A theory- and evidence-based approach to developing and piloting a cognitive-behavioural self-help intervention for anxiety in adults living with pulmonary hypertension

Gregg Harry Rawlings

A thesis submitted in partial fulfilment of the requirements for the award of Doctor of Clinical Psychology at the University of Sheffield

The University of Sheffield
Faculty of Science Clinical Psychology Unit
Department/School of Psychology

Submission Date: May 2021

Reserved for Access to Thesis form

Reserved for Access to Thesis form

Declaration

I, the author, confirm that the Thesis is my own work. I am aware of the University's Guidance on the Use of Unfair Means (www.sheffield.ac.uk/ssid/unfair-means). This work has not been previously been presented for an award at this, or any other, university.

Publications arising from the thesis:

Rawlings, G. H., Beail, N., Armstrong, I., Condliffe, R., Kiely, D.G., Sabroe, I., & Thompson, A.R. (2020). Adults' experiences of living with pulmonary hypertension: A thematic synthesis of qualitative studies. *BMJ Open,* 10, Article e041428. doi: 10.1136/bmjopen-2020-041428

Rawlings, G.H., Thompson, A.R., Armstrong, I., Kiely, D.G., & Beail, N. (in press). Development and pilot randomised controlled trial of a self-management intervention to help individuals with pulmonary hypertension live better with anxiety [conference abstract]. *Pulmonary Circulation*.

Funding

Costs associated with this project were covered by the Pulmonary Hypertension Association UK (https://www.phauk.org). The Open Access Fee (£1500 plus VAT) for the article entitled, "*Adults' experiences of living with pulmonary hypertension: A thematic synthesis of qualitative studies*" was kindly reduced by 20% by BMJ Open. The remaining fee was very kindly paid for by the Pulmonary Hypertension Association, UK and a personal research grant from Professor David G. Kiely.

Structure and Word Count

Section One: Literature Review

Excluding references and tables	7917
Including references and tables	12243

Section Two: Research Report

Excluding references and tables	8000
Including references and tables	13592

Total Word Count

Excluding references and tables	15917
Including references and tables	25835

Lay Summary

Pulmonary hypertension (PH) is a progressive and life-shortening cardiovascular disease associated with an increased risk of lower health-related quality of life (HRQoL) and psychological difficulties, including depression and anxiety. Anxiety in particular is an important issue given the prevalence, overlapping symptoms between anxiety and PH, and the physiological link between anxiety and the increased load on cardiovascular functioning.

While anxiety is a normal response to distressing stimuli, it can become unhelpful and impact on daily life. There is limited research examining treatments for anxiety in PH; however, evidence demonstrates the benefits of using a form of psychological therapy known as Cognitive Behavioural Therapy (CBT). CBT aims to help people change their attitudes, beliefs and actions. CBT is recommended by the National Institute for Health and Care Excellence for anxiety and panic disorders using a stepped care model, initially providing self-help interventions progressing to more intensive treatments.

The objective of the current thesis was to develop and investigate a self-help intervention for anxiety in adults with PH. Development of the intervention was guided by the Medical Research Council Framework for developing complex healthcare interventions. This involves four phases consisting of developing, piloting, evaluating and implementing the intervention. More specifically, the initial phase consists of identifying the evidence-base to inform intervention development. Therefore, a thematic synthesis was first undertaken of qualitative studies investigating adults' experiences of living with PH. In total, the data from 19 studies involving over 1900 adults from four continents were included. Studies generally were of high quality. Four analytical themes emerged reflecting the many uncertainties associated with the disease; that PH is often experienced somatically and misunderstood by others; participants' frustration and upset at the lack of awareness of PH; and the transitional

nature of life with PH. Results highlighted the numerous challenges that this clinical group experience and, in particular, the link between PH and anxiety.

A four-week self-help intervention was then developed based on the available literature and its acceptability, feasibility and preliminary effectiveness explored using a pilot randomised control trial (RCT). Individuals with PH self-reporting difficulties with anxiety were recruited from Pulmonary Hypertension Associations. Participants were randomised to the intervention (n=37) or a wait-list group (n=40). Participants were asked to complete a series of health-related measures before and after the intervention, and at one-month follow-up. Measures assessed anxiety, depression, HRQoL, dyspnoea, self-mastery and mood-related cognitions and behaviours. Participants in the intervention group were contacted part-way through and at the end of the study to investigate intervention adherence and acceptability. Overall 65/77 of participants completed the study. All participants in the self-help group agreed that the intervention was helpful. Compared to those in the control condition, participants reported a significant reduction in anxiety, depression and mood-related cognitions and behaviours. The relationship between intervention condition and change in anxiety and depression were mediated by changes in cognitions and behaviours, suggesting a mechanism of change. The intervention was acceptable and feasible. Findings support the next phase in the framework involving a definitive RCT to evaluate the intervention.

Acknowledgments

I would like to thank my primary supervisors, Professor Nigel Beail, Professor Andrew R. Thompson and Dr Iain Armstrong for their guidance and feedback throughout this thesis; it has been a pleasure working with you. I would like to thank Pulmonary Hypertension Association UK (PHA UK) for their support and also for funding the project and publishing fees; Shaun Clayton, Helen Crabtree, Mary Ferguson, Paul Sephton and John Smith, without your dedication, passion and creativity this project would not have been possible. I am also extremely grateful to Dr Robin Condliffe, Professor David Kiely and Professor Ian Sabroe for sharing their expertise, and also to Professor Kiely for funding publishing fees. I would like to thank members of the stakeholders' team for sharing their wisdom and insight; you have helped make this research far greater. I am appreciative of Pulmonary Hypertension Associations around the world for sharing the research project with their members. I would also like to thank all of the participants who taken part for their time and allowing me to learn from their experiences.

Finally and on a personal note, I am grateful for all the support and encouragement from my mam and dad, my partner, Barbora, and my friends.

Table of Contents

Page(s)

Declaration 4
Structure and Word Count 5
Lay Summary 6-7
Acknowledgements 8

Section One: Literature Review

Title 11
Abstract 12-13
Introduction 14-17
Methods 18-23
Results 24-45
Discussion 45-49
References 50-62

Appendices

Appendix A – PRISMA Standards Checklists 63-66
Appendix B - ENTREQ statement 67
Appendix C – Registration in Review Database 68-71
Appendix D – Published Manuscript 72-84
Appendix E - Critical Appraisal Skills Programme Checklist 85-93
Appendix F – List of Excluded Studies 94-102

Section Two: Research Report

Title 103
Abstract 104-105
Introduction 106-111
Methods 111-126
Results 126-149
Discussion 149-155
References 156-166

Appendices

Appendix A – Stakeholder's Questionnaire 167-168
Appendix B - Self-Help Intervention 169-195
Appendix C – CONSORT Guidelines Checklist 196-198
Appendix D – Ethical Approval Letter 199-200
Appendix E – Research Governance Sponsor Letter 201-202
Appendix F – Trial Registration Form 203
Appendix G – Consent Form 204-205
Appendix H – Screening Questionnaire 206
Appendix I – Study Advert Example 207
Appendix J – Participant Information Sheet 208-211
Appendix K– Adherence and Acceptability Check-In Questionnaire 212-213
Appendix L – Final Acceptability Questionnaire 214-216

Appendix M – Demographic Questionnaire 217
Appendix N – Generalised Anxiety Disorder-7 Questionnaire 218
Appendix O – Patient Health Questionnaire -9 219-220
Appendix P – emPHasis10 - Health-Related Quality of Life Measure 221
Appendix Q – Dyspnoea 12 questionnaire 222
Appendix R – Self-Mastery Scale 223
Appendix S – Cognitive and Behavioural Processes Questionnaire 224-228

Section One: Literature Review

Adults’ experiences of living with pulmonary hypertension: A thematic synthesis and systematic review of qualitative studies

Abstract

Objectives: Pulmonary hypertension (PH) is a life-shortening disease that has a considerable impact on quality of life. This review aims to synthesise the published qualitative research that has listened to adults (≥18 years old) discuss their experiences of living with PH. Understanding how individuals are affected and cope with the disease can help to improve services and treatment outcomes.

Method: 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 subject to a thematic synthesis.

Results: Nineteen articles were identified reflecting the experiences of over 1900 individuals. 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 PH. Four higher order analytical themes were developed from the descriptive themes, reflecting: (a) uncertainties encountered related to PH; (b) the somatic nature and lack of recognition of the impact of the disease; (c) frustration at the paucity of awareness of PH; and (d), participant's accounts of transitioning through stages of living with PH.

Conclusions: These findings form the first synthesis of experiences of life in individuals with PH illustrating the multifaceted impact of the condition. Further research is required to examine differences in accounts related to severity of the disease and underrepresented groups. The results have implications for clinical practice, emphasising the role of educational and psychological therapies.

Practitioner Points

- Examining individual's experiences of living with pulmonary hypertension can help to identify important therapeutic targets.
- Helping individuals with pulmonary hypertension develop strategies to better manage distress associated with uncertainty and anxiety are needed.
- People with pulmonary hypertension may focus on somatic sensations to monitor progression of the disease. As such, helping individuals to disengage their attention and develop strategies to differentiate between anxiety and pulmonary hypertension-related symptoms are likely to be beneficial.
- Pulmonary hypertension care should be responsive to the multifaceted and transitional nature of the disease, further highlighting the need for multidisciplinary informed treatments.

Key words

Pulmonary Arterial Hypertension; Anxiety; Quality of Life; Phenomenology; Thematic

Adults' experiences of living with pulmonary hypertension: A thematic synthesis and systematic review of qualitative studies

Pulmonary hypertension (PH) describes a group of serious conditions characterised by elevated pulmonary artery pressure, which if untreated results in right heart failure and premature death (Hoeper et al., 2013; Hoeper & Humbert, 2019). The most recent guidelines define PH as a mean pulmonary arterial pressure of ≥20 mmHg at rest (Thomas et al., 2020).

In PH, the arteries that supply blood to the lungs, known as the pulmonary arteries, become constricted disrupting blood flow. It is essential that blood travels to the lungs to pick up oxygen, which is then transported around the body to organs, muscles and other body tissue. The restriction observed in PH places an additional burden on the heart, which over time becomes less efficient, resulting in the progression of cardiorespiratory and other PH-related symptoms (Lai et al., 2014). Five groups of PH 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 (Hoeper et al., 2017).

PH is not common, but nor is it rare, for example the prevalence of PAH ranges from 10-52 individuals per one million (Marius & Gibbs, 2014). Anyone can develop PH however it is more common in females (Rich et al., 1987) and those who are middle aged (in the third or fourth decade of life) (Frost et al., 2011). Research is less clear on the prevalence of PH as a result of other demographic factors, including ethnicity or economic status (Frost et al., 2011).

The cause of PH can be idiopathic, related to hereditary factors, drug induced or secondary to comorbid disorders, such as other heart conditions, connective tissue disease and infection, including human immunodeficiency virus (HIV) (Hambly et al., 2016). The disease is associated with high mortality with long-term survival rates of 87.9% at one year, 72.5% at five years and 62.6% at ten years (Chang et al., 2016). Early diagnosis and appropriate treatment is vital; however, due to difficulties in differentiating between PH and other conditions, individuals can experience a delay of several years by which point, the disease may have progressed to a more advanced stage (Brown et al., 2011). It is recognised that PH care should be patient-centred and guided by the form and severity of the condition, and needs of the individual patient. Treatments can include medication, oxygen therapy, physiotherapy and psychological therapy – and if suitable, surgery. Interventions largely aim to stabilise the disease, halting its progression and maximise health-related quality of life (HRQoL) (Hoeper et al., 2017; Kiely et al., 2013).

The World Health Organisation (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 (McGoon et al., 2004). The cardinal symptom of PH is dyspnoea (breathing difficulties), although syncope, pain, edema, fatigue, irregular heart rate and palpitations, and sleep difficulties are common (Hoeper et al., 2017; Yorke et al., 2018). Patients can also exhibit cognitive difficulties associated with PH and side-effects of treatment, such as impaired memory and attention (White et al., 2006). The difficulties that patients experience can also be related to their life-stage or progression and nature of the disease. For instance, younger people may encounter challenges concerning family planning or childcare - as women are advised against pregnancy in PH due to the risks posed to the mother and child (Hemnes et al., 2015) - while those with severe PH may be receiving end of life support for the disease (Khirfan et al., 2018).

Despite improved outcomes in PH-related mortality associated with advancements in medical therapies, including the formation of designated PH centres providing specialist care (Galiè et al., 2016; Hoeper et al., 2017; Kiely et al., 2013), it remains a life-shortening condition with significant physical and psychological morbidity and a profound impact on HRQoL (Kiely et al., 2013). Indeed, research has explored patient's experiences of living with the disease (Keen et al., 2018; McGoon et al., 2019), demonstrating the importance of psychosocial factors in PH on HRQoL (Cole et al., 2016; Delcroix & Howard, 2015; Halimi et al., 2018) and engagement in care (Graarup et al., 2016). A greater understanding of how individuals are affected and cope with PH should help to improve treatment pathways and patient-reported outcomes (McGoon et al., 2019).

How an individual perceives, experiences and appraises different aspects of their illness can impact how they cope and self-reported outcomes (Leventhal et al., 2003). Coping has been conceptualised as cognitive and behavioural responses used by an individual with the aim of dealing with an internal or external stressor, which is subjectively perceived as threatening, distressing or uncomfortable (Lazarus & Folkman, 1984). As in other chronic and incurable conditions (Turner & Kelly, 2000), coping in PH is an important area of investigation: (a) given the increased risk of individuals experiencing many challenges linked to the disease (Wryobeck et al., 2007); (b) the association between coping strategies and psychological problems, such as anxiety and depression, and reduced HRQoL (Halimi et al., 2018; Rawlings et al., 2021); and (c) the psychological and physiological impact of distress, which has been shown to increase vasoconstriction in people with PH (Schachinger et al., 2000).

The aim of the current review was to systemically synthesise the published qualitative evidence examining adults' experiences of PH. The majority of researchers in this area have employed quantitative methodologies to explore experiences of living with PH (McGoon et al., 2019). These studies have investigated a specific difficulty, such as breathlessness, fatigue or mood, by asking individuals to answer a series of questions and selecting a structured response (Bonner et al., 2013; McKenna et al., 2006; Yorke et al., 2014). While this has produced large generalisable datasets, participants may not have been able to elaborate on their answer or discuss other, and potentially more, challenging difficulties. Qualitative methods allow individuals to discuss their experiences in their own words, meaning rich and fine-grained data can be collected. Moreover, a more-in-depth and nuanced understanding of experiences can be obtained, which can highlight any unmet needs and identify new areas of investigation (Harper & Thompson, 2012). Indeed, the current findings were used to help recognise the need and guide the development of a self-help intervention for individuals with PH to manage anxiety. The number of systematic synthesises of qualitative research investigating topics in healthcare are growing (Tong et al., 2012) and have already helped to examine the phenomenology of other cardiovascular conditions, such as chronic obstructive pulmonary disease (Russell et al., 2018) and stroke (McKevitt et al., 2004).

Methods

Search Strategy

This systematic review has been conducted in accordance with PRISMA (Page et al., 2021) and ENTREQ statements (Tong et al., 2012) (Appendix A and B). The protocol was registered at the Open Science Framework – DOI 10.17605/OSF.IO/TYNJD (Appendix C). A comprehensive search of four databases - Web of Science, PubMed, PsycINFO and Cochrane Library - was performed by the lead researcher in January 2020, which was repeated in May 2020 prior to submission for publication in the BMJ Open (Appendix D) (Rawlings et al., 2020). Search terms were informed by the authors' experience of previously published articles on PH. The term "pulmonary hypertension" (opposed to describing all of the five groups of PH) was used to remain inclusive as this was likely to also identify other groups of PH. Key words of suitable articles were checked to confirm salient words had not been omitted (Table 1).

Table 1

Search terms and boolean operators for all databases

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*"

The titles and abstracts of all articles identified by the systematic search were first screened by the lead researcher using the criteria presented in Table 2. Papers that met the criteria or when there was some ambiguity over eligibility, were subjected to a full-text review by the lead researcher. The references of suitable articles were searched for relevant studies, which helped to identify one additional article (Lo et al., 2019).

Table 2

Inclusion and exclusion criteria – based on the SPIDER tool (Cooke et al., 2012)

	Inclusion	Exclusion
Sample	Adults (18 years or older) diagnosed with PH; participants who self-reported being impacted by a diagnosis of PH. It was assumed participants were adults unless otherwise stated.	Children (<18 years old), individuals without a diagnosis of PH.
Phenomenon of interest	Exploring individual's experiences of PH i.e. diagnosis, treatment, psychosocial impact.	Exploring other cardiovascular conditions. Using qualitative methodology to address a quantitative research goal i.e. development of a quantitative clinical measure.
Design	Any study that reported utilising a qualitative research methodology i.e. open-ended question was considered. If participants with PH were grouped with individuals with a different diagnosis, their unique contribution must have been made explicit.	Solely quantitative research methodologies or converted experiences to numbers i.e. standardised psychometric measures. Participants with PH grouped with other individuals without PH and it was not clear from whom data was collected from.
Evaluation	Any form of qualitative analysis that aimed to report on subjective experiences i.e. 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 i.e. books, abstracts, blogs, unpublished studies, reviews.

PH = Pulmonary Hypertension

Quality Assessment

Relevant studies were evaluated using the ten-item Critical Appraisal Skills Programme for qualitative research (Critical Appraisal Skills Programme, 2018). All research papers were assessed whether the item was fulfilled (+) or not (-) (see Table 4 and Appendix E). Each article was given an overall score demonstrating that the study had achieved the majority or all (++), most (+) or few (-) of the criteria. This approach is in line with other qualitative reviews (Rawlings et al., 2019; Wray & Clarke, 2017). The lead author (GHR) assessed all articles. A second researcher (CG; a trainee clinical psychologist at the University of Sheffield) independently re-assessed approximately one-quarter of the papers (n=5) chosen as random. Following discussion, a 94% level of agreement per study was observed for all eleven-items per study (55 in total), compared to an 87% level of agreement prior to discussion. All findings, regardless of quality, were included in the synthesis to remain inclusive. The quality of papers was assessed to further help evaluate the evidence and for any interpretations to be made in light of its source.

Data Analysis

A range of approaches have been proposed to synthesis qualitative data with no general agreement on the most suitable method (Tong et al., 2012). Instead, methodology should be guided by the aim and question of the review (Dixon-Woods et al., 2005). Given that the current objective was to gain a greater understanding of the impact of PH and how people cope, and to help identify any unmet needs and possible therapeutic targets, a thematic synthesis was used to analyse the data (Thomas & Harden, 2008). This approach aims to develop higher order themes transparently, as authors explicitly report and distinguish between descriptive and analytical findings. Moreover, the novel analytical themes are designed to help inform future interventions and research, which is particularly important given that the current findings were used to guide the development of a self-help intervention for anxiety disorders in adults with PH.

While reflexivity is an important stage in empirical qualitative research, it is not discussed in the context of qualitative reviews; for instance, PRISMA and ENTREQ statements do not suggest that this process is required. Reflexivity involves the researcher attending to the wider content of knowledge acquisition and how the data was collection, for example, reflecting on the potential role of relationships between participants and themselves in their investigation (Dodgson, 2019). Given that the data were collected using a systematic method, the role of bias in data collection was low. Nevertheless, biases may have influenced data analysis and interpretations. Notwithstanding research statements, details has been reported below to provide additional information regarding data analysis. A four-staged approach to analysis was utilised:

1. The lead researcher (GHR) read each paper several times and extracted relevant information into Microsoft Word, which included authors, date, country, participant information, data collection and analysis methodologies, results, discussion and conclusions. The researcher was a trainee clinical psychologist with previous experience of having conducted primary and secondary qualitative research. The researcher aimed to be inclusive at this stage utilising an inductive process, which was not consciously guided by pre-existing theories. Data were collected regardless of whether saturation was achieved. During this process, the researcher was unaware that the findings

would be used to help inform the development of an anxiety-specific intervention.

2. Findings were coded by the lead researcher with the aim of developing descriptive themes. Themes were informed by the frequency and saliency of codes, as opposed to study characteristics (e.g., sample size). This was an iterative and data driven process, as the researcher worked closely with the data. Attention was given to n=1 or negative cases (i.e., accounts that contradict recurrent or strong narratives) with the aim of helping to provide alternative viewpoints, improve representativeness of the findings and reduce bias (Birks et al., 2014). The codes were then grouped thematically based on their similarities and differences, to create themes. Towards the end of this stage, anxiety was recognised as a prevalent issue in PH, which was discussed with the wider research team with the aim of developing a self-help intervention targeting anxiety disorders in this clinical group. However, the content of this work was not actively used to inform the production of findings during this process. Themes reported have been ordered as per participants' journey from diagnosis and interactions with healthcare professionals, to impact and coping.
3. Analytical themes were developed, which aimed to go beyond the original data and descriptive themes in context of the review's objective. This process is viewed as a defining feature of this approach. At this stage, themes were discussed amongst other members of the research team for further refinement. The lead researcher, who was most familiar with the raw data, first interpreted the findings before sharing the themes with members of the research team. The aim of this was to draw on the experience of researchers with extensive

qualitative experience and specialist PH clinicians to help consider reliability, application and alternative perspectives regarding the results. While the lead researcher had pre-existing specialist knowledge of anxiety disorders and treatments, which helped to guide the interpretation of the analytical themes given the saliency of anxiety in PH, a hybrid method was utilised based on a deductive and inductive approach. The order of themes is consistent with how they are reflected in the descriptive themes i.e., uncertainty was expressed first in response to symptom onset.

4. The lead researcher read each paper one final time to confirm that the findings were represented in the emergent themes. Moreover, the researcher aimed to identify evidence that contradicts the themes with the aim of assessing an important question concerning whether or not the findings had been influenced or shaped by the research teams' interest in developing an intervention for anxiety in PH. The researcher was satisfied that the main themes had captured participants' accounts and equal importance was given to the different descriptive and analytical themes. The final report was written, and quotations were identified. "" have been used when reporting participants words and ' ' for authors.

Patient Public Involvement

The review was developed with experts in PH and professionals from Pulmonary Hypertension Association, UK. There was no other 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 2005–2020; however, 18/19 were published in the last ten years (2010-2020) (Flattery et al., 2005). Data reflects over 1900 individuals across Europe, North and South America, and Asia. Ages ranged from 19-91 years old. In all but one study, the sample were predominantly female (Goddard et al., 2017) – likely reflecting the gender bias observed in PH (Hoeper & Gibbs, 2014). Only three studies provided sufficient detail regarding sample ethnicity (Carroll et al., 2012; Lo et al., 2019; McDonough et al., 2011). 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 because, as discussed later, experiences seemed to differ depending on condition-specific issues (Lo et al., 2019; Muntingh et al., 2017). The majority (n=14) of studies used research interviews for data collection, whereas a greater variation of analytical methods were utilised (Table 3).

Figure 1

PRISMA diagram (see Appendix F for list of excluded studies)

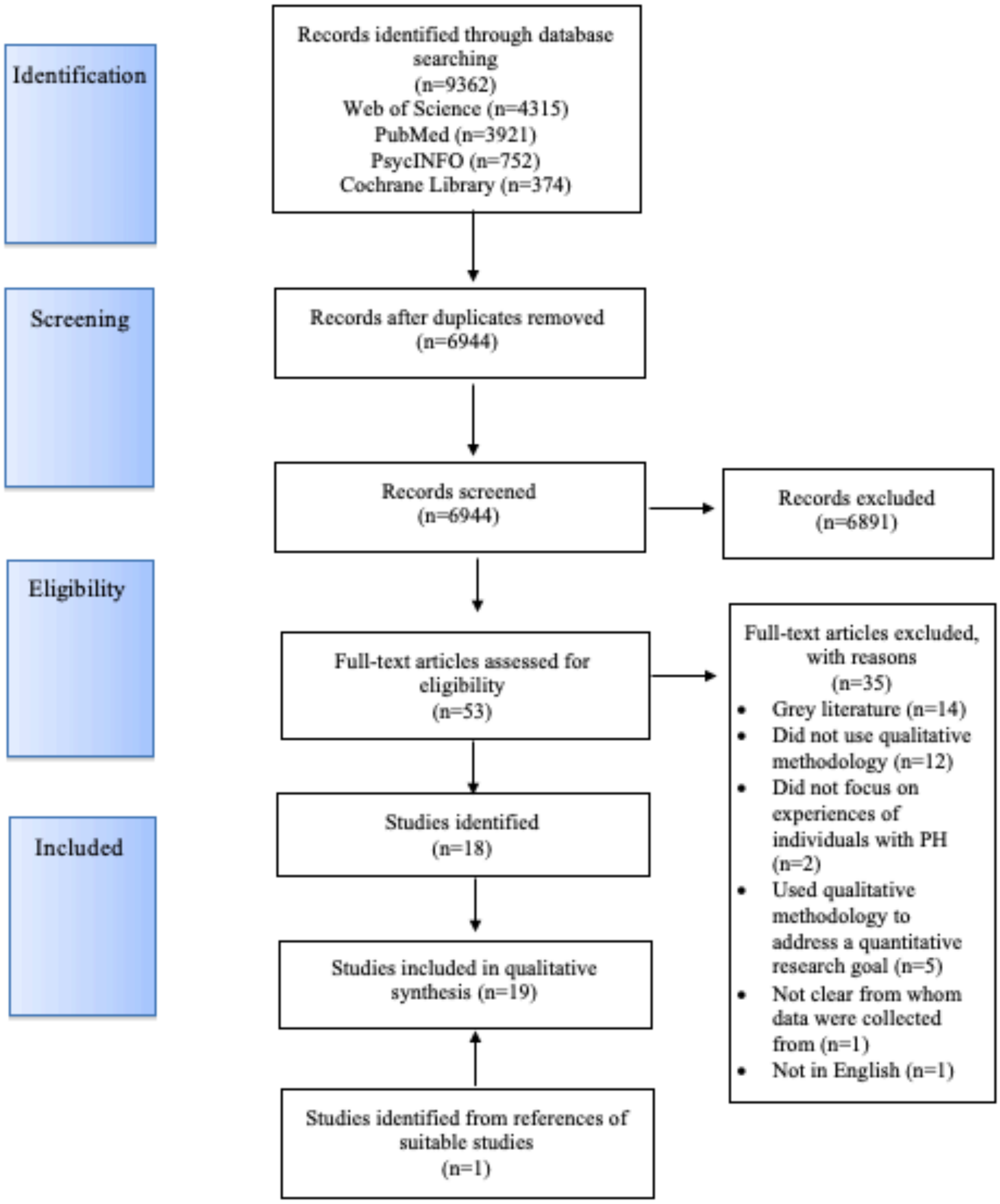


Table 3

*Summary of studies; *, +, ^ same sample investigated across studies*

Primary author and year	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 (2016)	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 (2012)*	UK	To investigate participant's experiences of being diagnosed with PH.	30	60	56.3 26-80	NR	<1 to >10	Group 1 PAH (86%) 37%=idiopathic PAH 23%=connective tissue disease-PAH 17%=congenital heart disease-PAH 3% =portopulmonary hypertension 3%=heritable PAH 3%=drugs/toxins-PAH	I=0% II=30% III=63% IV=7%	Semi-structured interviews; Thematic analysis

								Group 3 PH-lung disease (3%) Group 4 CTEPH (4%) NR (3%)		
Carroll (2012)	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 (2018)	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
Flattery (2005)	USA	To describe patients experiences of PAH and explore the impact.	11	73	58.5 40-72	“most were white” (pg. 101)	3.5 0.5-14	Group 1 PAH 55% = idiopathic PAH 18% = systemic lupus erythematosis-PAH 9% = mixed connective tissue disease	NR	Semi-structured interviews; Colaizzi’s approach to phenomenological analysis

								9% = portopulmonary hypertension; 9% = other PAH Not specified (45%)		
Goddard (2017)	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 (2012)	Canada	To describe patients 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 (2014)+	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-PAH 12%=PAH Group 4 CTEPH (29%) 11% treated by pulmonary endarterectomy	NR	Semi-structured interviews; Content analysis
Ivarsson (2016)+	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	NR	Semi-structured interviews; Content analysis

								24%= systemic sclerosis-PAH 12%=PAH Group 4 CTEPH (29%) 11% treated by pulmonary endarterectomy		
Keen (2018)	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
Kingman (2014)	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 (2019)	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 2018^	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	6.3 0.5-24	Group 1PAH (93%) 40% PAH associated with CTD	I=10% II=60% III=23% IV=7%	Semi-structured interview; thematic analysis

						, 13% other		Group 2 PH Left Heart Disease (7%)		
Matura (2013)	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 was collected from online a discussion board posted by individuals with PH; Qualitative descriptive methodology
Martin (2020)^	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 (2011)	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
Muntingh (2017)	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 (2019)	Germany	To explore burden of living with a	4	NR	NR	NR	NR	Group 1 PAH (100%)	NR	Focus groups; Content analysis

		rare disease, including PH								
Yorke (2014)*	UK	To explore participant's experiences of daily life with PH.	30	60	56.3 26 -80	NR	<1 to >10	Group 1 PAH (89%) 37%=idiopathic PAH 23%=connective tissue disease-PAH 17%=congenital heart disease-PAH 3%=heritable PAH 3% =portopulmonary hypertension 3%=drugs/toxins-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

PH = Pulmonary hypertension, PAH = Pulmonary Arterial Hypertension N = Number, NR = Not Reported, UK = United Kingdom, USA = United States of America, WHO = World Health Organisation

Quality Assessment

All studies met the majority of the ten quality assessment items (Table 4). Eleven articles scores ++ and seven +. Only one study scored – (Keen et al., 2018). This was because the purpose of the article was to examine physiotherapy practice in PH from a physiotherapist and patient perspective, and while six physiotherapists (and an additional 63 for the quantitative aspect of the study) were recruited, only three individuals with PH were recruited despite a framework analysis being utilised, which would typically require a far greater sample size (Fugard & Potts, 2015). Moreover, minimal information was provided on how patients' data were collected and analysed.

Seven studies failed to justify their decision for utilising a qualitative approach and therefore it is unclear how appropriate the method was to address the research aims. One study provided a limited descriptive of their data analysis method. Surprisingly, fifteen articles did not report on their method of reflexivity – this is discussed further in the limitations section.

Table 4

*Quality assessment using Critical Appraisal Skills Programme (Critical Appraisal Skills Programme, 2018) ; *, +, ^ same sample investigated across studies (+ = criteria met, - = criteria not met)*

Primary author and year	Criteria										Rating
	1	2	3	4	5	6	7	8	9	10	
Alami (2016)	+	+	+	+	+	+	+	+	+	+	++
Armstrong (2012)*	+	+	+	+	+	-	+	+	+	+	++
Carroll (2012)	+	+	-	+	+	-	+	+	+	+	++
Chiang (2018)	+	+	-	+	+	-	+	+	+	+	+
Flattery (2005)	+	+	+	+	+	-	+	+	+	+	+
Goddard (2017)	+	+	+	+	+	-	+	+	+	+	++
Hall (2012)	+	+	+	+	+	+	+	+	+	+	++
Ivarsson (2014)+	+	+	-	+	+	-	+	+	+	+	+
Ivarsson (2016)+	+	+	-	+	+	-	+	+	+	+	+
Keen (2018)	+	+	-	-	-	-	+	+	+	+	-
Kingman (2014)	+	+	+	+	+	-	+	+	+	+	++
Lee (2019)	+	+	+	+	+	-	+	-	+	+	+
Lo (2018)^	+	+	-	+	+	-	+	+	+	+	+
Matura (2013)	+	+	+	+	+	+	+	+	+	+	++
Martin (2020)^	+	+	-	+	-	-	+	+	+	+	+
McDonough (2011)	+	+	+	+	+	+	+	+	+	+	++
Muntingh (2017)	+	+	+	+	+	-	+	+	+	+	++
Uhlenbusch (2019)	+	+	+	+	+	-	+	+	+	+	++
Yorke (2014)*	+	+	+	+	+	-	+	+	+	+	++

Was the research aim(s) (1) clearly stated (2) and was a qualitative method appropriate. Evaluation of the: (3) research design given the aim(s), (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.

Descriptive Themes

Six descriptive themes emerged:

Diagnosis

Participants reported experiencing PH-related symptoms for a "*long*" time (Alami et al., 2016, p.E19) prior to obtaining a diagnosis (Alami et al., 2016; Goddard et al., 2017;

Ivarsson et al., 2014). This period was characterised by a decline in physical functioning, HRQoL, and emotional and social difficulties (Armstrong et al., 2012; Flattery et al., 2005; Kingman et al., 2014; Muntingh et al., 2017). During this time, and thereafter, individuals expressed confusion over the cause of their experiences (Armstrong et al., 2012) - worries that were not necessarily reduced by the diagnosis, which in itself was described as having posed "*more questions than it answers*" (Goddard et al., 2017, p.3). Participants tended to avoid, adapt to or explain away their symptoms: "[I] *just assumed that everything was down to smoking*", which often delayed seeking medical help (Armstrong et al., 2012, p.4).

Participants described undergoing a series of examinations, referrals and misdiagnoses to find an explanation for their experiences (Alami et al., 2016; Armstrong et al., 2012; Ivarsson et al., 2014; Kingman et al., 2014). This process left participants feeling '*frustrated*' (Flattery et al., 2005, p.103), *'uncertain'* (Muntingh et al., 2017, p.611) , angry and "*disillusioned*" with the perceived meaning of their symptoms (Armstrong et al., 2012, p.6). Receiving the diagnosis was described as '*life-changing'* (Martin et al., 2020, p.4). While people responded differently, for instance, expressing feelings of '*relief*' – as they can now receive treatment (Goddard et al., 2017, p.3) - '*shock*' (Hall et al., 2012, p.38), helplessness (Chiang et al., 2018) and confusion (Alami et al., 2016), a strong emotional response was evident (Hall et al., 2012). Following the diagnosis, individuals ruminated over the cause of their disease, often blaming themselves or worrying if it was heredity (Ivarsson et al., 2014).

Treatment

Participants discussed their reliance on specialist PH centres (Goddard et al., 2017; Keen et al., 2018), which attending helped to reduce feelings of uncertainty as staff answered questions and dispelled false narratives about PH that participants had often read on the internet: "*it* [the diagnosis] *wasn't as devastatingly unhopeful as first appeared*" (Armstrong et al., 2012, p.7). A disparity in care between services was described (Keen et al., 2018), specifically, between specialist PH centres and community services in terms of knowledge, treatments and approach to care (Armstrong et al., 2012; Goddard et al., 2017; Hall et al., 2012). Many participants expressed the need for greater collaboration between services (Alami et al., 2016; Goddard et al., 2017; Ivarsson et al., 2014; Keen et al., 2018).

Individuals often discussed how they felt their care could be improved, which included being offered regular check-ups (Carroll et al., 2012), receiving more information on PH (Ivarsson et al., 2014; Uhlenbusch et al., 2019), promoting shared-decision making (Alami et al., 2016) and holistic care (Lee et al., 2019), and involving family members in their care: "[my wife] *has more questions than I do* [about PH] *and she's never been given any information*" (Ivarsson et al., 2014, p.4).

Participants did not view medication as a "*cure*" (Chiang et al., 2018, p.4), but rather to alleviate symptoms, improve their health, and ultimately, survival: "[if medication was stopped] *Well, all of us would die. It's as simple as that"* (Alami et al., 2016; Carroll et al., 2012; Chiang et al., 2018; Ivarsson et al., 2016; Ivarsson et al., 2014; Kingman et al., 2014; Yorke et al., 2014, p.458). Many reported an almost immediate reduction in symptoms attributed to starting treatment (Ivarsson et al., 2016; Ivarsson et al., 2014) with a '*strong emotional attachmen*t' to medication being noted (Carroll et al., 2012; Kingman et al., 2014, p.5).

Side effects of treatments were common, which seemed to mostly manifest physically (Flattery et al., 2005; McDonough et al., 2011; Uhlenbusch et al., 2019). 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 (Lee et al., 2019; Matura et al., 2013). Side effects posed as a barrier to adherence, as while some viewed the negatives of treatment as a trade-off: "*it's added years to my life…but it's bittersweet"*(Flattery et al., 2005, p.102), others stopped despite the risk (Yorke et al., 2014).

Other barriers were discussed such as, accessibility and cost of treatment, emotional distress and problems with self-administration of some treatments (Carroll et al., 2012; Hall et al., 2012; Ivarsson et al., 2016; Lee et al., 2019; McDonough et al., 2011; Uhlenbusch et al., 2019; Yorke et al., 2014). Lack of perceived need also affected uptake of services as while many experienced emotional difficulties, some felt they did not need psychological support (Ivarsson et al., 2016), perceiving: "*the disease itself is the problem*" (Chiang et al., 2018, p.4). However, some expressed the need for help with managing anxiety and low mood (Muntingh et al., 2017).

Participants described a number of worries associated with their treatment, such as, whether it was effective (Flattery et al., 2005) and if so for how long (Yorke et al., 2014), the potential risks (Hall et al., 2012), what other options are available (Hall et al., 2012; Kingman et al., 2014; Yorke et al., 2014) and the wait to receive treatment (Lo et al., 2019). Visible treatments could also be a source of '*social stigma'* (Alami et al., 2016; Yorke et al., 2014, p.456), leaving participants feeling *'self-conscious'* (Kingman et al., 2014, p.4).

Prognosis

Participants were saddened (Chiang et al., 2018), "t*errified*" (Matura et al., 2013, p.162) and anxious over the prospect of their death (Lo et al., 2019; Muntingh et al., 2017). Individuals described many uncertainties associated with their prognosis including, how their disease would progress (Matura et al., 2013; McDonough et al., 2011), what their future would hold (Ivarsson et al., 2016), the impact of their death on loved ones (Goddard et al., 2017; Lo et al., 2019; Muntingh et al., 2017), and how long they had remaining: "*I am clueless as to how long my heart will hold out"* (Chiang et al., 2018, p.3). One women wondered if her death would make it "*easier*" due to the impact of PH on her family (Lo et al., 2019, p.4). The pervasive fear over when or how their symptoms would worsen '*plagued'* participants (McDonough et al., 2011, p.129), resulting in emotional and psychological difficulties (Ivarsson et al., 2016; McDonough et al., 2011).

Healthcare Professionals

Developing a therapeutic relationship with a PH "*expert"* was important (Kingman et al., 2014, p.5). Professionals were described as a source of support (Ivarsson et al., 2016), advice (Keen et al., 2018), knowledge (Hall et al., 2012), reassurance (Armstrong et al., 2012), motivation and '*hope*' (Chiang et al., 2018; Ivarsson et al., 2014, p.4). That said, participants reported some difficulties accessing or discussing their worries with some professionals (Alami et al., 2016; Ivarsson et al., 2014), as well as previous encounters in which professionals lacked competency (Uhlenbusch et al., 2019), empathy or sensitivity: one participant, for example, reported being told "*It's* [PH] *worse than cancer, we can't treat it*" (Armstrong et al., 2012, p.6).

Participants expressed their frustration over healthcare professionals' poor understanding of PH (Armstrong et al., 2012; Hall et al., 2012; Uhlenbusch et al., 2019). Individuals spoke of pleading (Goddard et al., 2017) or taking it upon themselves to explain the disease to care staff (Ivarsson et al., 2014). Such an experience was typified by a short narrative entitled "*Insider's knowledge*" describing an individual mentally preparing himself for an encounter he was having with a doctor who knew little about the disease (Goddard et al., 2017, p.3).

Impact

A range of symptoms were experienced by participants including, dyspnoea, pain, dizziness, fatigue, palpitations and cognitive difficulties (Alami et al., 2016; Flattery et al., 2005; Goddard et al., 2017; Matura et al., 2013; McDonough et al., 2011; Uhlenbusch et al., 2019). Breathing problems, however, were described as the "*main symptom*" (Alami et al., 2016, p.e16), which were associated with feelings of breathlessness, '*suffocating', 'choking'* and '*pain'* – this was closely interlinked with anxiety and panic (Alami et al., 2016, p.e16; McDonough et al., 2011). Breathing could become a '*conscious act'* (Alami et al., 2016, p.e16), as participants provided precise details over which activities would cause them shortness of breath (Alami et al., 2016). That said, some symptoms were '*unpredictable*' and likened to being on a *'rollercoaster'* (Hall et al., 2012; Yorke et al., 2014, p.456).

Decline in physical ability, mobility and energy levels were prevalent (Chiang et al., 2018; Muntingh et al., 2017), which could be caused by any form of activity (Alami et al., 2016; Hall et al., 2012; McDonough et al., 2011). Participants expressed '*fear'* of engaging in activities (Kingman et al., 2014, p.4) due to the onset of symptoms, pain (McDonough et al., 2011) or it requiring too much energy (Ivarsson et al., 2016). There seemed to be a discrepancy between participant's ideal and actual self, in terms of ability (Alami et al., 2016), which manifested in feelings of '*anger'* (McDonough et al., 2011, p.129), "*failing"* (Goddard et al., 2017, p.4), '*disappointment'* (Ivarsson et al., 2016, p.38) and shame: "*If I had to go up one floor and there was no one else in the lift hall to see me, then I preferred to take the lift, but I was ashamed of it*" (Ivarsson et al., 2016, p.38).

Individuals had to reduce or cease many activities (Alami et al., 2016; Lo et al., 2019; Martin et al., 2020), leading them to relate their situation to a "*shackle"* (Kingman et al., 2014, p.4). Reflected in this, was the feeling of restriction, the nature of which had a considerable impact on profession, family and social roles (Alami et al., 2016; Chiang et al., 2018; Goddard et al., 2017; Lee et al., 2019; Lo et al., 2019; Muntingh et al., 2017; Uhlenbusch et al., 2019). This appeared to be, somewhat, moderated by age, gender and stage of the disease (Lo et al., 2019; Muntingh et al., 2017). For example, younger participants had to limit themselves to activities that matched their '*activity tolerance'* opposed to interests (Kingman et al., 2014, p.4); some participants reported sexual difficulties largely due to fatigue and shortness of breath, which negatively affected their relationship (Alami et al., 2016; Ivarsson et al., 2016; Lo et al., 2019); financial problems associated with the cost of care and loss of earnings (Chiang et al., 2018; Muntingh et al., 2017; Uhlenbusch et al., 2019); practical and emotional problems caused by traveling and going on holiday (Hall et al., 2012; Ivarsson et al., 2016); and both male and female participants discussed difficulties associated with having or caring for their children resulting in emotional difficulties (Ivarsson et al., 2016; Ivarsson et al., 2014; Lo et al., 2019).

There was a general sense that the disease could be invisible and "*hidden"* from others (Yorke et al., 2014, p.458). 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 (Alami et al., 2016; Flattery et al., 2005; Ivarsson et al., 2016; Ivarsson et al., 2014; Lo et al., 2019; Uhlenbusch et al., 2019; Yorke et al., 2014). Participants found themselves having to '*battle'* for their illness to be recognised (Yorke et al., 2014, p.458), and justify their difficulties (Ivarsson et al., 2016; Ivarsson et al., 2014). This could leave participants feeling judged as "*lazy", "unfit*" and *"old"*(Goddard et al., 2017, p.4)

Life with PH could have a negative effect on participant's mental health; with anxiety, low mood, isolation (Kingman et al., 2014; Matura et al., 2013; McDonough et al., 2011; Muntingh et al., 2017; Uhlenbusch et al., 2019) and suicidal ideation being discussed:

"When I think about my disease, I am drawn to the idea of killing myself" (Chiang et al., 2018, p.4; Martin et al., 2020).

Coping

Participants required a period of time to adjust to life with PH (Flattery et al., 2005; McDonough et al., 2011; Yorke et al., 2014), which could be a '*painstaking'* process (Chiang et al., 2018, p.4). Individuals coped differently; for example, one 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 more of a social life (Kingman et al., 2014, p.4-5).

A common approach to coping was preparation (Goddard et al., 2017), '*extensive planning'* and developing '*daily routines'* (Hall et al., 2012; Kingman et al., 2014, p.5). Individuals discussed weighing up how activities would impact them, rather than "*act on impulse*" (McDonough et al., 2011, p.127). Participants reported feeling as if they had to '*hold back or take it* [daily activities] *slow*' (Flattery et al., 2005; McDonough et al., 2011, p.127). Personal limits were learnt through '*testing*' their own ability (Hall et al., 2012, p.39).

Many acknowledged the importance of developing a helpful or positive *"mindset*" (Chiang et al., 2018, p.4; Muntingh et al., 2017), for example, focusing on activities that were still within their ability (McDonough et al., 2011): "*I tell myself that I have to look at what I can still do…*" (Hall et al., 2012, p.39). Self-talk (Hall et al., 2012), focusing on having overcome previous adversity (Martin et al., 2020), faith (Flattery et al., 2005) and self-humour were also used to cope (Flattery et al., 2005; Goddard et al., 2017).

Altering expectations of their ability, recovery and future (Alami et al., 2016; Ivarsson et al., 2014; Muntingh et al., 2017; Yorke et al., 2014) allowed some to develop a new sense of purpose (Hall et al., 2012) or "*redefine'* their lives (Alami et al., 2016; Flattery et al., 2005; McDonough et al., 2011, p.128). Accepting their new limits gave participants the perception that they were not letting their disease "*dominate*" (Carroll et al., 2012; Kingman et al., 2014, p.4; Muntingh et al., 2017) and provided a sense of control (McDonough et al., 2011)*: "…it's just my life, it's just what I do"* (Flattery et al., 2005, p.102). Keeping active was described as helping to shift their focus away from their symptoms (Matura et al., 2013), control disease progression and have psychological benefits (Keen et al., 2018). Some reflected that through such change, they gained new meaning and personal growth (Chiang et al., 2018; Hall et al., 2012).

Family, friends and employers were described as offering practical and emotional support (Chiang et al., 2018; Ivarsson et al., 2016; Kingman et al., 2014; Matura et al., 2013; Muntingh et al., 2017). However, participants appeared conflicted, describing their own position as "*enforced dependency*" (Kingman et al., 2014, p.3; Lo et al., 2019). Social support could also be a barrier or counterproductive to coping, as how others dealt with the disease was not always helpful: "*I'm not allowed to look after the grandchildren, so I asked why. The children's answer was that they were afraid I would get sick, but I said that if I felt worse there were telephones"* (Ivarsson et al., 2016, p.38; Uhlenbusch et al., 2019).

Participants spoke about concealing, avoiding, diminishing and making excuses for their difficulties (Chiang et al., 2018; Goddard et al., 2017; Kingman et al., 2014; Lo et al., 2019; Martin et al., 2020; Yorke et al., 2014). This helped to reduce some of the emotional impact of PH, as well as feeling less of a burden or protecting others (Hall et al., 2012; Ivarsson et al., 2016; Martin et al., 2020). However, this could lead to social withdrawal and isolation, further straining relationships and causing psychological difficulties (Chiang et al., 2018; Kingman et al., 2014; Muntingh et al., 2017).

Peer supports groups and organisations were common sources of reassurance, support and advice (Ivarsson et al., 2016; Uhlenbusch et al., 2019). Individuals sought validation and normalisation of their symptoms from people with shared experience (Matura et al., 2013) and also looked to help others (Carroll et al., 2012).

Analytical Themes

Four key analytical themes emerged. Table 5 shows the association between the descriptive and analytical themes.

Table 5

Descriptive and analytical themes; "X" denotes that the descriptive theme contributed to the analytical theme

	Managing uncertainty	Physical nature of PH	Living with the rarity of PH	Transitional nature of PH
Diagnosis	X	X		
Treatment	X	X	X	X
Prognosis	X			
Healthcare Professionals	X		X	
Impact of PH	X	X	X	X
Coping with PH	X	X	X	X

PH = Pulmonary Hypertension

Managing Uncertainty

Participants acknowledged many uncertainties caused by internal-, such as bodily sensations, or external-events linked to PH, including the reaction of others (Alami et al., 2016) or what the future may hold (Lo et al., 2019). 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 (Flattery et al., 2005), in the hope of gaining knowledge, understanding and some sense of certainty (Armstrong et al., 2012). Unfortunately, given the complexity and lack of understanding of PH, this could result in even greater uncertainty (Armstrong et al., 2012).

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 (Yorke et al., 2014) or it removed *'spontaneity in their life'* (McDonough et al., 2011, p.127). Worrying was also closely linked with hypervigilance to the impact of everyday activities (McDonough et al., 2011) and somatic changes as an '*indicator*' to alter behaviour (Alami et al., 2016; Goddard et al., 2017). This resulted in anticipatory anxiety and panic (McDonough et al., 2011).

Some described avoiding (Martin et al., 2020; Muntingh et al., 2017) or concealing their disease, however, this often prolonged distress and resulted in emotional difficulties, such as anxiety, guilt and loneliness (Lo et al., 2019).

Physical Nature of PH

PH was described as a physically limiting disease (Uhlenbusch et al., 2019), as was there a tendency for individuals to report somatic experiences (Goddard et al., 2017; McDonough et al., 2011). Furthermore, despite experiencing emotional difficulties, participants often resisted the idea or need for psychological support (Martin et al., 2020; Muntingh et al., 2017; Uhlenbusch et al., 2019). A notion further reflected in the relief upon hearing that the disease was not psychosomatic in nature (Armstrong et al., 2012).

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 (Muntingh et al., 2017; Yorke et al., 2014, p.456). Discordance between the meaning and impact that 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 (Ivarsson et al., 2016; Muntingh et al., 2017). It seemed the support that participant's received was, at times, influenced by other's representation of the disease (Goddard et al., 2017). 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 many discussions (Alami et al., 2016; Muntingh et al., 2017), however, no more so than when individuals expressed their frustration at the lack of PH awareness in society and healthcare settings (Ivarsson et al., 2014; Uhlenbusch et al., 2019). Poor understanding and misconceptions over PH resulted in participants feeling stigmatised and discriminated against (Goddard et al., 2017; Muntingh et al., 2017; Uhlenbusch et al., 2019). Participants found themselves having to justify and legitimise their sick role, despite having an incurable disease (Uhlenbusch et al., 2019; Yorke et al., 2014) or withdrawing to avoid difficulties (Lo et al., 2019). 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 their care (Goddard et al., 2017; Ivarsson et al., 2014). Living with a rare condition left individuals feeling lonely and isolated: "*forced to* *carry a burden no one wants, or understands fully*" (Kingman et al., 2014, p.4). Participants found understanding from experts in PH and PH communities (Flattery et al., 2005).

