicts of interest to report. DGK has received honoraria from Actelion Bayer, GSK and MSD for participation as a speaker and consultancy work and his unit has received research and unrestricted educational grants from Actelion and GSK. IS's department receives an unrestricted educational grant from GSK for an annual clinical education meeting.

## ETHICS STATEMENT

Ethical approval was not required for this study.

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