Transitional Nature of PH

Participants progressed through different transitions living with PH (Muntingh et al., 2017). At first, individuals experienced a decline in general functioning, which appeared to improve following the diagnosis and administration of treatment (Armstrong et al., 2012). Participants next faced the process of adapting to life with PH and treatment (Flattery et al., 2005; Hall et al., 2012), the goal of which was to stabilise the disease (Keen et al., 2018), maintain a good HRQoL and survive (Muntingh et al., 2017). Participants had to find a balance between feeling underactive, in which they perceived their identity as changing (Lo et al., 2019) and themselves "*failing*" (Goddard et al., 2017, 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 (McDonough et al., 2011). This balance was often achieved through '*trial and error'* (Hall et al., 2012, 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 (Lippe et al., 2017). 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) (Dugas & Robichaud, 2007). In GAD, individuals find the feeling of uncertainty difficult. This distress is often managed by seeking reassurance from others or engaging in worry as a method to prepare for possible eventualities – which was also commonly reported in those with PH. However, this can have a consequential role on anxiety for example, such methods of coping can become compulsive and less effective overtime, and influence individuals to appraise and overestimate events as negative and threatening. This in turn, can generate greater uncertainty and anxiety (Bottesi et al., 2016; Dugas & Robichaud, 2007). A study investigating GAD in PH found 93% reported some symptoms (Harzheim et al., 2013). Symptoms of GAD in PH has been found to be associated with depression (Harzheim et al., 2013), which may also impact individuals in their ability to adapt to the challenges of PH (Wryobeck et al., 2007). Although anxiety and depression in PH is common, just over one-third of patients receive treatment (Somaini et al., 2015). Strategies aimed at helping individuals to cope better with distress associated with uncertainty are likely to be useful. A range of psychological treatments have been shown to be effective for GAD in the wider population (Hunot et al., 2007), however, there is paucity of evidence regarding psychological treatments for anxiety in PH (Bussotti & Sommaruga, 2018).

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 (White et al., 2010). 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

(Clark, 1986). As such, it may be beneficial to help individuals with PH to better understand and differentiate between the shared overlap between cardiorespiratory symptoms and anxiety (Fleet & Beitman, 1998). Interventions, such as mindfulness and relaxation techniques, that help individuals disengage their attention from actual, and the perceived meaning of, sensations may also be useful (Tulloh et al., 2018).

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 (Whitehead et al., 2013). Such incongruences have been suggested to influence how individuals present to services with their symptoms and adherence to treatment (Monzoni et al., 2011). 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 (Fenstad et al., 2014). 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 coping skills to manage conversations when other people are confused about PH is also important (Clarke et al., 2014).

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 (Halimi et al., 2015). 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 their concerns. For example, caregivers should help individuals to reflect on their difficulties and construct new meaning (Egnew, 2018). 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 that patients with PH present to services with (Fenstad et al., 2014).

Supporting individuals to manage fatigue through education, pacing and prioritising valued actions is likely to helpful - this may also reduce mood disorders (Connolly et al., 2013).

Limitations

Most studies did not discuss reflexivity. This is an important stage in some qualitative research approaches and also contributes to rigour and trustworthiness (Larkin & Thompson, 2012). The lack of this information means that it is not possible to ascertain or discuss the potential role of researcher bias in the results. For example, findings may have been influenced by characteristics of the researcher(s) such as their age, gender, profession or level of experience.

Due to a lack of reporting, the analysis was unable to distinguish between experiences based on PH-related factors, such as functional class or cause of PH, which has been shown to be associated with psychological distress (Yorke et al., 2015; Yorke et al., 2018). Grouping individuals based solely on their diagnosis of PH may have resulted in idiosyncratic findings being missed.

Regarding limitations of the review, only one researcher was involved in the screening and data extraction process. Moreover, while information has been provided concerning reflexivity, the data could still be influenced by an unintentional or unrecognised bias. Having multiple researchers identify and extract data on all, or a proportion, of the studies as well as have a greater number of discussions in stages two-three of the data analysis may have helped to reduce the impact of any bias and protect against human error. Furthermore, a member of the research team could have coded a number of articles, which had already been coded by the lead researcher to assess the reliability of coding.

The review protocol was registered at Open Science Framework (OSF) 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.

PH-related experiences, such as receiving palliative care, pregnancy, surgery and psychological interventions were largely unrepresented, despite being commonly encountered in practice. Indeed, representativeness of the findings may have been affected by the eligibility criteria. For example, research that was not written in English or published in scientific journals were excluded. This means that the narratives from non-Western countries and evidence exploring the relationship between PH and socio-cultural factors are missing. Furthermore, the review has failed to benefit from the vital work undertaken by PH Associations who play a key role in supporting this clinical group, and capturing the nature and impact of many challenges associated with the disease.

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 (Lee et al., 2019; Matura et al., 2013). The review finds numerous voices are missing, which needs addressing, including those from palliative care, non-white backgrounds 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 in coping with the disease. Indeed, findings from the current review were used to identify anxiety as a therapeutic target in this clinical group and inform the development of a self-help intervention for anxiety in PH.

References

Alami, S., Cottin, V., Mouthon, L., Desjeux, D., Quessette, E., Poiraudeau, S., & Sitbon, O. (2016). Patients', relatives', and practitioners' views of pulmonary arterial hypertension: A qualitative study. *La Presse Medicale*, *45*(2), e11-27. https://doi.org/10.1016/j.lpm.2015.06.017

Armstrong, I., Rochnia, N., Harries, C., Bundock, S., & Yorke, J. (2012). The trajectory to diagnosis with pulmonary arterial hypertension: a qualitative study. *BMJ Open*, *2*(2), Article e000806. https://doi.org/10.1136/bmjopen-2011-000806

Birks, Y., Harrison, R., Bosanquet, K., Hall, J., Harden, M., Entwistle, V., Watt, I., Walsh, P., Ronaldson, S., Roberts, D., Adamson, J., Wright, J., & Idema, R. An exploration of the implemention of open disclosure of adervse events in the UK: a scoping review and qualitative exploration. *Health Services and Delivery Research, 2*(20). https://doi.org/10.3310/hsdr02200

Bonner, N., Abetz, L., Meunier, J., Sikirica, M., & Mathai, S. C. (2013). Development and validation of the living with pulmonary hypertension questionnaire in pulmonary arterial hypertension patients. *Health and Quality of Life Outcomes*, *11*, Article 161. https://doi.org/10.1186/1477-7525-11-161

Bottesi, G., Ghisi, M., Carraro, E., Barclay, N., Payne, R., & Freeston, M. H. (2016). Revising the Intolerance of Uncertainty Model of Generalized Anxiety Disorder: Evidence from UK and Italian Undergraduate Samples. *Frontiers in Psychology*, *7*, Article 1723. 10.3389/fpsyg.2016.01723

Brown, L. M., Chen, H., Halpern, S., Taichman, D., McGoon, M. D., Farber, H. W., Frost, A. E., Liou, T. G., Turner, M., Feldkircher, K., Miller, D. P., & Elliott, C. G. (2011). Delay in recognition of pulmonary arterial hypertension factors identified from the REVEAL registry. *Chest*, *140*(1), 19-26. https://doi.org/10.1378/chest.10-1166

Bussotti, M., & Sommaruga, M. (2018). Anxiety and depression in patients with pulmonary hypertension: impact and management challenges. *Vascular Health and Risk Management*, *14*, 349-360. https://doi.org/10.2147/VHRM.S147173

Carroll, R., Antigua, J., Taichman, D., Palevsky, H., Forfia, P., Kawut, S., & Halpern, S. D. (2012). Motivations of patients with pulmonary arterial hypertension to participate in randomized clinical trials. *Clinical Trials*, *9*(3), 348-357. https://doi.org/10.1177/1740774512438981

Chang, W. T., Weng, S. F., Hsu, C. H., Shih, J. Y., Wang, J. J., Wu, C. Y., & Chen, Z. C. (2016). Prognostic factors in patients with pulmonary hypertension—A nationwide cohort study. *Journal of American Heart Association*, *5*(9), Article 2003579. https://doi.org/10.1161/JAHA.116.003579

Chiang, Y.-C., Hu, L.-Y., Couper, J., Cheng, C.-C., Wang, L.-H., Huang, W.-C., & Lu, T. (2018). Exploring the experiences and psychosocial stresses of Taiwanese patients with pulmonary hypertension: a qualitative interview study. *Pulmonary Circulation*, *8*(3), Article 2045894018787479. https://doi.org/10.1177/2045894018787479

Clark, D. M. (1986). A cognitive approach to panic. *Behaviour Research and Therapy*, *24*(4), 461-470. https://doi.org/10.1016/0005-7967(86)90011-2

Clarke, A., Thompson, A. R., Jenkinson, E., Rumsey, N., & Newell, R. (2014). *CBT for appearance anxiety: Psychosocial interventions for anxiety due to visible difference*. Wiley-Blackwell.

Cole, E., Armstrong, I., & Cutts, K. (2016). Links between psychological factors and health-related quality of life in pulmonary hypertension. *European Respiratory Journal*, *48*, PA1622. https://doi.org/10.1183/13993003.congress-2016.PA1622

Connolly, D., O'Toole, L., Redmond, P., & Smith, S. M. (2013). Managing fatigue in patients with chronic conditions in primary care. *Family Practice, 30*(2), 123-124). https://doi.org/10.1093/fampra/cmt005

Cooke, A., Smith, D., & Booth, A. (2012). Beyond PICO: The SPIDER tool for qualitative Evidence Synthesis. *Qualitative Health Research*, *22*(10), 1435-1443. https://doi.org/10.1177/1049732312452938

Critical Appraisal Skills Programme. (2018). *CASP qualitative checklist*. https://casp-uk.net/wp-content/uploads/2018/03/CASP-Qualitative-Checklist-2018_fillable_form.pdf.

Delcroix, M., & Howard, L. (2015). Pulmonary arterial hypertension: The burden of disease and impact on quality of life. *European Respiratory Review*, *24*(138), 621-629. https://doi.org/10.1183/16000617.0063-2015

Dodgson, J. E. (2019). Reflexivity in qualitative research. *Journal of Human Lactation*, *35*(2), 220-222. https://doi.org/10.1177/0890334419830990

Dugas, M. J., & Robichaud, M. (2007). *Practical clinical guidebooks. Cognitive-behavioral treatment for generalized anxiety disorder: From science to practice.* Routledge.

Egnew, T. R. (2018). A narrative approach to healing chronic illness. *Annals of Family Medicine*, *16*(2), 160-165. https://doi.org/10.1370/afm.2182

Fenstad, E. R., Shanafelt, T. D., Sloan, J. A., Novotny, P. J., Durst, L. A., Frantz, R. P., McGoon, M. D., & Swetz, K. M. (2014). Physician attitudes toward palliative care for patients with pulmonary arterial hypertension: Results of a cross- sectional survey. *Pulmonary Circulation*, *4*(3), 504-510. https://doi.org/10.1086/677365

Flattery, M. P., Pinson, J. M., Savage, L., & Salyer, J. (2005). Living with pulmonary artery hypertension: Patients’ experiences. *Heart & Lung*, *34*(2), 99-107. https://doi.org/10.1016/j.hrtlng.2004.06.010

Fleet, R. P., & Beitman, B. D. (1998). Cardiovascular death from panic disorder and panic-like anxiety: A critical review of the literature. *Journal of Psychosomatic Research, 44*(1), 71-80. doi: 10.1016/s0022-3999(97)00135-9.

Fugard, A.J.B., & Potts, H.W.W. Supporting thinking on sample sizes for thematic analyses: a quantitative tool. *International Journal of Social Research Methodology, 18*(6), 669-684. https://doi.org/10.1080/13645579.2015.1005453

Frost, A. E., Badesch, D. B., Barst, R. J., Benza, R. L., Elliott, C. G., Farber, H. W., Krichman, A., Liou, T. G., Raskob, G. E., Wason, P., Feldkircher, K., Turner, M., & McGoon, M. D. (2011). The changing picture of patients with pulmonary arterial hypertension in the United States: how REVEAL differs from historic and non- US Contemporary Registries. *Chest*, *139*(1), 128-137. https://doi.org/10.1378/chest.10-0075

Galiè, N., Humbert, M., Vachiery, J.-L., Gibbs, S., Lang, I., Torbicki, A., Simonneau, G., Peacock, A., Vonk Noordegraaf, A., Beghetti, M., Ghofrani, A., Gomez Sanchez, M. A., Hansmann, G., Klepetko, W., Lancellotti, P., Matucci, M., McDonagh, T., Pierard, L. A., Trindade, P. T., … Luis Zamorano, J. (2016). 2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension. *European Heart Journal*, *37*(1), 67-119. https://doi.org/10.1093/eurheartj/ehv317

Goddard, J. C., Armstrong, I. J., Kiely, D. G., Elliot, C. A., Charalampopoulos, A., Condliffe, R., Stone, B. J., & Sabroe, I. (2017). Combining creative writing and narrative analysis to deliver new insights into the impact of pulmonary hypertension. *BMJ Open Respiratory Research*, *4*(1), Article e000184. https://doi.org/10.1136/bmjresp-2017-000184

Graarup, J., Ferrari, P., & Howard, L. S. (2016). Patient engagement and self-management in pulmonary arterial hypertension. *European Respiratory Review*, *25*(142), 399-407. https://doi.org/10.1183/16000617.0078-2016

Halimi, L., Marin, G., Molinari, N., Gamez, A.-S., Boissin, C., Suehs, C. M., Vachier, I., & Bourdin, A. (2018). Impact of psychological factors on the health-related quality of life of patients treated for pulmonary arterial hypertension. *Journal of Psychosomatic Research*, *105*, 45-51. https://doi.org/10.1016/j.jpsychores.2017.12.003

Halimi, L., Molinari, N., Marin, G., Gamez, A. S., Boissin, C., Vachier, I., & Bourdin, A. (2015). Quality of life in worsening and non-worsening PAH patient's in a 1 year follow-up: What's going on? *European Respiratory Journal*, *46,* PA705. https://doi.org/10.1183/13993003.congress-2015.PA705

Hall, H., Cote, J., McBean, A., & Purden, M. (2012). The experiences of patients with pulmonary. arterial hypertension receiving continuous intravenous infusion of epoprostenol (Flolan) and their support persons. *Heart & Lung*, *41*(1), 35-43. https://doi.org/10.1016/j.hrtlng.2011.05.007

Hambly, N., Alawfi, F., & Mehta, S. (2016). Pulmonary hypertension: diagnostic approach and optimal management. *Canadian Medical Association Journal*, *188*(11), 804-812. https://doi.org/10.1503/cmaj.151075

Harper, D., & Thompson, A. R. (2012). *Qualitative research methods in mental health and psychotherapy: A guide for students and practitioners*. Wiley-Blackwell.

Harzheim, D., Klose, H., Pinado, F. P., Ehlken, N., Nagel, C., Fischer, C., Ghofrani, A., Rosenkranz, S., Seyfarth, H. J., Halank, M., Mayer, E., Grünig, E., & Guth, S. (2013). Anxiety and depression disorders in patients with pulmonary arterial hypertension and chronic thromboembolic pulmonary hypertension. *Respiratory Research*, *14*(1), Article 104. https://doi.org/10.1186/1465-9921-14-104

Hemnes, A. R., Kiely, D. G., Cockrill, B. A., Safdar, Z., Wilson, V. J., Hazmi, M. A., Preston, I. R., Maclean, M. R., & Lahm, T. (2015). Statement on pregnancy in pulmonary hypertension from the pulmonary vascular research institute. *Pulmonary Circulation*, *5*(3), 435-465. https://doi.org/10.1086/682230

Hoeper, M. M., Bogaard, H. J., Condliffe, R., Frantz, R., Khanna, D., Kurzyna, M., Langleben, D., Manes, A., Satoh, T., Torres, F., Wilkins, M. R., & Badesch, D. B. (2013). Definitions and diagnosis of pulmonary hypertension. *Journal of the American College of Cardiology*, *62*(25), D42-D50. https://doi.org/10.1016/j.jacc.2013.10.032

Hoeper, M. M., Ghofrani, H.-A., Grünig, E., Klose, H., Olschewski, H., & Rosenkranz, S. (2017). Pulmonary hypertension. *Deutsches Arzteblatt international*, *114*(5), 73-84. https://doi.org/10.3238/arztebl.2017.0073

Hoeper, M. M., & Gibbs, S. R. (2014). The changing landscape of pulmonary arterial hypertension and implications for patient care. *European Respiratory Review*, *23*(134), 450-457. https://doi.org/10.1183/09059180.00007814

Hoeper, M. M., & Humbert, M. (2019). The new haemodynamic definition of pulmonary hypertension: Evidence prevails, finally! *European Respiratory Journal*, *53*(3), Article 1900038. https://doi.org/10.1183/13993003.00038-2019

Hunot, V., Churchill, R., Silva de Lima, M., & Teixeira, V. (2007). Psychological therapies for generalised anxiety disorder. *Cochrane Database of Systematic Reviews*. https://doi.org/10.1016/j.rmed.2010.02.020

Ivarsson, B., Ekmehag, B., & Sjoberg, T. (2016). Support experienced by patients living with pulmonary arterial hypertension and chronic thromboembolic pulmonary hypertension. *Heart Lung and Circulation*, *25*(1), 35-40. https://doi.org/10.1016/j.hlc.2015.03.026

Ivarsson, B., Ekmehag, B., & Sjöberg, T. (2014). Information experiences and needs in patients with pulmonary arterial hypertension or chronic thromboembolic pulmonary hypertension. *Nursing Research and Practice*, *2014*, Article 704094. https://doi.org/10.1155/2014/704094

Keen, C., Fowler-Davis, S., McLean, S., & Manson, J. (2018). Physiotherapy practice in pulmonary hypertension: physiotherapist and patient perspectives. *Pulmonary Circulation*, *8*(3), Article 2045894018783738. https://doi.org/10.1177/2045894018783738

Khirfan, G., Tonelli, A. R., Ramsey, J., & Sahay, S. (2018). Palliative care in pulmonary arterial hypertension: an underutilised treatment. *European Respiratory Review*, *27*, Article 180069. https://doi.org/10.1183/16000617.0069-2018

Kiely, D., Elliot, C., Sabroe, I., & Condliffe, R. (2013). Pulmonary hypertension: diagnosis and management. *British Medical Journal*, *346*, Article f2028. https://doi.org/10.1136/bmj.f2028

Kingman, M., Hinzmann, B., Sweet, O., & Vachiéry, J. L. (2014). Living with pulmonary hypertension: unique insights from an international ethnographic study. *BMJ Open*, *4*(5), Article e004735. https://doi.org/10.1136/bmjopen-2013-004735

Larkin, M., & Thompson, A. (2012). Interpretative phenomenological analysis in mental health and psychotherapy research. In D. Harper & A. R. Thompson (Eds.), *Qualitative research methods in mental health and psychotherapy* (pp. 101-116). Wiley-Blackwell.

Lee, C., St Clair, C., Merenda, C. C., Araojo, C. R., Ray, S., Beasley, D., & Hinton, R. D. (2019). 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 : RSAP*. https://doi.org/10.1016/j.sapharm.2019.10.009

Lippe, C., Diesen, P. S., & Feragen, K. B. (2017). Living with a rare disorder: a systematic review of the qualitative literature. *Molecular Genetics & Genomic Medicine, 5*(6), 758-773. doi: 10.1002/mgg3.315.

Lo, C., Sarker, T., Canning, O., Martin, V., McCarthy, M. A., Granton, J., & Tan, A. (2019). Clinical presentation of existential distress in pulmonary arterial hypertension. *Canadian Journal of Respiratory, Critical Care, and Sleep Medicine*, *3*(1), 56-62. https://doi.org/10.1080/24745332.2018.1507614

Marius, M. H., & Gibbs, J. S. R. (2014). The changing landscape of pulmonary arterial hypertension and implications for patient care. *European Respiratory Review*, *23*(134), 450-457. https://doi.org/10.1183/09059180.00007814

Martin, V., Sarker, T., Slusarek, E., McCarthy, M. A., Granton, J., Tan, A., & Lo, C. (2020). Conversational avoidance during existential interviews with patients with progressive illness. *Psychology, Health & Medicine*, *25*(9), 1073-1082. https://doi.org/http://dx.doi.org/10.1080/13548506.2020.1719282

Matura, L. A., McDonough, A., Aglietti, L. M., Herzog, J. L., & Gallant, K. A. (2013). A virtual community: Concerns of patients with pulmonary hypertension. *Clinical Nursing Research*, *22*(2), 155-171. https://doi.org/10.1177/1054773812462867

McDonough, A., Matura, L. A., & Carroll, D. L. (2011). Symptom experience of pulmonary arterial hypertension patients. *Clinical Nursing Research*, *20*(2), 120-134. https://doi.org/10.1177/1054773810391249

McGoon, M., Gutterman, D., Steen, V., Barst, R., McCrory, D. C., Fortin, T. A., & Loyd, J. E. (2004). Screening, early detection, and diagnosis of pulmonary arterial hypertension: ACCP evidence-based clinical practice guidelines. *Chest*, *126*(1), 14S-34S. https://doi.org/10.1378/chest.126.1_suppl.14S

McGoon, M. D., Ferrari, P., Armstrong, I., Denis, M., Howard, L. S., Lowe, G., Mehta, S., Murakami, N., & Wong, B. A. (2019). The importance of patient perspectives in pulmonary hypertension. *European Respiratory Journal*, *53*(1), Article 1801919. https://doi.org/10.1183/13993003.01919-2018

McKenna, S. P., Doughty, N., Meads, D. M., Doward, L. C., & Pepke-Zaba, J. (2006). The Cambridge Pulmonary Hypertension Outcome Review (CAMPHOR): A measure of health-related quality of life and quality of life for patients with pulmonary hypertension [Health Psychology Testing 2226]. *Quality of Life Research*, *15*(1), 103-115. https://doi.org/http://dx.doi.org/10.1007/s11136-005-3513-4

McKevitt, C., Redfern, J., Mold, F., & Wolfe, C. (2004). Qualitative Studies of Stroke: A Systematic Review. *Stroke*, *35*(6), 1499-1505. https://doi.org/10.1161/01.STR.0000127532.64840.36

Monzoni, C. M., Duncan, R., Grünewald, R., & Reuber, M. (2011). 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 Education and Counseling*, *85*(3), e189-e200. https://doi.org/10.1016/j.pec.2011.07.014

Muntingh, A. D. T., Gerritsen, S., Batelaan, N. M., & Bogaard, H. J. (2017). Pathways to experiencing mental problems and need for mental support in patients with pulmonary arterial hypertension. *Pulmonary Circulation*, *7*(3), 609-616. https://doi.org/10.1177/2045893217724750

Page, M.J., McKenzie, J.E., Bossuyt, P.M., Boutron, I., Hoffmann, T.C., Mulrow, C.D., Shamseer, L., Brennan, S.E., Chou, R., Glanville, J., Grimshaw, J.M., Hrobjartsson, A., Lalu, M.M., Li, T., Loder, E.W., Mayo-Wilson, E., McDonald, S., McGuinness, L.A., Stewart, L.A., Thomas, J., … Moher, D. (2021). The PRISMA 2020 statement: an updated guideline for reporting systematic reviews. *BMJ, 372*, Article 71. doi: 10.1136/bmj.n71

Rawlings, G. H., Beail, N., Armstrong, I., Condliffe, R., Kiely, D.G., Sabroe, I., & Thompson, A.R. (2020). Adults' experiences of living with pulmonary hypertension: A thematic synthesis of qualitative studies. *BMJ Open,* 10, Article e041428. doi: 10.1136/bmjopen-2020-041428

Rawlings, G. H., Williams, R. K., Clarke, D. J., English, C., Fitzsimons, C., Holloway, I., Lawton, R., Mead, G., Patel, A., & Forster, A. (2019). Exploring adults' experiences of sedentary behaviour and participation in non-workplace interventions designed to reduce sedentary behaviour: a thematic synthesis of qualitative studies. *BMC Public Health, 19*, Article 1099. https://doi.org/10.1186/s12889-019-7365-1

Rich, S., Dantzker, D. R., Ayres, S. M., Bergofsky, E. H., Brundage, B. H., Detre, K. M., Fishman, A. P., Goldring, R. M., Groves, B. M., & Koerner, S. K. (1987). Primary pulmonary hypertension. A national prospective study. *Annals of Internal Medicine*, *107*(2), 216-223. https://doi.org/10.7326/0003-4819-107-2-216

Russell, S., Ogunbayo, O., Newham, J., Heslop-Marshall, K., Netts, P., Hanratty, B., Beyer, F., & Kaner, E. (2018). Qualitative systematic review of barriers and facilitators to self- management of chronic obstructive pulmonary disease: views of patients and healthcare professionals. *npj Primary Care Respiratory Medicine*, *28*, Article 2. https://doi.org/10.1038/s41533-017-0069-z

Somaini, G., Hasler, E., Muller-Mottet, S., Huber, L., Speich, R., Bloch, K., & Ulrich, S. (2015). Anxiety and depression are highly prevalent in pulmonary hypertension and might improve with target therapy. *European Respiratory Journal*, *46,* PA2112. https://doi.org/10.1183/13993003.congress-2015.PA2112

Thomas, J., & Harden, A. (2008). Methods for the thematic synthesis of qualitative research in systematic reviews. *BMC Medical Research Methodology*, *8*, Article 45. https://doi.org/10.1186/1471-2288-8-45

Tong, A., Flemming, K., McInnes, E., Oliver, S., & Craig, J. (2012). Enhancing transparency in reporting the synthesis of qualitative research: ENTREQ. *BMC Medical Research methodology*, *12*, Article 181. https://doi.org/10.1186/1471-2288-12-181

Tulloh, R. M. R., Garratt, V., Tagney, J., Turner-Cobb, J., Marques, E., Greenwood, R., Howard, L., Gin-Sing, W., Barton, A., Ewings, P., Craggs, P., & Hollingworth, W. (2018). A pilot randomised controlled trial investigating a mindfulness-based stress reduction (MBSR) intervention in individuals with pulmonary arterial hypertension (PAH): the PATHWAYS study. *Pilot and Feasibility Studies*, *4*, Article 78. https://doi.org/10.1186/s40814-018-0270-z

Uhlenbusch, N., Lowe, B., & Depping, M. K. (2019). Perceived burden in dealing with different rare diseases: a qualitative focus group study. *BMJ Open*, *9*(12), Article e033353. https://doi.org/10.1136/bmjopen-2019-033353

White, J., Hopkins, R. O., Glissmeyer, E. W., Kitterman, N., & Elliott, C. G. (2006). Cognitive, emotional, and quality of life outcomes in patients with pulmonary arterial hypertension. *Respiratory Research*, *7*(1), Article 55. doi: 10.1186/1465-9921-7-55.

White, K. S., Craft, J. M., & Gervino, E. V. (2010). Anxiety and hypervigilance to cardiopulmonary sensations in non- cardiac chest pain patients with and without psychiatric disorders. *Behaviour Research and Therapy*, *48*(5), 394-401. https://doi.org/10.1016/j.brat.2010.01.001

Whitehead, K., Kandler, R., & Reuber, M. (2013). Patients' and neurologists' perception of epilepsy and psychogenic nonepileptic seizures. *Epilepsia*, *54*(4), 708-717. https://doi.org/10.1111/epi.12087

Wray, F., & Clarke, D. (2017). Longer-term needs of stroke survivors with communication difficulties living in the community: A systematic review and thematic synthesis of qualitative studies. *BMJ Open, 7,* Article e017944. doi: 10.1136/bmjopen-2017-017944

Wryobeck, J. M., Lippo, G., McLaughlin, V., Riba, M., & Rubenfire, M. (2007). Psychosocial aspects of pulmonary hypertension: A review. *Psychosomatics: Journal of Consultation and Liaison Psychiatry*, *48*(6), 467-475. https://doi.org/http://dx.doi.org/10.1176/appi.psy.48.6.467

Yorke, J., Armstrong, I., & Bundock, S. (2014). Impact of living with pulmonary hypertension: A qualitative exploration. *Nursing & Health Sciences*, *16*(4), 454-460. https://doi.org/10.1111/nhs.12138

Yorke, J., Armstrong, I., Deaton, C., Campbell, M., Pilling, M., McGowan, L., & Sephton, P. (2015). Patient rated WHO functional class and psychological distress are strongly associated with dyspnoea in pulmonary hypertension. *European Respiratory Journal*, *46,* PA331. https://doi.org/10.1183/13993003.congress-2015.PA331

Yorke, J., Corris, P., Gaine, S., Gibbs, J., Kiely, D., Harries, C., Pollock, V., & Armstrong, I. (2014). emPHasis-10: development of a health-related quality of life measure in pulmonary hypertension. *European Respiratory Journal*, *43*(4), 1106-1113. https://doi.org/10.1183/09031936.00127113

Yorke, J., Deaton, C., Campbell, M., McGowen, L., Sephton, P., Kiely, D. G., & Armstrong, I. (2018). 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 Respiratory Research*, *5*(1), Article e000263. https://doi.org/10.1136/bmjresp-2017-000263

Appendix A: PRISMA Standards Checklist

Section and Topic	Item #	Checklist item	Location where item is reported
TITLE			
Title	1	Identify the report as a systematic review.	11
ABSTRACT			
Abstract	2	See the PRISMA 2020 for Abstracts checklist.	12-13
INTRODUCTION			
Rationale	3	Describe the rationale for the review in the context of existing knowledge.	14-17
Objectives	4	Provide an explicit statement of the objective(s) or question(s) the review addresses.	17
METHODS			
Eligibility criteria	5	Specify the inclusion and exclusion criteria for the review and how studies were grouped for the syntheses.	18-23
Information sources	6	Specify all databases, registers, websites, organisations, reference lists and other sources searched or consulted to identify studies. Specify the date when each source was last searched or consulted.	18
Search strategy	7	Present the full search strategies for all databases, registers and websites, including any filters and limits used.	18-19
Selection process	8	Specify the methods used to decide whether a study met the inclusion criteria of the review, including how many reviewers screened each record and each report retrieved, whether they worked independently, and if applicable, details of automation tools used in the process.	18-19
Data collection process	9	Specify the methods used to collect data from reports, including how many reviewers collected data from each report, whether they worked independently, any processes for obtaining or confirming data from study investigators, and if applicable, details of automation tools used in the process.	20-23
Data items	10a	List and define all outcomes for which data were sought. Specify whether all results that were compatible with each outcome domain in each study were sought (e.g. for all measures, time points, analyses), and if not, the methods used to decide which results to collect.	20-23
	10b	List and define all other variables for which data were sought (e.g. participant and intervention characteristics, funding sources). Describe any assumptions made about any missing or unclear information.	20-23
Study risk of bias assessment	11	Specify the methods used to assess risk of bias in the included studies, including details of the tool(s) used, how many reviewers assessed each study and whether they worked independently, and if applicable, details of automation tools used in the process.	20
Effect measures	12	Specify for each outcome the effect measure(s) (e.g. risk ratio, mean difference) used in the synthesis or presentation of results.	N/A
Synthesis methods	13a	Describe the processes used to decide which studies were eligible for each synthesis (e.g. tabulating the study intervention characteristics and comparing against the planned groups for each synthesis (item #5)).	18-23
	13b	Describe any methods required to prepare the data for presentation or synthesis, such as handling of missing summary statistics, or data conversions.	20-23

Section and Topic	Item #	Checklist item	Location where item is reported
	13c	Describe any methods used to tabulate or visually display results of individual studies and syntheses.	20-23
	13d	Describe any methods used to synthesize results and provide a rationale for the choice(s). If meta-analysis was performed, describe the model(s), method(s) to identify the presence and extent of statistical heterogeneity, and software package(s) used.	20-23
	13e	Describe any methods used to explore possible causes of heterogeneity among study results (e.g. subgroup analysis, meta-regression).	20-23
	13f	Describe any sensitivity analyses conducted to assess robustness of the synthesized results.	20-23
Reporting bias assessment	14	Describe any methods used to assess risk of bias due to missing results in a synthesis (arising from reporting biases).	20
Certainty assessment	15	Describe any methods used to assess certainty (or confidence) in the body of evidence for an outcome.	20-23
RESULTS			
Study selection	16a	Describe the results of the search and selection process, from the number of records identified in the search to the number of studies included in the review, ideally using a flow diagram.	24-25
	16b	Cite studies that might appear to meet the inclusion criteria, but which were excluded, and explain why they were excluded.	25, Appendix F
Study characteristics	17	Cite each included study and present its characteristics.	26-31
Risk of bias in studies	18	Present assessments of risk of bias for each included study.	32-33
Results of individual studies	19	For all outcomes, present, for each study: (a) summary statistics for each group (where appropriate) and (b) an effect estimate and its precision (e.g. confidence/credible interval), ideally using structured tables or plots.	26-31
Results of syntheses	20a	For each synthesis, briefly summarise the characteristics and risk of bias among contributing studies.	26-33
	20b	Present results of all statistical syntheses conducted. If meta-analysis was done, present for each the summary estimate and its precision (e.g. confidence/credible interval) and measures of statistical heterogeneity. If comparing groups, describe the direction of the effect.	33-45
	20c	Present results of all investigations of possible causes of heterogeneity among study results.	33-45
	20d	Present results of all sensitivity analyses conducted to assess the robustness of the synthesized results.	33-45
Reporting biases	21	Present assessments of risk of bias due to missing results (arising from reporting biases) for each synthesis assessed.	32-33
Certainty of evidence	22	Present assessments of certainty (or confidence) in the body of evidence for each outcome assessed.	33-45
DISCUSSION			

Section and Topic	Item #	Checklist item	Location where item is reported
Discussion	23a	Provide a general interpretation of the results in the context of other evidence.	45-47
	23b	Discuss any limitations of the evidence included in the review.	48-49
	23c	Discuss any limitations of the review processes used.	48-49
	23d	Discuss implications of the results for practice, policy, and future research.	45-49
OTHER INFORMATION			
Registration and protocol	24a	Provide registration information for the review, including register name and registration number, or state that the review was not registered.	18
	24b	Indicate where the review protocol can be accessed, or state that a protocol was not prepared.	18, Appendix C
	24c	Describe and explain any amendments to information provided at registration or in the protocol.	N/A, 48
Support	25	Describe sources of financial or non-financial support for the review, and the role of the funders or sponsors in the review.	4
Competing interests	26	Declare any competing interests of review authors.	82-83
Availability of data, code and other materials	27	Report which of the following are publicly available and where they can be found: template data collection forms; data extracted from included studies; data used for all analyses; analytic code; any other materials used in the review.	18

Section and Topic	Item #	Checklist item	Reported (Yes/No)
TITLE			
Title	1	Identify the report as a systematic review.	Yes
BACKGROUND			
Objectives	2	Provide an explicit statement of the main objective(s) or question(s) the review addresses.	Yes
METHODS			
Eligibility criteria	3	Specify the inclusion and exclusion criteria for the review.	Yes
Information sources	4	Specify the information sources (e.g. databases, registers) used to identify studies and the date when each was last searched.	Yes
Risk of bias	5	Specify the methods used to assess risk of bias in the included studies.	Yes
Synthesis of results	6	Specify the methods used to present and synthesise results.	Yes
RESULTS			
Included studies	7	Give the total number of included studies and participants and summarise relevant characteristics of studies.	Yes
Synthesis of results	8	Present results for main outcomes, preferably indicating the number of included studies and participants for each. If meta-analysis was done, report the summary estimate and confidence/credible interval. If comparing groups, indicate the direction of the effect (i.e. which group is favoured).	Yes
DISCUSSION			
Limitations of evidence	9	Provide a brief summary of the limitations of the evidence included in the review (e.g. study risk of bias, inconsistency and imprecision).	Yes
Interpretation	10	Provide a general interpretation of the results and important implications.	Yes
OTHER			
Funding	11	Specify the primary source of funding for the review.	Page 4
Registration	12	Provide the register name and registration number.	Page 18

Page, M.J., McKenzie, J.E., Bossuyt, P.M., Boutron, I., Hoffmann, T.C., Mulrow, C.D., Shamseer, L., Brennan, S.E., Chou, R., Glanville, J., Grimshaw, J.M., Hrobjartsson, A., Lalu, M.M., Li, T., Loder, E.W., Mayo-Wilson, E., McDonald, S., McGuinness, L.A., Stewart, L.A., … Moher, D. (2021). The PRISMA 2020 statement: an updated guideline for reporting systematic reviews. *BMJ, 372*, Article 71. doi: 10.1136/bmj.n71

Appendix B: ENTREQ statement

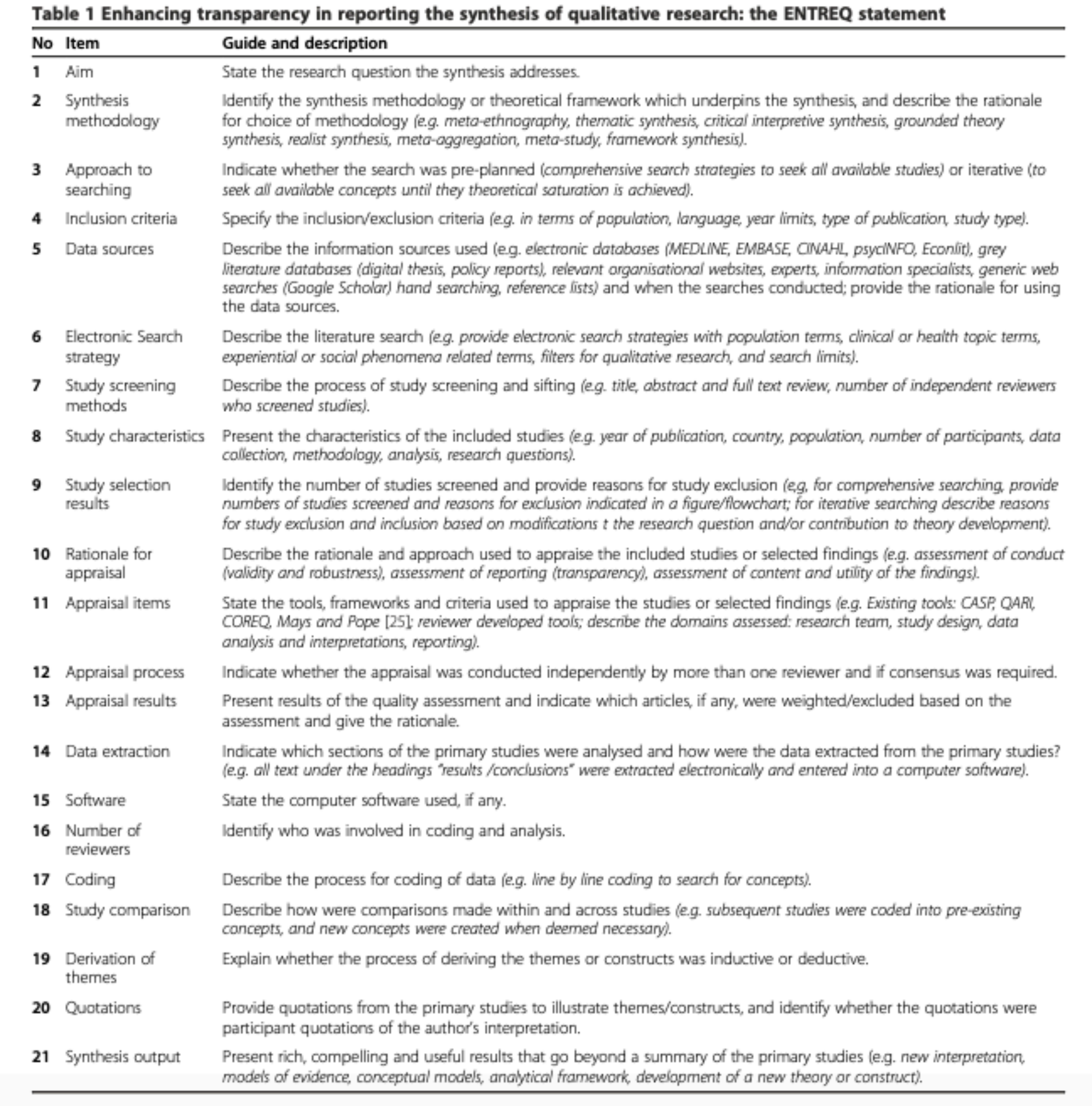
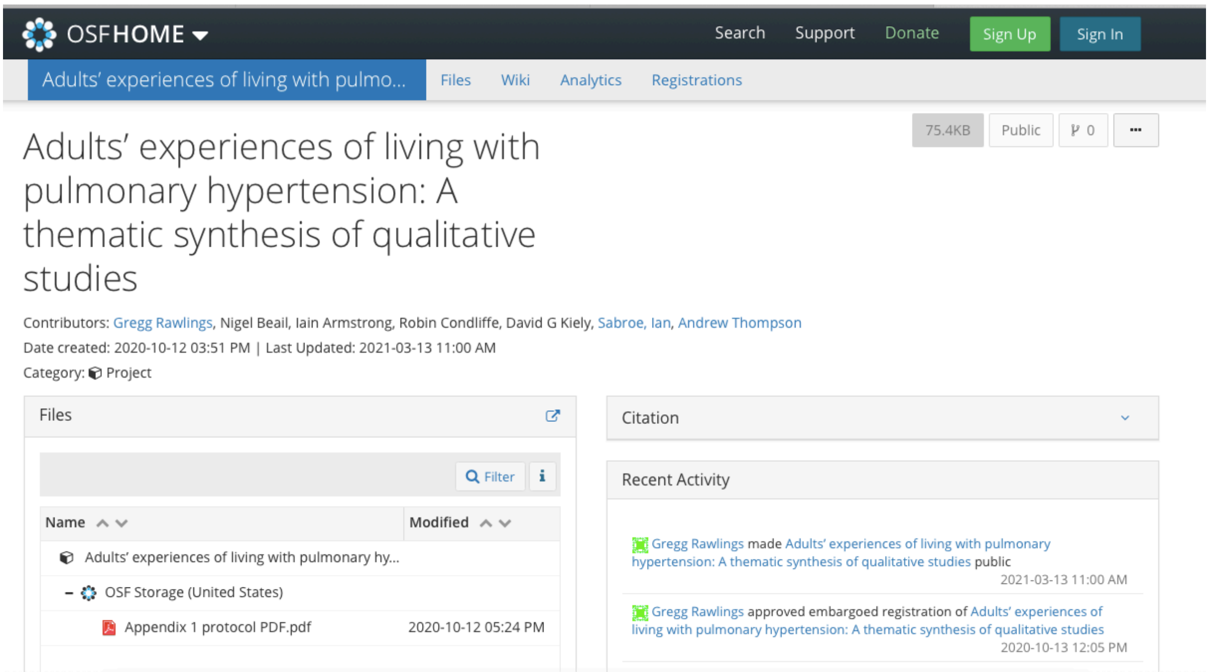


Table 1 Enhancing transparency in reporting the synthesis of qualitative research: the ENTREQ statement

No	Item	Guide and description
1	Aim	State the research question the synthesis addresses.
2	Synthesis methodology	Identify the synthesis methodology or theoretical framework which underpins the synthesis, and describe the rationale for choice of methodology *(e.g. meta-ethnography, thematic synthesis, critical interpretive synthesis, grounded theory synthesis, realist synthesis, meta-aggregation, meta-study, framework synthesis).*
3	Approach to searching	Indicate whether the search was pre-planned *(comprehensive search strategies to seek all available studies)* or iterative *(to seek all available concepts until they theoretical saturation is achieved).*
4	Inclusion criteria	Specify the inclusion/exclusion criteria *(e.g. in terms of population, language, year limits, type of publication, study type).*
5	Data sources	Describe the information sources used (e.g. *electronic databases (MEDLINE, EMBASE, CINAHL, psycINFO, Econlit), grey literature databases (digital thesis, policy reports), relevant organisational websites, experts, information specialists, generic web searches (Google Scholar) hand searching, reference lists)* and when the searches conducted; provide the rationale for using the data sources.
6	Electronic Search strategy	Describe the literature search *(e.g. provide electronic search strategies with population terms, clinical or health topic terms, experiential or social phenomena related terms, filters for qualitative research, and search limits).*
7	Study screening methods	Describe the process of study screening and sifting *(e.g. title, abstract and full text review, number of independent reviewers who screened studies).*
8	Study characteristics	Present the characteristics of the included studies *(e.g. year of publication, country, population, number of participants, data collection, methodology, analysis, research questions).*
9	Study selection results	Identify the number of studies screened and provide reasons for study exclusion *(e.g, for comprehensive searching, provide numbers of studies screened and reasons for exclusion indicated in a figure/flowchart; for iterative searching describe reasons for study exclusion and inclusion based on modifications t the research question and/or contribution to theory development).*
10	Rationale for appraisal	Describe the rationale and approach used to appraise the included studies or selected findings *(e.g. assessment of conduct (validity and robustness), assessment of reporting (transparency), assessment of content and utility of the findings).*
11	Appraisal items	State the tools, frameworks and criteria used to appraise the studies or selected findings *(e.g. Existing tools: CASP, QARI, COREQ, Mays and Pope* [25]*; reviewer developed tools; describe the domains assessed: research team, study design, data analysis and interpretations, reporting).*
12	Appraisal process	Indicate whether the appraisal was conducted independently by more than one reviewer and if consensus was required.
13	Appraisal results	Present results of the quality assessment and indicate which articles, if any, were weighted/excluded based on the assessment and give the rationale.
14	Data extraction	Indicate which sections of the primary studies were analysed and how were the data extracted from the primary studies? *(e.g. all text under the headings "results /conclusions" were extracted electronically and entered into a computer software).*
15	Software	State the computer software used, if any.
16	Number of reviewers	Identify who was involved in coding and analysis.
17	Coding	Describe the process for coding of data *(e.g. line by line coding to search for concepts).*
18	Study comparison	Describe how were comparisons made within and across studies *(e.g. subsequent studies were coded into pre-existing concepts, and new concepts were created when deemed necessary).*
19	Derivation of themes	Explain whether the process of deriving the themes or constructs was inductive or deductive.
20	Quotations	Provide quotations from the primary studies to illustrate themes/constructs, and identify whether the quotations were participant quotations of the author's interpretation.
21	Synthesis output	Present rich, compelling and useful results that go beyond a summary of the primary studies (e.g. *new interpretation, models of evidence, conceptual models, analytical framework, development of a new theory or construct).*

Tong, A., Flemming, K., McInnes, E., Oliver, S., & Craig, J. (2012). Enhancing transparency in reporting the synthesis of qualitative research: ENTREQ. *BMC medical research methodology*, *12*(1), Article 181. https://doi.org/10.1186/1471-2288-12-181

Appendix C – Registration in Review Database



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

Contributors: Gregg Rawlings, Nigel Beail, Iain Armstrong, Robin Condliffe, David G Kiely, Sabroe, Ian, Andrew Thompson

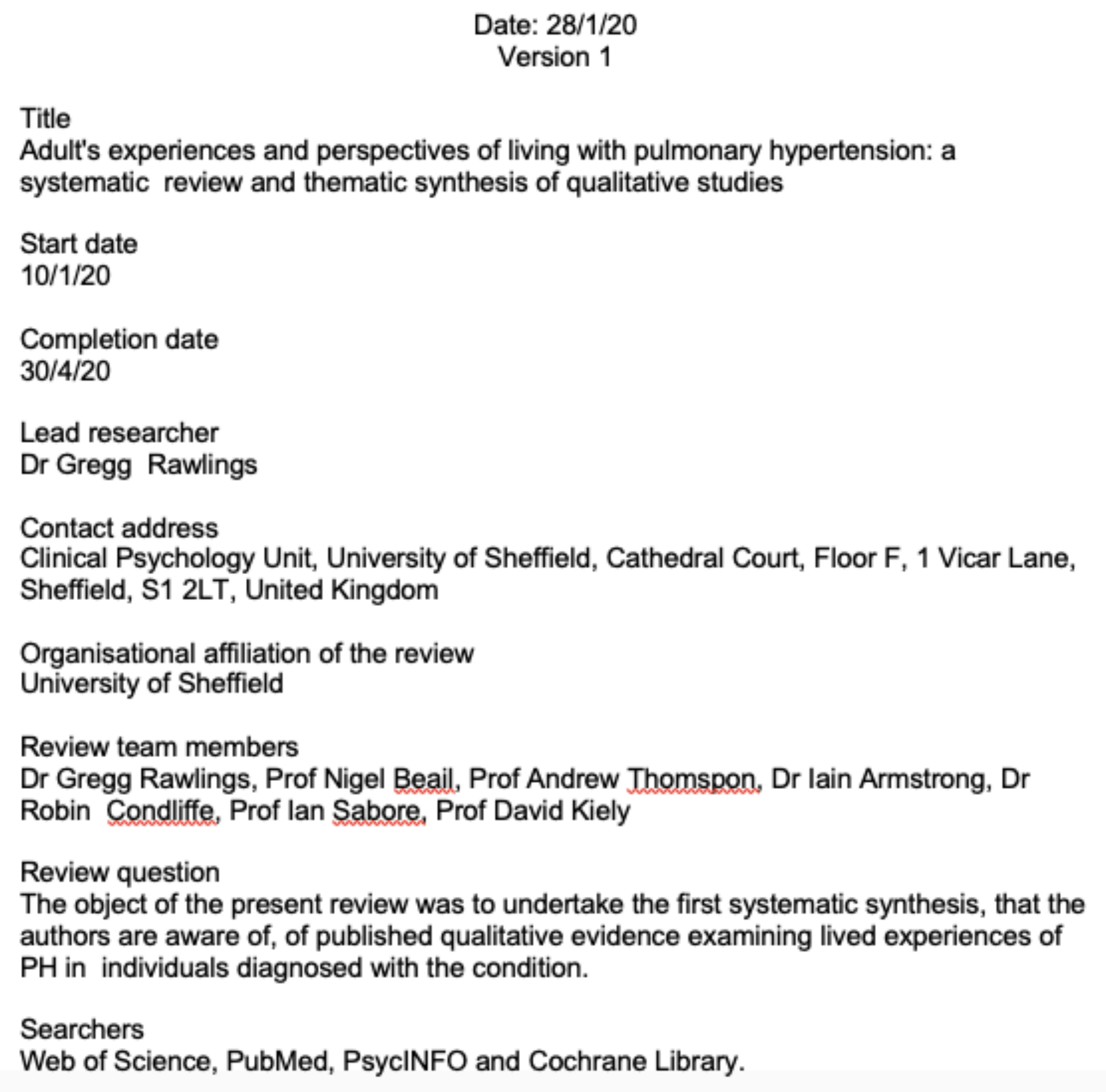
Date created: 2020-10-12 03:51 PM | Last Updated: 2021-03-13 11:00 AM

Category: Project

Name	Modified
Adults' experiences of living with pulmonary hy...	
– OSF Storage (United States)	
Appendix 1 protocol PDF.pdf	2020-10-12 05:24 PM

Gregg Rawlings made Adults' experiences of living with pulmonary hypertension: A thematic synthesis of qualitative studies public
2021-03-13 11:00 AM

Gregg Rawlings approved embargoed registration of Adults' experiences of living with pulmonary hypertension: A thematic synthesis of qualitative studies
2020-10-13 12:05 PM



Date: 28/1/20
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
Dr Gregg Rawlings

Contact address
Clinical Psychology Unit, University of Sheffield, Cathedral Court, Floor F, 1 Vicar Lane,
Sheffield, S1 2LT, United Kingdom

Organisational affiliation of the review
University of Sheffield

Review team members
Dr Gregg Rawlings, Prof Nigel Beail, Prof Andrew Thomspon, Dr Iain Armstrong, Dr Robin Condliffe, Prof Ian Sabore, Prof David Kiely

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 (Hoeper et al., 2013; 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 ap praise 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 D: Published Manuscript

Open access **Original research**

BMJ Open

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

Gregg Harry Rawlings,[1] Nigel Beail,[1] Iain Armstrong,[2]
Robin Condliffe,[2] David G Kiely,[2,3] Ian Sabroe,[4]
Andrew R Thompson[5]

To cite: Rawlings GH, Beail N, Armstrong I, *et al*. Adults' experiences of living with pulmonary hypertension: a thematic synthesis of qualitative studies. *BMJ Open* 2020;**10**:e041428. doi:10.1136/bmjopen-2020-041428

- Additional material is published online only. To view, please visit the journal online (http://dx.doi.org/10.1136/bmjopen-2020-041428).

Received 10 June 2020
Revised 27 October 2020
Accepted 15 November 2020

[1]Clinical Psychology Unit, The University of Sheffield, Sheffield, UK
[2]Sheffield Pulmonary Vascular Disease Unit, Sheffield Teaching Hospitals NHS Foundation Trust, Sheffield, UK
[3]Department of Infection, Immunity and Cardiovascular Disease, The University of Sheffield, Sheffield, UK
[4]School of Medicine and Biomedical Sciences, University of Sheffield, Sheffield, UK
[5]South Wales Clinical Psychology Training Programme, Cardiff University, Cardiff, UK

Correspondence to
Dr Gregg Harry Rawlings;
ghrawlings1@sheffield.ac.uk

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 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.

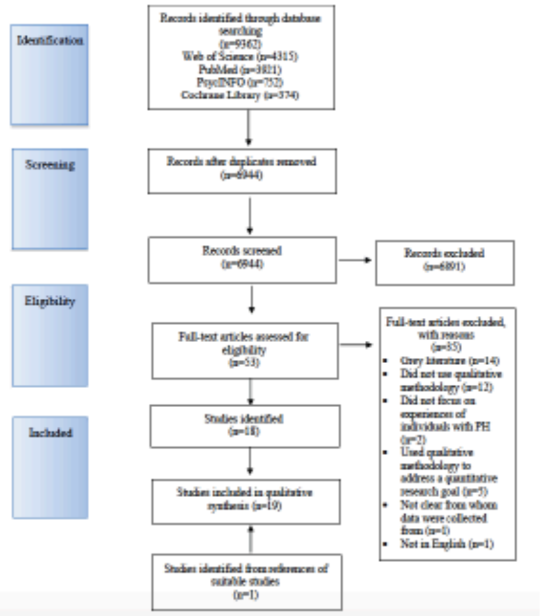


Figure 1 Preferred Reporting Items for Systematic Reviews and Meta-Analyses diagram. PH, pulmonary hypertension.

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.[20–29] 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	<1 to >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[34]*	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 1	2	3	4	5	6	7	8	9	10	Rating
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] "*terrified*"[35, p. 162] and anxious over the prospect of their death.[17 23] 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,[20] 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[30] 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.[53 55] 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.

Acknowledgements The authors would like to thank Christopher Gaskell (CK), a Trainee Clinical Psychologist from the University of Sheffield, for his help in rating the quality of included studies.

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.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests GHR, NB, ART, RC and IA have no conflict of interests 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.

Patient consent for publication Not required.

Provenance and peer review Not commissioned; externally peer reviewed.

Data availability statement No data are available. The current article was a systematic synthesis of data that had already been published.

ORCID iDs
Gregg Harry Rawlings http://orcid.org/0000-0003-4962-3551
Nigel Beail http://orcid.org/0000-0002-7916-9313
Iain Armstrong http://orcid.org/0000-0001-5898-3087
Robin Condliffe http://orcid.org/0000-0002-3492-4143
David G Kiely http://orcid.org/0000-0003-0184-6502
Ian Sabroe http://orcid.org/000-0001-9750-8975
Andrew R Thompson http://orcid.org/0000-0001-6788-7222

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.

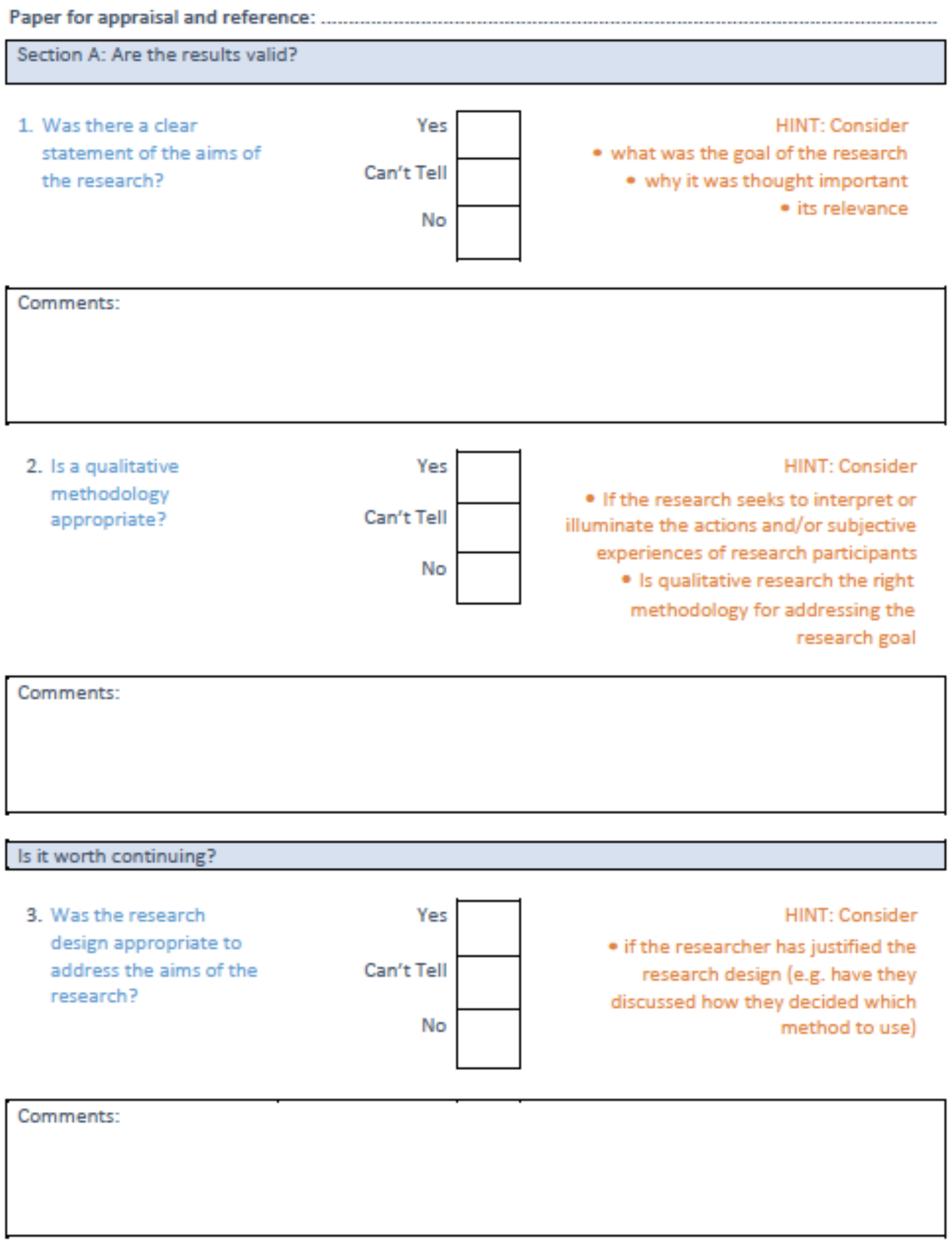
58 Larkin M, Thompson A. Interpretative Phenomenological Analysis in Mental Health and Psychotherapy Research. In: Harper D, Thompson AR, eds. *Qualitative research methods in mental health and psychotherapy*. Wiley-Blackwell, 2012: 101–16.

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.

Appendix E: Critical Appraisal Skills Programme Checklist



Paper for appraisal and reference: ....

Section A: Are the results valid?

1. Was there a clear statement of the aims of the research?

Yes ☐
Can't Tell ☐
No ☐

HINT: Consider
- what was the goal of the research
- why it was thought important
- its relevance

Comments:

2. Is a qualitative methodology appropriate?

Yes ☐
Can't Tell ☐
No ☐

HINT: Consider
- If the research seeks to interpret or illuminate the actions and/or subjective experiences of research participants
- Is qualitative research the right methodology for addressing the research goal

Comments:

Is it worth continuing?

3. Was the research design appropriate to address the aims of the research?

Yes ☐
Can't Tell ☐
No ☐

HINT: Consider
- if the researcher has justified the research design (e.g. have they discussed how they decided which method to use)

Comments:

4. Was the recruitment strategy appropriate to the aims of the research?

Yes ☐
Can't Tell ☐
No ☐

HINT: Consider
- If the researcher has explained how the participants were selected
- If they explained why the participants they selected were the most appropriate to provide access to the type of knowledge sought by the study
- If there are any discussions around recruitment (e.g. why some people chose not to take part)

Comments:

5. Was the data collected in a way that addressed the research issue?

Yes ☐
Can't Tell ☐
No ☐

HINT: Consider
- If the setting for the data collection was justified
- If it is clear how data were collected (e.g. focus group, semi-structured interview etc.)
- If the researcher has justified the methods chosen
- If the researcher has made the methods explicit (e.g. for interview method, is there an indication of how interviews are conducted, or did they use a topic guide)
- If methods were modified during the study. If so, has the researcher explained how and why
- If the form of data is clear (e.g. tape recordings, video material, notes etc.)
- If the researcher has discussed saturation of data

Comments:

6. Has the relationship between researcher and participants been adequately considered?

Yes ☐

Can't Tell ☐

No ☐

HINT: Consider

- If the researcher critically examined their own role, potential bias and influence during (a) formulation of the research questions (b) data collection, including sample recruitment and choice of location
- How the researcher responded to events during the study and whether they considered the implications of any changes in the research design

Comments:

Section B: What are the results?

7. Have ethical issues been taken into consideration?

Yes ☐

Can't Tell ☐

No ☐

HINT: Consider

- If there are sufficient details of how the research was explained to participants for the reader to assess whether ethical standards were maintained
- If the researcher has discussed issues raised by the study (e.g. issues around informed consent or confidentiality or how they have handled the effects of the study on the participants during and after the study)
- If approval has been sought from the ethics committee

Comments:

8. Was the data analysis sufficiently rigorous?

Yes ☐

Can't Tell ☐

No ☐

HINT: Consider

- If there is an in-depth description of the analysis process
- If thematic analysis is used. If so, is it clear how the categories/themes were derived from the data
- Whether the researcher explains how the data presented were selected from the original sample to demonstrate the analysis process
- If sufficient data are presented to support the findings
- To what extent contradictory data are taken into account
- Whether the researcher critically examined their own role, potential bias and influence during analysis and selection of data for presentation

Comments:

9. Is there a clear statement of findings?

Yes ☐

Can't Tell ☐

No ☐

HINT: Consider whether

- If the findings are explicit
- If there is adequate discussion of the evidence both for and against the researcher's arguments
- If the researcher has discussed the credibility of their findings (e.g. triangulation, respondent validation, more than one analyst)
- If the findings are discussed in relation to the original research question

Comments:

Section C: Will the results help locally?

10. How valuable is the research?

HINT: Consider

- If the researcher discusses the contribution the study makes to existing knowledge or understanding (e.g. do they consider the findings in relation to current practice or policy, or relevant research-based literature
- If they identify new areas where research is necessary
- If the researchers have discussed whether or how the findings can be transferred to other populations or considered other ways the research may be used

Comments:

Ratings from CG (second reviewer)

Article	Clear statement of the aims	Qualitative Methodology Appropriate	Research design appropriate to the aims/questions	Recruitment strategy appropriate to the aims of the research	Data collected in a way that addressed the research issue	Relationship between researcher and participants considered	Ethic issues taken into consideration	Data analysis sufficiently rigorous	Clear statement of findings	How valuable is the research?	Overall
Armstrong (2010)	Yes	Yes	Yes	Yes	Yes - The authors clarify the use of a semi-structured interview and discuss the use and justification of pictorial timeline; data material type is reported - there is however no mention of saturation	No - No mention of reflexivity methods and no consideration of relationship of those involved.	Yes - Ethics approval report; informed consent fine; however no mention of potential ethical issues (e.g. inviting someone on a register who may have be of very poor health or recently deceased)	Yes	Yes - Clear findings, multiple analysts and audited, linked back to question	Yes	++
Hall (2012)	Yes	Yes	Yes - Question is around experiences of patients and carers	Yes	Yes	No	Yes - Ethical Approval Reported;	Yes	Yes	Yes	++
Kingman (2014)	Yes	Yes	Yes	Yes	Yes	No	Yes - Ethical Approval Reported; Informed Consent reported;	Yes - but quite limited	Yes	Yes - Clinical and research suggested	++

Matura (2013)	Yes	Yes	Yes	Yes	Yes	No - Journal for reflections but nothing stated regarding its use or relationship	Yes - Ethical Approval Reported;	Yes	Yes	Yes	++
McDonough (2011)	Yes	Yes	Yes	Yes	Yes	No - Journal for reflections but nothing stated regarding its use or relationship	Yes - but only informed consent	Yes	Yes	Yes	++

Ratings from GHR (lead author) (* = non-consensus, ^ = altered following discussion, [ ] showing what it was changed to)

Article	Clear statement of the aims	Qualitative Methodology Appropriate	Research design appropriate to the aims/questions	Recruitment strategy appropriate to the aims of the research	Data collected in a way that addressed the research issue	Relationship between researcher and participants considered	Ethic issues taken into consideration	Data analysis sufficiently rigorous	Clear statement of findings	How valuable is the research?	Overall
Armstrong (2012)	Yes page 1- article summary provided	Yes page 2 – discussed why qualitative approach was used	Yes page 2 – research design was discussed and related to aim	Yes page 2-3 – discussed comprehensively	Yes page 2-3 – discussed data collection and provided interview guide	No NA	Yes Page 9 – ethical consent was obtained	Yes Page 2-3 – limited in description but makes reference to approach and cites relevant literature	Yes Page 1,7-9 – clear discussion and conclusions and summary box provided	Yes Page 1,9 – makes reference to future research and clinical implications	+^ [++]
Hall (2012)	Yes Page 36 – provided in the abstract and introduction	Yes Page 36-37 – provided a rationale for using qualitative research design	Yes Page 37 – discussed in study design section of methods	Yes Page 37 – provided a clear rationale including recruitment procedure and sample	Yes Page 37-38 – discussed in procedure section	Yes* Page 37-38 provided a section of trustworthiness and data analysis	Yes Page 37– ethical consent was obtained	Yes Page 37-38 – discussed methodology and cited relevant literature	Yes Page 40-42 – discussed in the conclusion and abstract	Yes Page 42 – authors suggested directions for future research and for nursing practice	++
Kingman (2014)	Yes Page 2 - aims are provided	Yes Page 2 - the authors discuss using an ethnographic approach	Yes Page 2, discussed in data collection section	Yes Page 2-3- Discussed method providing additional information in table 1	Yes Page 2-3 - Discussed using diaries and making observations providing examples in Box 1	No NA	Yes Page 7 – ethical consent was obtained	Yes Page 3 – discussed in analysis and findings further expanded on in Figure 1	Yes Page 1,6-7 – conclusion is provided and strengths and limitations	Yes Page 5-6 – specific section provided on meaning and implications	+ [++]

Matura (2013)	Yes Page 159 – specific section on aims provided	Yes Page 159 – discussed in the design section making reference to evidence base	Yes Page 159 – discussed in the aims and design section	Yes Page 158-159 discussed using online discussion boards and cited previous research using a similar design	Yes Page 160 – discussed in data collection design	Yes* Page 160-161, discussed credibility and the use of journal	Yes Page 159 – a section on ethical considerations provided	Yes Page 160-161 Approach is discussed including the use of NVIVO	Yes Page 167 – comprehensive conclusions section provided	Yes Page 166 – authors make reference to findings having implications for theory and practice	++
McDonough (2011)	Yes Page 123 – specific section on the aim is provided	Yes Page 124 – authors discuss rationale for using qualitative methods	Yes Page 124 – discussed at the start of the method section	Yes Page 124 – clearly discussed in participants and sampling section	Yes Page 124 – discussed using narrative interviews and provided interview questions	Yes* Page 125 – discussed using a journal and audit trial	No^ [Yes] Does not state ethical approval was obtained – although verbal consent was obtained	Yes Page 125 - discussed using NVIVO and method of analysis	Yes Page 132 – conclusion section provided in addition to clear discussion	Yes Page 131-132 specific section on implications for practice provided	+^[++]

Appendix F: List of Excluded Studies

Author	Title	Reason for exclusion
(Armstrong et al., 2019)	The patient experience of pulmonary hypertension: a large cross-sectional study of UK patients	Did not use qualitative methodology
(Armstrong et al., 2011)	Living with pulmonary hypertension: A qualitative perspective	Grey literature
(Awdish et al., 2015)	Development of a modified yoga program for pulmonary hypertension: a case series	Did not use qualitative methodology
(Bonner et al., 2013)	Development and validation of the living with pulmonary hypertension questionnaire in pulmonary arterial hypertension patients	Used qualitative methodology to address a quantitative research goal
(Caputo, 2014)	Exploring quality of life in Italian patients with rare disease: a computer-aided content analysis of illness stories	Not clear if data was collected from individuals with PH
(Chen et al., 2011)	A qualitative approach to understanding quality of life impairment in pulmonary arterial hypertension: getting beyond the numbers	Grey literature
(Ferrari & Skara, 2019)	My life with pulmonary arterial hypertension: a patient perspective	Grey literature
(Guillevin et al., 2013)	Understanding the impact of pulmonary arterial hypertension on patients' and carers' lives	Used qualitative methodology to address a quantitative research goal
(Gray et al., 2015)	Participant expectations in pulmonary hypertension-related research studies	Did not use qualitative methodology
(Grinnan et al., 2012)	The end-of-life experience for a cohort of patients with pulmonary arterial hypertension	Did not use qualitative methodology
(Halimi et al., 2018)	Impact of psychological factors on the health- related quality of life of patients treated for pulmonary arterial hypertension	Did not use qualitative methodology
(Hall et al., 2007)	The experiences of patients with pulmonary arterial hypertension (PAH) receiving continuous intravenous infusion of epoprostenol (Flolan (TM)), and their support persons	Grey literature
(Hall et al., 2017)	A survey of the impact of owning a service dog on quality of life for individuals with physical and hearing disability: A pilot study	Did not use qualitative methodology
(Ivarsson et al., 2014)	Perceptions of received information, social support, and coping in patients with pulmonary arterial hypertension or chronic thromboembolic pulmonary hypertension	Did not use qualitative methodology

(Ivarsson et al., 2019)	Everyday life experiences of spouses of patients who suffer from pulmonary arterial hypertension or chronic thromboembolic pulmonary hypertension	Not perspectives of individuals with PH
(Jones et al., 2008)	What patients and their relatives think about testing for BMPR2	Did not use qualitative methodology
(Khan, 2011)	Commentary on symptom experience of pulmonary arterial hypertension (PAH) patients	Grey literature
(Kobayashi et al., 2012)	Quantitative and qualitative analysis of health-related quality of life in patients with pulmonary arterial hypertension (CP)	Grey literature
(Lichenstein et al., 2013)	Cyber support describing concerns of caregivers of people with pulmonary hypertension	Not perspectives of individuals with PH
(McKenna et al., 2006)	Cambridge Pulmonary Hypertension Outcome Review (CAMPHOR): A measure of health-related quality of life and quality of life for patients with pulmonary hypertension	Used qualitative methodology to address a quantitative research goal
(MacKenzie & Johnson, 2015)	Patient perspectives on exercise training in pulmonary arterial hypertension (PAH)	Grey literature
(McGoon et al., 2019)	The importance of patient perspectives in pulmonary hypertension	Grey literature
(Monson & Pennell, 2012)	The experience of the older adults with pulmonary arterial hypertension	Grey literature
(Mulgirigama et al., 2012)	A slower life in a smaller world. Patients' perspective on living with pulmonary arterial hypertension	Grey literature
(Nakanishi, 2015)	Pulmonary arterial hypertension associated with congenital heart disease. Personal perspectives	Did not use qualitative methodology
(Peloquin et al., 2007)	Perception of quality of life by women with stage III or IV primary pulmonary hypertension and receiving treatment with prostacyclin	Not in English
(Quijano-Campos et al., 2018)	Understanding the experience of adults living with pulmonary hypertension	Grey literature
(Tyebally et al., 2016)	Intravenous prostacyclin for pulmonary hypertension: Patient's perspective on complications	Grey literature
(Waldreus et al., 2017)	Patient reported experience measurement of health-care for patients with pulmonary hypertension	Grey literature
(Waldreus et al., 2019)	Development and Validation of a Questionnaire to Measure Patient's Experiences of Health Care in Pulmonary Arterial Hypertension Outpatient Clinics	Used qualitative methodology to address a quantitative research goal

(Walker et al., 2006)	Temporal trends and drug exposures in pulmonary hypertension: an American experience	Did not use qualitative methodology
(Woolcock et al., 2018)	Exploring the delivery of palliative and end of life care in patients with pulmonary arterial hypertension; A U.K based qualitative interview study	Grey literature
(Yorke et al., 2014)	mPHasis- 10: development of a health-related quality of life measure in pulmonary hypertension	Used qualitative methodology to address a quantitative research goal
(Yorke et al., 2018)	Symptom severity and its effect on health-related quality of life over time in patients with pulmonary hypertension: a multisite longitudinal cohort study	Did not use qualitative methodology
(Zhai et al., 2017)	The impact and financial burden of pulmonary arterial hypertension on patients and caregivers: results from a national survey.	Did not use qualitative methodology

PH = Pulmonary Hypertension; UK = United Kingdom

References

Armstrong, I., Billings, C., Kiely, D. G., Yorke, J., Harries, C., Clayton, S., & Gin-Sing, W. (2019). The patient experience of pulmonary hypertension: a large cross-sectional study of UK patients. *BMC Pulmonary Medicine*, *19*, Article 67. https://doi.org/10.1186/s12890-019-0827-5

Armstrong, I., Bundock, S., Harries, C., & Yorke, J. (2011). Living with pulmonary hypertension: A qualitative perspective. *American Journal of Respiratory and Critical Care Medicine*, *183*, A5925. https://doi.org/10.1164/ajrccm-conference.2011.183.1_MeetingAbstracts.A5925

Awdish, R., Small, B., & Cajigas, H. (2015). Development of a modified yoga program for pulmonary hypertension: a case series. *Alternative Therapies in Health and Medicine*, *21*(2), 48-52.

Bonner, N., Abetz, L., Meunier, J., Sikirica, M., & Mathai, S. C. (2013). Development and validation of the living with pulmonary hypertension questionnaire in pulmonary arterial hypertension patients. *Health and Quality of Life Outcomes*, *11*, Article 161. https://doi.org/10.1186/1477-7525-11-161

Caputo, A. (2014). Exploring quality of life in Italian patients with rare disease: A computer-aided content analysis of illness stories. *Psychology Health & Medicine*, *19*(2), 211-221. https://doi.org/10.1080/13548506.2013.793372

Chen, H., Kobashigawa, E. A., Blanc, P. D., De Marco, T., & Napoles-Springer, A. (2011). A qualitative approach to understanding quality of life impairment in pulmonary arterial hypertension: Getting beyond the numbers. *American Journal of Respiratory and Critical Care Medicine*, *183*, A5926. https://doi.org/10.1164/ajrccm-conference.2011.183.1_MeetingAbstracts.A5926

Ferrari, P., & Skara, H. (2019). My life with pulmonary arterial hypertension: A patient perspective. *European Heart Journal Supplements, 21*, K54-K59. https://doi.org/10.1093/eurheartj/suz208

Gray, M. P., Aldrighetti, R., & Fagan, K. A. (2015). Participant expectations in pulmonary hypertension-related research studies. *Pulmonary Circulation*, *5*(2), 376-381. https://doi.org/10.1086/681271

Grinnan, D., Swetz, K., Pinson, J., Lyckholm, L., & Smith, T. (2012). The end-of-life experience of a cohort of patients with pulmonary arterial hypertension. *Journal of Pain and Symptom Management*, *43*(2), 445-446. https://doi.org/10.1016/j.jpainsymman.2011.12.221

Guillevin, L., Armstrong, I., Aldrighetti, R., Howard, L. S., Ryftenius, H., Fischer, A., Lombardi, S., Studer, S., & Ferrari, P. (2013). Understanding the impact of pulmonary arterial hypertension on patients' and carers' lives. *European Respiratory Review, 22*(130), 535-542. https://doi.org/10.1183/09059180.00005713

Halimi, L., Marin, G., Molinari, N., Gamez, A.-S., Boissin, C., Suehs, C. M., Vachier, I., & Bourdin, A. (2018). Impact of psychological factors on the health-related quality of life of patients treated for pulmonary arterial hypertension. *Journal of Psychosomatic Research*, *105*, 45-51. https://doi.org/10.1016/j.jpsychores.2017.12.003

Hall, H., Cote, J., & McBean, A. (2007). The experiences of patients with pulmonary arterial hypertension (PAH) receiving continuous intravenous infusion of epoprostenol (Flolan (TM)), and their support persons. *Canadian Journal of Cardiology*, *23*, 323C.

Hall, S. S., MacMichael, J., Turner, A., & Mills, D. S. (2017). A survey of the impact of owning a service dog on quality of life for individuals with physical and hearing disability: A pilot study. *Health and Quality of Life Outcomes*, *15,* Article 59. DOI 10.1186/s12955-017-0640-x

Ivarsson, B., Ekmehag, B., Hesselstrand, R., Radegran, G., & Sjöberg, T. (2014). Perceptions of received information, social support, and coping in patients with pulmonary arterial hypertension or chronic thromboembolic pulmonary hypertension. *Clinical Medicine Insights: Circulatory, Respiratory and Pulmonary Medicine*, *8*, 21-28. https://doi.org/10.4137/CCRPM.S18586

Ivarsson, B., Sjoberg, T., Hesselstrand, R., Radegran, G., & Kjellstrom, B. (2019). Everyday life experiences of spouses of patients who suffer from pulmonary arterial hypertension or chronic thromboembolic pulmonary hypertension. *European Respiratory Journal Open Research*, *5*, 00218-2018. https://doi.org/10.1183/23120541.00218-2018

Jones, D. L., Sandberg, J. C., Rosenthal, M. J., Saunders, R. C., Hannig, V. L., & Clayton, E. W. (2008). What patients and their relatives think about testing for BMPR2. *Journal of Genetic Counseling*, *17*(5), 452-458. https://doi.org/10.1007/s10897-008-9172-1

Khan, E. U. (2011). Commentary on symptom experience of pulmonary arterial hypertension (PAH) patients. *Clinical Nursing Research*, *20*(2), 135-143. https://doi.org/http://dx.doi.org/10.1177/1054773810395112

Kobayashi, N., Tamura, Y., Takeda, M., Yasuda, K., Kurihara, H., Satoh, R., Aihara, Y., Kayama, R., Murata, T., Satoh, T., & Fukuda, K. (2012). Quantitative and qualitative analysis of health related quality of life in patients with pulmonary arterial hypertension (CP). *Quality of Life Research*, *20*, 63.

Lichenstein, S., McDonough, A., & Matura, L. A. (2013). Cyber support: Describing concerns of caregivers of people with pulmonary hypertension. *Computers Informatic Nursing*, *31*(12), 581-588. https://doi.org/10.1097/cin.0000000000000008

MacKenzie, A., & Johnson, M. (2015). Patient perspectives on exercise training in pulmonary arterial hypertension (PAH). *European Respiratory Journal*, *46*, PA3779 https://doi.org/10.1183/13993003.congress-2015.PA3779

McGoon, M. D., Ferrari, P., Armstrong, I., Denis, M., Howard, L. S., Lowe, G., Mehta, S., Murakami, N., & Wong, B. A. (2019). The importance of patient perspectives in pulmonary hypertension. *European Respiratory Journal*, *53*(1), Article 1801919. https://doi.org/10.1183/13993003.01919-2018

McKenna, S. P., Doughty, N., Meads, D. M., Doward, L. C., & Pepke-Zaba, J. (2006). The Cambridge Pulmonary Hypertension Outcome Review (CAMPHOR): A measure of health-related quality of life and quality of life for patients with pulmonary hypertension. *Quality of Life Research, 15*(1), 103-115. https://doi.org/http://dx.doi.org/10.1007/s11136-005-3513-4

Monson, A., & Pennell, S. (2012). The experience of the older adult with pulmonary arterial hypertension. *Gerontologist*, *52*, 91-91.

Mulgirigama, A., Hunt, D., Wilson, R., & Pepke-Zaba, J. (2012). A slower life in a smaller world. Patients' perspective on living with pulmonary arterial hypertension. *European Respiratory Journal*, *40*(65)*, P966.*

Nakanishi, T. (2015). Pulmonary arterial hypertension associated with congenital heart disease. Personal perspectives. *International Heart Journal*, *56*, S1-S3. https://doi.org/10.1536/ihj.15-064

Peloquin, J., Robichaud-Ekstran, S., & Pepin, J. (2007). Perception of quality of life by women with stage III or IV primary pulmonary hypertension and receiving treatment with prostacyclin. *Recherche en Soins Infirmiers,* (88), 51-63.

Quijano-Campos, J. C., Doughty, N., & Cross, J. (2018). Understanding the experience of auts living with pulmonary hypertension. *Thorax*, *73*, A207. https://doi.org/10.1136/thorax-2018-212555.353

Tyebally, S., Sturdy, A., Khokhar, A., Parfitt, L., Harries, C., Price, L., Wort, J., Alonso, R., Dimopoulos, K., & Gatzoulis, M. (2016). Intravenous prostacyclin for pulmonary hypertension: Patient's perspective on complications. *European Respiratory Journal*, *48,* PA2481. https://doi.org/10.1183/13993003.congress-2016.PA2481

Waldreus, N., Jaarsma, T., Ivarsson, B., Stromberg, A., Arestedt, K., & Kjellstrom, B. (2019). Development and validation of a questionnaire to measure patient's experiences of health care in pulmonary arterial hypertension outpatient clinics. *Heart Lung and Circulation*, *28*(7), 1074-1081. https://doi.org/10.1016/j.hlc.2018.07.011

Waldreus, N., Jaarsma, T., Ivarsson, B., Stromberg, A., & Kjellstrom, B. (2017). Patient reported experience measurement of health-care for patients with pulmonary hypertension. *European Journal of Cardiovascular Nursing*, *16*, S95-S96.

Walker, A. M., Langleben, D., Korelitz, J. J., Rich, S., Rubin, L. J., Strom, B. L., Gonin, R., Keast, S., Badesch, D., Barst, R. J., Bourge, R. C., Channick, R., Frost, A., Gaine, S., McGoon, M., McLaughlin, V., Murali, S., Oudiz, R. J., Robbins, I. M., Tapson, V., Abenhaim, L., & Constantine, G. (2006). Temporal trends and drug exposures in pulmonary hypertension: an American experience. *American Heart Journal*, *152*(3), 521-526. https://doi.org/10.1016/j.ahj.2006.02.020

Woolcock, S., Hamilton, D. W., Corris, P. A., & Exley, C. (2018). Exploring the delivery of palliative and end of life care in patients with pulmonary arterial hypertension; A U.K based qualitative interview study. *American Journal of Respiratory and Critical Care Medicine*, *197*, A7594

Yorke, J., Corris, P., Gaine, S., Gibbs, S. S., Kiely, D. G., Harries, C., Pollock, V., & Armstrong, I. (2014). emPHasis- 10: development of a health- related quality of life measure in pulmonary hypertension. *European Respiratory Journal, 43,* 1106-1113. 10.1183/09031936.00127113

Yorke, J., Deaton, C., Campbell, M., McGowen, L., Sephton, P., Kiely, D. G., & Armstrong, I. (2018). 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 Respiratory Research*, *5*, Article e000263. https://doi.org/10.1136/bmjresp-2017-000263

Zhai, Z., Zhou, X., Zhang, S., Xie, W., Wan, J., Kuang, T., Yang, Y., Huang, H., & Wang, C. (2017). The impact and financial burden of pulmonary arterial hypertension on patients and caregivers: results from a national survey. *Medicine*, *96*(39), Article e6783. https://doi.org/10.1097/md.0000000000006783

Section Two: Research Report

Development and pilot randomised controlled trial of a self-help intervention, based on Cognitive Behavioural Therapy, for anxiety in adults with pulmonary hypertension

Abstract

Objective: People with pulmonary hypertension (PH) are at an increased risk of experiencing anxiety disorders. This study developed and tested the acceptability, feasibility and preliminary effectiveness of a cognitive behavioural self-help intervention for anxiety in adults with PH using a pilot randomised control trial design.

Methods: Guided by the Medical Research Council framework for developing complex healthcare interventions, a four-week self-help intervention was first developed. Next, individuals with PH impacted by anxiety recruited from Pulmonary Hypertension Associations were randomised to either receive the self-help intervention (n=37) or a wait-list condition (n=40). Acceptability was explored using mixed-methods questionnaires. A 2x3 repeated-measures analysis of variance was used to explore anxiety (GAD-7), depression (PHQ-9), health-related quality of life (emPHAsis-10), dyspnoea (D-12), self-mastery (Self-Mastery Scale) and mood-related cognitions and behaviours (CBP-Q) at baseline, post-intervention and one-month follow up. A mediation analysis was performed to examine potential mechanisms of change.

Results: Dropout was low at 15.6%. All participants were likely to recommend the intervention to another person with PH and felt it had helped with their anxiety. Participants in the intervention group reported a significant reduction in anxiety, depression and cognitive and behavioural processes linked with mood disorders, compared to the control group. Change in unhelpful cognitions and behaviours mediated the relationship between intervention condition and change in anxiety and depression scores.

Discussion: The intervention was found to be acceptable and feasible. Findings add to the growing evidence supporting the use of psychological therapies in people with PH.

Practitioner points

- Mood disorders are prevalent in people with pulmonary hypertension.
- Cognitive behavioural therapy can be helpful for anxiety in people with cardiovascular conditions.
- A cognitive behavioural therapy informed self-help intervention for anxiety in pulmonary hypertension was found to be acceptable and feasible.
- Preliminary evidence suggests that the intervention was associated with improvements in anxiety and depression.

Key words

CBT; Self-Management; Pulmonary Arterial Hypertension; Mental Health; Psychological therapy

Development and pilot randomised controlled trial of a self-help intervention, based on Cognitive Behavioural Therapy, for anxiety in adults with pulmonary hypertension

Pulmonary hypertension (PH) is a life-altering disease associated with high rates of morbidity and mortality (Halimi et al., 2018; Hoeper, Kramer, et al., 2017). There are five different groups of the disease, each associated with different aetiological factors manifesting in cardiorespiratory symptoms, such as breathlessness and fatigue (Hoeper, Ghofrani, et al., 2017), which are typically more debilitating in those with severe PH (Yorke et al., 2018). PH severity is measured using the World Health Organisation (WHO) functional classification system. This ranges from class I, in which mild symptoms may be experienced during physical activity, to IV suggesting symptoms will be reported at rest (McGoon et al., 2004).

Individuals with PH are at an increased risk of experiencing lower health-related quality of life (HRQoL) and psychological difficulties (Lowe et al., 2004; Wryobeck et al., 2007; Yorke et al., 2014; Yorke et al., 2018). Research has identified depression and anxiety disorders in up to 50% of participants with PH (Bussotti & Sommaruga, 2018). Moreover, such figures may be underestimated as mental health conditions are often underrecognised in this group (Wryobeck et al., 2007); one study found less than one-quarter (n=14/58) of individuals with PH were receiving medication (n=8), psychological treatment (n=2) or both (n=4) for difficulties identified using the Patient Health Questionnaire-9 (PHQ-9) (Kroenke et al., 2001; Lowe et al., 2004).

Anxiety is a pertinent issue in PH due to its prevalence, the physiological effects on cardiovascular functioning which could exacerbate difficulties

(Schachinger et al., 2000) and overlapping symptoms; for instance, breathlessness, dizziness, fatigue and chest pain could be caused by anxiety or PH. Anxiety has been conceptualised as a psychological, physiological and behavioural response to stimuli perceived by the individual as aversive or threatening. It is viewed as an adaptive function to help manage a stressor (Steimer, 2002) and therefore, it can be important to normalise anxiety as a healthy reaction to certain challenges, such as living with a chronic illness (Lebel et al., 2020). However, anxiety can become unhelpful and pathological; for example, when the intensity, duration or frequency interferes with everyday life (Steimer, 2002). Anxiety in PH is associated with reduced HRQoL and depression (Rawlings, Thompson, et al., 2021; Yorke et al., 2018) – and mortality risk in those with other heart and lung conditions (Panagioti et al., 2014; Watkins et al., 2013).

There is limited research examining treatments for anxiety in PH. In terms of psychological interventions, a systematic review investigating a sub-group of PH, pulmonary arterial hypertension (PAH), found only three trials, all of which used Cognitive behavioural Therapy (CBT) or strategies commonly associated with CBT, such as psychoeducation, relaxation techniques and guided breathing (Bussotti & Sommaruga, 2018). CBT aims to help people alter their attitudes and behaviours by exploring their cognitions, emotions, physiological reactions and behaviours, and develop adaptive coping skills (Shafran et al., 2013). CBT informed interventions were associated with significant improvements in anxiety, depression and HRQoL in PH. The authors concluded while CBT is associated with benefits in other cardiovascular conditions (Johnston, 2000), there is a paucity of evidence regarding CBT in PH (Bussotti & Sommaruga, 2018). Furthermore, recent evidence suggests psychological treatments targeting mood may also have a preventative function in PH.

A randomised control trial (RCT) compared unspecified psychological counselling for anxiety and depression in people with PAH versus no counselling finding those receiving counselling reported a reduction in anxiety and depression, while mood symptomatology increased in the other group (Tarantino et al., 2020).

The National Institute for Health and Care Excellence (NICE) recommend stepped care for treating anxiety and panic disorders (National Institute for Health and Care Excellence, 2019). For those with less severe symptoms, self-help approaches, informed by CBT, are recommended. If individuals are experiencing difficulties following treatment or their initial needs are too severe, care should be stepped up to more intensive interventions, including psychopharmacology. This approach means that individuals receive a level of care appropriate to their needs, while at the same time optimising resource (Webster et al., 2014).

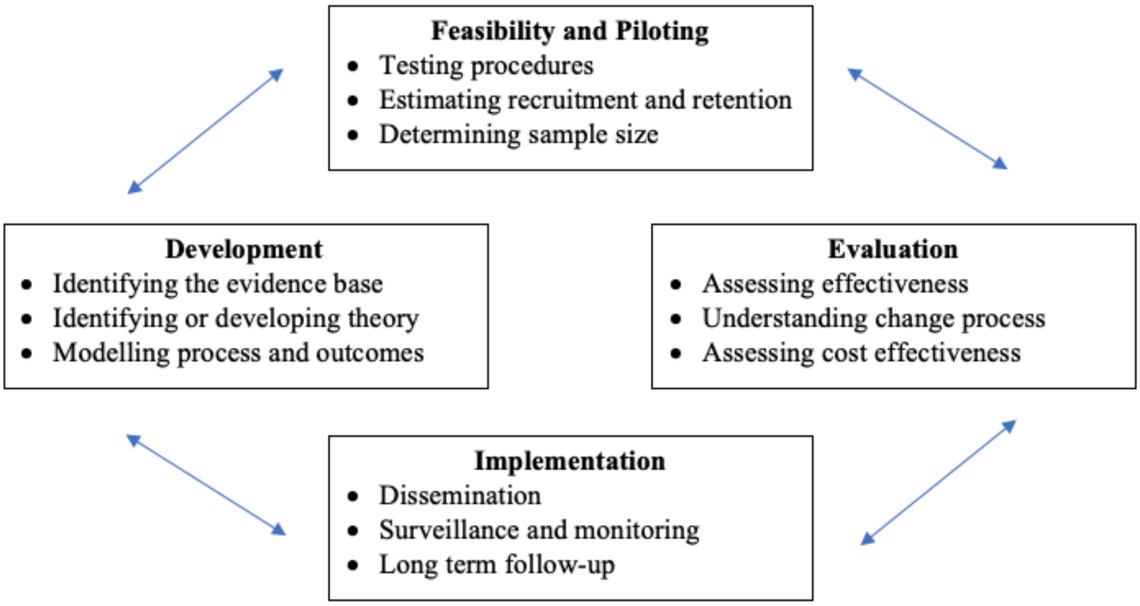
Self-help interventions typically involve individuals working through a series of materials discussing psychoeducation and self-management techniques (Cuijpers & Schuurmans, 2007). There is growing evidence suggesting self-management interventions offer an accessible and effective form of treatment (Webster et al., 2014). What is more, condition-specific self-help interventions for psychological difficulties have been developed, which have the benefit of addressing general and particular challenges observed within clinical groups (Howard & Dupont, 2014; Pasterfield et al., 2019; Novakova et al., 2019).

Aims

The primary aim of the current study was to develop a CBT informed self-help intervention targeting anxiety in adults with PH. A theory- and evidence-based approach was used, utilising developmental guidelines for healthcare interventions (O'Cathain et al., 2019; Yardley et al., 2015), and more precisely, the Medical Research Council (MRC) Framework (Craig et al., 2008) (Figure 1). This framework has four iterative phases: developing an intervention informed by the available evidence-base; next, aspects of the intervention and proposed method of evaluation should be tested using feasibility, acceptability or pilot studies; after necessary refinements based on findings from previous phases and if suitable, effectiveness can be investigated using systematic methodologies; and finally, the intervention can be implemented, if appropriate. The framework has been used to develop self-help interventions for people living with chronic conditions (Bobrow et al., 2018).

Figure 1

Medical Research Council Framework



A secondary aim was to conduct a pilot RCT investigating the newly developed self-help intervention against a wait-list control condition, with the intention of informing a future definitive trial.

The terms feasibility and pilot studies are often erroneously used interchangeably (Whitehead et al., 2014). A conceptual framework has been developed defining a feasibility study as investigating whether an element of a future RCT is suitable, whereas a pilot study is a miniature version of a definitive trial, which may also investigate feasibility (Eldridge, Lancaster, et al., 2016). A pilot RCT does not hypothesis-test, but rather explores proposed procedures and obtains estimates for sample size calculations for a larger trial (Leon et al., 2010; Whitehead et al., 2016).

The design, method for recruitment and data collection for a definitive study had already been identified, as the final self-help intervention was to be made available to all 4500 members of a United Kingdom (UK)-based charity for people with PH, Pulmonary Hypertension Association (PHA UK), who also funded the current project. Therefore, a pilot study was selected to first test this approach on a smaller scale.

Objectives

The objective of the pilot RCT was to explore the feasibility and acceptability of the proposed design of a definitive trial. More precisely, examining recruitment methods, randomisation, attrition, implementation of a novel self-help intervention for anxiety in PH, and completion of health-related questionnaires.

As a secondary objective, preliminary effectiveness of the intervention was investigated to help inform a sample size calculation for a definitive trial.

Furthermore, potential active mechanisms of change associated with the trial were explored to help understand change processes. More specifically, a mediation analysis was used to examine whether changes in cognitions and behaviours related to mood explained the relationship between intervention condition and significant changes in health-related outcomes.

Methods

Phase One: Intervention Development

Identifying Existing Evidence

To identify the existing evidence-base regarding what is currently known about the phenomenology of living with PH and how people cope, a systematic review of qualitative studies examining adults' accounts was performed (Rawlings et al., 2020). The findings first helped to identify anxiety as a possible therapeutic target in PH; for example, participants reported uncertainty in response to different PH-related challenges. This included the often-lengthy delay in obtaining a diagnosis of PH, experience of PH-related symptoms; for example, breathlessness and panic were closely linked; the meaning, cause and progression of symptoms; treatment options; prognosis; and the reactions of others to the disease. Individuals engaged in worrying to plan or problem solve, and sought reassurance as a method of coping. Unfortunately, due to the unpredictable and complex nature of PH, this was not always possible or helpful. Participants described focusing on somatic symptoms as a method of monitoring the progression of PH; however, this resulted in anticipatory anxiety and given the overlap between symptoms of PH and anxiety, it may be that individuals found it difficult to differentiate between normal and potentially harmful responses.

The link between anxiety and PH was then further examined by investigating quantitative findings previously discussed regarding the prevalence, nature and treatment. Finally, the aforementioned evidence was used to select appropriate theories (see below) and inform the intervention development. For instance, helping to develop case examples.

Identifying Theory

As evidence suggests anxiety in PH appears responsive to psychological treatment, particularly CBT (Bussotti & Sommaruga, 2018), theories related to CBT for anxiety were selected to guide the intervention. More specifically, theories of generalised anxiety disorder (GAD), panic disorder and health anxiety, which have all previously been documented as prevalent in this group (Bussotti & Sommaruga, 2018; Rawlings et al., 2021) were used. GAD can be viewed as a strong reaction to uncertainty, which is often managed through excessive worry and reassurance seeking (Dugas & Robichaud, 2007); panic disorders are formulated as a catastrophic misinterpretation of bodily sensations (Clark, 1986); and similarly, misinterpretation and hypervigilance of somatic experiences are believed to maintain health anxiety (Salkovskis et al., 2002).

The content of CBT can differ depending on the client's needs and available resource as treatment can use a longitudinal perspective, focusing on how past events may have influenced current difficulties, or adopt a specific focus on experiences related to the here and now. A longitudinal approach can be more intensive requiring additional facilitation; for example, discussing early life experiences. Therefore, a here and now perspective was utilised. Generally speaking, CBT for anxiety has three main cyclic phases (assessment, the use of cognitive and behavioural strategies, and relapse prevention) (Shafran et al., 2013), which were all represented in the current intervention:

(a) Participants first explored their anxiety and identified targets for treatment. The four-areas model was used to help facilitate understanding and socialise participants to key principles of CBT. This is a trans-diagnostic cross-sectional CBT formulation conceptualising the complex interaction between cognitive, emotional, physiological and behavioural factors (Padesky & Mooney, 1990). Additional psychoeducational resources were discussed with the aim of helping individuals to develop a greater and non-judgemental understanding of their difficulties.

(b) Individuals were asked to focus on cognitive and behavioural processes that may be maintaining their current difficulties. Participants practiced challenging their unhelpful thinking styles and behaviours, and developed more helpful ways of coping. Typically, graded exposure is an important mechanism in CBT in which the individual repeatedly engages in anxiety-provoking activities until habituation. This strategy may also have a bi-directional impact on anxiety; for instance, depression is highly prevalent and related to anxiety in PH (Rawlings, Thompson, et al., 2021) with behavioural engagement being an important element in depression-related treatments (Ekers et al., 2014). Moreover, given fatigue is a common and debilitating symptom in people with PH (Tartavoulle et al., 2018), the current intervention was adapted using principles of pacing, which aimed to help participants build stamina and prevent exhaustion from performing behaviours (Connolly et al., 2013).

(c) To conclude, participants focused on consolidating therapeutic gains and identifying possible setbacks. Individuals took an active role in devising strategies for prevention of relapse. Further psychoeducation was used; for example, focusing on other behavioural strategies to help with anxiety such as diet and sleep.

Modelling Process and Outcomes

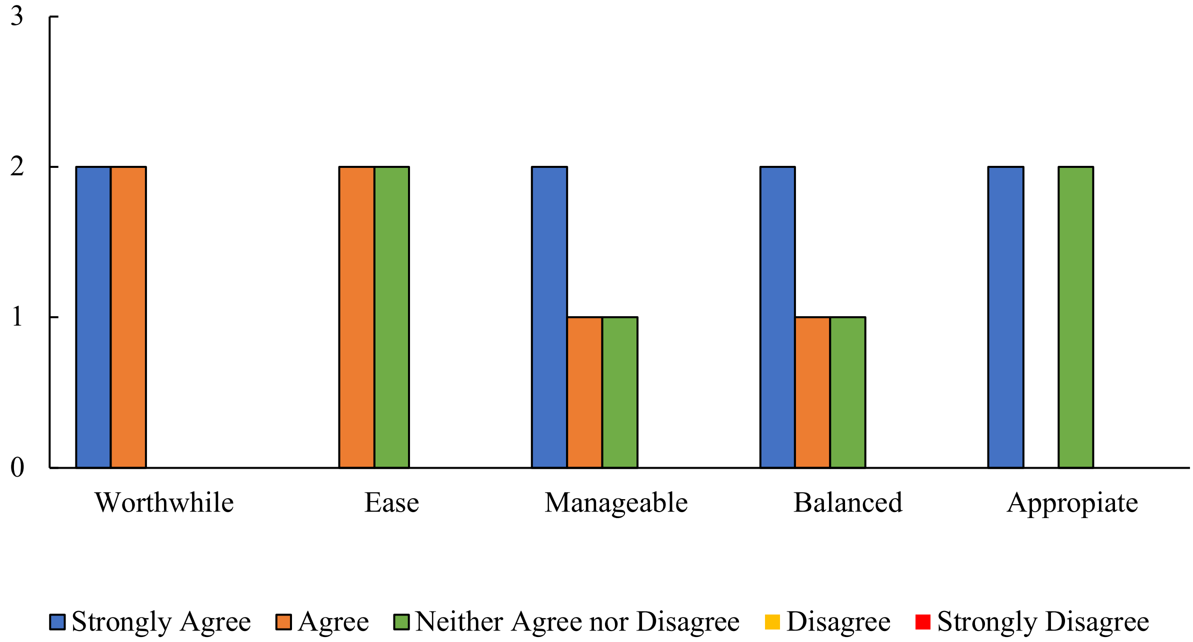
A multi-disciplinary stakeholders' group was established in collaboration with PHA UK to develop and model the intervention. The group comprised of adults living with PH and caregivers (n=4), clinical psychologists (n=2), PH nurse specialists (n=2), professionals from PHA UK (n=3), a cognitive behavioural therapist and graphic designer. The team agreed on the target problem, and approach to developing the intervention. The lead researcher developed the intervention guided by clinical experience and the aforementioned evidence-base, including previous condition-specific self-help interventions (Anderson et al., 2011; Anxiety Canada, n.d.; Khazan, 2013; Mander & Cameron, 2013.; Psychology Tools Limited, n.d.; Pasterfield & Thompson, 2013; Shafran et al., 2013; Therapist Aid, 2017; Whalley & Kaur, 2020). The lead researcher kept in regular contact with members of the stakeholders' group through a series of meetings.

To further refine the intervention, four individuals with PH and their caregivers, who were members of the stakeholders' group, were asked to review the intervention. A questionnaire was developed by the lead researcher for this purpose collecting quantitative and qualitative information (Appendix A). Group members were asked to rate, on a 5-point Likert scale ("*Strong Agree*" to "*Strongly Disagree*"), whether the intervention was worthwhile, easy to read and understand, manageable given the workload, had a good balance between text and images, and the examples were appropriate (Figure 2). Members were asked what they liked and disliked about the intervention, and for any other comments.

All members reported "*Agree*" or "*Strongly Agree*" that the intervention was worthwhile, manageable and well-balanced. Members valued that the different phases of CBT were split up and people could work through it step by step. There was an equal split between members when asked about its ease and appropriateness of examples. As such, a number of refinements were made, which included reducing long sentences and modifying case vignettes. Once the content was finalised, the layout was designed by a designer as part of the stakeholders' group.

Figure 2

Results from stakeholder's questionnaire(n=4)



Final Intervention

The final intervention consisted of a four-week programme corresponding to four individual booklets (Table 1 and Appendix B). The length of each booklet varied between 19–27 A5 pages. It was recommended to participants to read each booklet per week and in order.

Table 1

Outline of the intervention

Booklet	Content	Exercises
1 'Anxiety and PH'	• Introduction to CBT model • Introduction to the intervention • Normalise difficulties with anxiety through case studies • Psychoeducation on psychosocial impact of PH, anxiety (panic, fear, stress and worry) and anxiety in PH • Highlight the importance of coping strategies	•Explore and identify common symptoms of anxiety •Complete the four-areas model – develop a mini-formulation of difficulties •Slow breathing exercise
2 'What keeps anxiety going'	• Psychoeducation on anxiety – focusing on avoidance, reassurance seeking, safety behaviours, symptoms and unhelpful thoughts about health – all specific to experiences of PH	• Explore and recognise maintenance cycles • Develop a here and now formulation of difficulties recognising triggers, reactions, unhelpful coping strategies and short- and long-term consequences • Two mindfulness exercises – focusing on bodily symptoms and everyday events
3 'The power of thoughts'	• Psychoeducation on cognitive biases grounded in the four-factor model: predicting the future, mind reading, over-generalisation, all-or-nothing thinking and self-criticism • Challenging unhelpful thoughts – evidence for and against • Normalise difficulties with anxiety through case studies • Develop techniques to help manage worry	• Recognise unhelpful thinking biases • Thought challenging • Worry tree • Worry delay • Thought monitoring exercise

4 ‘The power of your actions’	• Psychoeducation on graded exposure to manage anxiety; pacing, and boom and bust cycle to manage pain and fatigue • Psychoeducation on other factors affecting anxiety: routine, diet, sleep, social support, keeping physically active and confidence/assurance • Relapse prevention reviewing what participants have learnt, skills they have developed, and identifying and managing setbacks • Signpost to other services for mental health and PH-related problems	• Development of anxiety hierarchy • Development of pacing hierarchy • Create a plan to cope with setbacks • Progressive muscle relaxation exercise

CBT = Cognitive Behavioural Therapy; PH = Pulmonary Hypertension

Phase Two: Piloting

Trial Design

The trial has been conducted in accordance with CONSORT guidelines (Eldridge, Chan, et al., 2016) (Appendix C). Data was collected between August 2020 – April 2021 with recruitment ending in February 2021, when the sample size was achieved. Participants were block randomised 1:1 (first in a block of 64, then in blocks of ten to account for dropout (see Sample Size Analysis)), using an online randomiser (https://www.random.org), to one of two groups: CBT self-help intervention for anxiety in PH or a wait-list condition. All participants were asked to complete a series of measures (see measures) at baseline, post-intervention and one-month post-intervention. Participants and researchers were not blinded to condition allocation.

Ethical approval was obtained from the Department of Psychology Ethics Committee at the University of Sheffield (034442) (Appendix D and E). For any safeguarding concerns, participants were signposted to other services and the event would be discussed with the research team with the option of stopping the trial if necessary- however none of these steps were required. The study was registered at clinicaltrials.gov: NCT04450862 (Appendix F).

Sample Size Analysis

As a pilot study does not test hypotheses, traditional sample size calculations are not appropriate (Bell et al., 2018). However, Whitehead et al., propose an optimal solution if the effect size of a larger trial is known, to justify sample sizes of a pilot study (Whitehead et al., 2016). A meta-analysis of 54 self-help interventions for anxiety reported an average effect size of 0.53 (Hedges' *g)* for clinical samples (95% Confidence Interval (CI)=0.31-0.76) (Haug et al., 2012). Therefore, utilising a RCT design with a standardised effect size of 0.5, 90% power and 5% type I error rate, a sample size of 32 was required in each arm. However, an average dropout rate of 16.5% (range 0-54.8%) was reported, therefore this was revised to n=76.

Eligibility Criteria

To be eligible, participants were required to: be aged 18 years or over; have a diagnosis of PH; provide informed consent (Appendix G); complete a series of self-report questionnaires independently in English; and have a clinical need. To examine clinical need, participants were required to respond "yes" (from a "yes" or "no" option) when asked if they had difficulty with anxiety (Appendix H). A standardised screening method could not be used as no anxiety-specific measure has been in PH.

NICE guidelines recommend people with severe symptoms of anxiety may not be suitable for self-help interventions; therefore, individuals were not eligible if they self-reported experiencing thoughts of self-harm or suicide. Initially, people who had received treatment for anxiety in last twelve-months were not eligible; however, after receiving feedback from PH-clinicians, this stipulation was removed in November 2020 (Appendix D and E). Individuals who were not eligible were given the option of contacting the lead researcher for further signposting – no individuals made contact.

Procedure

A convenience sampling method was used. Participants were recruited via advertisements on social media platforms (Appendix I) of Pulmonary Hypertension Associations in the UK, Canada and India. The advert directed people to a webpage containing the participant information sheet (Appendix J). If individuals were still interested after reading the information sheet, they were asked to complete the eligibility form (Appendix H). If suitable, participants were then asked to complete a consent form, and demographic and health-related questionnaires (see Measures). Participants were then randomised by the lead researcher.

Those in the intervention condition were sent the intervention. Initially, all participants were sent a paper-version. However, as discussed, the intention was to make the final intervention available to all PHA UK members. Although this is a UK based charity, they are contacted by people outside of the UK, with whom it is more practical to correspond electronically. In accordance with how the intervention would be used in the real world and in a definitive trial and to help maximise recruitment, from November 2020, UK participants continued to receive a paper version, while international participants received an electronic version (Appendix D) – at this stage, only two international participants were enrolled, neither of whom had received a paper version. We also approached international PH organisations to support with advertising the study. Results were not stratified based on type of intervention.

Two weeks after the intervention was sent, UK participants were contacted by telephone and international participants by email, by the lead researcher to assess adherence and acceptability, using a semi-structured questionnaire (see Measures, Table 5 and Appendix K). Participants were informed the phone call should last approximately ten minutes. Overall, n=10 were successfully contacted via email and n=22 via telephone and responded with calls lasting on average 12.36 minutes (Standard Deviation (SD)=12.25 minutes). Given the aim of this contact, the researcher did not provide any recommendations or advice, and instead, normalised or summarised participants' experiences related to the intervention.

Participants randomised to the wait-list condition were informed that they would be contacted again in four-weeks.

After four weeks, participants in both conditions were asked to complete the health-related questionnaires, which were repeated one-month later. Participants in the intervention group were then asked to complete a semi-structured acceptability questionnaire (see Measures, Tables 6-9 and Appendix L). Participants in the control group were informed if the intervention was acceptable, they would receive a copy in the future. Questionnaires were hosted by Qualtrics (https://www.qualtrics.com).

Measures

To obtain demographic and clinical factors, participants were asked to record their name, date of birth, gender, contact details, employment status, years of education, ethnicity, diagnosis of PH, PH functional class, duration of PH and whether they had received treatment for anxiety in the last twelve-months – this was to assess whether participants may have received a similar intervention as to the one being examined, which could influence their responses (Appendix M).

Primary Outcome Measure

Anxiety was measured using the GAD-7 questionnaire (Spitzer et al., 2006) (Appendix N). Participants were asked to report on a Likert scale ("*Not at all*", "*Several days*", "*More than half the days*", "*Nearly every day*"), how often over the last two weeks had they been bothered by seven anxiety-related difficulties. A score of 0–4 indicates "*minimal difficulties*", 5-9 "*mild*", 10–14 "*moderate*" and 15–21 "*severe*" anxiety. The clinical cut-off score is ≥8 with a reliable change of ≥4 (Clark & Oates, 2014). The measure has been used in people with PH (Harzheim et al., 2013). In the current study, excellent internal consistency was observed (Cronbach's Alpha=0.91) – ≥0.9=excellent, ≥0.8=good, ≥0.7=acceptable, ≥0.6=poor, ≤0.5=unacceptable (George & Mallery, 2003).

Secondary Outcome Measures

Depression is related to anxiety in PH (Rawlings, Thompson et al., 2021) and was measured using the PHQ-9 (Kroenke et al., 2001) (Appendix O). Respondents were asked to rate on a four-item Likert scale ("*Not at all*", "*Several days*", "*More than half the days*", "*Nearly every day*") how often over the last two weeks had they been bothered by nine depression-related difficulties. A score of 0-4 indicates "*minimal difficulties*", 5-9 "*mild*", 10-14 "*moderate*", 15-19 "*moderately severe*" and ≥20 "*severe*" depression. The clinical cut-off score is ≥10, with a reliable change of ≥6 (Clark & Oates, 2014). This measure has been used in people with PH. It had good internal consistency (Cronbach's Alpha=0.84).

HRQoL is associated with anxiety in PH (Yorke et al., 2018), and was examined using a condition-specific measure, emPHAsis-10 (Yorke et al., 2014) (Appendix P). Participants were asked to rate on a six item Likert scale, how much they had recently experienced ten PH-related challenges. Scores range from 0-50, with a higher score indicative of lower HRQoL. The measure had good internal consistency (Cronbach's Alpha=0.89).

Breathing difficulties are a cardinal symptom of PH and linked to panic (Rawlings et al., 2020); therefore this was explored using the Dyspnoea 12 (D12) (Yorke et al., 2010) (Appendix Q). Participants were asked to rate twelve questions in relation to their breathlessness "*these days*", on a Likert scale ("*minimal*", "*mild*", "*moderate*" or "*severe*"). Total scores range from 0-36 with a higher score indicating greater difficulty. The measure has been used in people with PH (Yorke et al., 2018). It had excellent internal consistency (Cronbach's Alpha=0.95).

Self-mastery reflects a person's perception of their own ability to cope (Pearlin & Schooler, 1978). Given the relationship between coping and anxiety in PH (Rawlings et al., 2021), coping was measured using the Self-Mastery Scale (Pearlin & Schooler, 1978)

(Appendix R). Respondents were asked to rate seven coping-related questions on a seven-point Likert scale ranging from "*Strongly Agree*" to "*Strongly Disagree*". Total scores range from 7-49 with a higher score suggesting a greater perceived capacity to cope. Good internal consistency was reported (Cronbach's Alpha=0.8).

To measure changes in how participants respond to unhelpful cognitions, emotions and behaviours, which may underlie anxiety and therefore act as a mechanism of change, the Cognitive and Behavioural Processes Questionnaire (CBP-Q) was used (Patel et al., 2015) (Appendix S). This is a 15-item transdiagnostic measure asking eight questions related to cognitions and seven to behaviours. Participants were asked to rate questions using a nine-item Likert scale, which different depending on the item. Total scores range from 0-120. Higher scores are associated with more unhelpful responses, which have been found to positively relate to anxiety and depression symptomatology (Patel et al., 2015). Internal consistency was good (Cronbach's Alpha=0.86).

Adherence and Acceptability Questionnaires

Two mixed-methods questionnaires (Check-In Questionnaire and Final Acceptability Questionnaire) were developed by the lead researcher to collect information concerning adherence and acceptability part-way through and at the end of the study (Appendix K and L). Participants were asked about their experiences of anxiety in PH, the different aspects of the intervention and their engagement in the research trial. Participants were asked to respond to structured questions using a five-item Likert scale and were given the space to write their answers to open-ended questions (Tables 5-9).

Data Analysis

Quantitative Data. A series of independent samples *t*-tests and Pearson's chi-square tests were performed to investigate group differences between CBT vs. control group to explore randomisation; UK vs. international (non-UK) participants to examine whether it was suitable to group participants together; and completers vs. non-completers investigating possible variables impacting attrition – non-completers were defined as participants who did not complete measures at post-intervention or follow-up.

Results from the adherence and acceptability questionnaires were summarised using descriptive statistics (e.g. mean and standard deviations).

To examine preliminary effectiveness, a series of two-way mixed analysis of variance (ANOVA) were ran for each outcome measure examining the interaction, with condition representing the between subjects factor and time the within subjects factor. Regarding assumptions: outliers were explored but retained to increase the number of data points. Five variables violated the assumption of normality (Shapiro-Wilk's test $p<0.05$), however as there is not a non-parametric alternative, violations have just been reported. Levene's test of homogeneity of variance and Mauchly's test of Sphericity were performed and violations reported. If sphericity was violated, Huynh-Feldt correction was used as all epsilon corrections were >0.75 (Laerd Statistics, 2018). Data were not transformed to preserve the clarity of original data and given that the study was not designed to test effectiveness.

Where the interaction (group x time) was significant, a series of independent samples *t*-tests were performed to compare differences at each time point between the two groups.

A series of repeated measures ANOVAs was then used to investigate effects of time for each condition for all outcome measures. Bonferroni correction was used for post-hoc pairwise comparisons to prevent type 1 errors given the number of tests performed. The alpha level (0.05) was accepted for all other tests.

G*Power-3 (2020) was used to perform the sample size calculation to inform a future trial, using the effect size of the primary outcome measure. Partial Eta Squared was converted to Cohen's *f* using G*Power-3, which were interpreted using Cohens' benchmarks (f=0.1 small, 0.25 medium and 0.4 large) (Cohen, 1988).

To explore mechanisms of change, a series of mediated regression analyses were performed to investigate whether the effects of condition on significant changes in health-outcomes were mediated by change in CBP-Q scores. Regarding assumptions, a linear relationship was observed, no multicollinearity was reported (<0.8), residuals were independent (Durbin-Watson=1.61-1.78), homoscedasticity was met and data was normally distributed using plots. To assess significance, 95% bootstrapped confidence intervals were assessed (5000 bootstrap samples). Change in scores was calculated by subtracting scores at baseline from one-month follow-up. An online calculator was used to perform a Sobel test (Preacher & Leonardelli, 2021). Cohen's benchmarks were used to interpret effect sizes (r^2=0.02 small, 0.15 medium and 0.35 large) (Cohen, 1988).

SPSS25 (IBM Corp. 2017) was used for all other statistical analysis with Hayes' process macro extension (Hayes, 2021) for the mediation analyses.

Qualitative Data. Qualitative data collected from the adherence and acceptability questionnaires were analysed using quantitative content analysis (Huxley, 2020). This approach aims to identify patterns and recurring categories within datasets. Overriding categories were first identified by the lead researcher, which corresponded to the open-ended questions (Table 9). After reading the full dataset, a coding scheme was developed by the lead researcher based on responses to the Acceptability Questionnaire. This was shared amongst members of the research team to achieve consensus. The lead researcher proceeded to use the coding scheme to code all of the qualitative data before quantifying participants' responses. Suitable quotes were selected as examples.

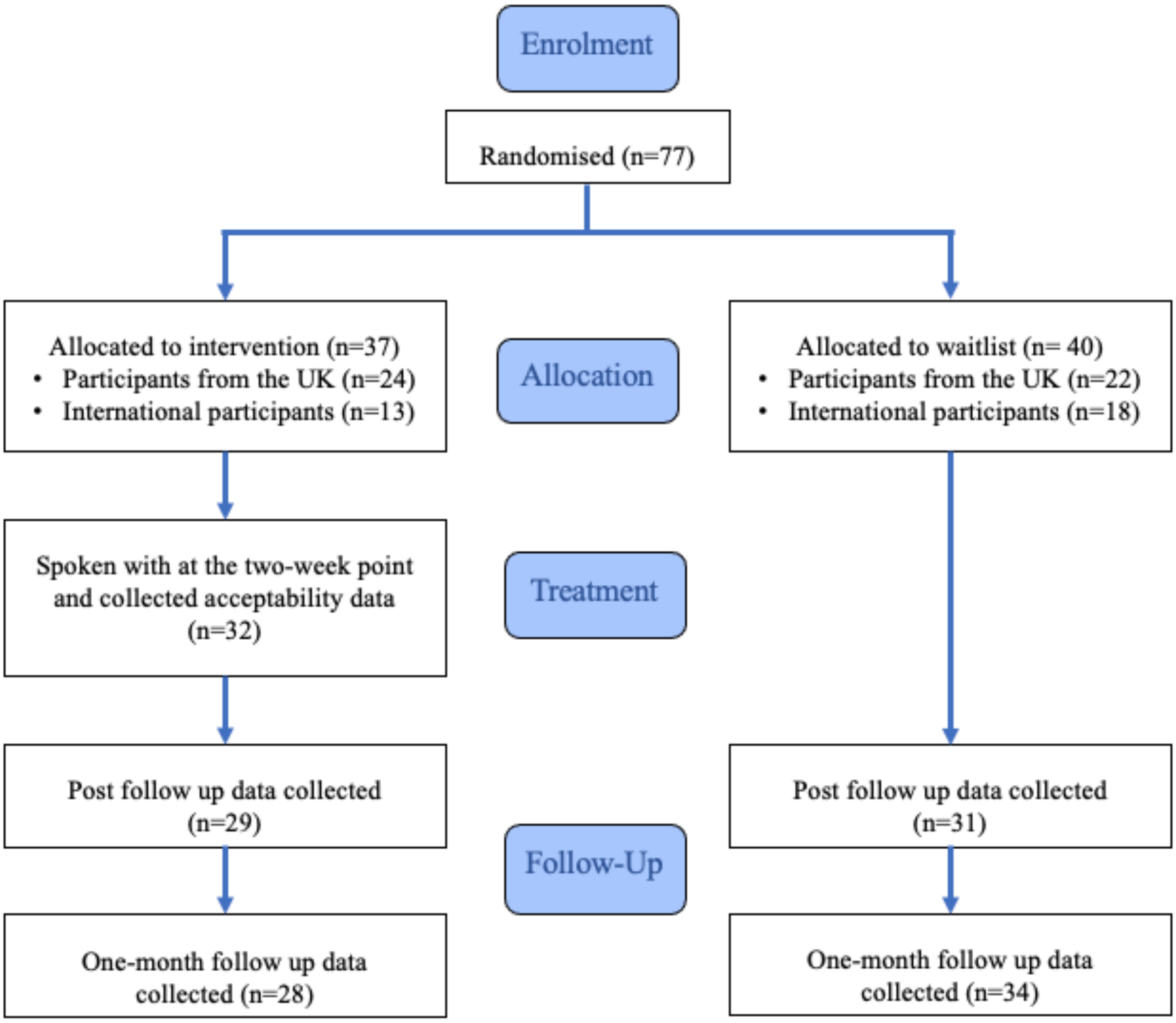
Results

Recruitment

Overall, n=138 individuals accessed the link to read the participant information sheet, of whom n=77 (55.8%) consented to take part and completed the baseline measures (Figure 3). Reasons for individuals not choosing to take part are unknown. It is also unknown how many people viewed the study advert.

Figure 3

CONSORT flow diagram



Randomisation

A series of *t*-tests and chi-squared tests revealed no significant differences in demographic or health-related outcomes between participants in the self-help or control group (Table 2).

Table 2

Baseline data for participants randomised to the CBT self-help or control group. Means and (standard deviations) unless otherwise stated

Characteristics	CBT self-help	Control	*p* value
Number of participants	n=37	n=40	0.73
Demographics			
Age (years)	48.2 (14.03)	47.5 (13.1)	0.83
Gender			0.4
Male	n=1	n=3	
Female	n=35	n=37	
Other	n=1	n=0	
Ethnicity			0.57
White	n=25	n=10	
Not reported or not clear	n=12	n=12	
Asian	n=4	n=5	
Hispanic	n=0	n=1	
Black	n=0	n=1	
Latina	n=1	n=1	
Education (years)	15.3 (4.7)	15.6 (3.8)	0.77
Employment			0.73
Employed	n=14	n=13	
Not employed	n=8	n=14	
Retired	n=9	n=9	
Student	n=2	n=1	
Other	n=4	n=3	
PH factors			
PH Type			0.83
Idiopathic PH	n=19	n=16	
Chronic thromboembolic PH	n=7	n=7	
Connective tissue disease	n=2	n=4	
Congenital PH	n=2	n=2	
Familial PH	n=0	n=1	
Other	n=3	n=6	
Not sure	n=4	n=4	
PH Class			0.27
I	n=6	n=2	
II	n=7	n=10	
III	n=12	n=12	
IV	n=2	n=0	
Not sure	n=10	n=13	
Years since diagnosis	9.48 (10.02)	7.84 (7.6)	0.42
Psychological factors			
Prescribed medication for anxiety	n= 9 (24.3%)	n=13 (32.5%)	0.43
Received therapy for anxiety	n=7 (18.9%)	n=9 (22.5%)	0.7
Anxiety	10.43 (5.11)	11.75 (5.78)	0.29
Depression	12.38 (5.12)	12.7 (6)	0.8
HRQoL	38.49 (10.57)	41.28 (11.39)	0.27
Dyspnoea	14.38 (9.91)	16.55 (8.96)	0.32
Self-mastery	25.95 (7.08)	24.33 (7.75)	0.34
CBP-Q	69.59 (13.43)	68.55 (19.67)	0.79

n = Number of participants; PH = Pulmonary Hypertension; HRQoL = Health-Related Quality of Life; CBP-Q = Cognitive and Behavioural Processes Questionnaire

UK vs. International Participants

International participants lived in Canada (n=17), India (n=7), United States (n=5), the Netherlands (n=1) and Australia (n=1). No significant differences were observed between the two groups with the exception of ethnicity, as a greater number of UK participants self-identified as white. This difference may be expected, given the ethnic composition of countries from where individuals were recruited (e.g., UK vs. India) Therefore, it was deemed appropriate to group participants (Table 3).

Table 3

Baseline data for participants living in the UK or outside of the UK (international). Means and (standard deviations) unless otherwise stated

Characteristics	UK	International	*p* value
Number of participants	n=46	n=31	0.09
Condition			0.38
CBT Self-help	n=24	n=13	0.07
Control	n=22	n=18	0.53
Demographics			
Age (years)	47.91 (12.59)	47.67 (14.91)	0.94
Gender			
Male	n=2	n=2	0.43
Female	n=44	n=28	
Other	n=0	n=1	
Ethnicity			0.004
White	n=29	n=16	
Not reported or not clear	n=15	n=4	
Asian	n=1	n=8	
Hispanic	n=0	n=1	
Black	n=1	n=0	
Latina	n=0	n=2	
Education (years)	14.54 (5.07)	16.42 (2.75)	0.07
Employment			0.07
Employed	n=16	n=11	
Not employed	n=14	n=8	
Retired	n=12	n=6	
Student	n=3	n=0	
Other	n=1	n=6	
PH factors			
Type			0.08
Idiopathic PH	n=17	n=18	
Chronic thromboembolic PH	n=12	n=2	
Connective tissue disease	n=5	n=1	
Congenital PH	n=1	n=3	
Familial PH	n=0	n=1	
Other	n=5	n=4	
Not sure	n=6	n=2	
Class			0.38
I	n=4	n=4	
II	n=8	n=9	
III	n=19	n=8	
IV	n=2	n=0	
Not sure	n=13	n=10	
Years since diagnosis	8.21 (8.55)	9.25 (9.33)	0.62
Psychological factors			
Prescribed medication for anxiety	n=12	n=10	0.56
Received therapy for anxiety	n=20	n=13	0.89
Anxiety	11.74 (5.23)	10.19 (5.78)	0.23
Depression	13.13 (5.32)	11.68 (5.88)	0.26
HRQoL	40.5 (11.15)	39.1 (10.95)	0.59

Dyspnoea	16.5 (9.79)	14.03 (8.81)	0.26
Self-mastery	24.52 (7.58)	25.97 (7.23)	0.4
CBP-Q	69.59 (16.51)	68.32 (17.4)	0.75

n = Number of participants; PH = Pulmonary Hypertension; HRQoL = Health-Related Quality of Life; CBP-Q = Cognitive and Behavioural Processes Questionnaire; UK = United Kingdom

Completers vs. Non-Completers

A significantly greater number of participants completed the intervention. An overall dropout rate of 15.6% was observed (self-help=18.9%, control=12.5%). On average, those classified as non-completers were significantly younger and reported lower levels of anxiety. There was a significant difference in gender between the two groups, with a greater ratio of males in those who completed. Notwithstanding this difference, males overall were greatly underrepresented accounting for 5% of the total sample (Table 4).

Table 4

Baseline data for participants defined as completers vs. non-completers. Means and (standard deviations) unless otherwise stated

Characteristics	Completers	Non-completers	*p* value
Number of participants	n=65	n=12	<0.001
Condition			0.44
CBT Self-help	n=30	n=7	
Control	n=35	n=5	
Demographics			
Age (years)	49.27 (13.11)	40.08 (13.17)	0.03
Gender			0.046
Male	n=4	n=0	
Female	n=61	n=11	
Other	n=0	n=1	
Country			0.17
UK	n=41	n=5	
International	n=24	n=7	
Ethnicity			0.43
White	n=29	n=6	
Not reported or not clear	n=17	n=2	
Asian	n=6	n=3	
Hispanic	n=1	n=0	
Black	n=1	n=0	
Latina	n=1	n=1	
Education (years)	15.47 (4.23)	15.18 (4.26)	0.84
Employment			0.39
Employed	n=25	n=2	
Not employed	n=17	n=5	
Retired	n=16	n=2	
Student	n=2	n=1	
Other	n=5	n=2	
PH factors			
PH Type			0.4
Idiopathic PH	n=30	n=5	
Chronic thromboembolic PH	n=14	n=0	
Connective tissue disease	n=5	n=1	
Congenital PH	n=3	n=1	
Familial PH	n=1	n=0	
Other	n=7	n=2	
Not sure	n=5	n=3	
PH Class			0.86
I	n=7	n=1	
II	n=15	n=2	
III	n=23	n=4	
IV	n=2	n=0	
Not sure	n=18	n=5	
Years since diagnosis	7.93 (8.36)	12.42 (10.64)	0.11

Psychological factors			
Prescribed medication for anxiety	n=18	n=4	0.69
Received therapy for anxiety	n=12	n=2	0.24
Anxiety	11.75 (5.44)	7.67 (4.4)	0.02
Depression	2.95 (5.54)	10.33 (5.37)	0.13
HRQoL	40.63 (10.89)	36.17 (11.4)	0.2
Dyspnoea	15.92 (9.57)	13.25 (8.65)	0.37
Self-mastery	24.35 (7.4)	29.17 (6.38)	0.04
CBP-Q	69.95 (17.2)	63.9 (13.58)	0.27

n = Number of participants; PH = Pulmonary Hypertension; HRQoL = Health-Related Quality of Life; CBP-Q = Cognitive and Behavioural Processes Questionnaire; UK = United Kingdom

Acceptability

Quantitative Findings. Overall, 32/37 (86.5%) of participants in the intervention condition completed the Check-in Questionnaire at two-weeks. All participants had received and started the intervention. The majority of participants were working on booklet one (n=7) or two (n=17); booklet three (n=6) and four (n=2). The majority (>70%, n=23/32) understood most of the content, felt it could help with their anxiety and other areas of their life, and did not find the intervention difficult or distressing (Table 5). All participants intended to finish the intervention.

Table 5

Check-in Questionnaire (n=32); values represent n unless otherwise stated

Question	Great Deal – A Lot	Moderate Amount	None at All – A Little	Mean (SD)
How much have you looked at the intervention?	15 (46.9%)	13 (40.6%)	4 (12.5%)	3.6 (0.97)
How much of the intervention have you understood?	27 (84.4%)	3 (9.3%)	2 (6.3%)	4.35 (1.02)
How distressing is the intervention?	2 (6.3%)	5 (15.6%)	25 (78.1%)	1.55 (0.74)
How difficult is the intervention?	3 (9.4%)	6 (18.7%)	23 (71.9%)	1.97 (1.08)
How often have you been using the skills?	16 (50%)	10 (31.2%)	6 (18.8%)	3.29 (1.19)
How much **<u>is</u>** the intervention helping with your anxiety?	15 (46.9%)	10 (31.2%)	7 (21.9%)	3.23 (1.02)
How much **<u>can</u>** the intervention help with your anxiety?	30 (93.8%)	1 (3%)	1 (3%)	4.23 (0.68)
How much **<u>is</u>** the intervention helping in other areas of your life?	10 (31.3%)	14 (43.7%)	8 (25%)	2.97 (0.95)
How much **<u>can</u>** the intervention help in other areas of your life?	26 (81.3%)	6% (18.7%)	0 (0%)	4.06 (0.68)

SD = Standard Deviation

In total, 25/32 (78.1%) of participants completed the Final Acceptability Questionnaire. Ten participants rated the intervention as "*Excellent*", fourteen "*Good*", and one "*Fair*" – no participant reported it "*Poor*" or "*Very Poor*". Nearly half of the participants reported the intervention as "*Moderately*" helpful for anxiety and other areas of their life. All participants would recommend the intervention to another person with PH (Table 6). Most (>68%,n=17/25) found the different booklets "*Extremely*" – "*Very Helpful*", in particular, booklet two which focused on maintenance factors of anxiety (Table 7). At the end of the study, >80% (n=20/25) felt more in control of their anxiety, supported during the intervention, valued that the treatment was specific to PH and felt others affected by PH would benefit from the resource (Table 8).

Table 6

Final Acceptability Questionnaire post-intervention (n=25). Values represent n (%) unless otherwise stated

Question	Great Deal – A Lot	Moderate Amount	A Little	Mean (SD)
How much did the intervention help you with your anxiety?	9 (36%)	12 (48%)	4 (16%)	3.3 (0.84)
How much did the intervention help in other areas of your life?	10 (40%)	11 (44%)	4 (16%)	3.3 (0.85)
How likely are you to recommend the intervention?	21 (84%)	3 (12%)	1 (4%)	4.3 (0.84)

No participants reported "Not at All Helpful"

Table 7

Results from the Final Acceptability Questionnaire continued.

Item	Extremely - Very Helpful	Somewhat Helpful	Slightly Unhelpful	Mean (SD)
Booklet 1 (Anxiety and PH)	18 (72%)	6 (24%)	1 (4%)	3.8 (0.75)
Booklet 2 & 3 (What keeps anxiety going and The power of thoughts)	23 (92%)	2 (8%)	0 (0%)	4.1 (0.49)
Booklet 4 (The power of your actions)	17 (68%)	8 (32%)	0 (0%)	3.9 (0.74)
Booklet 4 – (Other factors affecting anxiety)	18 (72%)	7 (28%)	0 (0%)	3.9 (0.7)
Homework tasks	19 (76%)	6 (24%)	0 (0%)	3.9 (0.65)

No participant reported "Not at All Helpful"

Table 8

Results from the final acceptability questionnaire continued.

Question	Strongly-Somewhat Agree	Neither Agree nor Disagree	Strongly – Somewhat Disagree	Mean (SD)
I feel more in control of my anxiety	22 (88%)	3 (12%)	0 (0%)	4.1 (0.6)
People diagnosed with PH would benefit from the intervention	25 (100%)	0 (0%)	0 (0%)	4.8 (0.37)
Family and friends would benefit from this intervention	23 (92%)	2 (8%)	0 (0%)	4.4 (0.65)
I valued the intervention was specific to PH	24 (96%)	1 (4%)	1 (4%)	4.7 (0.85)
I valued being contacted part-way through	24 (96%)	1 (4%)	0 (0%)	4.6 (0.58)
The level of support I received was sufficient `	24 (96%)	1 (4%)	0 (0%)	4.6 (0.58)
I was given enough time to work through the intervention	24 (96%)	1 (4%)	0 (0%)	4.3 (0.95)
Health outcomes were relevant to my difficulties	20 (80%)	3 (12%)	2 (8%)	4 (1)

PH = Pulmonary Hypertension

Qualitative Findings. Thirty-two participants contributed qualitative data. Five main categories were identified reflective of the questions asked (Table 9).

Participants discussed wanting to take part in the intervention due to difficulties with their anxiety, which for some was linked to COVID-19 (n=10). The majority of participants reported the benefits of learning more about their anxiety (n=17) and developing strategies to manage (n=22). The intervention also helped to normalise participant's difficulties, and while reading the case studies increased anxiety in some (n=3), this was often viewed as "*manageable"* and a "*necessary evil*". Participants recognised the importance of continuing to practice the strategies and valued the ability to refer back to the booklets at a later date. The largest group (n=9) felt there was "*nothing*" they did not like about the intervention. For others, it seemed external sources of motivation could have helped them to engage better.

The majority of participants (n=22) found the intervention user friendly and informative. Individuals also valued the emphasis on developing coping strategies and how it was specific to PH.

Table 9

Results from the quantitative content analysis investigating acceptability (n=32)

Categories	Responses	Examples	n
Motivation to take part	Affected by anxiety and/or mental health	*"I have struggled with anxiety for over 15 years but have had times where it has been worse since my PH diagnosis"* (P31)	15
	Learn how to better understand and manage anxiety	*"To learn how to help manage my own anxiety and stress & I was so amazed that the help was specified towards PH."* (P67)	14
	COVID, lockdown or shielding made anxiety worse	*"Due to the COVID-19 situation my mental health was getting worse."* (P43)	10
	To help other people through participating in a trial	*"I wanted to help other sufferers"* (P22)	4
	Like to participate in trials for PH	*"I want to take part in any research that can help pulmonary hypertension patients"* (P20)	2
	Better understand PH	"*To better understand pulmonary hypertension as I was only diagnosed this year… it was very scary getting told that and that I have blood clots in lungs"* (P28)	1
Result of taking part	Developed coping strategies to manage and feel more in control of anxiety	*"Thank you for helping to give tools to help people dealing with PH and many other challenges!!!!!!"* (P67)	22
	Improved how I understand anxiety	*"It also made me aware of things that I do when feeling anxious that I wasn't even aware of."* (P17)	17
	Normalised my experiences	*"…the true-life scenarios were very helpful as you do tend to feel that sometimes it must be only you that feels like this"* (P33)	10
	Made me confront my difficulties	*"It helped me look at my anxiety in a new way and helped me to confront it."* (P20)	5
	Induced my anxiety in the short-term	*"The women in the study makes me quite worried but I feel it is useful, but not a criticism"* (P2)	3
	Helped in other areas, such as self esteem	*"Improved my self-esteem and motivation."* (P18)	2

How will you continue to use what you learnt in the study?	Keep practicing and using helpful coping strategies	*"Day by day putting in the work"* (P62)	21
	Going back to the booklets to recap	*"I plan to keep going over the materials, and I hope to form habits that will help with anxiety and stress."* (P67)	16
	Share the booklets and my anxiety difficulties with others	*"I plan to continue using the breathing exercises and plan on letting my partner read my responses in the booklets to better understand what worries me when it comes to my health."* (P20)	1
Barriers (what did you like the least?)	None	*"I found it all useful."* (P14)	9
	Struggled to stay motivated, keep focused or keep on track	*"Nothing really, the only thing maybe would be a reminder to ensure you know what week you're on. For example a reminder text"* (P52)	5
	Takes time to remember all the recommendations	*"It's difficult making them a habit when you have years of over thinking."* (P80)	4
	Questionnaires were not always relevant or forgetting to do them	"*I probably found the questionnaires quite difficult to answer as the choice of boxes to tick did not really relate…"* (P17)	4
	Being asked to face to my fears	*"Having to confront the situations that made me feel anxious and worried"* (P26)	3
	More information about the symptoms and more resources	*"Maybe more info could be good? That said I found what it contained specifically helpful"* (P74)	3
	Some of my difficulties were not represented	"*In particular, the booklets didn't help me to work out how to help my family"* (P24)	2
	Doing it by yourself	*"It's good that we do this on our own, but it would've been nice to interact with someone." (P67)*	2
	Case studies were anxiety provoking	*"The case studies with other people who have PH"* (P20)	1
	Font was very small – electronic version	*"I was only able to read the booklets online because the font in the printed version was too small and almost impossible to read."* (P77)	1
	Needed longer than one-week	*"I think a two-week interval between starting the other booklets would have been more helpful."* (P18)	1

Facilitators (What did you like the best?)	The information was user friendly and informative	*"It was straightforward and easy to work through."* (P14)	22
	Booklets helped promote self-evaluation	*"It made me think about my situation and how I manage my reactions." (P22)*	8
	You can work at your own pace	*"Letting you work through one stage at a time and not being bombarded with information."* (P38)	7
	It was specific to PH	*"I was very happy that the material was geared to people with pulmonary hypertension because it is rare to hear of information specific to PH and even more rare to have help with the issues we face."* (P67)	6
	The promotion of coping strategies	*"Gaining skills and techniques from the booklets. Better understanding why I react the way I do in some situations and being able to cope with it."* (P43)	5
	Homework tasks	*"I like the practical strategies that are suggested and I have found them useful."* (P17)	4
	Case studies	*"The testimonies, to know others feel the same and have similar experiences."* (P34)	4
	I feel less alone	*"Felt less alone and isolated when dealing with anxiety."* (P54)	3
	It was effective	*"It was simple and effective and support was given."* (P52)	1
	Support from the researcher	*"Having a call during the intervention was helpful to discuss matters."* (P52)	1
	I felt empowered to help myself	*"The self-help exercises. Found it quite empowering to feel that I was able to help myself cope and manage my anxiety."* (P18)	1
	Not having to wait for treatment	*"I think it is a very worthwhile study, as mental health services are so underfunded you can wait ages to access any support."* (P38)	1
	Relevant to everyday life	*"Interventions were realistic."* (P80)	1

P = Participant number; PH = Pulmonary Hypertension

Preliminary Effectiveness

Reliable Change. Both groups scored above the clinical cut-off on the GAD-7 and PHQ-9 measures at baseline, scoring within the "*moderate*" range (Figure 4 and 5). While anxiety and depression symptomatology reduced below the clinical level and to the "*mild*" range following the self-help intervention and remained at this level at follow-up, those in the control group continued to report "*moderate"* and clinical levels throughout.

Figure 4

Anxiety mean scores for groups across time. Error bars represent standard error.

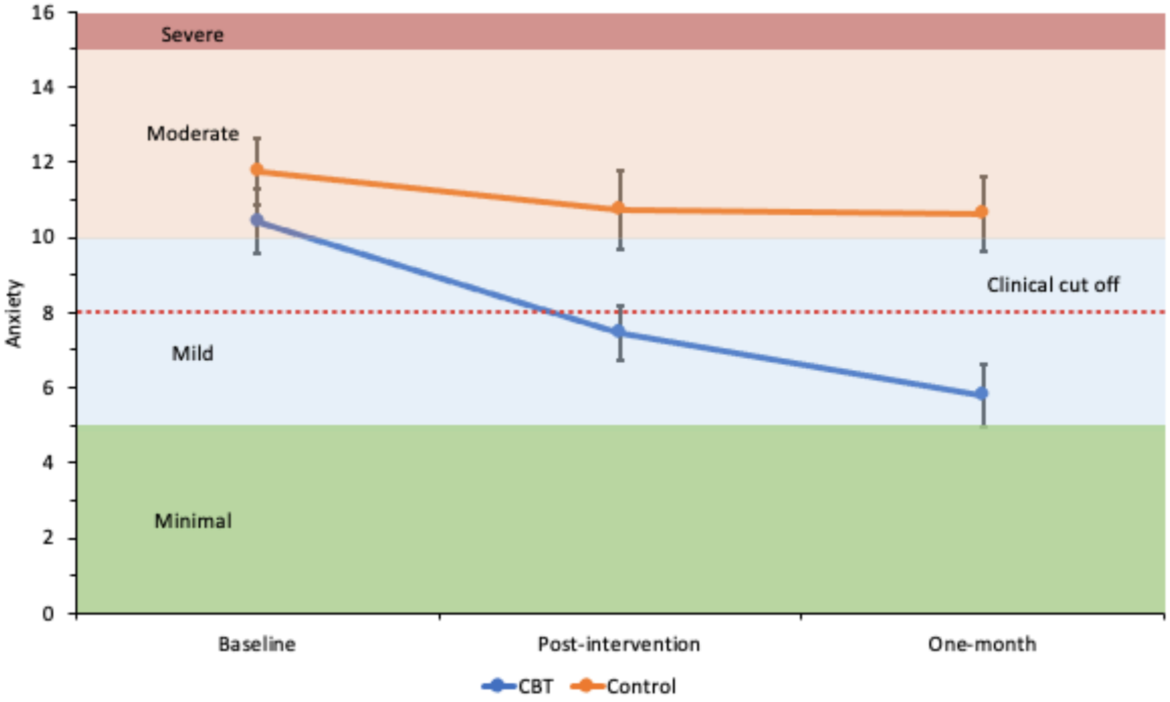
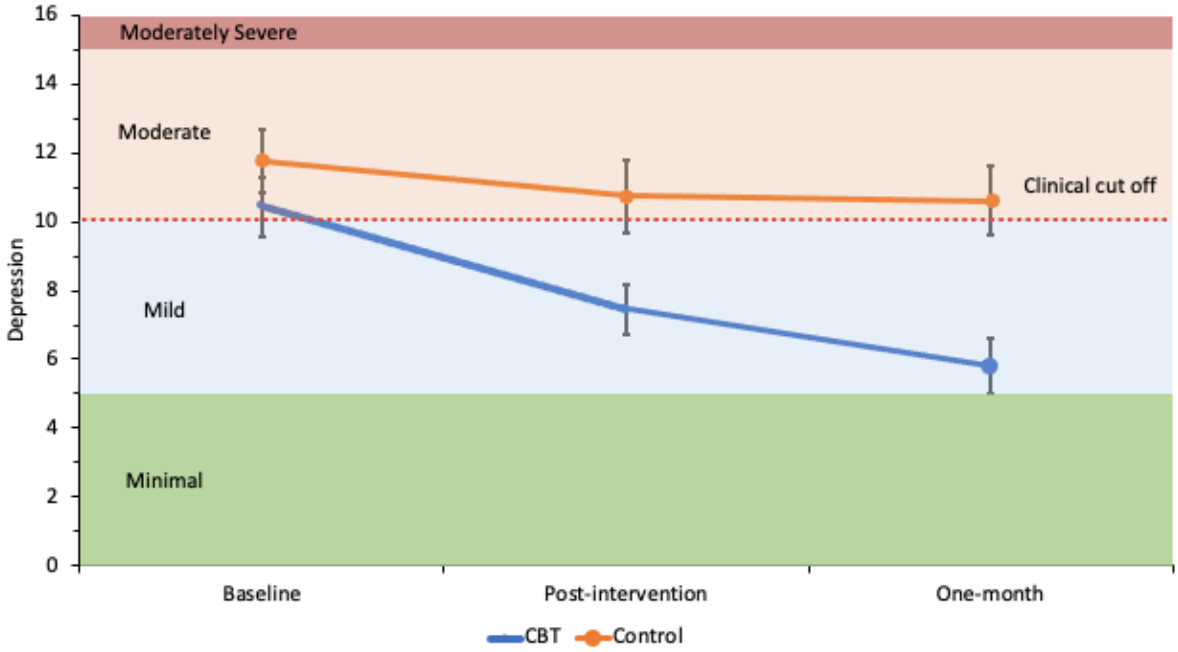


Figure 5

Depression mean scores for groups across time. Error bars represent standard error.



To examine change in symptomatology at an individual level, participants were stratified based on their scores on the GAD-7 and PHQ-9. In the self-help group, 64.9% and 67.6% of participants scored above the clinical level on anxiety and depression, respectively, which by the end of treatment, reduced to 25% and 32.1%. Overall, 71.4% and 39.3% of participants reported a reliable change in anxiety and depression, respectively, at the end of the study, compared to baseline or post-intervention scores.

In the control group, 44.1% of individuals reported a reliable change in anxiety at one-month follow up, although nearly two-thirds remained above the clinical cut-off (Table 10). Similarly, over half of the participants in the control group experienced clinical levels of depression at the end of the study.

Table 10

Anxiety and depression scores for participants in the CBT (n=37) and control group (n=40), values = n (%)

Group	Measure		Baseline	Post-intervention	One-month follow up
CBT	Anxiety	0-4 Minimal	4 (10.8%)	8 (27.6%)	15 (53.5%)
		5-9 Mild	16 (43.2%)	11 (37.9%)	6 (21.4%)
		10-14 Moderate	7 (18.9%)	9 (31%)	6 (21.4%)
		15-21 Severe	10 (27%)	1 (3.4%)	1 (3.6%)
		Above cut off (≥8)	24 (64.9%)	16 (55.2%)	7 (25%)
		Reliable change (≥4)	-	13 (44.8%)	20 (71.4%)
	Depression	0-4 Minimal	3 (8.1%)	10 (34.5%)	11 (39.3%
		5-9 Mild	9 (24.4%)	6 (20.7%)	8 (28.6%)
		10-14 Moderate	8 (21.6%)	12 (41.4%)	7 (25%)
		15-19 Moderately Severe	15 (40.5%)	1 (3.4%)	1 (3.6%)
		20-27 Severe	2 (5.4%)	0 (0%)	1 (3.6%)
		Above cut off (≥10)	25 (67.6%)	13 (44.8%)	9 32.1%)
		Reliable change (≥6)	-	13 (44.8%)	11 (39.3%)
Control	Anxiety	0-4 Minimal	3 (7.5%)	4 (12.9%)	5 (14.7%)
		5-9 Mild	13 (32.5%)	10 (32.3%)	11 (32.4%)
		10-14 Moderate	11 (27.5%)	7 (22.6%)	8 (23.5%)
		15-21 Severe	13 (32.5%)	10 (32.3%)	10 (29.4%)
		Above cut off (≥8)	30 (75%)	18 (58.1%)	22 (64.7%)
		Reliable change (≥4)	-	9 (29%)	15 (44.1%)
	Depression	0-4 Minimal	3 (7.5%)	4 (12.9%)	6 (15.7%)
		5-9 Mild	12 (30%)	9 (29%)	10 (29.4%)
		10-14 Moderate	6 (15%)	8 (25.8%)	5 (14.7%)
		15-19 Moderately Severe	15 (37.%%)	9 (29%)	7 (20.5%)
		20-27 Severe	4 (10%)	1 (32.2%)	6 (17.26%)
		Above cut off (≥10)	25 (62.5%)	18 (58.1%)	18 (52.9%)
		Reliable change (≥6)	-	7 (22.6%)	6 (17.6%)

CBT = Cognitive Behavioural Therapy

Statistical Change. A series of two-way mixed ANOVAs revealed statistically significant interactions between group and time for anxiety (medium effect), depression (small-medium effect) and CBP-Q (medium-large effect) (Table 11).

Table 11

Means and (Standard deviations) in the CBT (n=27) and control (n=30) group for health-outcomes and results of 2x3 mixed ANOVAs

					Two-way mixed ANOVA (group x time)			
Measure	Group	Baseline	Post	One-month	F	*p*	η^2	*Cohens' f (observed power)*
Anxiety	CBT	11 (5.03)	7.56 (3.93)*	5.89 (4.4)	F(2,110)= 4.23	0.017	0.07	0.28 (72.8%)
	Control	11.9 (5.29)	11.03 (5.78)*	10.23 (5.62)				
Depression	CBT	12.41 (5.47)	8.11 (4.21)	7.48 (5.11)*	F(1.76,97.71) =3.26 HF	0.048	0.06	0.24 (61%)
	Control	13.5 (5.4)	11.3 (5.47)	11.7 (7.1)*				
HRQoL	CBT	37.7 (11.16)	35.59 (10.69)	34.37 (10.14)	F(2,110)= 0.1	0.9	0.002	0.04 (6.6%)
	Control	42.07 (11.27)	41.67 (11.06)	39.5 (11.81)				
Dyspnoea	CBT	14.93 (10.6)	14.85 (10.07)	12.63 (8.54)	F(1.85, 101.67) =0.77 HF	0.46	0.01	0.12 (18%)
	Control	16.77 (8.74)	15.77 (8.92)	15.3 (9.29)				
Self-Mastery	CBT	25.22 (6.7)	26.41 (5.66)	27.63 (4.79)	^F(2,110) =0.24	0.79	0.004	0.06 (8.4%)
	Control	24.2 (7.59)	25.63 (5.3)	25.67 (4.73)				
CBP-Q	CBT	69.56 (14.3)	57.59 (15.93)	50.26 (18.8)	^F(1.67,91.74) =8.55 HF	0.001	0.13	0.39 (95.7%)
	Control	67.77 (17.88)	63.87 (16.47)	64.17 (22.28)				

CBT = Cognitive Behavioural Therapy; η^2 = Partial Eta Squared; HF = Huynh-Feldt; ^ = Test of Equality of Covariance significant; *f* = ANOVA effect size; ANOVA= Analysis of Variance; CBP-Q = Cognitive Behavioural Processes Questionnaire; *= Levene's Test of Equality not met (p>0.05) for mixed ANOVA

A series of independent samples *t*-tests were performed to examine simple effects between the two groups on the aforementioned measures. For anxiety, while no statistical difference was observed at baseline (*t*(55)=-0.66, *p*=0.51), there was a statistical difference at the post-intervention (*t*(51.31)=-2.68, p=0.01) and follow-up stage (*t*(55)=-3.33, *p*=0.002), with those in the CBT group reporting lower scores. Similarly, for depression, differences at baseline was not significant (*t*(55)=-0.76, *p*=0.45), but was at post-intervention (*t*(55)=-2.45, *p*=0.018) and follow-up (*t*(52.56)=-2.55, p=0.01) in the same direction. Finally, for the CBP-Q, differences were non-significant at baseline (*t*(55)=0.41, *p*=0.68) and post-intervention (*t*(55)=-1.46, *p*=0.15), however, at the one one-month stage there was a significant difference (*t*(55)=-2.53, *p*=0.014), with those in the control group experiencing a greater number of unhelpful thoughts and behaviours.

A series of repeated measures ANOVAs were used to explore simple main effects of time in each condition. A statistically significant effect of time on anxiety, depression and CBP-Q was observed in the CBT group, indicating reductions across time, with large effect sizes. A significant effect was also observed for depression in the control group (large effect size), however, post-hoc tests revealed this reduction was only temporary as there was no significant difference between baseline and one-month follow-up scores (Table 12).

No significant differences were observed in the other health-related measures.

Table 12

Mean differences in the CBT (n=27) and control (n=30) group for health-outcomes and results of repeated measures ANOVAs and pairwise comparisons corrected using Bonferroni

Measure	Group	Repeated measures ANOVA				Pairwise comparison		
		F	p	η^2	*Cohens' f observed (power)*	Baseline vs Post	Baseline vs. One-month	Post vs. One-month
Anxiety	CBT	F(2,52) =18.54	<0.001	0.42	0.84 (100%)	-3.44 (p=0.003)	-5.11 (p<0.001)	-1.67 (p=0.045)
	Control	F(2,58)=1.84	0.17	0.06	0.25 (37%)	-0.87 (p=0.92)	-1.67 (p=0.29)	-0.8 (p=1)
Depression	CBT	F(1.63,42.25) =13.02 HF	<0.001	0.33	0.71 (98.7%)	-4.3 (p=0.002)	-4.93 (p=0.002)	-0.63 (p=1)
	Control	F(2,58)= 5.43	0.007	0.16	0.43 (83.4%)	-2.2 (p=0.004)	-1.8 (p=0.11)	0.4 (p=1)
HRQoL	CBT	F(2,52)=2.54	0.09	0.09	0.31 (49.1%)	-1.11 (p=1)	-3.33 (p=0.2)	-2.22 (p=0.34)
	Control	F(2,58)=2.85	0.07	0.09	0.31 (54.1%)	-0.4 (p=1)	-2.57 (p=0.15)	-2.17 (p=0.16)
Dyspnoea	CBT	F(2,52)=2.45	0.1	0.09	0.31 (49.1%)	-0.07 (p=1)	-2.3 (p=0.34)	-2.22 (p=0.13)
	Control	F(2,58)=1.64	0.2	0.05	0.24 (31.2%)	-1 (p=0.67)	-1.47 (p=0.39)	-0.47 (p=1)
Self-mastery	CBT	F(2,52)=1.25	0.3	0.05	0.22 (28.2%)	1.19 (p=1)	2.41 (p=0.54)	1.22 (p=1)
	Control	F(2,58)=0.3	0.29	0.04	0.21 (25.4%)	1.43 (p=0.79)	1.47 (p=0.38)	0.03 (p=1)
CBP-Q	CBT	F(1.63,42.3)= 18.38 HF	<0.001	0.41	0.83 (100%)	-11.9 (p=0.001)	-19.3 (p<0.001)	-7.33 (p=0.03)
	Control	F(2,58)=2.04	0.14	0.07	0.27 (42.9%	-3.9 (p=0.15)	-3.6 (p=0.52)	0.3 (p=1)

CBT = Cognitive Behavioural Therapy; η^2 = Partial Eta Squared; HF = Huynh-Feldt; *f* = ANOVA effect size; ANOVA= Analysis of Variance; CBP-Q = Cognitive Behavioural Processes Questionnaire

Sample Size Analysis for Definitive Trial

A sample size analysis was calculated using an observed effect size of *f*=0.28 for the primary outcome measure, GAD-7. Accepting 0.05 as the alpha level, a level of statistical power of 80%, two groups (self-help and wait-list) and three measurements (baseline, post-intervention and one-month) and sphericity not violated, a final sample size of n=68 is required. A post-hoc analysis revealed the current study had a power of 72.8%. Taking into consideration a dropout rate of 15.6%, 80 participants are required for a definitive trial.

Mechanisms of Change

Intervention group was significantly related to change in anxiety scores (*b*=3.62(standard error(se))=1.35), *t*(59)=2.69, *p*=0.009) and change in CBP-Q at one-month (*b*=14.86(se=4.4), *t*(59)=3.37, *p*=0.001). Change in CBP-Q scores significantly predicted change in anxiety (*b*=0.13(se=0.04), *t*(58)=3.52, $p<0.001$). Controlling for change in CBP-Q as a mediator, condition was not a significant predictor of change in anxiety (*b*=1.72(se=1.35), *t*(58)=1.27, *p*=0.21). Overall, change in CBP-Q was a significant mediator of change in anxiety (effect=1.91(se=0.82), 95% CI=0.56 – 3.67), which was confirmed by a Sobel test (Z=2.34(se=0.82), *p*=0.02). A post-hoc power analysis was computed using the observed effect size of n=61, *R*=0.33, R^2=0.11, f^2=0.12 (revealing a small-medium effect size), significant level of 0.05 and with two predictors, revealing a power of 66.6%.

For depression, intervention group significantly predicted change in depression scores (*b*=3.31(se=1.38), *t*(59)=2.4, *p*=0.02) and change in CBP-Q (*b*=14.86(se=4.4), *t*(59)=3.37, *p*=0.001). Change in CBP-Q was a significant predictor of change in depression (*b*=0.19(se=0.03), *t*(58)=5.87, $p<0.001$). When change in CBP-Q was controlled, condition was not a significant predictor of change in depression (*b*=0.46(se=1.2), *t*(58)=0.38,*p*=0.7). As in anxiety, change in CBP-Q was a significant mediator of change in depression (effect=2.84(se=1.23), 95% CI=0.78 – 5.6), which was further assessed using a Sobel test (Z=2.98(se=0.95), p=0.003). A post-hoc power analysis using the observed effect size of n=61, R=0.3, R^2=0.09, f^2=0.1 (revealing a small-medium effect size) and same parameters as above, demonstrated a power of 56.3%.

In summary, change in CBP-Q fully mediated the relationship between intervention group and change in anxiety and depression.

Discussion

This is the first study investigating a self-help intervention for anxiety in PH. Participants were recruited within the community across six countries and all causes and severity of PH were included, thus increasing the sample's diversity. The majority of participants were working-age females, diagnosed with idiopathic PH and reported a WHO functional class II or III, which is generally representative of the larger population (Hoeper & Gibbs, 2014); therefore improving the findings generalisability. A wait-list group was used as a control condition, which is characteristic of the current support available as information for anxiety in PH is limited.

Findings suggest that the intervention was feasible, acceptable, and while not the aim of a pilot study, safe, as no adverse events were reported. The required sample size was achieved, indicating the proposed method of recruitment is suitable for a large-scale trial. Fidelity was high as all participants who were successfully contacted part-way through the intervention had commenced with the booklets. Furthermore, an overall attrition rate of 15.6% was observed, which is comparable to rates reported by a meta-analysis of 54 self-help interventions for anxiety (16.5%) (Haug et al., 2012), as well as a meta-analysis of 45 studies investigating face-to-face psychological therapies for adults with GAD (16.99%) (Gersh et al., 2017). The current rate should also be interpreted in light of the minimal contact that participants had with members of the research team, for example, compared to facilitated interventions requiring greater involvement from professionals (Cuijpers & Schuurmans, 2007).

Overall, 24 out of 25 participants valued that the intervention was specific to PH, therefore providing additional evidence for the need of condition-specific interventions. The majority of participants reported the intervention as "*Excellent*" or "*Good*" and would recommend it to someone with PH. This further supports the assertion that the intervention was acceptable. Nevertheless, based on participants' feedback, a number of improvements could be proposed. Given that an electronic version of the intervention was found acceptable, and this format is generally more effective than bibliotherapy (Haug et al., 2012), an online or blended approach may be utilised. This would have the advantage of being able to direct participants to a greater number of non-PH or specifically developed resources, for example, videos on relaxed-breathing and mindfulness. Moreover, as some participants found it difficult to stay motivated or on track and disliked completing the booklets alone, the intervention could be supplemented by an online group allowing participants to share their experiences and support others. However, such an addition would need to be investigated, for instance, examining its feasibility.

Participants who completed the study were significantly older and reported a greater level of anxiety, than non-completers. This finding is consistent with research suggesting younger adults are more likely to dropout of psychological therapies (Matthew Prina et al., 2014) – although based on the current dataset, no strong conclusions can be posed. It is plausible that people who were less affected by anxiety had lower motivation to engage. However, preliminary results suggest that the intervention was helpful in reducing symptoms of anxiety in those with mild to severe symptoms. Combined with previous findings demonstrating psychological interventions can serve a preventative role in PH (Tarantino et al., 2020), supporting individuals with all levels of anxiety to engage with psychological care may be beneficial. Similarly, while it has been suggested that anxiety is greater in those recently diagnosed with PH - due to the emotional impact of the diagnosis and initial period of adjustment (Bussotti & Sommaruga, 2018) - as participants had been living with PH for an average of seven-nine years, interventions targeting anxiety may be helpful and of interest to people across the clinical group. However, our current ability to explore anxiety in people with PH is likely to be impaired by the lack of standardised screening assessments. Indeed, given that one-fifth of participants here did not agree that the outcome measures were relevant to their difficulties, further research is required to investigate the suitability, reliability and adaptation of non-PH and generic health-related questionnaires in this population.

At baseline, 70.1% (n=54/77) and 64.9% (n=50/77) of participants scored above the clinical level on anxiety and depression, respectively, which is greater than previous studies reporting rates of 50% (Bussotti & Sommaruga, 2018). Given that participants here had to be experiencing difficulties with anxiety, it is reasonable to suggest that rates are higher compared to the wider PH population. Nevertheless, it could also reflect the impact of the COVID-19 pandemic, as highlighted by ten participants who discussed the pandemic as a reason for their participation. There is little research investigating the additional burden COVID-19 has had on this clinical group, despite their clinical vulnerability. A qualitative investigation examined experiences of adults with PH during the pandemic, finding anxiety was commonly reported and associated with fear of infection, especially given the risk of increased mortality, isolation and reduced sense of safety (Rawlings, Armstrong, et al., 2021). While participants in the current study found the intervention was helpful in managing their anxiety during the pandemic, it is not known whether this was translated to COVID-19-

related anxieties. Further research is required examining the psychosocial effects of the pandemic in people with PH and how best to support this group.

Notwithstanding that this study was underpowered to detect a between-group difference, nor was it the primary aim, those in the CBT group reported a significant reduction in anxiety, depression and unhelpful cognitions and behaviours over time, compared to participants in the control group. As such, the results further justify a large scale RCT of the current intervention, which would require a sample size of n=80. No significant differences were observed in the other measures; however, it is unclear whether this is because the intervention does not help to alleviate these symptoms, or such difficulties need longer than two-months to improve. Within-group differences revealed a large and significant reduction in the CBT group in symptoms of anxiety and depression, with participants scoring below the clinical level at the post-intervention stage with gains maintained, and significantly improved upon for anxiety, at follow-up. Taken together, the findings support the evidence concerning the benefits of psychological therapy, specifically CBT, in this population.

Interestingly, those in the control group experienced an improvement in depression at the post-intervention stage, with some also reporting a reliable change for anxiety and depression. It is not unusual for participants in control groups to report therapeutic gains; this is sometimes referred to as the Hawthorne effect, in which people change their behaviour in response to being observed (Sedgwick & Greenwood, 2015). Nevertheless, the majority of participants remained above the clinical level and gains were only temporary.

Interventions are often criticised for failing to report or explore mechanisms of change, which can impact the reliability and replicability of treatments (Hoffmann et al., 2014; Michie et al., 2015). Therefore, a series of mediation analyses were performed revealing changes in cognitions and behaviours fully explained the relationship between intervention group and change in anxiety and depression at one-month. Notwithstanding the fact that such analyses were underpowered and the CBP-Q has not been validated in people with PH, the results indicate that the intervention had an indirect effect on improvement in mood outcomes, through changing cognitions and behaviours. Further research is required to examine the relationship between the cognitive and behavioural strategies that individuals use to cope, and health-related outcomes in PH. What is more, insights gained from longitudinal studies are likely to be necessary, as the current findings suggest a significant change in unhelpful thoughts and behaviours related to mood required longer than four-weeks. Indeed, identifying factors associated with positive health outcomes can help to recognise important therapeutic targets, and guide the development and improvement of treatment (Rawlings, Thompson, et al., 2021; Wryobeck et al., 2007; York et al., 2014).

Limitations

Due to the modest sample size, differences in outcomes between participants receiving paper vs. electronic versions of the intervention were not examined. Therefore, it is not clear what impact (if any) this had on observed effect sizes, especially given the difference in effectiveness between the two approaches observed (Haug et al., 2012). The reported sample size will likely need to be revised should a definitive trial aim to differentiate between the two.

Results were not stratified by functional impairment of PH, despite this being linked to health-outcomes elsewhere (Yorke et al., 2018). However, in those who knew their functional WHO class, class II and III were the most common which is representative of the larger population (Wijeratne et al., 2018).

Notwithstanding the female preponderance reported in PH (Hoeper & Gibbs, 2014), men were considerably underrepresented in the current sample, which is consistent with previous evidence suggesting females with PH are more likely to take part in research

(Halimi et al., 2018). Further investigation is required to explore such gender differences (including the likelihood of males with PH engaging with psychological therapies) and develop strategies to help overcome barriers for men.

While efforts were taken to include common challenges in PH within the booklets, participants reported a number of experiences that were unintentionally overlooked, such as end-of-life challenges. The difficulties addressed in the intervention largely reflect what is discussed in the PH literature as identified by the systematic review (see Rawlings et al., 2020). More research is required to investigate experiences of PH, which will help to improve our understanding of the disease and inform the development of services.

As discussed, the study was conducted during the COVID-19 pandemic, which may have affected aspects of the trial; for example, social restrictions are likely to have made it difficult to engage in certain strategies, such as increasing behaviour. Whilst it cannot be concluded with certainty whether (or to what extent) this has influenced the current outcomes, this should be considered in a future trial.

Finally, steps involved in the quantitative content analysis were performed by one researcher and not subjected to a test of inter-rater reliability. Such measures could have helped to improve the reliability of this analysis and protect against bias.

Conclusion

There is growing evidence demonstrating condition-specific self-help interventions targeting psychological difficulties can be acceptable and effective forms of treatment. In the current study, a self-help intervention for anxiety in PH was developed and examined using a pilot RCT. The intervention was found to be acceptable, feasible and safe. Attrition was comparable to other psychological therapies, including face-to-face treatments. While only preliminary, participants in the self-help condition reported a significant and reliable reduction in symptoms of anxiety and depression, which were fully mediated by a change in cognitions and behaviours related to the intervention. While no prespecified criteria was proposed to decide whether to proceed with a definitive trial based on the findings, it is concluded that the overall results support the next steps of developing and evaluating the intervention.

References

Anderson, R., Saulsman, L., & Nathan, P. (2011). *Helping health anxiety*. Centre for Clinical Interventions.

Anxiety Candada (n.d.). *Calm breathing. https://www.anxietycanada.com/sites/default/files/CalmBreathing.pdf*

Bell, M. L., Whitehead, A. L., & Julious, S. A. (2018). Guidance for using pilot studies to inform the design of intervention trials with continuous outcomes. *Clinical Epidemiology*, *10*, 153-157. https://doi.org/10.2147/CLEP.S146397

Bobrow, K., Farmer, A., Cishe, N., Nwagi, N., Namane, M., Brennan, T. P., Springer, D., Tarassenko, L., & Levitt, N. (2018). Using the Medical Research Council framework for development and evaluation of complex interventions in a low resource setting to develop a theory-based treatment support intervention delivered via SMS text message to improve blood pressure control. *BMC Health Service Research*, *18*(1), Article 33. https://doi.org/10.1186/s12913-017-2808-9

Bussotti, M., & Sommaruga, M. (2018). Anxiety and depression in patients with pulmonary hypertension: impact and management challenges. *Vascular Health and rRisk Management*, *14*, 349-360. https://doi.org/10.2147/VHRM.S147173

Clark, D., & Oates, M. (2014). *Improving Access to Psychological Therapies: Measuring improvement and recovery adult services (Version 2)*. http://www.oxfordahsn.org/wp-content/uploads/2015/11/measuring-recovery-2014.pdf

Clark, D. M. (1986). A cognitive approach to panic. *Behaviour Research and Therapy*, *24*(4), 461-470. https://doi.org/10.1016/0005-7967(86)90011-2

Cohen, J. (1988). *Statistical power analysis for the behavioral sciences* (2nd ed.). Academic Press.

Connolly, D., O'Toole, L., Redmond, P., & Smith, S. M. (2013). Managing fatigue in patients with chronic conditions in primary care. *Family Practice, 30*(2), 123-124). https://doi.org/10.1093/fampra/cmt005

Craig, P., Dieppe, P., Macintyre, S., Mitchie, S., Nazareth, I., & Petticrew, M. (2008). Developing and evaluating complex interventions: The new Medical Research Council Guidance. *BMJ*, *337*, Article a1655. https://doi.org/10.1136/bmj.a1655

Cuijpers, P., & Schuurmans, J. (2007). Self-help interventions for anxiety disorders: An overview. *Current Psychiatry Reports*, *9*(4), 284-290. https://doi.org/10.1007/s11920-007-0034-6

Dugas, M. J., & Robichaud, M. (2007). *Practical clinical guidebooks. Cognitive-behavioral treatment for generalized anxiety disorder: From science to practice.* Routledge.

Ekers, D., Webster, L., Cuijpers, P., Richards, D., & Gilbody, S. (2014). Behavioural activation for depression; An update of meta-analysis of effectiveness and sub group analysis. *PLoS One*, *9*(6), Article e100100. https://doi.org/10.1371/journal.pone.0100100

Eldridge, S. M., Chan, C. L., Campbell, M. J., Bond, C. M., Hopewell, S., Thabane, L., & Lancaster, G. A. (2016). CONSORT 2010 statement: extension to randomised pilot and feasibility trials. *BMJ*, *355*, Article i5239. https://doi.org/10.1136/bmj.i5239

Eldridge, S. M., Lancaster, G. A., Campbell, M. J., Thabane, L., Hopewell, S., Coleman, C. L., & Bond, C. M. (2016). Defining feasibility and pilot studies in preparation for randomised controlled trials: Development of a conceptual framework. *PLoS One*, *11*(3), Article e0150205. https://doi.org/10.1371/journal.pone.0150205

George, D., & Mallery, P. (2003). *SPSS for Windows step by step: A simple guide and reference 11.0 update*. Allyn & Bacon.

Gersh, E., Hallford, D. J., Rice, S. M., Kazantzis, N., Gersh, H., Gersh, B., & McCarty, C. A. (2017). Systematic review and meta-analysis of dropout rates in individual psychotherapy for generalized anxiety disorder. *Journal of Anxiety Disorders*, *52*, 25-33. https://doi.org/10.1016/j.janxdis.2017.10.001

G*Power (2020). *G*Power 3.1.9.6.* https://www.psychologie.hhu.de/arbeitsgruppen/allgemeine-psychologie-und-arbeitspsychologie/gpower

Halimi, L., Marin, G., Molinari, N., Gamez, A.-S., Boissin, C., Suehs, C. M., Vachier, I., & Bourdin, A. (2018). Impact of psychological factors on the health-related quality of life of patients treated for pulmonary arterial hypertension. *Journal of Psychosomatic Research*, *105*, 45-51. https://doi.org/10.1016/j.jpsychores.2017.12.003

Harzheim, D., Klose, H., Pinado, F. P., Ehlken, N., Nagel, C., Fischer, C., Ghofrani, A., Rosenkranz, S., Seyfarth, H. J., Halank, M., Mayer, E., Grünig, E., & Guth, S. (2013). Anxiety and depression disorders in patients with pulmonary arterial hypertension and chronic thromboembolic pulmonary hypertension. *Respiratory Research*, *14*(1), Article 104. https://doi.org/10.1186/1465-9921-14-104

Haug, T., Nordgreen, T., Öst, L. G., & Havik, O. E. (2012). Self- help treatment of anxiety disorders: A meta- analysis and meta- regression of effects and potential moderators. *Clinical Psychology Review*, *32*(5), 425-445. https://doi.org/10.1016/j.cpr.2012.04.002

Hayes, A.F. (2012). *Introduction to Mediation, Moderation, and Conditional Process Analysis.* https://processmacro.org/index.html

Huxley, K. (2020). *Content analysis, quantitative*. SAGE Publications Ltd.

Hoeper, M. M., Ghofrani, H.-A., Grünig, E., Klose, H., Olschewski, H., & Rosenkranz, S. (2017). Pulmonary Hypertension. *Deutsches Arzteblatt international*, *114*(5), 73-84. https://doi.org/10.3238/arztebl.2017.0073

Hoeper, M. M., & Gibbs, S. R. (2014). The changing landscape of pulmonary arterial hypertension and implications for patient care. *European Respiratory Review*, *23*(134), 450-457. https://doi.org/10.1183/09059180.00007814

Hoeper, M. M., Kramer, T., Pan, Z., Eichstaedt, C. A., Spiesshoefer, J., Benjamin, N., Olsson, K. M., Meyer, K., Vizza, C. D., Vonk-Noordegraaf, A., Distler, O., Opitz, C., Gibbs, J. S. R., Delcroix, M., Ghofrani, H. A., Huscher, D., Pittrow, D., Rosenkranz, S., & Grünig, E. (2017). Mortality in pulmonary arterial hypertension: prediction by the 2015 European pulmonary hypertension guidelines risk stratification model. *European Respiratory Journal*, *50*(2), Article 1700740. https://doi.org/10.1183/13993003.00740-2017

Hoffmann, T. C., Glasziou, P. P., Boutron, I., Milne, R., Perera, R., Moher, D., Altman, D. G., Barbour, V., Macdonald, H., Johnston, M., Lamb, S. E., Dixon-Woods, M., McCulloch, P., Wyatt, J. C., Chan, A.-W., & Michie, S. (2014). Better reporting of interventions: template for intervention description and replication (TIDieR) checklist and guide. *BMJ*, *348*, Article 1687. https://doi.org/10.1136/bmj.g1687

Howard, C., & Dupont, S. (2014). 'The COPD breathlessness manual': a randomised controlled trial to test a cognitive-behavioural manual versus information booklets on health service use, mood and health status, in patients with chronic obstructive pulmonary disease. *Npj Primary Care Respiratory Medicine*, *24*, Article 14076. https://doi.org/10.1038/npjpcrm.2014.76

IBM Corp. Released 2017. *IBM SPSS Statistics for Macintosh, Version 25.0*. IBM Corp

Johnston, D. W. (2000). Cognitive behaviour therapy for cardiovascular diseases. *Zeitschrift fur Kardiologie, 89*, S9. https://doi.org/10.1007/s003920070035

Khazan, I.Z. (2013). *The clinical handbook of biofeedback: A step-by-step guide for training and practice with mindfulness*. John Wiley & Sons Ltd.

Kroenke, K., Spitzer, R. L., & Williams, J. B. W. (2001). The PHQ-9. *Journal of General Internal Medicine*, *16*(9), 606-613. https://doi.org/10.1046/j.1525-1497.2001.016009606.x

Laerd Statistics (2021). *Mixed ANOVA using SPSS Statistics*. https://statistics.laerd.com/spss-tutorials/mixed-anova-using-spss-statistics.php

Lebel, S., Mutsaers, B., Tomei, C., Leclair, C. S., Jones, G., Petricone-Westwood, D., Rutkowski, N., Ta, V., Trudel, G., Laflamme, S. Z., Lavigne, A.-A., & Dinkel, A. (2020). Health anxiety and illness-related fears across diverse chronic illnesses: A systematic review on conceptualization, measurement, prevalence, course, and correlates. *PloS one*, *15*(7), Article e0234124. https://doi.org/10.1371/journal.pone.0234124

Leon, A. C., Davis, L. L., & Kraemer, H. C. (2010). The role and interpretation of pilot studies in clinical research. *Journal of Psychiatric Research*, *45*(5), 626-629. https://doi.org/10.1016/j.jpsychires.2010.10.008

Lowe, B., Grafe, K., Ufer, C., Kroenke, K., Grunig, E., Herzog, W., & Borst, M. M. (2004). Anxiety and depression in patients with pulmonary hypertension. *Psychosomatic Medicine*, *66*(6), 831-836. https://doi.org/10.1097/01.psy.0000145593.37594.39

Mander, L, & Camerson, L. (2013). *Anxiety: A self help guide.* http://www.selfhelpguides.ntw.nhs.uk/iaptsheffield/leaflets/selfhelp/Anxiety.pdf

Matthew Prina, A., Marioni, R. E., Hammond, G. C., Jones, P. B., Brayne, C., & Dening, T. (2014). Improving access to psychological therapies and older people: Findings from the Eastern Region. *Behaviour Research and Therapy*, *56*(1), 75-81. https://doi.org/10.1016/j.brat.2014.03.008

McGoon, M., Gutterman, D., Steen, V., Barst, R., McCrory, D. C., Fortin, T. A., & Loyd, J. E. (2004). Screening, early detection, and diagnosis of pulmonary arterial hypertension: ACCP evidence-based clinical practice guidelines. *Chest*, *126*(1), 14S-34S. https://doi.org/10.1378/chest.126.1_suppl.14S

Michie, S., Wood, C. E., Johnston, M., Abraham, C., Francis, J. J., & Hardeman, W. (2015). Behaviour change techniques: The development and evaluation of a taxonomic method for reporting and describing behaviour change interventions (a suite of five studies involving consensus methods, randomised controlled trials and analysis of qualitative data). *Health Technology Assessment*, *19*(99), 1-187. https://doi.org/10.3310/hta19990

Muntingh, A. D. T., Gerritsen, S., Batelaan, N. M., & Bogaard, H. J. (2017). Pathways to experiencing mental problems and need for mental support in patients with pulmonary arterial hypertension. *Pulmonary Circulation*, *7*(3), 609-616. https://doi.org/10.1177/2045893217724750

National Institute for Health and Care Excellence. (2019). *Generalised anxiety disorder and panic disorder in adults: management Clinical guideline [CG113]*. https://www.nice.org.uk/guidance/cg113/chapter/1-Guidance.

Novakova, B., Harris, P. R., Rawlings, G. H., & Reuber, M. (2019). Coping with stress: A pilot study of a self-help stress management intervention for patients with epileptic or psychogenic nonepileptic seizures. *Epilepsy & Behavior*, *94*, 169-177. https://doi.org/10.1016/j.yebeh.2019.03.002

O'Cathain, A., Croot, L., Duncan, E., Rousseau, N., Sworn, K., Turner, K. M., Yardley, L., & Hoddinott, P. (2019). Guidance on how to develop complex interventions to improve health and healthcare. *BMJ Open*, *9*(8), Article e029954. https://doi.org/10.1136/bmjopen-2019-029954

Padesky, C. A., & Mooney, K. A. (1990). Presenting the cognitive model to clients. *International Cognitive Therapy Newsletter*, *6*, 13-14.

Panagioti, M., Scott, C., Blakemore, A., & Coventry, P. A. (2014). Overview of the prevalence, impact, and management of depression and anxiety in chronic obstructive pulmonary disease. *International Journal of Chronic Obstructive Pulmonary Disease*, *9*(1), 1289-1306. https://doi.org/10.2147/COPD.S72073

Pasterfield M. & Thompson, A.R. (2013). *Building Confidence in Social Situations: A guide for people living with a skin condition, including scars.* http://iaptsheffield.shsc.nhs.uk/wp-content/uploads/2019/05/Building-Confidence-in-Social-Situations-Skin-Conditions.pdf

Pasterfield, M., Clarke, S. A., & Thompson, A. R. (2019). The development of a self-help intervention to build social confidence in people living with visible skin conditions or scars: a think-aloud study. *Scars, Burns & Healing*, *5*, Article 2059513118822954. https://doi.org/10.1177/2059513118822954

Patel, T., Mansell, W., & Veale, D. (2015). The ognitive Behavioural Processes Questionnaire: A preliminary analysis within student, mixed clinical and community samples and the identification of a core transdiagnostic process. *Cognitive Therapy and Research*, *39*(2), 193-203. https://doi.org/10.1007/s10608-014-9641-9

Pearlin, L. I., & Schooler, C. (1978). The structure of coping. *Journal of Health and Social Behavior*, *19*(1), 2-21. https://doi.org/10.2307/2136319

Preacher, K.J & & Leonardelli, G.J. (2021, April 20). *Calculation for the Sobel test: An interactive calculation tool for mediation tests*. http://quantpsy.org/sobel/sobel.htm

Psychology Tools Limited, (n.d.). *Pacing for pain and fatigue.* https://www.psychologytools.com/resource/pacing-for-pain-and-fatigue/

Rawlings, G. H., Armstrong, I., Beail, N., & Thompson, A. R. (2021). *The impact of the coronavirus pandemic on people with pulmonary hypertension in the UK and how they are coping? A mixed-methods investigation* [Manuscript submitted for publication]. Clinical Psychology Unit, University of Sheffield.

Rawlings, G. H., Beail, N., Armstrong, I., Condliffe, R., Kiely, D. G., Sabroe, I., & Thompson, A. R. (2020). Adults' experiences of living with pulmonary hypertension: a thematic synthesis of qualitative studies. *BMJ Open*, *10*(12), Article e041428. https://doi.org/10.1136/bmjopen-2020-041428

Rawlings, G. H., Thompson, A. R., Armstrong, I., Novakova, B., & Beail, N. (2021). *Coping styles associated with health-anxiety, depression and health-related quality of life in pulmonary hypertension* [Manuscript submitted for publication]. Clinical Psychology Unit, University of Sheffield.

Salkovskis, P. M., Rimes, K. A., Warwick, H. M. C., & Clark, D. M. (2002). The Health Anxiety Inventory: Development and validation of scales for the measurement of health anxiety and hypochondriasis. *Psychological Medicine*, *32*(5), 843-853. https://doi.org/10.1017/S0033291702005822

Schachinger, H., Grob, M., Ritz, R., & Soler, M. (2000). Mental stress increases right heart afterload in severe pulmonary hypertension. *Clinical Physiology*, *20*(6), 483-487. https://doi.org/10.1046/j.1365-2281.2000.00287.x

Sedgwick, P., & Greenwood, N. (2015). Understanding the Hawthorne effect. *BMJ*, *351*, Article h4672. https://doi.org/10.1136/bmj.h4672

Shafran, R., Cooper , P., & Brosan, L. (2013). *The Complete CBT Guide for Anxiety*. Robinson.

Spitzer, R. L., Kroenke, K., Williams, J. B. W., & Löwe, B. (2006). A brief measure for assessing generalized anxiety disorder: the GAD-7. *Archives of Internal Medicine*, *166*(10), 1092-1097.

Steimer, T. (2002). The biology of fear- and anxiety-related behaviors. *Dialogues in Clinical Neuroscience*, *4*(3), 231-249.

Tarantino, E., Crescenzo, P., Campobasso, M., D'Alessandro, F., Argiento, P., Romeo, E., Ghio, S., Vitulo, P., Parisi, F., Mule, M., Albera, C., Vizza, C. D., & D'Alto, M. (2020). Psychological counselling interventions to improve perceived quality of life and counter anxiety and depression in pulmonary-arterial-hypertension patients: A clinical trial. *Journal of Health and Social Sciences*, *5*(4), 533-542. https://doi.org/10.19204/2020/psyc4

Tartavoulle, T. M., Karpinski, A. C., Aubin, A., Kluger, B. M., Distler, O., & Saketkoo, L. A. (2018). Multidimensional fatigue in pulmonary hypertension: prevalence, severity and predictors. *European Respiratory Journal Open Research*, *4*(1), Article 00079-2017. 10.1183/23120541.00079-2017

Therapist Aid, (2017). *Progressive muscle relaxation script*. https://www.therapistaid.com/worksheets/progressive-muscle-relaxation-script.pdf

Watkins, L. L., Koch, G. G., Sherwood, A., Blumenthal, J. A., Davidson, J. R. T., O'Connor, C., & Sketch, M. H. (2013). Association of anxiety and depression with all- cause mortality in individuals with coronary heart disease. *Journal of the American Heart Association*, *2*(2), Article e000068. https://doi.org/10.1161/JAHA.112.000068

Webster, R., Thompson, A., Webb, T. L., & Sheeran, P. (2014). *Self-help treatments and stepped care*. In P. Emmelkamp & T. Ehring (Eds.), *The Wiley handbook of anxiety disorders* (pp. 1242–1255). John Wiley & Sons.

Whalley, M. & Kaur, D. (2020, 19th March). *Living With Worry And Anxiety Amidst Global Uncertainty*. https://www.psychologytools.com/articles/free-guide-to-living-with-worry-and-anxiety-amidst-global-uncertainty/

Whitehead, A. L., Julious, S. A., Cooper, C. L., & Campbell, M. J. (2016). Estimating the sample size for a pilot randomised trial to minimise the overall trial sample size for the external pilot and main trial for a continuous outcome variable. *Statistical Methods in Medical Research*, *25*(3), 1057-1073. https://doi.org/10.1177/0962280215588241

Whitehead, A. L., Sully, B. G. O., & Campbell, M. J. (2014). Pilot and feasibility studies: Is there a difference from each other and from a randomised controlled trial? *Contemporary Clinical Trials*, *38*(1), 130-133. https://doi.org/10.1016/j.cct.2014.04.001

Wijeratne, T. D., Lajkosz, B. K., Brogly, D. S., Lougheed, M. M., Jiang, L. L., Housin, L. A., Barber, L. D., Johnson, L. A., Dolisczny, L. K., & Archer, L. S. (2018). Increasing incidence and prevalence of World Health Organization groups 1 to 4 pulmonary hypertension: A population-based cohort study in Ontario, Canada. *Circulation: Cardiovascular Quality and Outcomes*, *11*(2), Article e003973. https://doi.org/10.1161/CIRCOUTCOMES.117.003973

Wryobeck, J. M., Lippo, G., McLaughlin, V., Riba, M., & Rubenfire, M. (2007). Psychosocial aspects of pulmonary hypertension: A review [Physical & Somatoform & Psychogenic Disorders 3290]. *Psychosomatics: Journal of Consultation and Liaison Psychiatry*, *48*(6), 467-475. https://doi.org/http://dx.doi.org/10.1176/appi.psy.48.6.467

Yardley, L., Morrison, L., Bradbury, K., & Muller, I. (2015). The person-based approach to intervention development: Application to digital health-related behavior change interventions. *J Med Internet Res*, *17*(1), Article e30. https://doi.org/10.2196/jmir.4055

Yorke, J., Corris, P., Gaine, S., Gibbs, J., Kiely, D., Harries, C., Pollock, V., & Armstrong, I. (2014). emPHasis-10: development of a health-related quality of life measure in pulmonary hypertension. *European Respiratory Journal*, *43*(4), 1106-1113. https://doi.org/10.1183/09031936.00127113

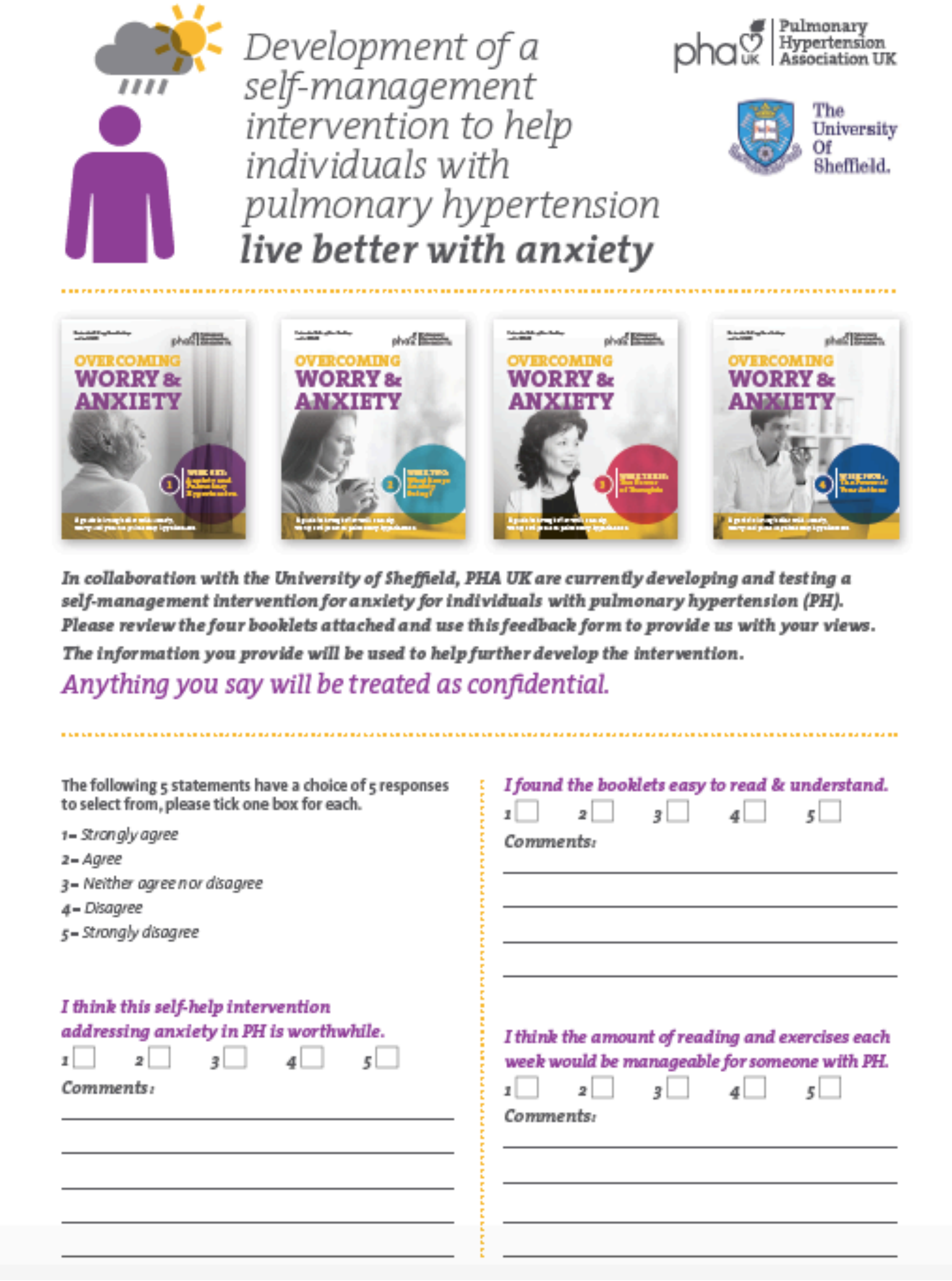
Yorke, J., Corris, P., Gaine, S., Gibbs, S. S., Kiely, D. G., Harries, C., Pollock, V., & Armstrong, I. (2014). emPHasis- 10: development of a health- related quality of life measure in pulmonary hypertension. *European Respiratoy Journal, 43,* 1106-1113. doi: 10.1183/09031936.00127113

Yorke, J., Deaton, C., Campbell, M., McGowen, L., Sephton, P., Kiely, D. G., & Armstrong, I. (2018). 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 Respiratory Research*, *5*, Article e000263. https://doi.org/10.1136/bmjresp-2017-000263

Yorke, J., Moosavi, S. H., Shuldham, C., & Jones, P. W. (2010). Quantification of dyspnoea using descriptors: development and initial testing of the Dyspnoea- 12. *Thorax*, *65*, 21-26. https://doi.org/10.1136/thx.2009.118521

Appendices

Appendix A: Stakeholders' Questionnaire



Development of a self-management intervention to help individuals with pulmonary hypertension

live better with anxiety

pha UK | Pulmonary Hypertension Association UK

The University Of Sheffield.

OVERCOMING WORRY & ANXIETY

OVERCOMING WORRY & ANXIETY

OVERCOMING WORRY & ANXIETY

OVERCOMING WORRY & ANXIETY

In collaboration with the University of Sheffield, PHA UK are currently developing and testing a self-management intervention for anxiety for individuals with pulmonary hypertension (PH). Please review the four booklets attached and use this feedback form to provide us with your views. The information you provide will be used to help further develop the intervention.

Anything you say will be treated as confidential.

The following 5 statements have a choice of 5 responses to select from, please tick one box for each.

1 – *Strongly agree*
2 – *Agree*
3 – *Neither agree nor disagree*
4 – *Disagree*
5 – *Strongly disagree*

I think this self-help intervention addressing anxiety in PH is worthwhile.

1 ☐ 2 ☐ 3 ☐ 4 ☐ 5 ☐

Comments:

____
____
____
____
____

I found the booklets easy to read & understand.

1 ☐ 2 ☐ 3 ☐ 4 ☐ 5 ☐

Comments:

____
____
____
____

I think the amount of reading and exercises each week would be manageable for someone with PH.

1 ☐ 2 ☐ 3 ☐ 4 ☐ 5 ☐

Comments:

____
____
____
____

There was a good balance between the amount of text and number of images.

1☐ 2☐ 3☐ 4☐ 5☐

Comments:

The quotations, and stories of Nadia and John were suitable and appropriate.

1☐ 2☐ 3☐ 4☐ 5☐

Comments:

Was there anything that you did not like about the booklets?

What did you like most about the booklets?

Do you have any other comments?

Thank you for your time.

FURTHER SUPPORT FROM THE PHA UK

A range of FREE resources are available on a variety of topics to help inform and advise people affected by PH.

If you would like copies of any of our publications go to **www.phauk.org/publications** *call us on* **01709 761450** *or email us at* **office@phauk.org**

pha UK Influence. Hope. Integrity.

Registered Charity No. 1120756

Appendix B: Self-Help Intervention

Week One:


Produced by Dr Gregg Harry Rawlings
and the PHA UK

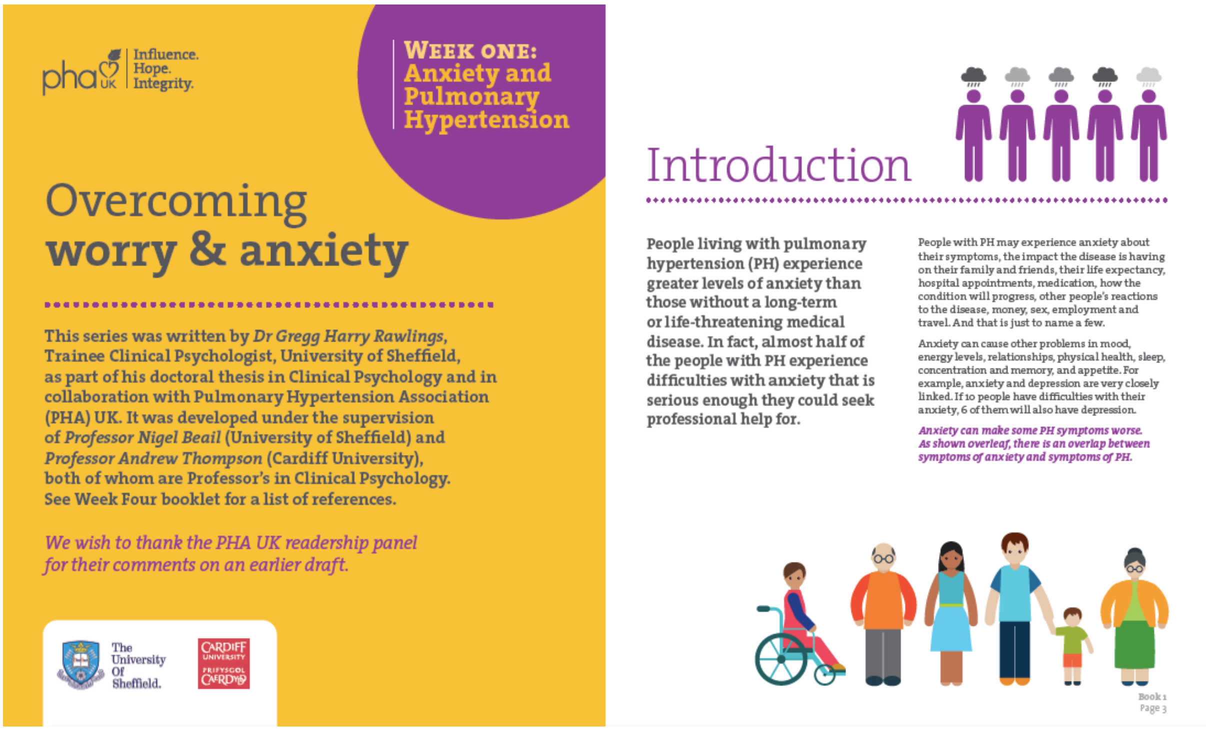
Pulmonary
Hypertension
Association UK

OVERCOMING
WORRY &
ANXIETY

1

WEEK ONE:
Anxiety and
Pulmonary
Hypertension

A guide to living better with anxiety,
worry and panic in pulmonary hypertension


Influence.
Hope.
Integrity.

WEEK ONE:
Anxiety and
Pulmonary
Hypertension

Overcoming
worry & anxiety

This series was written by *Dr Gregg Harry Rawlings*, Trainee Clinical Psychologist, University of Sheffield, as part of his doctoral thesis in Clinical Psychology and in collaboration with Pulmonary Hypertension Association (PHA) UK. It was developed under the supervision of *Professor Nigel Beail* (University of Sheffield) and *Professor Andrew Thompson* (Cardiff University), both of whom are Professor's in Clinical Psychology. See Week Four booklet for a list of references.

We wish to thank the PHA UK readership panel for their comments on an earlier draft.

The University Of Sheffield.

Introduction

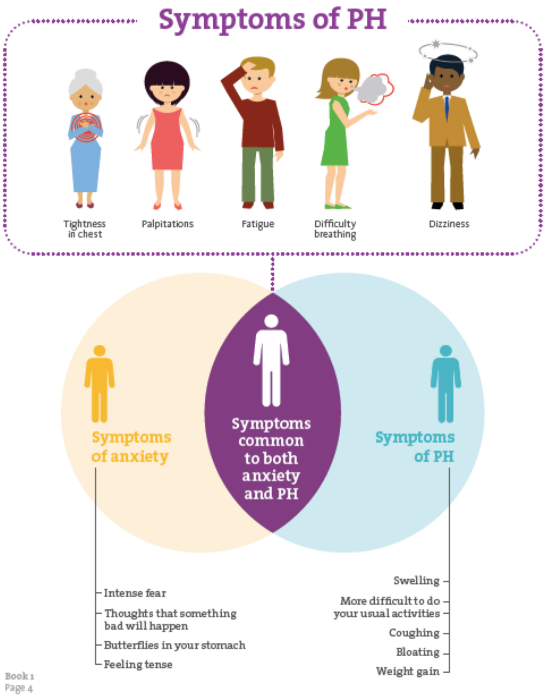
People living with pulmonary hypertension (PH) experience greater levels of anxiety than those without a long-term or life-threatening medical disease. In fact, almost half of the people with PH experience difficulties with anxiety that is serious enough they could seek professional help for.

People with PH may experience anxiety about their symptoms, the impact the disease is having on their family and friends, their life expectancy, hospital appointments, medication, how the condition will progress, other people's reactions to the disease, money, sex, employment and travel. And that is just to name a few.

Anxiety can cause other problems in mood, energy levels, relationships, physical health, sleep, concentration and memory, and appetite. For example, anxiety and depression are very closely linked. If 10 people have difficulties with their anxiety, 6 of them will also have depression.

Anxiety can make some PH symptoms worse. As shown overleaf, there is an overlap between symptoms of anxiety and symptoms of PH.

Book 1
Page 3

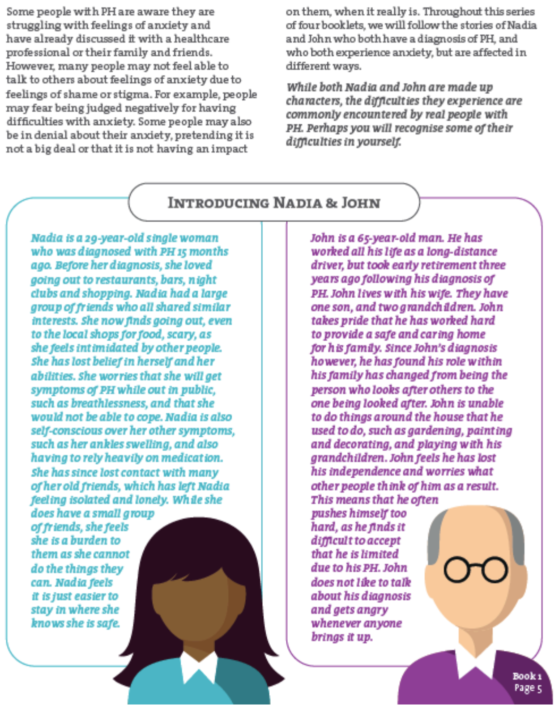


Symptoms of PH

- Intense fear
- Thoughts that something bad will happen
- Butterflies in your stomach
- Feeling tense

- Swelling
- More difficult to do your usual activities
- Coughing
- Bloating
- Weight gain

Book 1
Page 4



Some people with PH are aware they are struggling with feelings of anxiety and have already discussed it with a healthcare professional or their family and friends. However, many people may not feel able to talk to others about feelings of anxiety due to feelings of shame or stigma. For example, people may fear being judged negatively for having difficulties with anxiety. Some people may also be in denial about their anxiety, pretending it is not a big deal or that it is not having an impact on them, when it really is. Throughout this series of four booklets, we will follow the stories of Nadia and John who both have a diagnosis of PH, and who both experience anxiety, but are affected in different ways.

While both Nadia and John are made up characters, the difficulties they experience are commonly encountered by real people with PH. Perhaps you will recognise some of their difficulties in yourself.

INTRODUCING NADIA & JOHN

Nadia is a 29-year-old single woman who was diagnosed with PH 15 months ago. Before her diagnosis, she loved going out to restaurants, bars, night clubs and shopping. Nadia had a large group of friends who all shared similar interests. She now finds going out, even to the local shops for food, scary, as she feels intimidated by other people. She has lost belief in herself and her abilities. She worries that she will get symptoms of PH while out in public, such as breathlessness, and that she would not be able to cope. Nadia is also self-conscious over her other symptoms, such as her ankles swelling, and also having to rely heavily on medication. She has since lost contact with many of her old friends, which has left Nadia feeling isolated and lonely. While she does have a small group of friends, she feels she is a burden to them as she cannot do the things they can. Nadia feels it is just easier to stay in where she knows she is safe.

John is a 65-year-old man. He has worked all his life as a long-distance driver, but took early retirement three years ago following his diagnosis of PH. John lives with his wife. They have one son, and two grandchildren. John takes pride that he has worked hard to provide a safe and caring home for his family. Since John's diagnosis however, he has found his role within his family has changed from being the person who looks after others to the one being looked after. John is unable to do things around the house that he used to do, such as gardening, painting and decorating, and playing with his grandchildren. John feels he has lost his independence and worries what other people think of him as a result. This means that he often pushes himself too hard, as he finds it difficult to accept that he is limited due to his PH. John does not like to talk about his diagnosis and gets angry whenever anyone brings it up.

Book 1
Page 5

OVERCOMING
WORRY &
ANXIETY
1 WEEK ONE: Anxiety and Pulmonary Hypertension
A guide to living better with anxiety, worry and panic in pulmonary hypertension

OVERCOMING
WORRY &
ANXIETY
2 WEEK TWO: What keeps Anxiety Going?
A guide to living better with anxiety, worry and panic in pulmonary hypertension

OVERCOMING
WORRY &
ANXIETY
3 WEEK THREE: The Power of Thoughts
A guide to living better with anxiety, worry and panic in pulmonary hypertension

OVERCOMING
WORRY &
ANXIETY
4 WEEK FOUR: The Power of Your Actions
A guide to living better with anxiety, worry and panic in pulmonary hypertension

Book 1
Page 6

How to use these booklets

The goal of this series of four weekly booklets is to help you to live better with your anxiety. The information is based on a type of psychological treatment called Cognitive Behavioural Therapy, or CBT for short.

CBT can help you manage your anxiety by changing the way you think (the cognitive part) and what you do (the behavioural part).

As you go through each booklet, it will help you to better understand anxiety, how anxiety and PH can be connected, and how anxiety can become a problem. It will then help you to develop useful strategies aimed at better managing your anxiety.

You should read each booklet in order, giving yourself time to understand the different sections before moving onto the next booklet. Each booklet should take about one week to read and practice the different strategies. There is at least one strategy at the end of the booklets for you to practice over the following week, which aims to help you manage your anxiety.

We would recommend that you tell a family member or a friend that you are working through these booklets. Ask them if they would like to also read it along with you and provide support when you are making changes.

If you want to better manage your anxiety, it will not be enough to just read these booklets. You will need to commit yourself to the tasks and *practice*, *practice*, *practice*.

The more effort you put into each booklet, the more likely your anxiety will get better. It can be useful to think of mental health like physical health. If someone sets out to walk 3 miles without stopping, then they will need to train often. Once they are physically fit enough, then they can achieve their goal. However, if they then stopped practicing, their fitness would reduce and soon they would be unable to walk 3 miles without stopping. Mental health is very similar. The more you practice activities designed to improve your mental health, the better it will be. As soon as you stop doing helpful activities, your mental health may begin to suffer.

If you need further support after reading these booklets, we recommend that you contact your healthcare provider. We have also provided a list of contacts at the end of each booklet who may be able to provide you with further support.

Book 1
Page 7

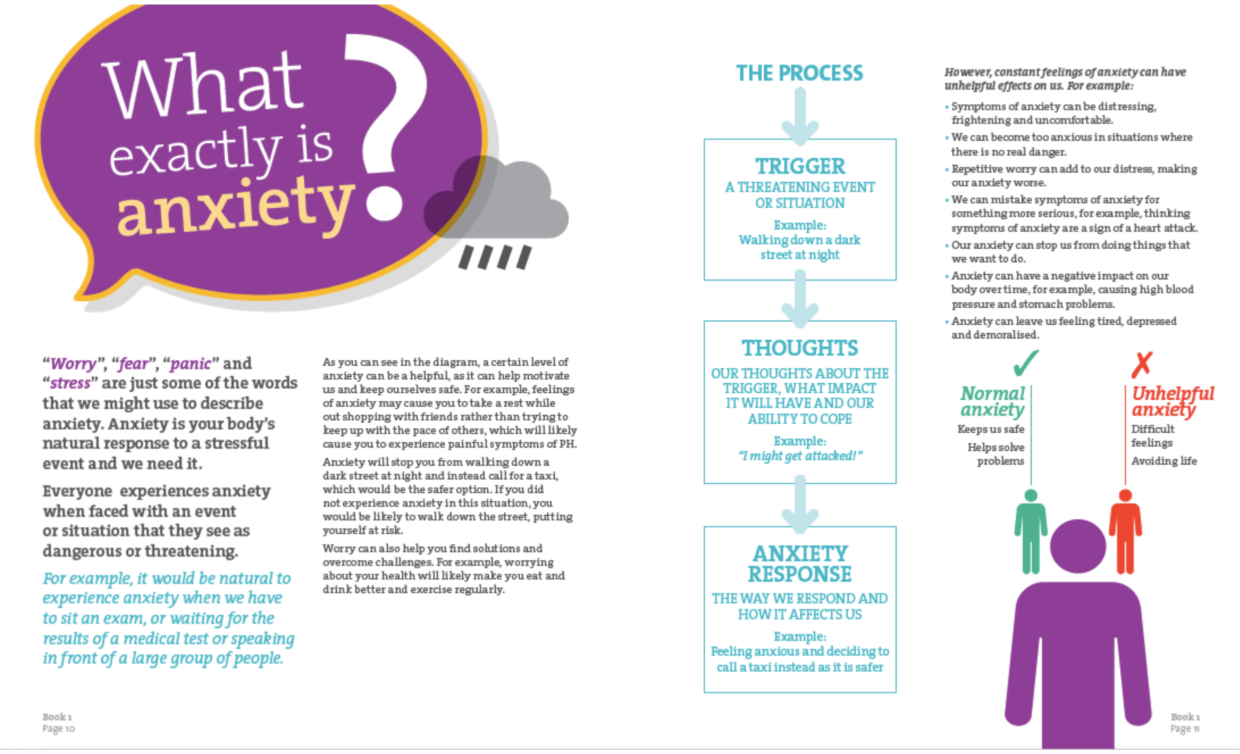
Learning from others

Remember, you are not alone in your difficulties with PH. Here is a testimony from Sarah who has PH and also struggles with anxiety.

You can also read Eileen's and Keith's testimonies in week two and three.

> *I've had anxiety for a long period of my life, but it worsened when I was diagnosed with Chronic Thromboembolic Pulmonary Hypertension (CTEPH) following a pulmonary embolism.*
>
> *I experience feelings of stress and panic, and I get physical symptoms of anxiety including heart palpitations and a rapid heartbeat. I also feel dizzy and nauseous. The worst part is the rapid heartbeat and palpitations because this was also a symptom of my embolism (and can be PH) and therefore it increases the worry and fear.*
>
> *In relation to the PH, I think the anxiety comes from constantly worrying that I could get ill again. According to my specialists, my embolism was not discovered as early as it should have been, so I feel like my symptoms may come back fiercely and it could potentially be too late.*
>
> *Listening to relaxing sounds and meditation helps me feel calmer and thankfully I still manage to go to work and have a fairly 'normal' lifestyle, but anxiety can occur randomly with seemingly no cause.*
>
> *It's so hard to understand something you can't see; even I don't always understand it, so I don't expect other people to.*
>
> **Sarah**

> *Cognitive Behavioural Therapy (CBT) has been shown to help people manage their anxiety, as well as improve their mood and overall quality of life.*
>
> **Dr Gregg Harry Rawlings**
> *Trainee Clinical Psychologist*
> *University of Sheffield*



What exactly is anxiety?

"*Worry*", "*fear*", "*panic*" and "*stress*" are just some of the words that we might use to describe anxiety. Anxiety is your body's natural response to a stressful event and we need it.

Everyone experiences anxiety when faced with an event or situation that they see as dangerous or threatening.

For example, it would be natural to experience anxiety when we have to sit an exam, or waiting for the results of a medical test or speaking in front of a large group of people.

As you can see in the diagram, a certain level of anxiety can be a helpful, as it can help motivate us and keep ourselves safe. For example, feelings of anxiety may cause you to take a rest while out shopping with friends rather than trying to keep up with the pace of others, which will likely cause you to experience painful symptoms of PH.

Anxiety will stop you from walking down a dark street at night and instead call for a taxi, which would be the safer option. If you did not experience anxiety in this situation, you would be likely to walk down the street, putting yourself at risk.

Worry can also help you find solutions and overcome challenges. For example, worrying about your health will likely make you eat and drink better and exercise regularly.

THE PROCESS

TRIGGER
A THREATENING EVENT OR SITUATION
Example:
Walking down a dark street at night

THOUGHTS
OUR THOUGHTS ABOUT THE TRIGGER, WHAT IMPACT IT WILL HAVE AND OUR ABILITY TO COPE
Example:
"I might get attacked!"

ANXIETY RESPONSE
THE WAY WE RESPOND AND HOW IT AFFECTS US
Example:
Feeling anxious and deciding to call a taxi instead as it is safer

However, constant feelings of anxiety can have unhelpful effects on us. For example:

- Symptoms of anxiety can be distressing, frightening and uncomfortable.
- We can become too anxious in situations where there is no real danger.
- Repetitive worry can add to our distress, making our anxiety worse.
- We can mistake symptoms of anxiety for something more serious, for example, thinking symptoms of anxiety are a sign of a heart attack.
- Our anxiety can stop us from doing things that we want to do.
- Anxiety can have a negative impact on our body over time, for example, causing high blood pressure and stomach problems.
- Anxiety can leave us feeling tired, depressed and demoralised.

✓ ***Normal anxiety***
Keeps us safe
Helps solve problems

✗ ***Unhelpful anxiety***
Difficult feelings
Avoiding life

What are the common symptoms of anxiety?

The first step in learning to manage your anxiety better is to recognise your symptoms. As you do this, you will become more confident at telling apart which symptoms are caused by anxiety, which are caused by PH and which could be both.

Anxiety can impact you in four main ways. You may have negative ***feelings***, worrying ***thoughts***, uncomfortable ***physical reactions***, all of which influence your ***behaviour***. Below is a list of common symptoms of anxiety. Please tick any that you have experienced when feeling anxious. Remember, anxiety is normal and just because you experience one of these symptoms does not mean it is bad. You may want to revisit this section as you work through the booklets and become more aware of your symptoms:

HOW YOU THINK

Constant worrying
Racing thoughts
Difficulty concentrating
Always thinking the worst will happen
Dwelling on the past
Worrying to find solutions to problems in life
Going over and over in your mind what you think will happen in the future
Self-critical thoughts, for example "I *can't cope*" or "*I'm useless*"
Replaying negative events in your mind
Trying to guess what other people are thinking of you
Thinking how other people will react
Distressing thoughts that you cannot stop coming into your mind
Becoming more aware of your own body sensations, such as breathing and heart rate
Other(s)

HOW YOU FEEL

Nervous
Worried
Frightened
Scared
Feeling something terrible is going to happen - dread
Tense
Stressed
Irritated
Agitated
Angry
On edge
Panicked
Strange
Out of sorts
Detached
Embarrassed
Other(s)

PHYSICAL REACTIONS

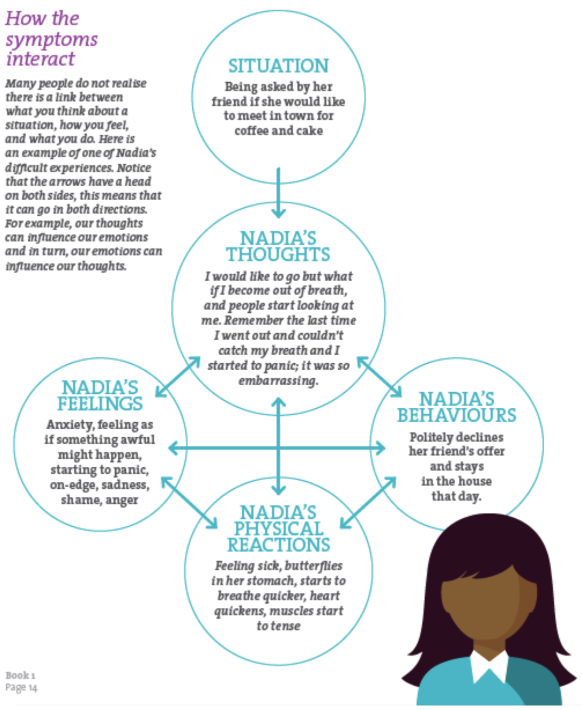
Pins and needles
Heart pounds/palpitations
Tightness and/or pain in your chest
Butterflies in your stomach
Breathing change i.e. breaths become quicker and more shallow
Sweating
Aches and pains
Having to use the toilet
Tense muscles
Struggling to rest or relax
Feeling surges of energy
Dizzy
Nausea/feeling sick
Woozy
Other(s)

YOUR BEHAVIOUR

Cannot sit still or relax
Finding it hard to switch off
Trouble starting and completing jobs
Become easily irritated or quick to anger
Finding it difficult to fall asleep
Eat more or less
Drink more or less
Smoke more
Drink alcohol more
Take recreational drugs
Withdraw from people
Prefer your own company
Talk more or less than usual
Avoid situations that you perceive as dangerous or difficult
Seeking reassurance from others
Other(s)

Book 1
Page 12

Book 1
Page 13



How the symptoms interact

Many people do not realise there is a link between what you think about a situation, how you feel, and what you do. Here is an example of one of Nadia's difficult experiences. Notice that the arrows have a head on both sides, this means that it can go in both directions. For example, our thoughts can influence our emotions and in turn, our emotions can influence our thoughts.

SITUATION

Being asked by her friend if she would like to meet in town for coffee and cake

NADIA'S THOUGHTS

I would like to go but what if I become out of breath, and people start looking at me. Remember the last time I went out and couldn't catch my breath and I started to panic; it was so embarrassing.

NADIA'S FEELINGS

Anxiety, feeling as if something awful might happen, starting to panic, on-edge, sadness, shame, anger

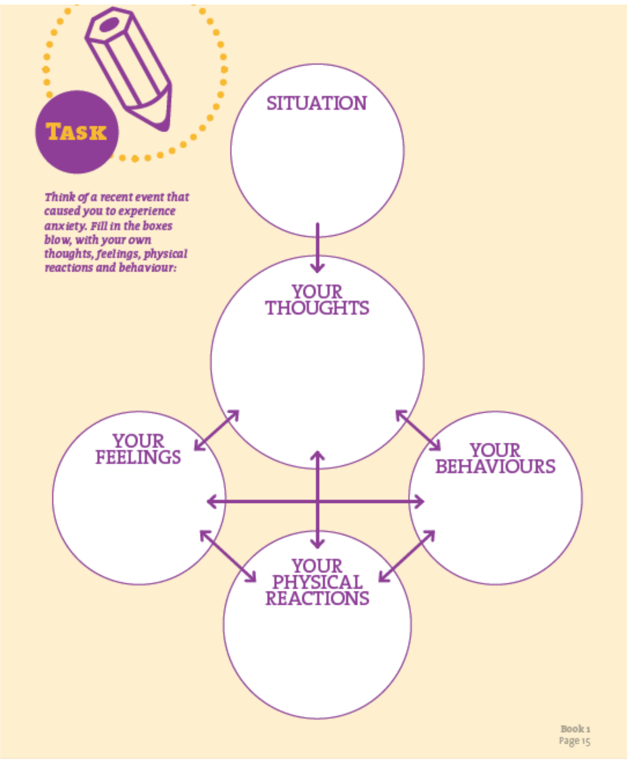
NADIA'S BEHAVIOURS

Politely declines her friend's offer and stays in the house that day.

NADIA'S PHYSICAL REACTIONS

Feeling sick, butterflies in her stomach, starts to breathe quicker, heart quickens, muscles start to tense

Book 1
Page 14



Task

Think of a recent event that caused you to experience anxiety. Fill in the boxes blow, with your own thoughts, feelings, physical reactions and behaviour:

Book 1
Page 15

Where does anxiety come from?

Anxiety is our body's natural alarm system, triggered by experiences that we perceive as threatening or dangerous. This response is known as the *"Fight-Flight-Freeze response"*.

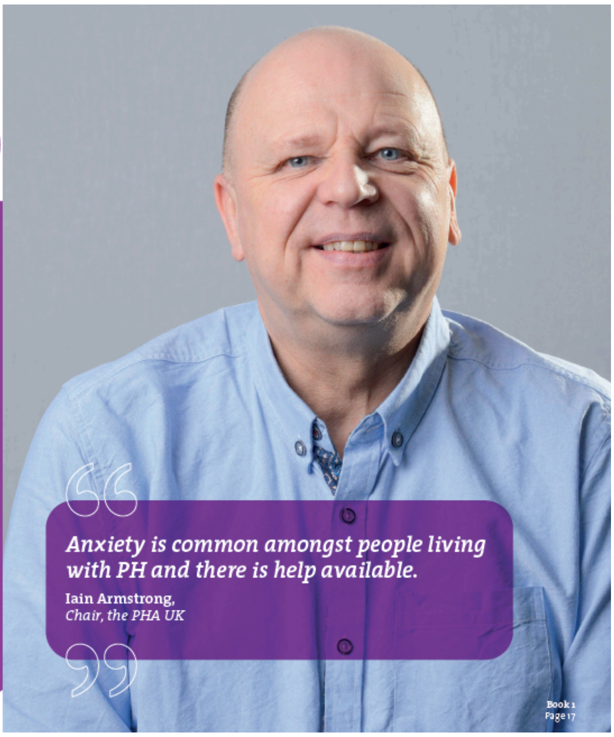
Imagine if an attacker suddenly bursts into the room you are in. Your body would have a sudden reaction without you even having to think about it. As the list right shows, your muscles would tense ready for action. Your breathing would increase to get more oxygen into your body for energy. Your heart would beat quicker to pump blood to your muscles and brain. You would start sweating to prevent your body from overheating. Your mind would start to race and you may get a surge of energy (also known as adrenaline) motivating you to act.

If the attacker gets a shock at how you react and runs away, your *"Fight-Flight-Freeze"* response would start to reduce. During this time however, you would still feel on edge and tense. Once your anxiety levels have returned to normal, you may be left feeling tired and upset.

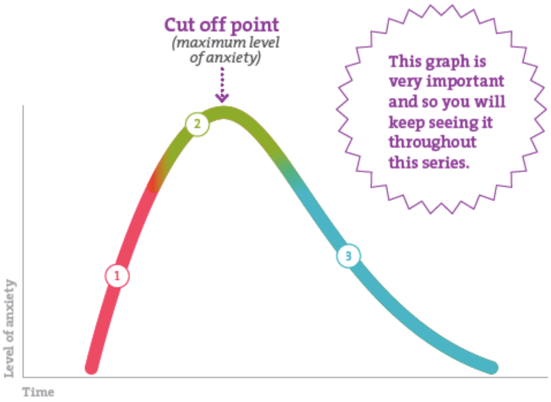
While Fiight or Flight is a response where your body prepares to fight or flee, freeze can help because it can give you more time to evaluate the situation before acting. If you freeze, you may hold your breath, feel unable to move or have an "out of body" experience.

- Release of adrenalin
- Breathing faster
- Churning stomach
- Tense muscles
- Start to sweat
- Mind starts to race
- Dry mouth
- Heart rate quickens
- Blood pressure increase
- Feeling sick
- Shaky hands
- Legs feel weak

This response is shown in the diagram on the next spread.



> *Anxiety is common amongst people living with PH and there is help available.*
>
> **Iain Armstrong,**
> *Chair, the PHA UK*



This graph is very important and so you will keep seeing it throughout this series.

① Your levels of anxiety will rise as your brain and body response to the situation

② Your anxiety will continue to rise until it reaches its "cut-off" point". At this stage, it can feel very distressing and scary. Your "Fight-Flight-Freeze" response will be making you experience the symptoms discussed previously. It is important to remember that your anxiety cannot get any worse as it has a "cut-off" point. This amount of anxiety is normal and harmless, although it may not feel like it in the moment.

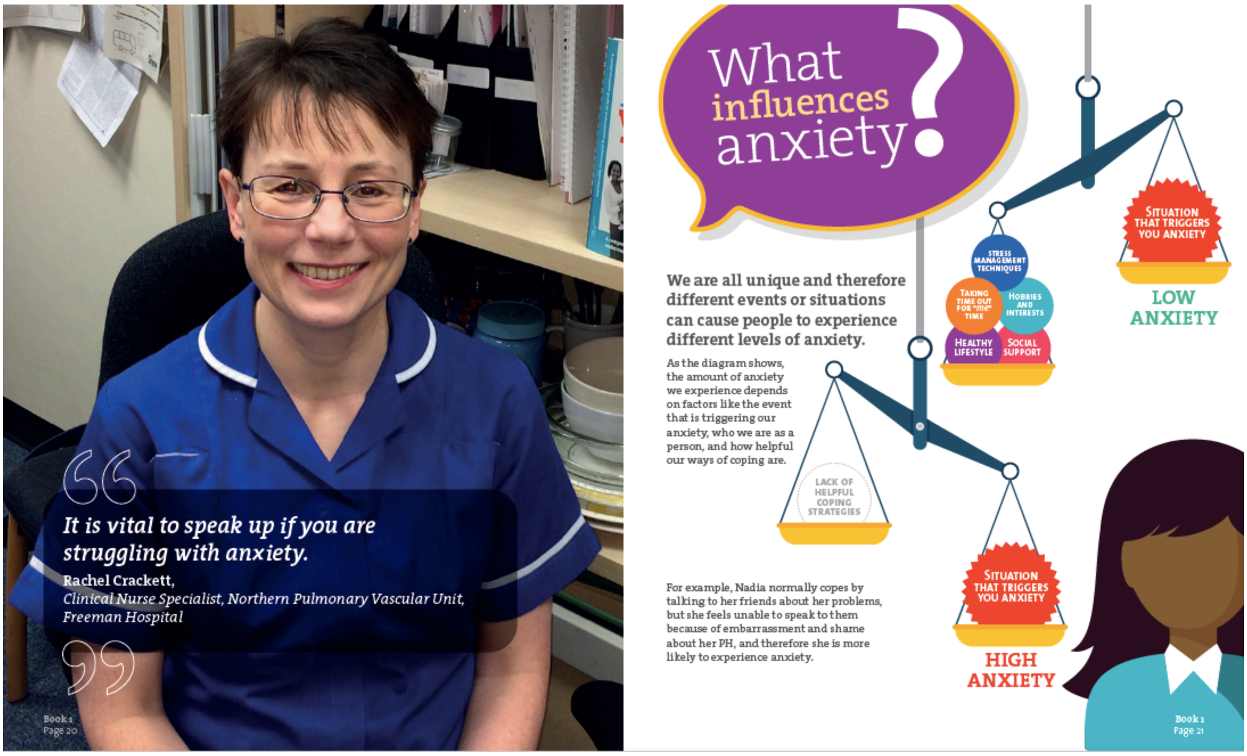
③ You can only experience this level of anxiety for a short period of time. This is because your body becomes tired and exhausted by the energy it takes to be so anxious. This means that your anxiety will gradually reduce overtime, until it returns to normal. This can be a quick or a more gradual reduction, but it will go down. For some people, their anxiety can surge again as it is reducing or soon after it has returned to normal. This is known as "riding the waves of panic". The idea is to ride the waves until the anxiety is more manageable.

Unhelpful anxiety

The *"Fight-Flight-Freeze"* response has been passed on from our ancestors for millions of years. During that time, the "Fight-Flight-Freeze " response has helped people to escape from life threatening situations. If our ancestors did not have this response, they would not have survived.

Today our survival rarely depends on fighting or running away from a situation. Instead, our anxiety response helps us to problem solve, concentrate and be motivated. However, most of the time, the physical symptoms of anxiety do not help and instead cause us problems.

For example, talking about your medication with your doctor might cause you to experience some anxiety. In this situation, you would want your mind to be clear and focused, allowing you to listen to your doctor and ask any questions. However, instead you may become distracted by your rapid heart rate and surge of energy making you get up and leave the room quickly, and before getting the chance to ask your questions.



> *It is vital to speak up if you are struggling with anxiety.*
>
> **Rachel Crackett,**
> *Clinical Nurse Specialist, Northern Pulmonary Vascular Unit, Freeman Hospital*

Book 1
Page 20

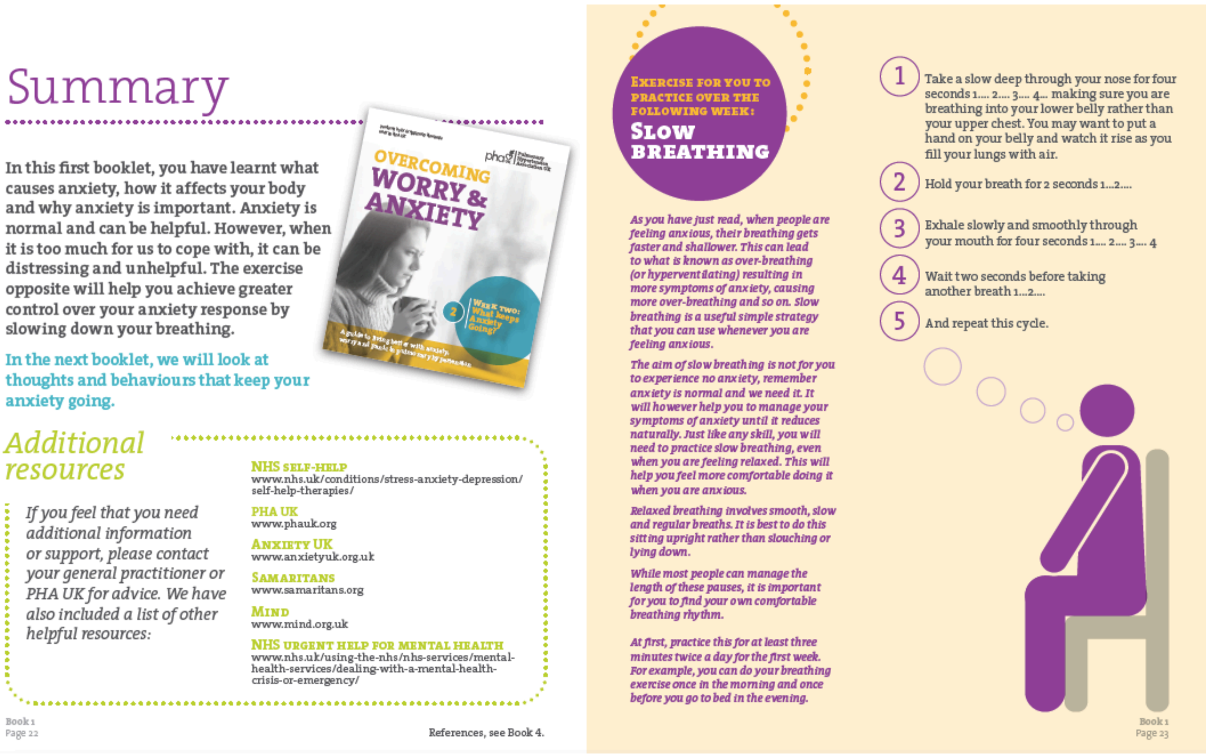
What influences anxiety?

We are all unique and therefore different events or situations can cause people to experience different levels of anxiety.

As the diagram shows, the amount of anxiety we experience depends on factors like the event that is triggering our anxiety, who we are as a person, and how helpful our ways of coping are.

For example, Nadia normally copes by talking to her friends about her problems, but she feels unable to speak to them because of embarrassment and shame about her PH, and therefore she is more likely to experience anxiety.

Book 1
Page 21



Summary

In this first booklet, you have learnt what causes anxiety, how it affects your body and why anxiety is important. Anxiety is normal and can be helpful. However, when it is too much for us to cope with, it can be distressing and unhelpful. The exercise opposite will help you achieve greater control over your anxiety response by slowing down your breathing.

In the next booklet, we will look at thoughts and behaviours that keep your anxiety going.

Additional resources

If you feel that you need additional information or support, please contact your general practitioner or PHA UK for advice. We have also included a list of other helpful resources:

NHS SELF-HELP
www.nhs.uk/conditions/stress-anxiety-depression/self-help-therapies/

PHA UK
www.phauk.org

ANXIETY UK
www.anxietyuk.org.uk

SAMARITANS
www.samaritans.org

MIND
www.mind.org.uk

NHS URGENT HELP FOR MENTAL HEALTH
www.nhs.uk/using-the-nhs/nhs-services/mental-health-services/dealing-with-a-mental-health-crisis-or-emergency/

Book 1
Page 22

References, see Book 4.

EXERCISE FOR YOU TO PRACTICE OVER THE FOLLOWING WEEK:
SLOW BREATHING

As you have just read, when people are feeling anxious, their breathing gets faster and shallower. This can lead to what is known as over-breathing (or hyperventilating) resulting in more symptoms of anxiety, causing more over-breathing and so on. Slow breathing is a useful simple strategy that you can use whenever you are feeling anxious.

The aim of slow breathing is not for you to experience no anxiety, remember anxiety is normal and we need it. It will however help you to manage your symptoms of anxiety until it reduces naturally. Just like any skill, you will need to practice slow breathing, even when you are feeling relaxed. This will help you feel more comfortable doing it when you are anxious.

Relaxed breathing involves smooth, slow and regular breaths. It is best to do this sitting upright rather than slouching or lying down.

While most people can manage the length of these pauses, it is important for you to find your own comfortable breathing rhythm.

At first, practice this for at least three minutes twice a day for the first week. For example, you can do your breathing exercise once in the morning and once before you go to bed in the evening.

① Take a slow deep through your nose for four seconds 1.... 2.... 3.... 4... making sure you are breathing into your lower belly rather than your upper chest. You may want to put a hand on your belly and watch it rise as you fill your lungs with air.

② Hold your breath for 2 seconds 1...2....

③ Exhale slowly and smoothly through your mouth for four seconds 1.... 2.... 3.... 4

④ Wait two seconds before taking another breath 1...2....

⑤ And repeat this cycle.

Book 1
Page 23

Influence.
Hope.
Integrity.

These booklets have been created with experts by education and experts by experience, with the aim of empowering people with pulmonary hypertension to develop skills to live better with anxiety.

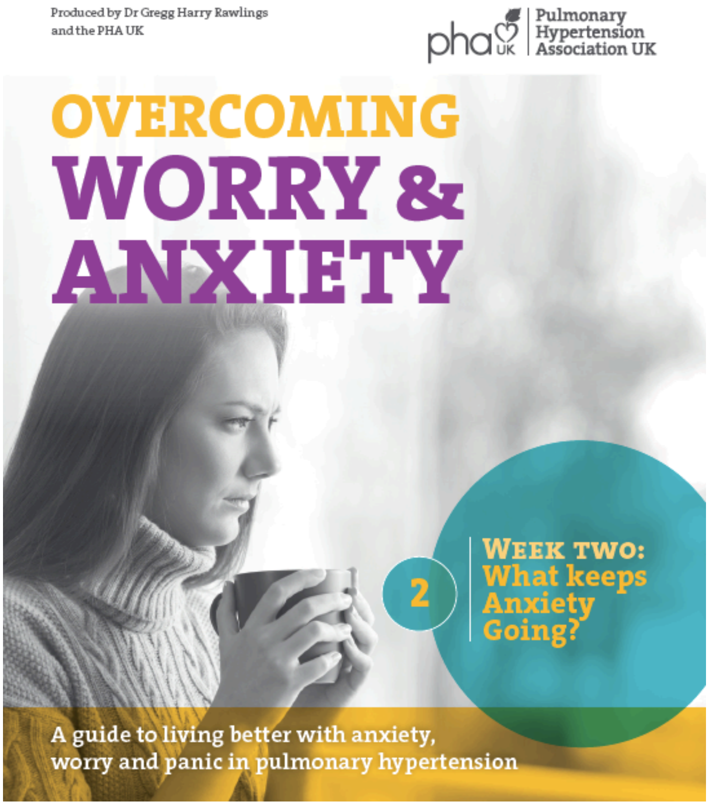
Dr Gregg Harry Rawlings, Trainee Clinical Psychologist

To find out more about the PHA UK, visit **www.phauk.org**

Pulmonary Hypertension Association UK
PHA UK Resource Centre, Unit 1, Newton Business Centre, Newton Chambers Road,
Thorncliffe Park, Chapeltown, Sheffield, England S35 2PH
T: 01709 761450 E: office@phauk.org @PULHAUK @PHA_UK @PHA ORG UK

Registered Charity No. 1120756. Copyright © 2020 PHA UK.

Week Two:



Produced by Dr Gregg Harry Rawlings
and the PHA UK

Pulmonary
Hypertension
Association UK

OVERCOMING
WORRY &
ANXIETY

2

WEEK TWO:
What keeps
Anxiety
Going?

A guide to living better with anxiety,
worry and panic in pulmonary hypertension

Influence.
Hope.
Integrity.

WEEK TWO: What keeps Anxiety Going?

Overcoming worry & anxiety

Without always knowing it, we all have thoughts and behaviours that can keep us stuck in our struggle with anxiety. In this booklet, we look at five of the most common traps that get in the way of us managing our anxiety more helpfully.

As you will see, these traps help to deal with anxiety in the short-term, however, they are not helpful in the longer-term and instead just keep the anxiety going.

The University Of Sheffield.

Avoidance

People can avoid situations in the hope of not experiencing any symptoms of anxiety. For example, people with PH may avoid doing activities that lead to uncomfortable physical sensations.

Such as, reducing the amount of exercise they do, walking upstairs or long distances, going out, having sex and eating certain foods. Individuals may also avoid people, places or situations that they feel may trigger their anxiety. This might include things like avoiding reading information on PH, going to places where they do not feel comfortable, writing a will, and not having certain conversations, such as those about funerals and death.

People with PH may also be avoidant of their symptoms. For example, they may avoid going to the doctors for a check-up or to ask about a cough that will not go away. Unfortunately, this means that they cannot find out if they have a health problem and they do not receive the correct treatment for their cough if it is needed. This leaves the person with the cough and also the worry about what the cough means.

In the short-term, avoidance can be very effective as it means you do not have to face situations that causes you anxiety, and this can give you a temporary sense of relief. However, in the long-term, avoidance keeps anxiety going or can make it even worse This is because by avoiding things that make you anxious, you miss out on the opportunity to develop more helpful skills to cope. You also become more likely to worry and feel anxious the next time you face a similar situation. And as you avoid activities, your world and the number of people in it will shrink, which may make you feel low and isolated.

It is important to gradually face your fears and build your confidence. Through repeatedly facing your fears and putting yourself in situations that you find difficult, you will find that it will cause you less anxiety over time.

The avoidance **merry-go-round**

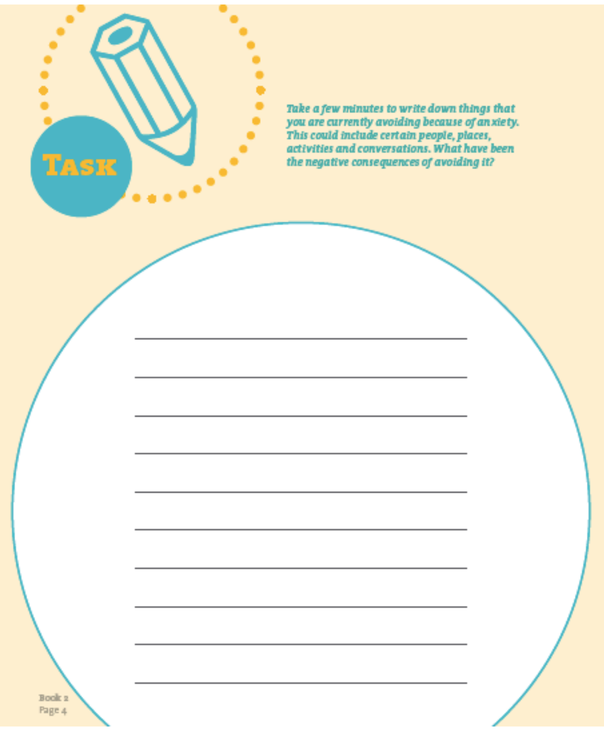
1 Situation that causes you anxiety

2 Avoidance or escape as a way to cope

3 *Short-term:* Relief as anxiety reduces

4 *Long-term:* Loss of confidence, reduced coping skills, not learning that anxiety can be tolerated and will eventually reduce, increased symptoms of anxiety, worry more, low mood

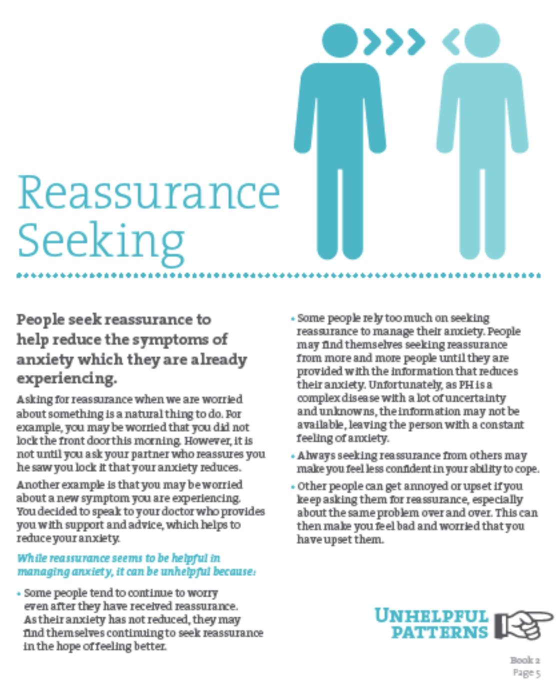
Book 2
Page 3



TASK

Take a few minutes to write down things that you are currently avoiding because of anxiety. This could include certain people, places, activities and conversations. What have been the negative consequences of avoiding it?

Book 2
Page 4



Reassurance Seeking

People seek reassurance to help reduce the symptoms of anxiety which they are already experiencing.

Asking for reassurance when we are worried about something is a natural thing to do. For example, you may be worried that you did not lock the front door this morning. However, it is not until you ask your partner who reassures you he saw you lock it that your anxiety reduces.

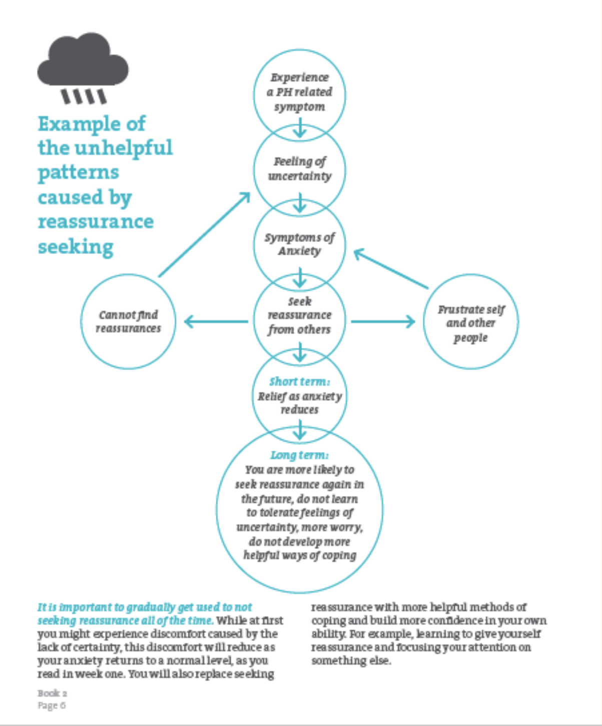
Another example is that you may be worried about a new symptom you are experiencing. You decided to speak to your doctor who provides you with support and advice, which helps to reduce your anxiety.

While reassurance seems to be helpful in managing anxiety, it can be unhelpful because:

- Some people tend to continue to worry even after they have received reassurance. As their anxiety has not reduced, they may find themselves continuing to seek reassurance in the hope of feeling better.
- Some people rely too much on seeking reassurance to manage their anxiety. People may find themselves seeking reassurance from more and more people until they are provided with the information that reduces their anxiety. Unfortunately, as PH is a complex disease with a lot of uncertainty and unknowns, the information may not be available, leaving the person with a constant feeling of anxiety.
- Always seeking reassurance from others may make you feel less confident in your ability to cope.
- Other people can get annoyed or upset if you keep asking them for reassurance, especially about the same problem over and over. This can then make you feel bad and worried that you have upset them.

UNHELPFUL PATTERNS

Book 2
Page 5



Example of the unhelpful patterns caused by reassurance seeking

It is important to gradually get used to not seeking reassurance all of the time. While at first you might experience discomfort caused by the lack of certainty, this discomfort will reduce as your anxiety returns to a normal level, as you read in week one. You will also replace seeking reassurance with more helpful methods of coping and build more confidence in your own ability. For example, learning to give yourself reassurance and focusing your attention on something else.

Book 2
Page 6

List any situations in which you recently sought reassurance. What were you looking for? What did you find? What were the negative consequences of seeking reassurance?

Book 2
Page 7

> *In my experience people often manage feelings of anxiety by developing lots of 'subtle' strategies which we tend to call 'safety behaviours'. Whilst these keep people 'safe' from the anxiety, they tend to prevent engagment in activities that will ultimately build confidence as shown in the 'cycle' opposite. So identifying and then revising safety behaviours is an important step in cognitive behavioural therapy.*
>
> **Prof Andrew Thompson**
> *Consultant Clinical Psychologist*
> *Cardiff University*

Book 2
Page 8

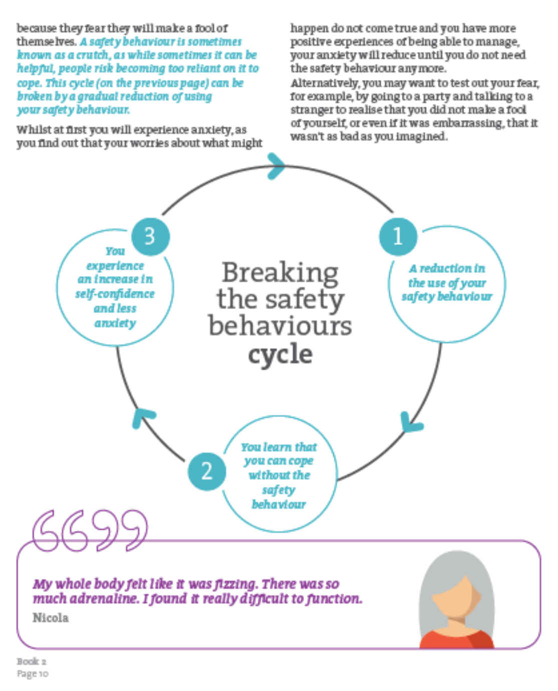
Safety Behaviours

Safety behaviours are things that we do that give us the feeling of safety.

For example, some people with PH are scared of getting an infection as it could make their symptoms worse. To help cope with this anxiety, when they leave the house, they must always have some hand sanitiser with them. Another example is some people with PH are anxious over having chest pains and so they must make sure they have a mobile phone or a personal alarm with them at all times, in case they need to contact someone for help.

Like avoidance, safety behaviours can help in the short-term as they reduce anxiety and provide you with a sense of control and relief. While these strategies may make sense - for example, having hand sanitiser to protect yourself from germs is sensible - the problem is that people often experience intense anxiety when they find themselves without their safety behaviour and can become unable to cope without it. In the long-term, people do not learn they can manage without the safety behaviour and gain confidence in their own ability to manage. Also, people may not learn what they are specifically worried about and so never learn to cope with it. For example, by never going to a party with friends, they may not realise that they are actually worried about talking to strangers

Book 2
Page 9



because they fear they will make a fool of themselves. *A safety behaviour is sometimes known as a crutch, as while sometimes it can be helpful, people risk becoming too reliant on it to cope. This cycle (on the previous page) can be broken by a gradual reduction of using your safety behaviour.*

Whilst at first you will experience anxiety, as you find out that your worries about what might happen do not come true and you have more positive experiences of being able to manage, your anxiety will reduce until you do not need the safety behaviour anymore.

Alternatively, you may want to test out your fear, for example, by going to a party and talking to a stranger to realise that you did not make a fool of yourself, or even if it was embarrassing, that it wasn't as bad as you imagined.

My whole body felt like it was fizzing. There was so much adrenaline. I found it really difficult to function.

Nicola

Book 2
Page 10

Write down any of your safety behaviours? You may not have realised a behaviour you do is a safety behaviour until now. How could you start to reduce your safety behaviours? What fear is your safety behaviour protecting you from?

Book 2
Page 11

Learning from others

Remember, you are not alone in your difficulties with PH. Here is a testimony from Eileen who has PH and also struggles with anxiety.

I have had Pulmonary Arterial Hypertension (PAH) since 2007. After an initial panic I decided I was going to live with the disease and not let it control me, and for around ten years that's what I did.

But as the drugs I was on stopped working so well I started experiencing more symptoms and that brought on anxiety. I stopped sleeping, fearing I wouldn't wake up. I felt I was a burden to my family and friends because of my moods and my lack of ability to do things.

About three years ago I realised I needed help, which was very hard to ask for as I pride myself on being the 'rock' in my family. It felt like a sign of weakness.

I bit the bullet and contacted my GP who thankfully was fantastic, and I was able to access counselling.

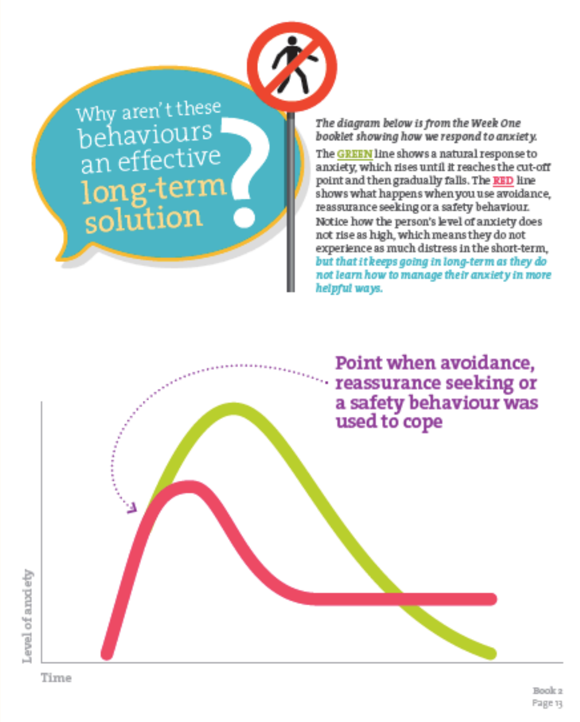
As time went on the fog started to lift and I decided to tell my family what was going on. They showed me so much love and support and it taught me that talking is a positive thing and I should have spoken to them sooner.

My illness took another turn last year and I ended up having a Hickman line fitted, which is where a small hollow tube is placed into my vein and is used for receiving treatment, and being listed for transplant. This caused me great distress and I recognised the deterioration in my mental health – I knew if I didn't get help again, I would be back on a downward spiral.

I accessed counselling again and I speak to my husband more about my thoughts now, which helps massively.

Eileen

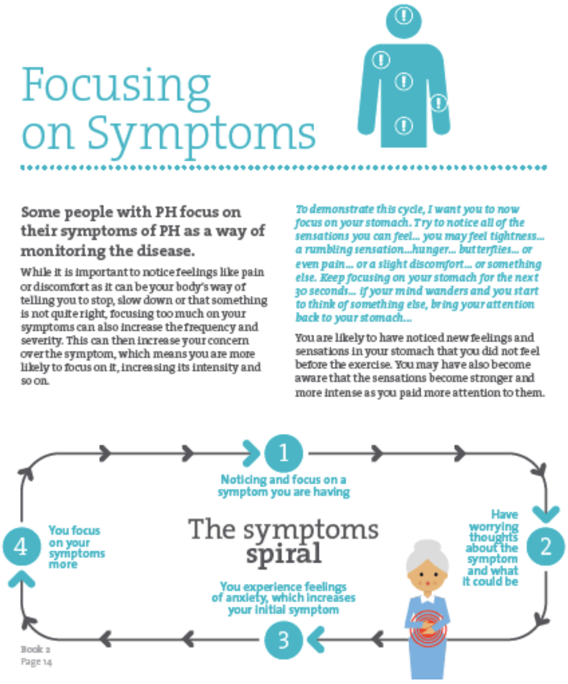
Book 2
Page 12



The diagram below is from the Week One booklet showing how we respond to anxiety.

The GREEN line shows a natural response to anxiety, which rises until it reaches the cut-off point and then gradually falls. The RED line shows what happens when you use avoidance, reassurance seeking or a safety behaviour. Notice how the person's level of anxiety does not rise as high, which means they do not experience as much distress in the short-term, *but that it keeps going in long-term as they do not learn how to manage their anxiety in more helpful ways.*

Book 2
Page 13



Focusing on Symptoms

Some people with PH focus on their symptoms of PH as a way of monitoring the disease.

While it is important to notice feelings like pain or discomfort as it can be your body's way of telling you to stop, slow down or that something is not quite right, focusing too much on your symptoms can also increase the frequency and severity. This can then increase your concern over the symptom, which means you are more likely to focus on it, increasing its intensity and so on.

To demonstrate this cycle, I want you to now focus on your stomach. Try to notice all of the sensations you can feel... you may feel tightness... a rumbling sensation...hunger... butterflies... or even pain... or a slight discomfort... or something else. Keep focusing on your stomach for the next 30 seconds... if your mind wanders and you start to think of something else, bring your attention back to your stomach...

You are likely to have noticed new feelings and sensations in your stomach that you did not feel before the exercise. You may have also become aware that the sensations become stronger and more intense as you paid more attention to them.

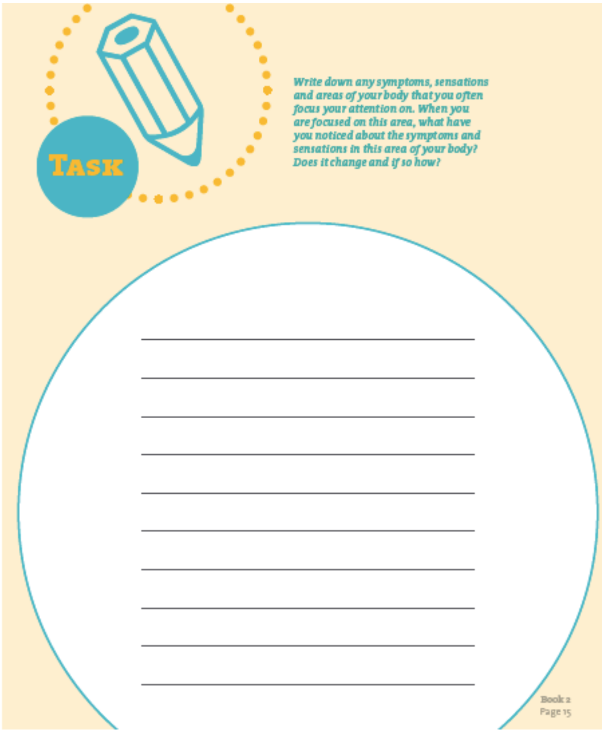
The symptoms spiral

1 Noticing and focus on a symptom you are having

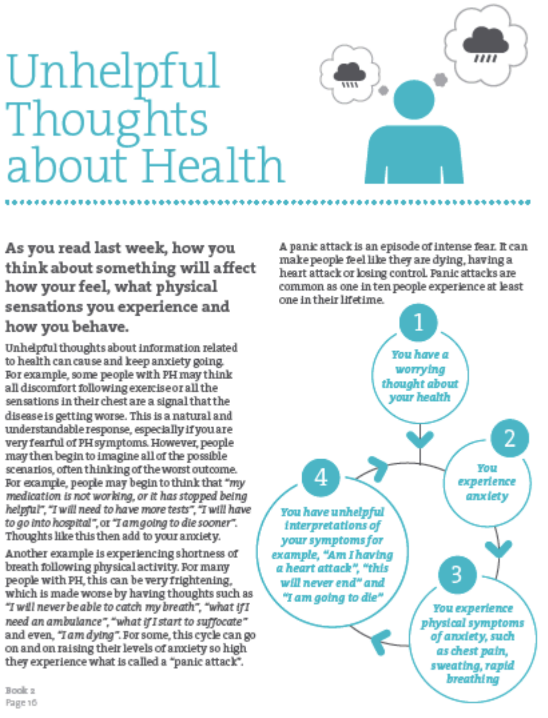
2 Have worrying thoughts about the symptom and what it could be

3 You experience feelings of anxiety, which increases your initial symptom

4 You focus on your symptoms more



Write down any symptoms, sensations and areas of your body that you often focus your attention on. When you are focused on this area, what have you noticed about the symptoms and sensations in this area of your body? Does it change and if so how?



Unhelpful Thoughts about Health

As you read last week, how you think about something will affect how your feel, what physical sensations you experience and how you behave.

Unhelpful thoughts about information related to health can cause and keep anxiety going. For example, some people with PH may think all discomfort following exercise or all the sensations in their chest are a signal that the disease is getting worse. This is a natural and understandable response, especially if you are very fearful of PH symptoms. However, people may then begin to imagine all of the possible scenarios, often thinking of the worst outcome. For example, people may begin to think that *"my medication is not working, or it has stopped being helpful"*, *"I will need to have more tests"*, *"I will have to go into hospital"*, or *"I am going to die sooner"*. Thoughts like this then add to your anxiety.

Another example is experiencing shortness of breath following physical activity. For many people with PH, this can be very frightening, which is made worse by having thoughts such as *"I will never be able to catch my breath"*, *"what if I need an ambulance"*, *"what if I start to suffocate"* and even, *"I am dying"*. For some, this cycle can go on and on raising their levels of anxiety so high they experience what is called a "panic attack".

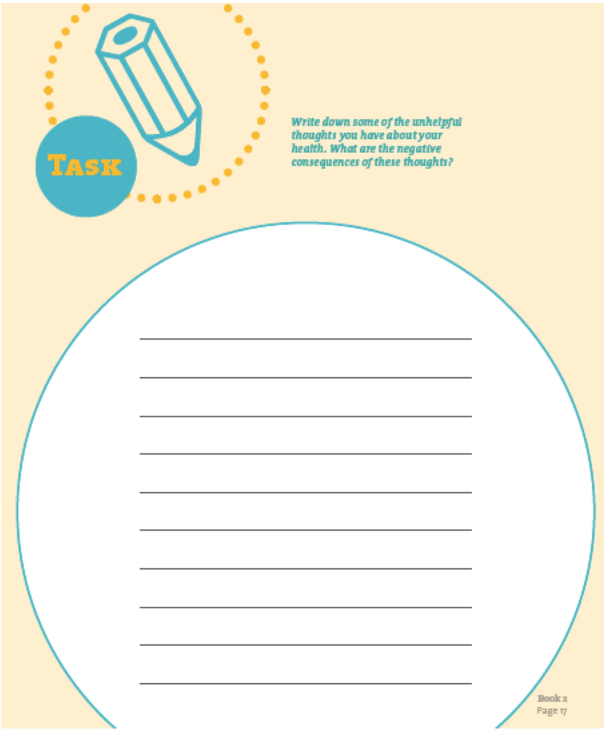
A panic attack is an episode of intense fear. It can make people feel like they are dying, having a heart attack or losing control. Panic attacks are common as one in ten people experience at least one in their lifetime.

1 *You have a worrying thought about your health*

2 *You experience anxiety*

3 *You experience physical symptoms of anxiety, such as chest pain, sweating, rapid breathing*

4 *You have unhelpful interpretations of your symptoms for example, "Am I having a heart attack", "this will never end" and "I am going to die"*



Write down some of the unhelpful thoughts you have about your health. What are the negative consequences of these thoughts?

How John's anxiety is maintained

John was told by his doctor that his test results showed a change. This was probably because John had a cold, he was going through a stressful time at home and he had recently changed his medication.

However, John went home and started to research his test results on the internet, looking for information that would provide an answer and reassurance *[Seeking Reassurance]*.

Reading about what his results could mean made him start to focus more on his bodily sensations. He began to experience his symptoms more intensely as well as new ones that he had not noticed before *[Focusing on Symptoms]*.

He began recording his symptoms in a notebook to help keep track of everything *[Safety Behaviour]*. As he did this, he began to have worried thoughts, such as *"I will probably need more invasive types of treatment"*, *"I need to stop doing the things I enjoy such as walking my dog" [Unhelpful Thoughts about Health]*.

John decided he was going to avoid his original doctor and ask to be seen by another expert in the hope of finding an answer *[Avoidance]*.

John then spent the next two weeks avoiding all forms of exercise and made sure he rested instead *[Safety Behaviour]*.

He continued to experience symptoms of anxiety until his next medical appointment.

Book 2
Page 18

How Nadia's anxiety is maintained

Triggers:
Nadia is watching a movie, which shows a hospital scene where someone is struggling to breathe, so they need to be put on a ventilator. Nadia starts to notice unusual sensations in her chest.

Thoughts:
I am struggling to breathe as well, I must take deeper breaths to get more oxygen, what if something is wrong with me, what if I end up on a ventilator too.

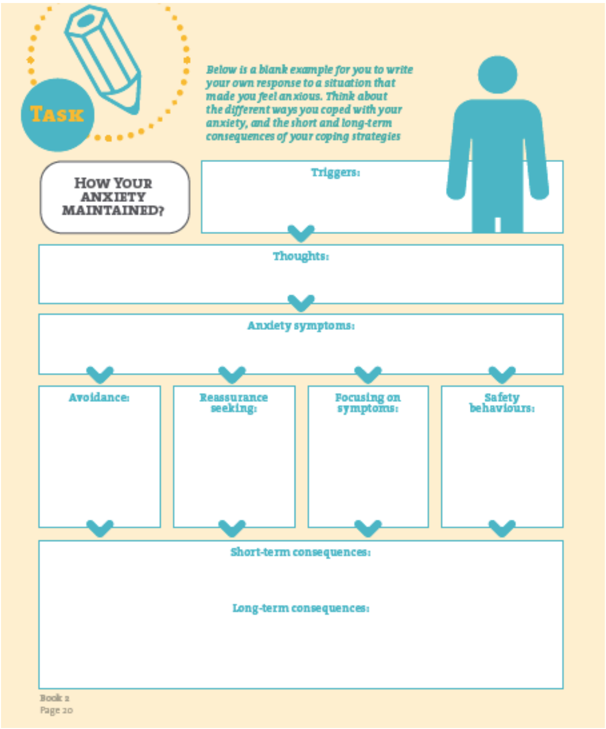
Anxiety symptoms:
Racing heart, dry mouth, feeling dizzy, feeling panicked, breathing rapidly, getting a surge of energy, tightness in chest.

Avoidance:	Reassurance seeking:	Focusing on symptoms:	Safety behaviours:
Stops watching medical movies.	Asks her parents if she will be fine, makes an appointment with a doctor and searches her symptoms online.	Pays close attention to her breathing, notices all changes in her body.	Nadia must read all about a movie before she will watch it to make sure there is no medical scenes.

Short-term consequences:
Symptoms of anxiety and PH reduce.

Long-term consequences:
Reduced self-esteem.
Feeling like it's another thing she cannot do.
Focusing on symptoms may make her symptoms worse.
Doctor and the internet cannot give complete reassurance.
Worrying increases as Nadia is concerned what triggered her shortness of breath.
Because avoidance, reassurance seeking, focusing on symptoms and safety behaviours helped reduced her anxiety, she is more likely to do them in the future meaning the vicious cycle keeps going.

Book 2
Page 19



Task

Below is a blank example for you to write your own response to a situation that made you feel anxious. Think about the different ways you coped with your anxiety, and the short and long-term consequences of your coping strategies

How your anxiety maintained?

Triggers:

Thoughts:

Anxiety symptoms:

Avoidance:	Reassurance seeking:	Focusing on symptoms:	Safety behaviours:

Short-term consequences:

Long-term consequences:

Book 2
Page 20

Summary

This week we learned that while anxiety is a normal response, it can be very unpleasant and frightening so it makes sense that we do our best to cope.

However, some of the ways we cope can cause difficulties for us in the long-term, as it keeps the problem going and us stuck in a vicious cycle. It is important to understand what is maintaining your anxiety, as it allows you to recognise where you can make changes to break your vicious cycles.

Activities such as relaxed breathing and others that you will learn about in this series will help you to develop more helpful ways of coping with anxiety.

Please remember, *Dr Gregg Rawlings* may be contacting you this week via email to see how you are getting on with this series. He will be asking you a few questions about your experience of the booklet so far.

Book 2
Page 21

TWO EXERCISES IN MINDFULNESS
FOR YOU TO PRACTICE OVER THE NEXT WEEK

As you will know by now, where you focus your attention can influence symptoms of anxiety. Rather than thinking about what happened in the past or worrying about future events, mindfulness is an exercise that helps you to stay in the present moment.

Here are two mindfulness exercises for you to practice over the next week. Please take your time going through the exercises, taking long pauses between each sentence. You could ask a friend or a family member to read the instructions out to you so you can focus on the exercise fully, if this is something you would feel comfortable with.

EXERCISE ONE
FOCUSING ON YOUR BODY

Get yourself into a comfortable position as you read this. Ideally, sitting upright in a chair with your feet flat on the floor, and your back feeling supported by the back of the chair.

Take a few minutes to focus on your breathing. Feel the rhythm of your breathing as you fill and empty your lungs with air, like the waves on the beach coming in before going back out... Focus on your inhale and exhale noticing the changes in the feelings of your body... Focus your awareness on the physical sensations in your body... Focus on the feelings of pressure where your body comes into contact with the chair and floor... If your mind wanders, just acknowledge your thought without judgment before bringing your attention back to your breathing...

Begin to change your thoughts to allowing yourself to be sitting here focusing on your breathing. There is no particular goal you need to achieve in this moment. Simply just allow the experience to be without needing it to be anything more than what it is. Do not try to control your breathing, just let your body breathe naturally. If you mind wanders away from your breath to worries, thoughts, concerns or mental images, it is okay. It is normal. When it happens, just notice that your mind has wandered and where it has wandered to. You may want to acknowledge the feeling or sensation you experienced as your mind wandered. Then gently guide your attention back to your breathing. Be kind and compassionate to your mind as it wanders.

When you become aware of any discomfort, tension or other physical sensations, just notice them, acknowledge them and see if you can make space for them. Do not try to control them or make them go away. See if you can make room for them, just allowing them to be there. Notice the sensations change from each passing moment. Sometimes the sensations may get stronger, while other times, weaker and sometimes they just stay the same. Just notice the change and allow them to be, just as they are. Breathe calmly into and out from the sensations of discomfort, gently guiding yourbreath towards where the sensation is in your body.

The goal of this is not to make you feel better, but to get better at feeling.

If you notice you are unable to focus on your breathing, because of an intense physical sensation or intense emotion, let go of the focus on your breath and move your attention to the place of the physical discomfort, the exact place where you experience the feeling the most strongly.

Continue overleaf... ☞

EXERCISE ONE
CONTINUED

Gently move your attention to the discomfort and stay with it no matter how bad it seems. Acknowledge the sensation... take a look at it.... what does it really feel like? Is it hot or cold? Does it feel sharp or smooth? Does it feel heavy or light? While doing this see if you can make room for the discomfort, allowing it to be there and be willing to stay with it.

You may also notice thoughts about the sensations. You may notice that you're judging or coming up with ideas about what the sensation means. You may think it is something dangerous. You may start to think about what will happen next. When you notice these thoughts, just acknowledge the thought before bringing your attention back...", not as your mind says it is, but just noticing your thoughts as thoughts, feelings as feelings, sensations as sensations, nothing more, nothing less. If you notice that you're asking yourself questions, just gently answer them with "I don't know" and return to your present experience, just as it is.

To help you to distance yourself from your thoughts and feelings, you may want to start to label them as you begin to notice them. For example, if you notice yourself starting to worry, say to yourself out loud or silently, "worry, that is a worry". Notice the worry without engaging with it, just allowing it to stay by making room for it. If you find yourself judging, just notice that and label it, "judging, there is judging". Observe these thoughts with kindness and compassion. You can do this with other thoughts and feelings, just naming them, such as "there is planning", "there is remembering", "there is wishing" or any another label you may notice. Label your emotions and thoughts and move on without judgment. Notice how your thoughts and feelings come and go in your mind and body. You are not what those thoughts and feelings say, no matter how intense or loud they seem.

Spend the next few minutes practicing this exercise. When you are ready to end the exercise, gradually widen your attention, taking in the noises, smells and sights around you. Allow the awareness of the present moment to stay with you throughout the day.

EXERCISE TWO
MINDFULNESS IN EVERYDAY LIFE

This exercise is designed to help you be more aware and appreciate simple and everyday tasks in your life.

Rather than being on autopilot and giving yourself time to worry, the idea is to focus your attention on the activity you are doing. Think of an activity that happens every day more than once. Something that you take for granted, like doing the washing up.

At the very moment you submerge your hands in the hot soapy water, stop for a moment and be mindful of where you are and how you are feeling in that moment. Bring you attention to the temperature of the water on your hands. Notice the sensations and smell of the soap and bubbles. Pick up the dirty dishes and gently place them in the water. Watch as the dirt is washed and scrubbed away leaving a clean smooth surface. Lift the dish out of the water noticing the water falling off it and splashing back down into the sink. If you notice your mind has wandered, just acknowledge the thought before brining your attention back to the feeling of water on your hands.

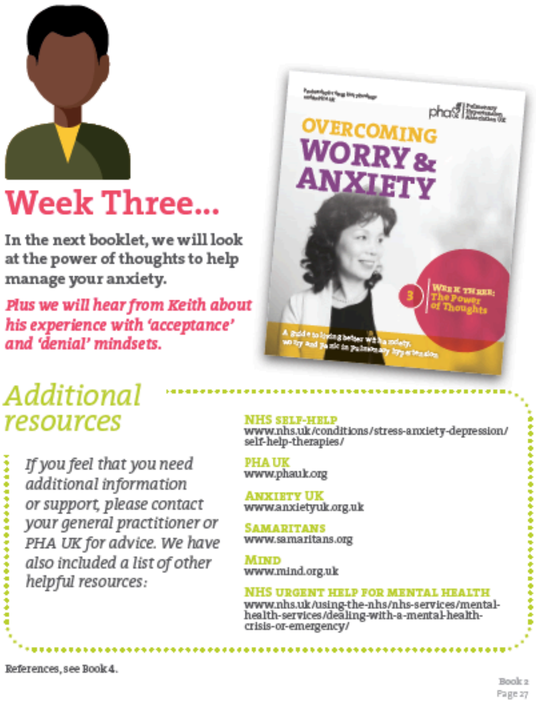
When people wash their dishes mindfully, as described above, they reported an improvement in how nervous they felt, compared to people who just washed the dishes normally.

These mindfulness exercises can be done for a range of everyday activities such as hoovering, sweeping, eating, showering and doing the laundry. A good activity to do, but very challenging for those of us with a sweet tooth, is try being mindful while eating a piece of chocolate. Just noticing the texture and taste of it on your tongue before taking a bite.

Try to practice at least one mindfulness exercise each day over the next week.

> *It's not a case mind over matter. The mind does matter, and mental health matters just as much as physical health.*
>
> **Rachel Crackett,**
> *Clinical Nurse Specialist, Northern Pulmonary Vascular Unit, Freeman Hospital*

Book 2
Page 26



Week Three...

In the next booklet, we will look at the power of thoughts to help manage your anxiety.

Plus we will hear from Keith about his experience with 'acceptance' and 'denial' mindsets.

Additional resources

If you feel that you need additional information or support, please contact your general practitioner or PHA UK for advice. We have also included a list of other helpful resources:

NHS SELF-HELP
www.nhs.uk/conditions/stress-anxiety-depression/self-help-therapies/

PHA UK
www.phauk.org

ANXIETY UK
www.anxietyuk.org.uk

SAMARITANS
www.samaritans.org

MIND
www.mind.org.uk

NHS URGENT HELP FOR MENTAL HEALTH
www.nhs.uk/using-the-nhs/nhs-services/mental-health-services/dealing-with-a-mental-health-crisis-or-emergency/

References, see Book 4.

Book 2
Page 27

> ***It is vital to speak up if you are struggling with anxiety.***
>
> **Rachel Crackett, Clinical Nurse Specialist in PH**

To find out more about the PHA UK, visit **www.phauk.org**

Pulmonary Hypertension Association UK
PHA UK Resource Centre, Unit 1, Newton Business Centre, Newton Chambers Road,
Thorncliffe Park, Chapeltown, Sheffield, England S35 2PH
T: 01709 761450 E: office@phauk.org @PULHAUK @PHA_UK @PHA ORG UK

Week Three:

Produced by Dr Gregg Harry Rawlings
and the PHA UK

pha UK | Pulmonary Hypertension Association UK

OVERCOMING
WORRY &
ANXIETY

3 | WEEK THREE:
The Power
of Thoughts

A guide to living better with anxiety,
worry and panic in pulmonary hypertension

pha UK | Influence. Hope. Integrity.

WEEK THREE:
The Power
of Thoughts

Overcoming
worry & anxiety

As you will know by now, when people are feeling anxious, they often experience thoughts that are unhelpful. These thoughts are also known as unhelpful thinking patterns. It is important to remember that thoughts are not facts and they are not always true. For example, you may think that you made a fool of yourself on a romantic date, and it is not until you speak to your date later on who explains he had a lovely time and would like to go out with you again that you stop worrying.

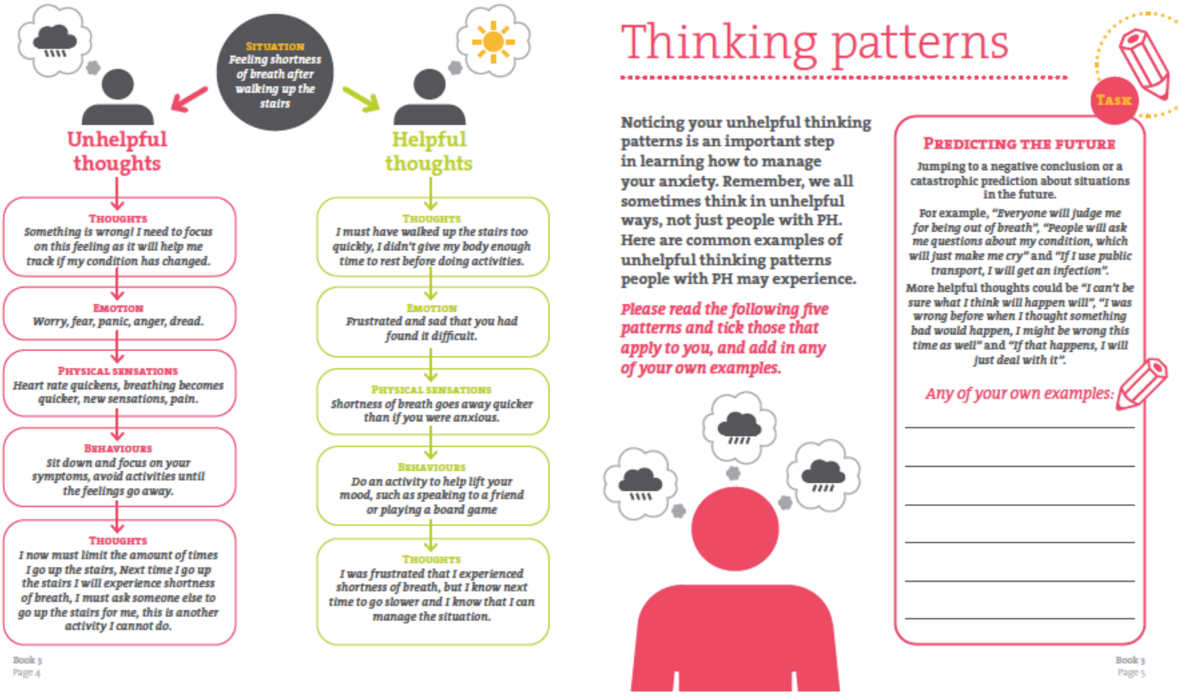
Unhelpful thinking patterns can fuel anxiety causing people to get stuck a vicious cycle. As you can see on the next page, the thoughts we have about an event can make a big difference in how we experience something and the impact it has on us.

The University Of Sheffield.
Cardiff University Prifysgol Caerdydd

A diagnosis of PH often goes hand-in-hand with feelings of anxiety. If you are struggling, you are not on your own.

Iain Armstrong,
Chair, the PHA UK

Book 3
Page 3



SITUATION
Feeling shortness of breath after walking up the stairs

Unhelpful thoughts

THOUGHTS
Something is wrong! I need to focus on this feeling as it will help me track if my condition has changed.

EMOTION
Worry, fear, panic, anger, dread.

PHYSICAL SENSATIONS
Heart rate quickens, breathing becomes quicker, new sensations, pain.

BEHAVIOURS
Sit down and focus on your symptoms, avoid activities until the feelings go away.

THOUGHTS
I now must limit the amount of times I go up the stairs, Next time I go up the stairs I will experience shortness of breath, I must ask someone else to go up the stairs for me, this is another activity I cannot do.

Helpful thoughts

THOUGHTS
I must have walked up the stairs too quickly, I didn't give my body enough time to rest before doing activities.

EMOTION
Frustrated and sad that you had found it difficult.

PHYSICAL SENSATIONS
Shortness of breath goes away quicker than if you were anxious.

BEHAVIOURS
Do an activity to help lift your mood, such as speaking to a friend or playing a board game

THOUGHTS
I was frustrated that I experienced shortness of breath, but I know next time to go slower and I know that I can manage the situation.

Book 3
Page 4

Thinking patterns

Noticing your unhelpful thinking patterns is an important step in learning how to manage your anxiety. Remember, we all sometimes think in unhelpful ways, not just people with PH. Here are common examples of unhelpful thinking patterns people with PH may experience.

Please read the following five patterns and tick those that apply to you, and add in any of your own examples.

TASK

PREDICTING THE FUTURE

Jumping to a negative conclusion or a catastrophic prediction about situations in the future.

For example, *"Everyone will judge me for being out of breath"*, *"People will ask me questions about my condition, which will just make me cry"* and *"If I use public transport, I will get an infection"*.

More helpful thoughts could be *"I can't be sure what I think will happen will"*, *"I was wrong before when I thought something bad would happen, I might be wrong this time as well"* and *"If that happens, I will just deal with it"*.

Any of your own examples:

Book 3
Page 5

TASK

MIND READING

Worrying about and guessing what other people are thinking.
For example, *"That person is thinking my swollen ankles look funny"* and *"I bet my friends think I am a burden now because we cannot do the things we used to"*.
More helpful thoughts could be *"I don't actually know what they are thinking"* and *"They will have their own things to worry about rather than judging me"*.

Any of your own examples:

OVER-GENERALISATION

Viewing a single negative event as affecting everything.
For example, *"My bus didn't arrive on time, which means nothing else will go right today"* or *"I struggled to catch my breath in the supermarket, which means it will always happen now when I go out"*.
More helpful thoughts could be *"The bus is always running late but I am in no rush, people will understand if I am little late"* and *"It doesn't really mean anything that I had difficulty, it doesn't mean it will definitely happen again but even if it does, I will manage it like I did before"*.

Any of your own examples:

Book 3
Page 6

ALL OR NOTHING THINKING

Only seeing the extremes and not the grey areas in between. For example, *"If I cannot walk up the stairs every day without being out of breath it means I have failed"* and *"I feel guilty that my partner has to do everything for me"*. More helpful thoughts could be *"I may struggle with stairs now and again but at least I am able to do it"* and *"Although I cannot do some things, I am still able to do a lot and I now have more time to support my partner in other ways"*.

Any of your own examples:

People also use all or nothing thinking to judge the behaviour of others. For example, *"My partner should know I cannot do that, he is always getting things wrong"* and *"My doctor said my tests were unclear, she is useless"*. More helpful thoughts could be, *"My partner gets most things right and he didn't mean to upset me"* and *"My doctor has arranged for more tests and explained she will speak to her colleagues about my symptoms"*.

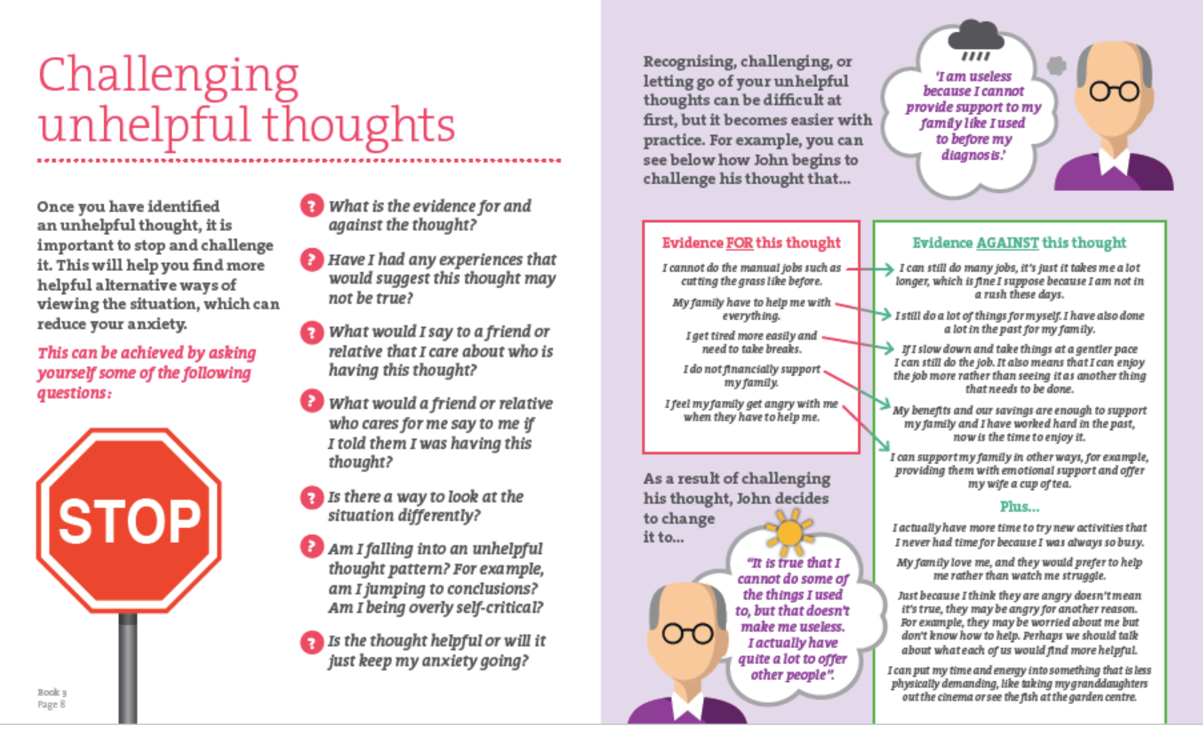
Any of your own examples:

SELF-CRITICISM

Calling yourself critical names or words. People can fall into patterns of self-bullying and having negative thoughts about themselves. For example, *"I am useless"*, *"I am boring"* or *"Why me, I used to be able to do all those things and now I can't do anything"*. More helpful thoughts could be *"Everyone has something different about them"*, *"No one is perfect"* and *"I am putting myself down"*.

Any of your own examples:

Book 3
Page 7



Challenging unhelpful thoughts

Once you have identified an unhelpful thought, it is important to stop and challenge it. This will help you find more helpful alternative ways of viewing the situation, which can reduce your anxiety.

This can be achieved by asking yourself some of the following questions:

- ? *What is the evidence for and against the thought?*
- ? *Have I had any experiences that would suggest this thought may not be true?*
- ? *What would I say to a friend or relative that I care about who is having this thought?*
- ? *What would a friend or relative who cares for me say to me if I told them I was having this thought?*
- ? *Is there a way to look at the situation differently?*
- ? *Am I falling into an unhelpful thought pattern? For example, am I jumping to conclusions? Am I being overly self-critical?*
- ? *Is the thought helpful or will it just keep my anxiety going?*

Book 3
Page 8

Recognising, challenging, or letting go of your unhelpful thoughts can be difficult at first, but it becomes easier with practice. For example, you can see below how John begins to challenge his thought that...

'I am useless because I cannot provide support to my family like I used to before my diagnosis.'

Evidence FOR this thought

I cannot do the manual jobs such as cutting the grass like before.

My family have to help me with everything.

I get tired more easily and need to take breaks.

I do not financially support my family.

I feel my family get angry with me when they have to help me.

Evidence AGAINST this thought

I can still do many jobs, it's just it takes me a lot longer, which is fine I suppose because I am not in a rush these days.

I still do a lot of things for myself. I have also done a lot in the past for my family.

If I slow down and take things at a gentler pace I can still do the job. It also means that I can enjoy the job more rather than seeing it as another thing that needs to be done.

My benefits and our savings are enough to support my family and I have worked hard in the past, now is the time to enjoy it.

I can support my family in other ways, for example, providing them with emotional support and offer my wife a cup of tea.

Plus...

I actually have more time to try new activities that I never had time for because I was always so busy.

My family love me, and they would prefer to help me rather than watch me struggle.

Just because I think they are angry doesn't mean it's true, they may be angry for another reason. For example, they may be worried about me but don't know how to help. Perhaps we should talk about what each of us would find more helpful.

I can put my time and energy into something that is less physically demanding, like taking my granddaughters out the cinema or see the fish at the garden centre.

As a result of challenging his thought, John decides to change it to...

"It is true that I cannot do some of the things I used to, but that doesn't make me useless. I actually have quite a lot to offer other people".

Task

Using the questions on the previous spread, begin to challenge some of the unhelpful thoughts you identified and try to come up with more helpful thoughts. *Make sure to think of responses that you believe in and are more compassionate alternatives.*

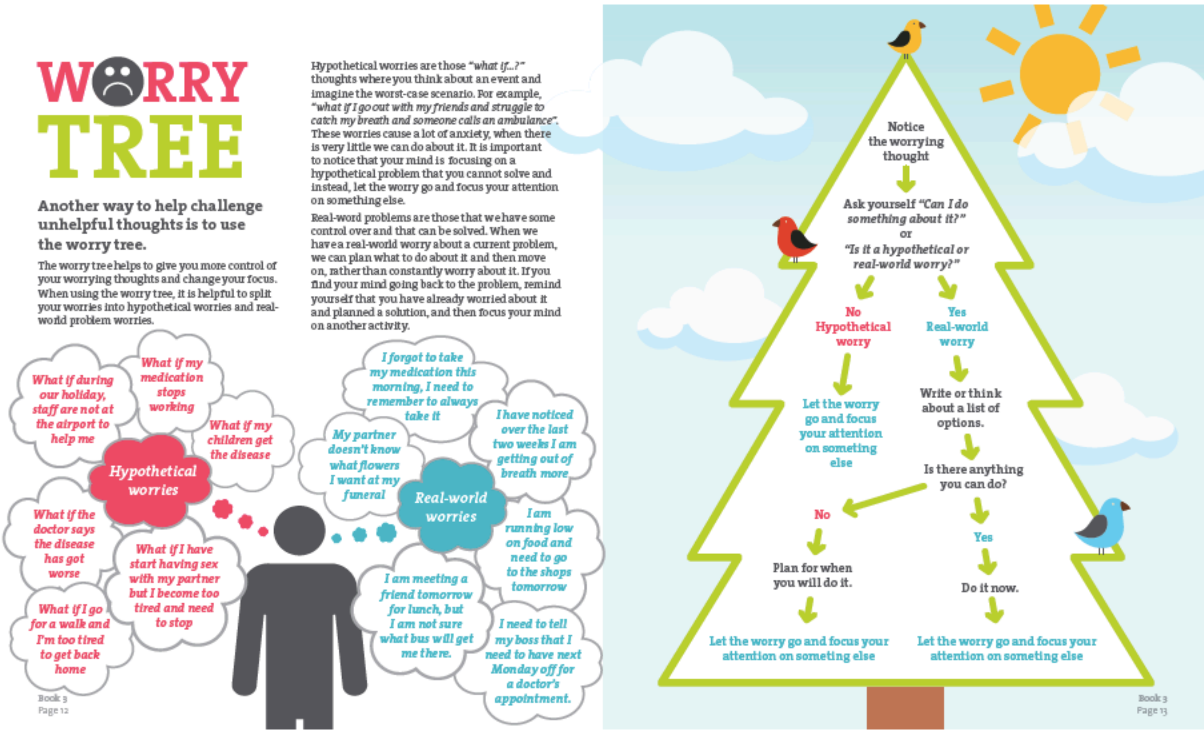
Book 3
Page 10

Learning from others

Remember, you are not alone in your difficulties with PH. Here is a testimony from Keith who has PH and also struggles with anxiety.

> *I was diagnosed with Pulmonary Arterial Hypertension (PAH) in 2015 and my problems with anxiety began about a year later when I was trying to get back to employment.*
>
> *I swing daily between acceptance and denial of having the disease. I frequently procrastinate between planning for the future when in the 'acceptance' mindset and then ripping the plans up when I flip to the 'denial' mindset.*
>
> *I am restless, my breathing is shorter and faster, and I have hot flushes and spikes in temperature. I have a continuous sense of dread; I can't concentrate, and I have lost confidence.*
>
> *I am anxious about nearing the average lifespan for a person with PAH. I fear the inevitably of declining to a point where I will have to go back onto oxygen and / or intravenous therapy.*
>
> *The faster and shorter breathing, hot flushes and spikes in temperature themselves bring an increase in anxiety as they can be symptoms of PH and underlying conditions. So, at times, I can't be sure if what I'm experiencing is anxiety or a flare-up of my physical conditions. That brings anxiety in itself!*
>
> *I have had several prolonged periods of times when anxiety has had a massive detrimental impact on my day-to-day life, affecting relationships and my work. At other times, my anxiety can be more subtle and manageable - and can actually have a positive impact if I'm able to control and channel the symptoms.*
>
> *As well as medication and counselling, I've found breathing techniques, understanding mindfulness, gardening, walking and maintaining social contact with friends and family all helpful in easing the symptoms of my anxiety.*
>
> **Keith**

Book 3
Page 11



WORRY TREE

Another way to help challenge unhelpful thoughts is to use the worry tree.

The worry tree helps to give you more control of your worrying thoughts and change your focus. When using the worry tree, it is helpful to split your worries into hypothetical worries and real-world problem worries.

Hypothetical worries are those *"what if...?"* thoughts where you think about an event and imagine the worst-case scenario. For example, *"what if I go out with my friends and struggle to catch my breath and someone calls an ambulance"*. These worries cause a lot of anxiety, when there is very little we can do about it. It is important to notice that your mind is focusing on a hypothetical problem that you cannot solve and instead, let the worry go and focus your attention on something else.

Real-word problems are those that we have some control over and that can be solved. When we have a real-world worry about a current problem, we can plan what to do about it and then move on, rather than constantly worry about it. If you find your mind going back to the problem, remind yourself that you have already worried about it and planned a solution, and then focus your mind on another activity.

Hypothetical worries

What if during our holiday, staff are not at the airport to help me

What if my medication stops working

What if my children get the disease

What if the doctor says the disease has got worse

What if I have start having sex with my partner but I become too tired and need to stop

What if I go for a walk and I'm too tired to get back home

Real-world worries

I forgot to take my medication this morning, I need to remember to always take it

My partner doesn't know what flowers I want at my funeral

I have noticed over the last two weeks I am getting out of breath more

I am running low on food and need to go to the shops tomorrow

I am meeting a friend tomorrow for lunch, but I am not sure what bus will get me there.

I need to tell my boss that I need to have next Monday off for a doctor's appointment.

Book 3
Page 12

Notice the worrying thought

Ask yourself *"Can I do something about it?"* or *"Is it a hypothetical or real-world worry?"*

No Hypothetical worry → Let the worry go and focus your attention on someting else

Yes Real-world worry → Write or think about a list of options. → Is there anything you can do?

No → Plan for when you will do it. → Let the worry go and focus your attention on someting else

Yes → Do it now. → Let the worry go and focus your attention on someting else

Book 3
Page 13

Worry delay

Worry and unhelpful thinking patterns can make it difficult to think about anything else. You may also feel like you have to engage in your worry right now, right in this moment. With practice, many people have found it useful to delay their worry.

This means setting some time aside each day to let yourself worry. For example, 15 minutes at 7pm each day. Whenever you being to worry about something or notice that you are worrying outside of this time, say to yourself, *"it is not my time to worry about that now"*.

Alternatively, use the worry tree to help decide whether it is something you need to manage now (a real-world worry) or if the worry can be delayed (hypothetical worry). Some people find it useful to write down their worries throughout the day and put it in a jar, until it is their time to worry.

What you will find is that some of the worries may have already sorted themselves out and those that have not may not feel as worrying anymore, as you are in a different state of mind to think about them (if you even want to think about them at all).

This exercise also allows you to expose yourself to your fears and anxious thoughts. Many people try not to think about their distressing thoughts and instead, pushes them away. However, as this is a form of avoidance, not thinking about your anxious thoughts will keep your anxiety going. By exposing yourself to your anxious thoughts, overtime they will become easier to cope with.

Worry delay can help you spend the rest of your day worry-free until your worry time.

Book 3
Page 14

Summary

In this chapter you have read about the impact your thoughts can have on how you feel and behave.

You identified some of your own unhelpful thinking patterns and started to challenge them using a range of different approaches.
With practice, this will become easier and it will help you break out of your vicious cycles.

As the diagram below shows, while the situation triggered an unhelpful thought, it can be challenged to something more positive, stopping it in its tracks from having a negative impact on your emotions, physical sensations and behaviours.

THOUGHT CHALLENGE: *I am trying to predict the future, whenever I am at my son's house no one cares about the oxygen cylinder, my family love me and want me there, he is my son and I deserve to be at his party to celebrate, If I avoid the party I will only be maintaining my anxiety.*

SITUATION: *Being invited to a restaurant for my son's birthday.*

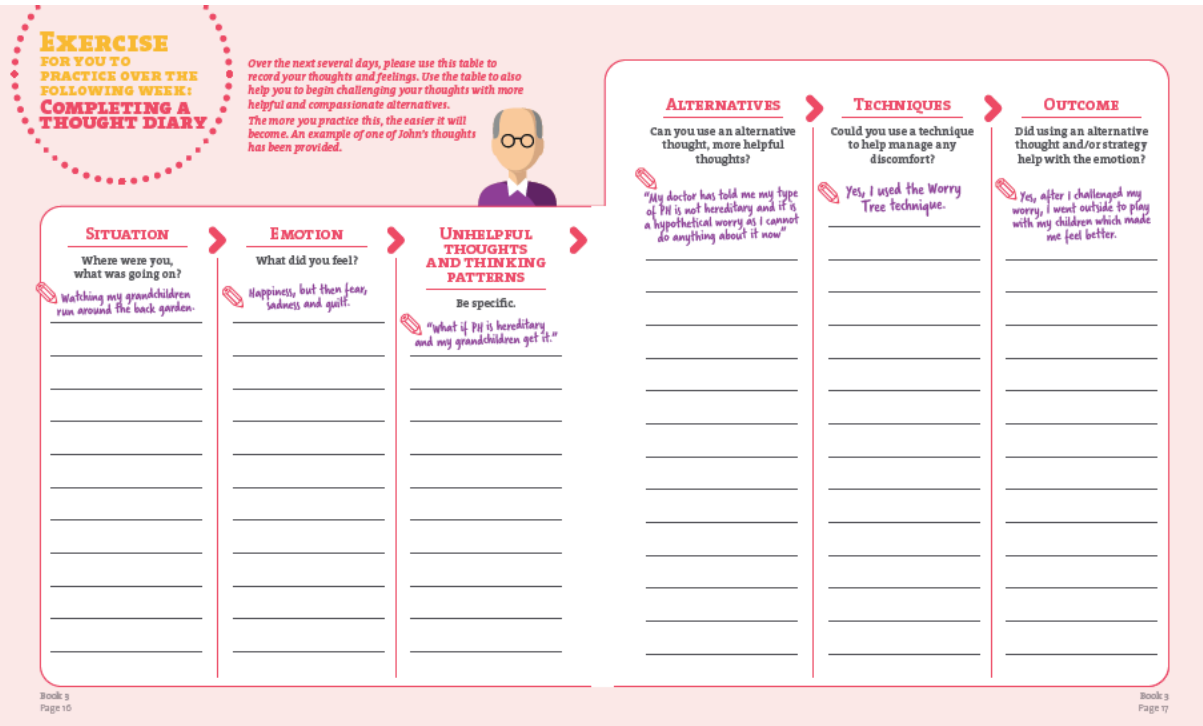
THOUGHT: *I will embarrass myself and make other people feel uncomfortable because I will need to take my oxygen cylinder.*

EMOTION: *Shame, embarrassment, fear, sadness, frustration.*

BEHAVIOUR: *Repeatedly check with my son that it will be okay, get in the taxi to go but then don't feel very well so I go back home.*

PHYSICAL SENSATION: *Churning stomach, tense muscles, experiencing more pain.*

Book 3
Page 15



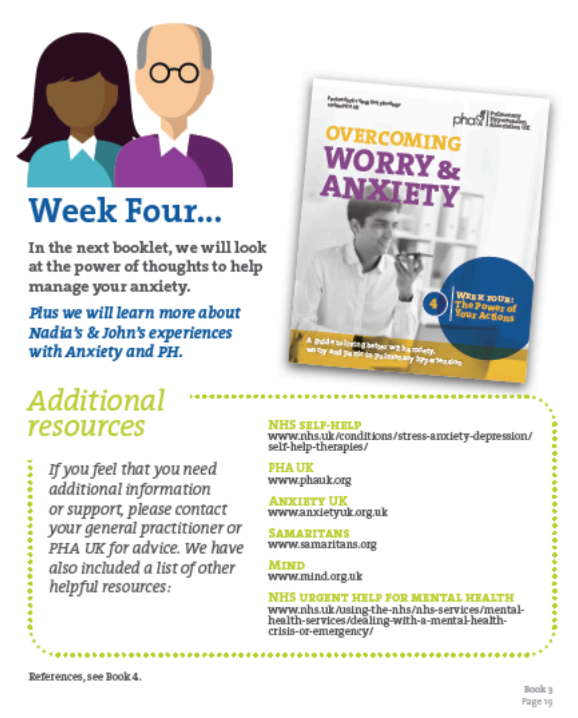
EXERCISE FOR YOU TO PRACTICE OVER THE FOLLOWING WEEK: COMPLETING A THOUGHT DIARY

Over the next several days, please use this table to record your thoughts and feelings. Use the table to also help you to begin challenging your thoughts with more helpful and compassionate alternatives.

The more you practice this, the easier it will become. An example of one of John's thoughts has been provided.

SITUATION	EMOTION	UNHELPFUL THOUGHTS AND THINKING PATTERNS	ALTERNATIVES	TECHNIQUES	OUTCOME
Where were you, what was going on?	What did you feel?	Be specific.	Can you use an alternative thought, more helpful thoughts?	Could you use a technique to help manage any discomfort?	Did using an alternative thought and/or strategy help with the emotion?
Watching my grandchildren run around the back garden.	Happiness, but then fear, sadness and guilt.	"What if PH is hereditary and my grandchildren get it."	"My doctor has told me my type of PH is not hereditary and it is a hypothetical worry as I cannot do anything about it now"	Yes, I used the Worry Tree technique.	Yes, after I challenged my worry, I went outside to play with my children which made me feel better.

> *Good mental well-being is important in every aspect of life. It effects the way we think and feel. Anxiety, if left untreated, can cause untold symptoms and can greatly decrease the ability to cope with your disease.*
>
> **Karon Carson,**
> *Pulmonary Hypertension Nurse Specialist,*
> *Scottish Pulmonary Vascular Unit*



Week Four...

In the next booklet, we will look at the power of thoughts to help manage your anxiety.

Plus we will learn more about Nadia's & John's experiences with Anxiety and PH.

Additional resources

If you feel that you need additional information or support, please contact your general practitioner or PHA UK for advice. We have also included a list of other helpful resources:

NHS SELF-HELP
www.nhs.uk/conditions/stress-anxiety-depression/self-help-therapies/

PHA UK
www.phauk.org

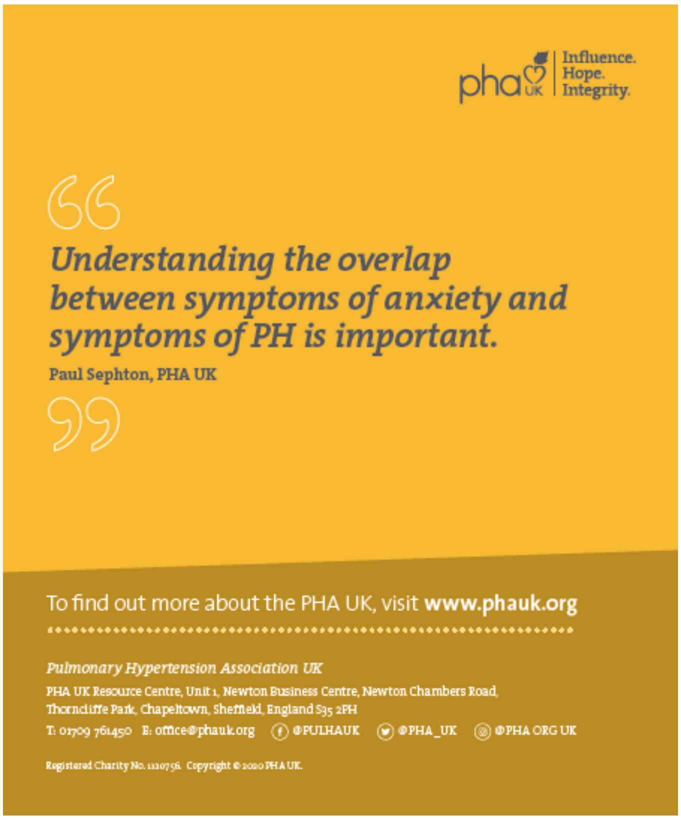
ANXIETY UK
www.anxietyuk.org.uk

SAMARITANS
www.samaritans.org

MIND
www.mind.org.uk

NHS URGENT HELP FOR MENTAL HEALTH
www.nhs.uk/using-the-nhs/nhs-services/mental-health-services/dealing-with-a-mental-health-crisis-or-emergency/

References, see Book 4.



Influence.
Hope.
Integrity.

Understanding the overlap between symptoms of anxiety and symptoms of PH is important.

Paul Sephton, PHA UK

To find out more about the PHA UK, visit **www.phauk.org**

Pulmonary Hypertension Association UK
PHA UK Resource Centre, Unit 1, Newton Business Centre, Newton Chambers Road,
Thorncliffe Park, Chapeltown, Sheffield, England S35 2PH
T: 01709 761450 E: office@phauk.org @PULHAUK @PHA_UK @PHA ORG UK

Registered Charity No. 1120756. Copyright © 2020 PHA UK.

Week Four:

Produced by Dr Gregg Harry Rawlings
and the PHA UK

Pulmonary
Hypertension
Association UK

OVERCOMING
WORRY &
ANXIETY

4 WEEK FOUR:
The Power of
Your Actions

A guide to living better with anxiety,
worry and panic in pulmonary hypertension

pha UK | Influence. Hope. Integrity.

Week four: The Power of Your Actions

Overcoming worry & anxiety

As you read in the previous weeks, certain things that people do to help them cope, such as avoiding situations that feel frightening, can help reduce anxiety in the short-term but can lead to more difficulties in the long-term because the person will:

- *Never get used to the situation and so will find it more difficult to overcome their worries.*
- *Miss out on activities that they may really want to do.*
- *Lose confidence in themselves and their ability.*
- *Never get a chance to learn how to cope with the situation in a more helpful way.*
- *Do not find out how the event compares to what they imagined it will be.*
- *Find the situation more difficult the next time.*
- *Experience a reduction in mood, making them more likely to have unhelpful thoughts that are more difficult to challenge.*
- *Be more likely to use avoidance as a coping technique to manage other difficult situations.*

The University Of Sheffield. Cardiff University Prifysgol Caerdydd

The best way to overcome a situation that triggers anxiety is to face it.

To achieve this, it is useful to break the situation up into smaller, more manageable tasks.

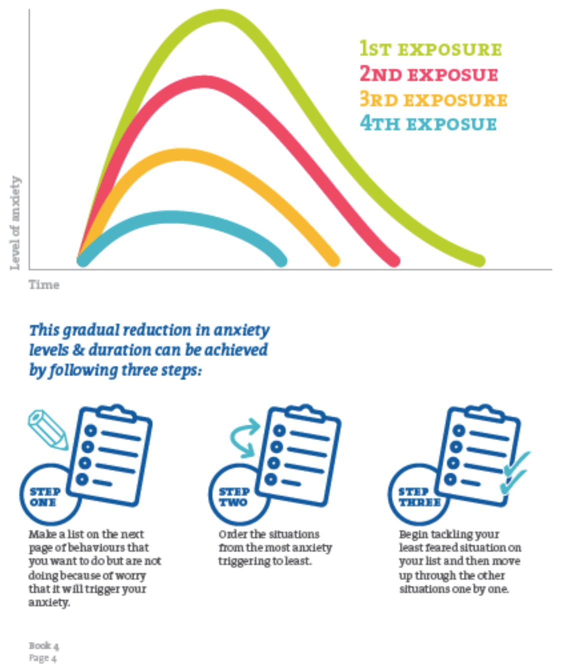
Although at first this is likely to make you feel more anxious, remember that as you learnt in the first week, if you stick with your anxiety, it will gradually reduce naturally, and you will feel more comfortable. Once you have mastered the smaller task and you can manage the anxiety it causes you more easily, you can move on to the next task until you have achieved them all.

The diagram below is from Week One. As you can remember, the green line shows a natural response to anxiety, which rises until it reaches the cut-off point and then gradually falls. As you can see from the red, yellow and blue lines, the more times you expose yourself to the experience, with time your brain and body gets used to the event and you feel less anxiety.

To help make this clearer, think about the first time you started a new job. On your first day you may have felt very anxious and worried, but with each passing day you felt more confident and less anxiety, until you did not experience any anxiety about going.

See overleaf how that looks... ☞

Book 4
Page 3



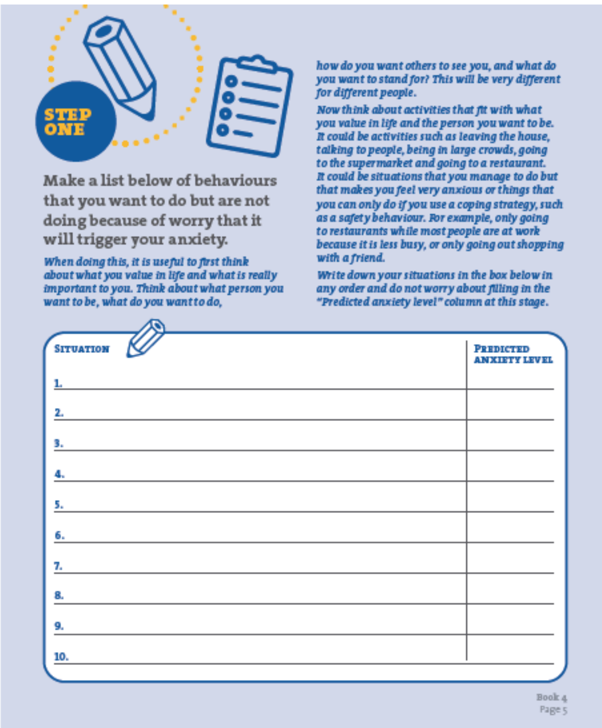
This gradual reduction in anxiety levels & duration can be achieved by following three steps:

Step one Make a list on the next page of behaviours that you want to do but are not doing because of worry that it will trigger your anxiety.

Step two Order the situations from the most anxiety triggering to least.

Step three Begin tackling your least feared situation on your list and then move up through the other situations one by one.

Book 4
Page 4



Step one

Make a list below of behaviours that you want to do but are not doing because of worry that it will trigger your anxiety.

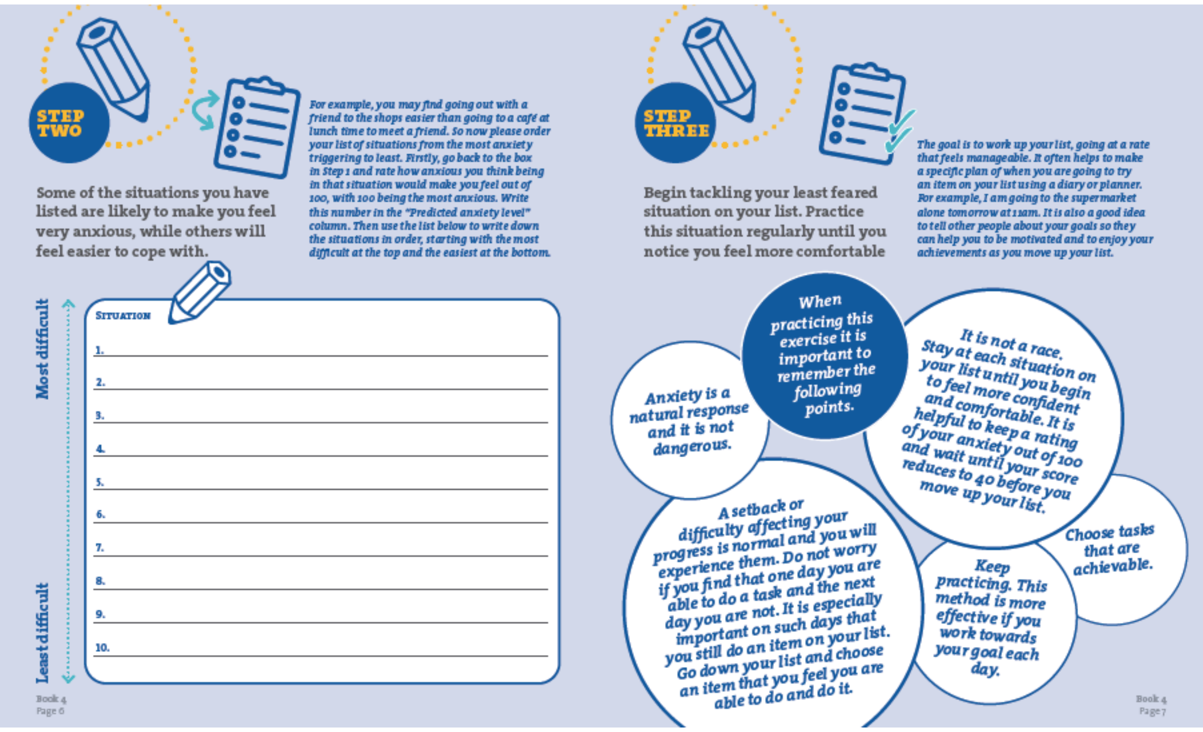
When doing this, it is useful to first think about what you value in life and what is really important to you. Think about what person you want to be, what do you want to do, how do you want others to see you, and what do you want to stand for? This will be very different for different people.

Now think about activities that fit with what you value in life and the person you want to be. It could be activities such as leaving the house, talking to people, being in large crowds, going to the supermarket and going to a restaurant. It could be situations that you manage to do but that makes you feel very anxious or things that you can only do if you use a coping strategy, such as a safety behaviour. For example, only going to restaurants while most people are at work because it is less busy, or only going out shopping with a friend.

Write down your situations in the box below in any order and do not worry about filling in the "Predicted anxiety level" column at this stage.

Situation	Predicted anxiety level
1.	
2.	
3.	
4.	
5.	
6.	
7.	
8.	
9.	
10.	

Book 4
Page 5



STEP TWO

Some of the situations you have listed are likely to make you feel very anxious, while others will feel easier to cope with.

For example, you may find going out with a friend to the shops easier than going to a café at lunch time to meet a friend. So now please order your list of situations from the most anxiety triggering to least. Firstly, go back to the box in Step 1 and rate how anxious you think being in that situation would make you feel out of 100, with 100 being the most anxious. Write this number in the "Predicted anxiety level" column. Then use the list below to write down the situations in order, starting with the most difficult at the top and the easiest at the bottom.

Most difficult

SITUATION

1. ____________________
2. ____________________
3. ____________________
4. ____________________
5. ____________________
6. ____________________
7. ____________________
8. ____________________
9. ____________________
10. ____________________

Least difficult

Book 4
Page 6

STEP THREE

Begin tackling your least feared situation on your list. Practice this situation regularly until you notice you feel more comfortable

The goal is to work up your list, going at a rate that feels manageable. It often helps to make a specific plan of when you are going to try an item on your list using a diary or planner. For example, I am going to the supermarket alone tomorrow at 11am. It is also a good idea to tell other people about your goals so they can help you to be motivated and to enjoy your achievements as you move up your list.

When practicing this exercise it is important to remember the following points.

Anxiety is a natural response and it is not dangerous.

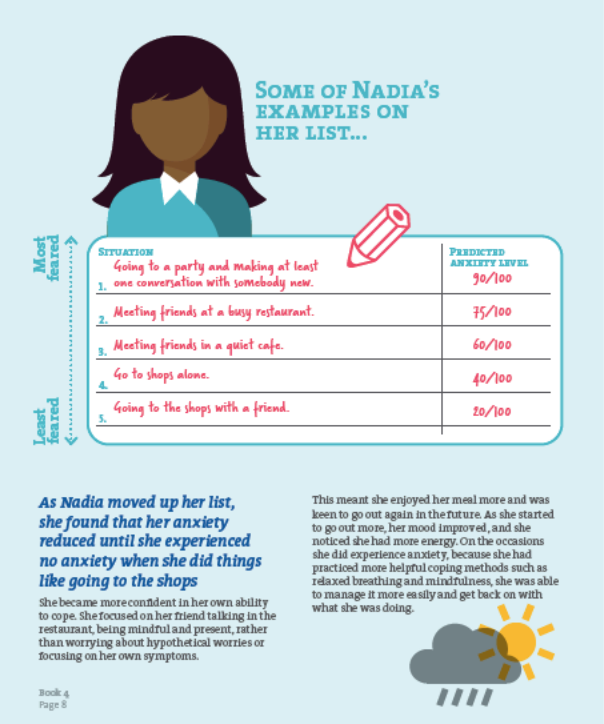
It is not a race. Stay at each situation on your list until you begin to feel more confident and comfortable. It is helpful to keep a rating of your anxiety out of 100 and wait until your score reduces to 40 before you move up your list.

A setback or difficulty affecting your progress is normal and you will experience them. Do not worry if you find that one day you are able to do a task and the next day you are not. It is especially important on such days that you still do an item on your list. Go down your list and choose an item that you feel you are able to do and do it.

Keep practicing. This method is more effective if you work towards your goal each day.

Choose tasks that are achievable.

Book 4
Page 7



SOME OF NADIA'S EXAMPLES ON HER LIST...

Most feared

SITUATION	PREDICTED ANXIETY LEVEL
1. Going to a party and making at least one conversation with somebody new.	90/100
2. Meeting friends at a busy restaurant.	75/100
3. Meeting friends in a quiet cafe.	60/100
4. Go to shops alone.	40/100
5. Going to the shops with a friend.	20/100

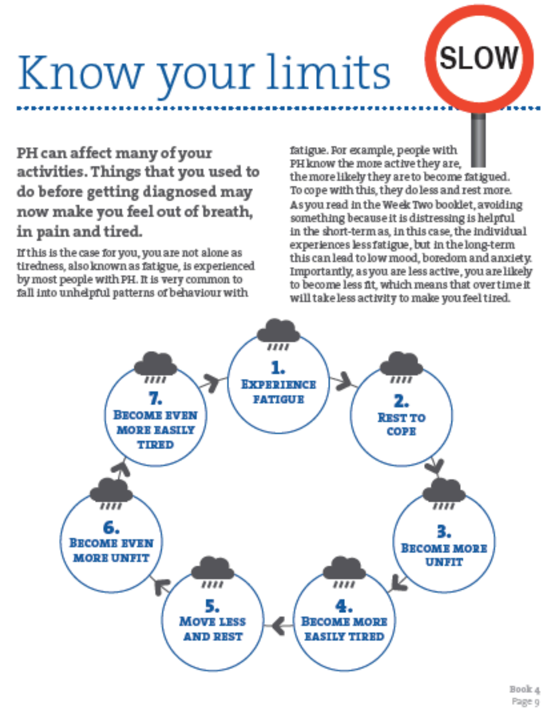
Least feared

As Nadia moved up her list, she found that her anxiety reduced until she experienced no anxiety when she did things like going to the shops

She became more confident in her own ability to cope. She focused on her friend talking in the restaurant, being mindful and present, rather than worrying about hypothetical worries or focusing on her own symptoms.

This meant she enjoyed her meal more and was keen to go out again in the future. As she started to go out more, her mood improved, and she noticed she had more energy. On the occasions she did experience anxiety, because she had practiced more helpful coping methods such as relaxed breathing and mindfulness, she was able to manage it more easily and get back on with what she was doing.

Book 4
Page 8



Know your limits

PH can affect many of your activities. Things that you used to do before getting diagnosed may now make you feel out of breath, in pain and tired.

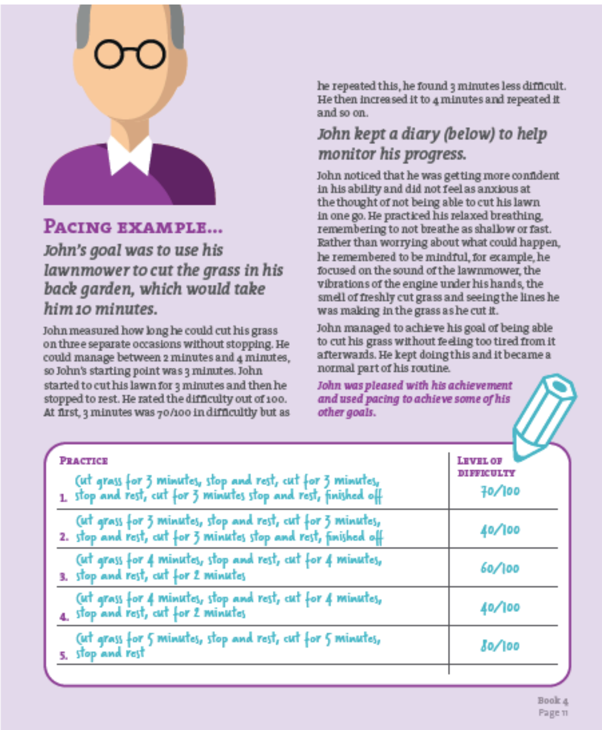
If this is the case for you, you are not alone as tiredness, also known as fatigue, is experienced by most people with PH. It is very common to fall into unhelpful patterns of behaviour with fatigue. For example, people with PH know the more active they are, the more likely they are to become fatigued. To cope with this, they do less and rest more. As you read in the Week Two booklet, avoiding something because it is distressing is helpful in the short-term as, in this case, the individual experiences less fatigue, but in the long-term this can lead to low mood, boredom and anxiety. Importantly, as you are less active, you are likely to become less fit, which means that over time it will take less activity to make you feel tired.

Book 4
Page 9

Another unhelpful cycle is called *"boom and bust"*, where on days people feel they have more energy, they push themselves into doing more work to make up for what they were unable to do on days when they had little energy or greater symptoms of PH.

Then they find they need to spend the next few days recovering as a result of over-exertion (or pushing themselves too hard), until they feel like they have more energy and any PH-related symptoms have reduced. Then they start the cycle all over again. What can also happen is as the individual becomes less fit and more easily tired, they can achieve less over time as shown by the diagram above.

Pacing is a coping method that, with practice and time, allows you to do activities in a way that causes you less fatigue. Pacing helps you find a balance between doing too much and doing too little. It means that you spend just enough time on an activity without pushing yourself to the point of exhaustion. This approach allows you to engage in more activities and do more overtime. This is achieved by increasing your level of activity in small, planned stages at your own pace, starting with the minimum you can achieve comfortably and working your way up.



PACING EXAMPLE...

John's goal was to use his lawnmower to cut the grass in his back garden, which would take him 10 minutes.

John measured how long he could cut his grass on three separate occasions without stopping. He could manage between 2 minutes and 4 minutes, so John's starting point was 3 minutes. John started to cut his lawn for 3 minutes and then he stopped to rest. He rated the difficulty out of 100. At first, 3 minutes was 70/100 in difficultly but as he repeated this, he found 3 minutes less difficult. He then increased it to 4 minutes and repeated it and so on.

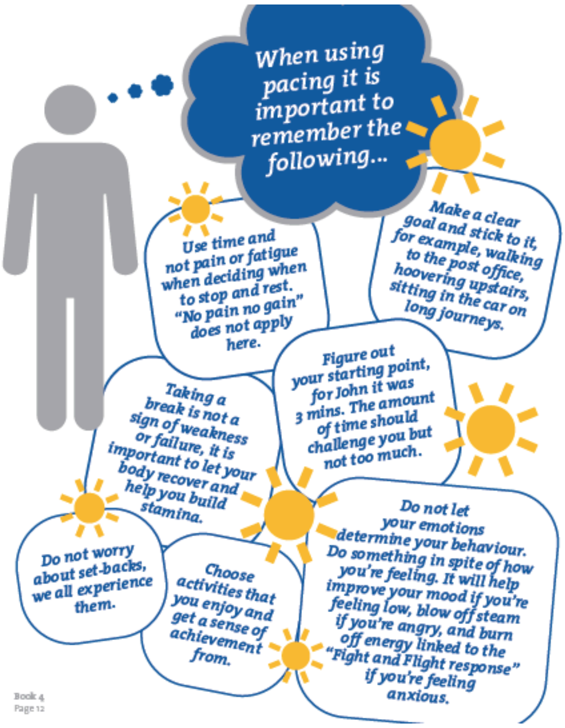
John kept a diary (below) to help monitor his progress.

John noticed that he was getting more confident in his ability and did not feel as anxious at the thought of not being able to cut his lawn in one go. He practiced his relaxed breathing, remembering to not breathe as shallow or fast. Rather than worrying about what could happen, he remembered to be mindful, for example, he focused on the sound of the lawnmower, the vibrations of the engine under his hands, the smell of freshly cut grass and seeing the lines he was making in the grass as he cut it.

John managed to achieve his goal of being able to cut his grass without feeling too tired from it afterwards. He kept doing this and it became a normal part of his routine.

John was pleased with his achievement and used pacing to achieve some of his other goals.

PRACTICE	LEVEL OF DIFFICULTY
1. Cut grass for 3 minutes, stop and rest, cut for 3 minutes, stop and rest, cut for 3 minutes stop and rest, finished off	70/100
2. Cut grass for 3 minutes, stop and rest, cut for 3 minutes, stop and rest, cut for 3 minutes stop and rest, finished off	40/100
3. Cut grass for 4 minutes, stop and rest, cut for 4 minutes, stop and rest, cut for 2 minutes	60/100
4. Cut grass for 4 minutes, stop and rest, cut for 4 minutes, stop and rest, cut for 2 minutes	40/100
5. Cut grass for 5 minutes, stop and rest, cut for 5 minutes, stop and rest	80/100



When using pacing it is important to remember the following...

Use time and not pain or fatigue when deciding when to stop and rest. "No pain no gain" does not apply here.

Make a clear goal and stick to it, for example, walking to the post office, hoovering upstairs, sitting in the car on long journeys.

Figure out your starting point, for John it was 3 mins. The amount of time should challenge you but not too much.

Taking a break is not a sign of weakness or failure, it is important to let your body recover and help you build stamina.

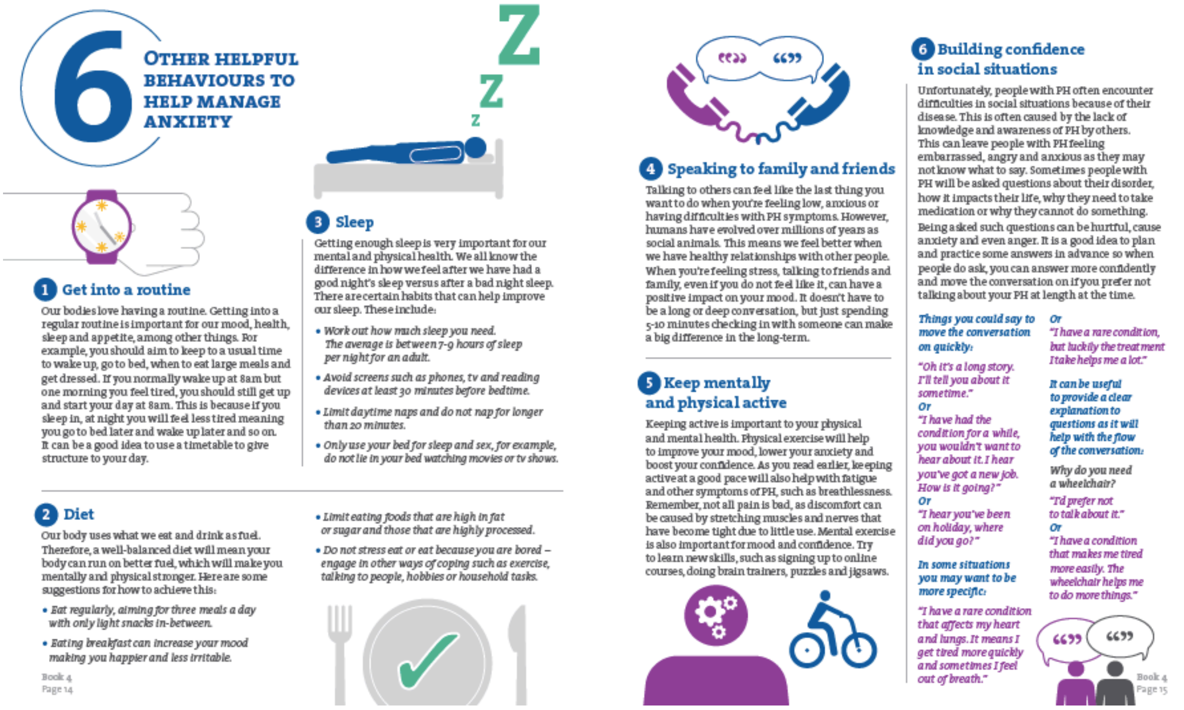
Do not worry about set-backs, we all experience them.

Choose activities that you enjoy and get a sense of achievement from.

Do not let your emotions determine your behaviour. Do something in spite of how you're feeling. It will help improve your mood if you're feeling low, blow off steam if you're angry, and burn off energy linked to the "Fight and Flight response" if you're feeling anxious.

Many people find CBT helps them to understand and respond to difficult experiences in a new way. This guide will be invaluable for anyone struggling with the challenge of PH.

Dr Louise Waddington
Consultant clinical psychologist and accredited CBT practitioner



6 OTHER HELPFUL BEHAVIOURS TO HELP MANAGE ANXIETY

1 Get into a routine

Our bodies love having a routine. Getting into a regular routine is important for our mood, health, sleep and appetite, among other things. For example, you should aim to keep to a usual time to wake up, go to bed, when to eat large meals and get dressed. If you normally wake up at 8am but one morning you feel tired, you should still get up and start your day at 8am. This is because if you sleep in, at night you will feel less tired meaning you go to bed later and wake up later and so on. It can be a good idea to use a timetable to give structure to your day.

3 Sleep

Getting enough sleep is very important for our mental and physical health. We all know the difference in how we feel after we have had a good night's sleep versus after a bad night sleep. There are certain habits that can help improve our sleep. These include:

- *Work out how much sleep you need. The average is between 7-9 hours of sleep per night for an adult.*
- *Avoid screens such as phones, tv and reading devices at least 30 minutes before bedtime.*
- *Limit daytime naps and do not nap for longer than 20 minutes.*
- *Only use your bed for sleep and sex, for example, do not lie in your bed watching movies or tv shows.*

2 Diet

Our body uses what we eat and drink as fuel. Therefore, a well-balanced diet will mean your body can run on better fuel, which will make you mentally and physical stronger. Here are some suggestions for how to achieve this:

- *Eat regularly, aiming for three meals a day with only light snacks in-between.*
- *Eating breakfast can increase your mood making you happier and less irritable.*
- *Limit eating foods that are high in fat or sugar and those that are highly processed.*
- *Do not stress eat or eat because you are bored – engage in other ways of coping such as exercise, talking to people, hobbies or household tasks.*

Book 4
Page 14

4 Speaking to family and friends

Talking to others can feel like the last thing you want to do when you're feeling low, anxious or having difficulties with PH symptoms. However, humans have evolved over millions of years as social animals. This means we feel better when we have healthy relationships with other people. When you're feeling stress, talking to friends and family, even if you do not feel like it, can have a positive impact on your mood. It doesn't have to be a long or deep conversation, but just spending 5-10 minutes checking in with someone can make a big difference in the long-term.

5 Keep mentally and physical active

Keeping active is important to your physical and mental health. Physical exercise will help to improve your mood, lower your anxiety and boost your confidence. As you read earlier, keeping active at a good pace will also help with fatigue and other symptoms of PH, such as breathlessness. Remember, not all pain is bad, as discomfort can be caused by stretching muscles and nerves that have become tight due to little use. Mental exercise is also important for mood and confidence. Try to learn new skills, such as signing up to online courses, doing brain trainers, puzzles and jigsaws.

6 Building confidence in social situations

Unfortunately, people with PH often encounter difficulties in social situations because of their disease. This is often caused by the lack of knowledge and awareness of PH by others. This can leave people with PH feeling embarrassed, angry and anxious as they may not know what to say. Sometimes people with PH will be asked questions about their disorder, how it impacts their life, why they need to take medication or why they cannot do something.

Being asked such questions can be hurtful, cause anxiety and even anger. It is a good idea to plan and practice some answers in advance so when people do ask, you can answer more confidently and move the conversation on if you prefer not talking about your PH at length at the time.

Things you could say to move the conversation on quickly:

"Oh it's a long story. I'll tell you about it sometime."
Or
"I have had the condition for a while, you wouldn't want to hear about it. I hear you've got a new job. How is it going?"
Or
"I hear you've been on holiday, where did you go?"

In some situations you may want to be more specific:

"I have a rare condition that affects my heart and lungs. It means I get tired more quickly and sometimes I feel out of breath."
Or
"I have a rare condition, but luckily the treatment I take helps me a lot."

It can be useful to provide a clear explanation to questions as it will help with the flow of the conversation:

Why do you need a wheelchair?

"I'd prefer not to talk about it."
Or
"I have a condition that makes me tired more easily. The wheelchair helps me to do more things."

Book 4
Page 15

Although anxiety can feel overwhelming at times, there are tools and techniques to help you control it.

Carl Harries,
Clinical Nurse Specialist, Royal Brompton Hospital

Book 4
Page 16

Summary

This week you learnt about the importance of not avoiding your fears, but instead exposing yourself to them gradually.

Breaking up bigger, more frightening tasks into smaller ones will make them more achievable and give you a sense of accomplishment and confidence. Remember, it is not a race to achieve your task. Instead, go at your own pace, being aware of any unhelpful patterns that could contribute to fatigue.

Anxiety can affect various aspects of your life, not just how you think and behave. It can impact on your sleep, your appetite, how you spend your days and your relationships with others. Therefore, it makes sense to also make changes to these areas of your life and introduce healthy habits in your sleep, diet, routine and how you interact with others, as this will also ***help you to live better with anxiety.***

EXERCISE

FOR YOU TO PRACTICE OVER THE FOLLOWING WEEK:

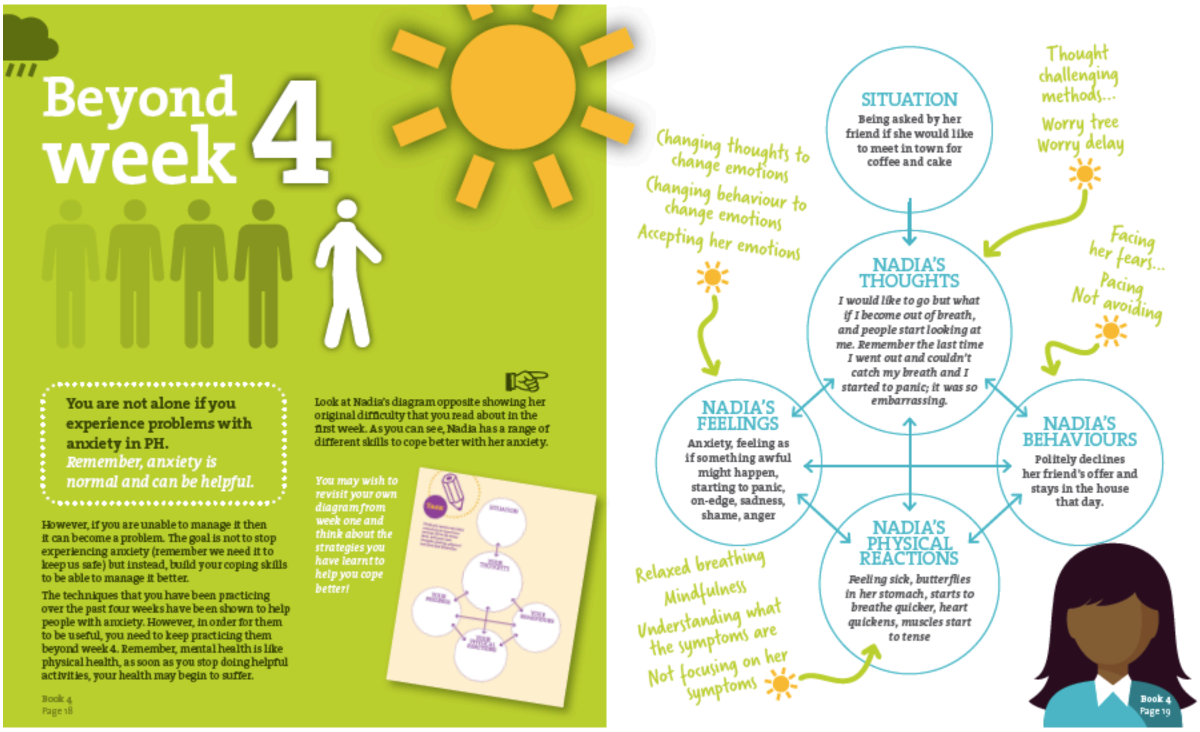
FACE SITUATIONS THAT TRIGGER YOUR ANXIETY

Over the next week, your task is to begin working up your list. Remember to use the skills you have already learnt and have been practicing helping you manage any anxiety you may experience before, during and after the task.

SITUATION

1. ______
2. ______
3. ______
4. ______
5. ______
6. ______
7. ______
8. ______
9. ______
10. ______

Book 4
Page 17



Beyond week 4

You are not alone if you experience problems with anxiety in PH.
Remember, anxiety is normal and can be helpful.

However, if you are unable to manage it then it can become a problem. The goal is not to stop experiencing anxiety (remember we need it to keep us safe) but instead, build your coping skills to be able to manage it better.

The techniques that you have been practicing over the past four weeks have been shown to help people with anxiety. However, in order for them to be useful, you need to keep practicing them beyond week 4. Remember, mental health is like physical health, as soon as you stop doing helpful activities, your health may begin to suffer.

Look at Nadia's diagram opposite showing her original difficulty that you read about in the first week. As you can see, Nadia has a range of different skills to cope better with her anxiety.

You may wish to revisit your own diagram from week one and think about the strategies you have learnt to help you cope better!

Book 4
Page 18

SITUATION
Being asked by her friend if she would like to meet in town for coffee and cake

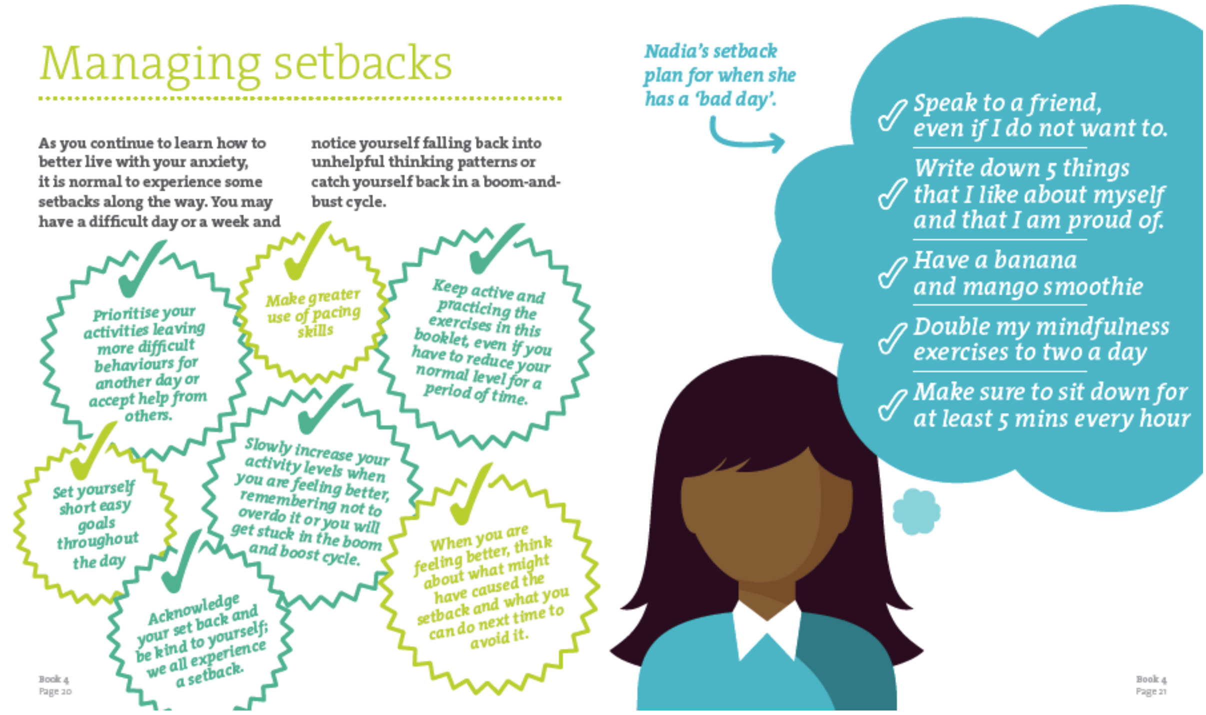
NADIA'S THOUGHTS
I would like to go but what if I become out of breath, and people start looking at me. Remember the last time I went out and couldn't catch my breath and I started to panic; it was so embarrassing.

NADIA'S FEELINGS
Anxiety, feeling as if something awful might happen, starting to panic, on-edge, sadness, shame, anger

NADIA'S BEHAVIOURS
Politely declines her friend's offer and stays in the house that day.

NADIA'S PHYSICAL REACTIONS
Feeling sick, butterflies in her stomach, starts to breathe quicker, heart quickens, muscles start to tense

Book 4
Page 19



Managing setbacks

As you continue to learn how to better live with your anxiety, it is normal to experience some setbacks along the way. You may have a difficult day or a week and notice yourself falling back into unhelpful thinking patterns or catch yourself back in a boom-and-bust cycle.

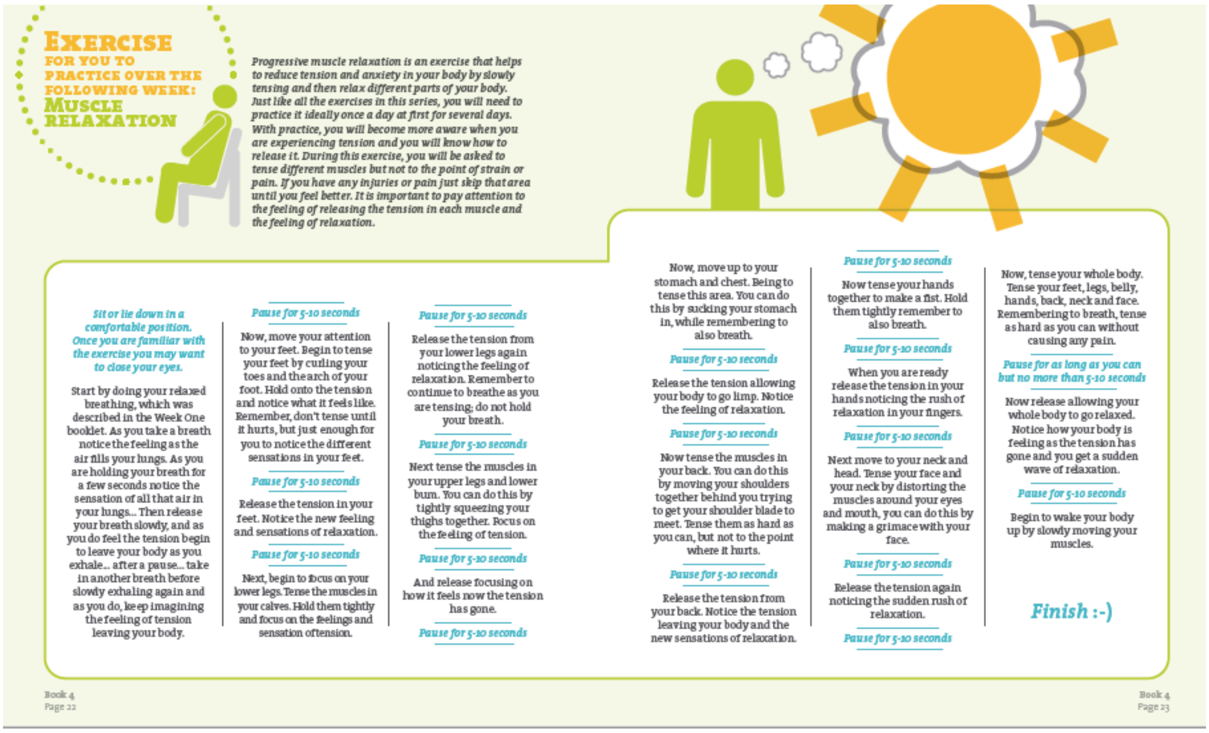
- Prioritise your activities leaving more difficult behaviours for another day or accept help from others.
- Make greater use of pacing skills
- Keep active and practicing the exercises in this booklet, even if you have to reduce your normal level for a period of time.
- Set yourself short easy goals throughout the day
- Slowly increase your activity levels when you are feeling better, remembering not to overdo it or you will get stuck in the boom and boost cycle.
- Acknowledge your set back and be kind to yourself; we all experience a setback.
- When you are feeling better, think about what might have caused the setback and what you can do next time to avoid it.

Book 4
Page 20

Nadia's setback plan for when she has a 'bad day'.

- ✓ Speak to a friend, even if I do not want to.
- ✓ Write down 5 things that I like about myself and that I am proud of.
- ✓ Have a banana and mango smoothie
- ✓ Double my mindfulness exercises to two a day
- ✓ Make sure to sit down for at least 5 mins every hour

Book 4
Page 21



EXERCISE FOR YOU TO PRACTICE OVER THE FOLLOWING WEEK: MUSCLE RELAXATION

Progressive muscle relaxation is an exercise that helps to reduce tension and anxiety in your body by slowly tensing and then relax different parts of your body. Just like all the exercises in this series, you will need to practice it ideally once a day at first for several days. With practice, you will become more aware when you are experiencing tension and you will know how to release it. During this exercise, you will be asked to tense different muscles but not to the point of strain or pain. If you have any injuries or pain just skip that area until you feel better. It is important to pay attention to the feeling of releasing the tension in each muscle and the feeling of relaxation.

Sit or lie down in a comfortable position. Once you are familiar with the exercise you may want to close your eyes.

Start by doing your relaxed breathing, which was described in the Week One booklet. As you take a breath notice the feeling as the air fills your lungs. As you are holding your breath for a few seconds notice the sensation of all that air in your lungs... Then release your breath slowly, and as you do feel the tension begin to leave your body as you exhale... after a pause... take in another breath before slowly exhaling again and as you do, keep imagining the feeling of tension leaving your body.

Pause for 5-10 seconds

Now, move your attention to your feet. Begin to tense your feet by curling your toes and the arch of your foot. Hold onto the tension and notice what it feels like. Remember, don't tense until it hurts, but just enough for you to notice the different sensations in your feet.

Pause for 5-10 seconds

Release the tension in your feet. Notice the new feeling and sensations of relaxation.

Pause for 5-10 seconds

Next, begin to focus on your lower legs. Tense the muscles in your calves. Hold them tightly and focus on the feelings and sensation of tension.

Pause for 5-10 seconds

Release the tension from your lower legs again noticing the feeling of relaxation. Remember to continue to breathe as you are tensing; do not hold your breath.

Pause for 5-10 seconds

Next tense the muscles in your upper legs and lower bum. You can do this by tightly squeezing your thighs together. Focus on the feeling of tension.

Pause for 5-10 seconds

And release focusing on how it feels now the tension has gone.

Pause for 5-10 seconds

Book 4
Page 22

Now, move up to your stomach and chest. Being to tense this area. You can do this by sucking your stomach in, while remembering to also breath.

Pause for 5-10 seconds

Release the tension allowing your body to go limp. Notice the feeling of relaxation.

Pause for 5-10 seconds

Now tense the muscles in your back. You can do this by moving your shoulders together behind you trying to get your shoulder blade to meet. Tense them as hard as you can, but not to the point where it hurts.

Pause for 5-10 seconds

Release the tension from your back. Notice the tension leaving your body and the new sensations of relaxation.

Pause for 5-10 seconds

Now tense your hands together to make a fist. Hold them tightly remember to also breath.

Pause for 5-10 seconds

When you are ready release the tension in your hands noticing the rush of relaxation in your fingers.

Pause for 5-10 seconds

Next move to your neck and head. Tense your face and your neck by distorting the muscles around your eyes and mouth, you can do this by making a grimace with your face.

Pause for 5-10 seconds

Release the tension again noticing the sudden rush of relaxation.

Pause for 5-10 seconds

Now, tense your whole body. Tense your feet, legs, belly, hands, back, neck and face. Remembering to breath, tense as hard as you can without causing any pain.

Pause for as long as you can but no more than 5-10 seconds

Now release allowing your whole body to go relaxed. Notice how your body is feeling as the tension has gone and you get a sudden wave of relaxation.

Pause for 5-10 seconds

Begin to wake your body up by slowly moving your muscles.

Finish :-)

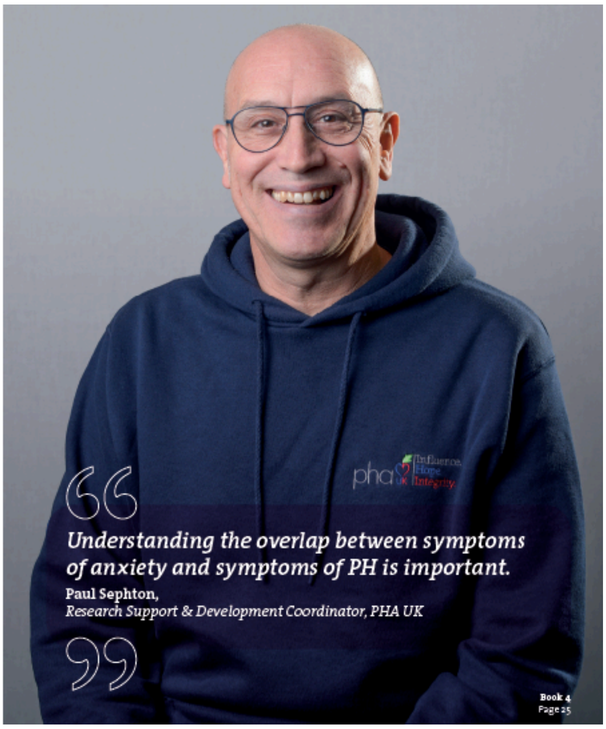
Book 4
Page 23

FURTHER SUPPORT FROM THE PHA UK

A range of FREE resources are available on a variety of topics to help inform and advise people affected by PH.

If you would like copies of any of our publications go to **www.phauk.org/publications** *call us on* **01709 761450** *or email us at* **office@phauk.org**

Book 4
Page 24



> *Understanding the overlap between symptoms of anxiety and symptoms of PH is important.*
>
> **Paul Sephton,**
> *Research Support & Development Coordinator, PHA UK*

Book 4
Page 25

A final word from Dr Rawlings...

This is the last booklet in this series. While this means that our time together has ended, I hope that your journey continues as you keep practicing the techniques in these booklets to help you live better with anxiety. I wish to thank you for all of your hard work over these last four weeks.

We will be sending you some questionnaires over the next four weeks. Please remember to complete these to help us evaluate how helpful these booklets have been.

Finally, remember to be kind and compassionate to yourself.
It is normal to feel angry, sad and upset once in a while. Do not bottle your feelings up or beat yourself up about having them. Instead, acknowledging them to yourself and remember to share them with other people can make you feel a lot better.

Additional resources

If you feel that you need additional information or support, please contact your general practitioner or PHA UK for advice. We have also included a list of other helpful resources:

NHS SELF-HELP
www.nhs.uk/conditions/stress-anxiety-depression/self-help-therapies/

PHA UK
www.phauk.org

ANXIETY UK
www.anxietyuk.org.uk

SAMARITANS
www.samaritans.org

MIND
www.mind.org.uk

NHS URGENT HELP FOR MENTAL HEALTH
www.nhs.uk/using-the-nhs/nhs-services/mental-health-services/dealing-with-a-mental-health-crisis-or-emergency/

Book 4
Page 26

References

The contents of this booklet have been adapted to make the information specific to pulmonary hypertension for the purpose of this series on anxiety from the following sources:

Anderson, R., Saulsman, L., & Nathan P. (2011).
Helping health anxiety.
Pert, Western Australia: Centre for Clinical interventions. Retrieved from: https://www.cci.health.wa.gov.au/~/media/CCI/Consumer%20Modules/Helping%20Health%20Anxiety/Helping%20Health%20Anxiety%20-%2003%20-%20What%20Keeps%20Health%20Anxiety%20Going.pdf

Anonymous. (date unknown).
Pacing for pain and fatigue [leaflet].
Psychology Tools Limited. Retrieved from: https://www.psychologytools.com/resource/pacing-for-pain-and-fatigue/

Anonymous, (2017).
Progressive muscle relaxation script [Guide].
Therapist Aid LLC. Retrieved from: https://www.therapistaid.com/worksheets/progressive-muscle-relaxation-script.pdf

Anxiety Canada (Unknown).
Calm Breathing [Leaflet].
Retrieved from https://www.anxietycanada.com/sites/default/files/CalmBreathing.pdf

Khazan, I. Z. (2013).
The clinical handbook of biofeedback: A step-by-step guide for training and ractice with mindfulness.
West Sussex, UK: John Wiley & Sons Ltd.

Maunder, L., & Cameron, L. (Unknown).
Anxiety: A self help guide [Guide].
Sheffield IAPT: Sheffield Health and Social Care. Retrieved from: http://www.selfhelpguides.ntw.nhs.uk/iaptsheffield/leaflets/selfhelp/Anxiety.pdf

Pasterfield, M., & Thompson A. (2013).
Building confidence in social situations: A guide for people living with a skin condition, including scars [Leaflet].
Sheffield IAPT: Sheffield Health and Social Care. Retrieved from: http://iaptsheffield.shsc.nhs.uk/wp-content/uploads/2019/05/Building-Confidence-in-Social-Situations-Skin-Conditions.pdf

Shafran, R., Brosa, L., Cooper, P. (2013).
The complete CBT guide for anxiety. A self-help guide for anxiety, panic, social anxiety, phobias, health anxiety and obsessive-compulsive disorder.
London: Constable & Robinson

Whalley, M., & Kaur, H., (2020).
Living with worry and anxiety amidst global uncertainty [Guide].
Psychology Tools Limited. Retrieved from: https://www.psychologytools.com/articles/free-guide-to-living-with-worry-and-anxiety-amidst-global-uncertainty/

Book 4
Page 27

pha UK | Influence. Hope. Integrity.

> ***Although anxiety can feel overwhelming at times, there are tools and techniques to help you control it.***
>
> **Carl Harries, Clinical Nurse Specialist in PH**

To find out more about the PHA UK, visit **www.phauk.org**

Pulmonary Hypertension Association UK
PHA UK Resource Centre, Unit 1, Newton Business Centre, Newton Chambers Road,
Thorncliffe Park, Chapeltown, Sheffield, England S35 2PH
T: 01709 761450 E: office@phauk.org @PULHAUK @PHA_UK @PHA ORG UK

Registered Charity No. 1120756. Copyright © 2020 PHA UK.

Appendix C: CONSORT Guidelines Checklist

CONSORT 2010 checklist when reporting a pilot trial*

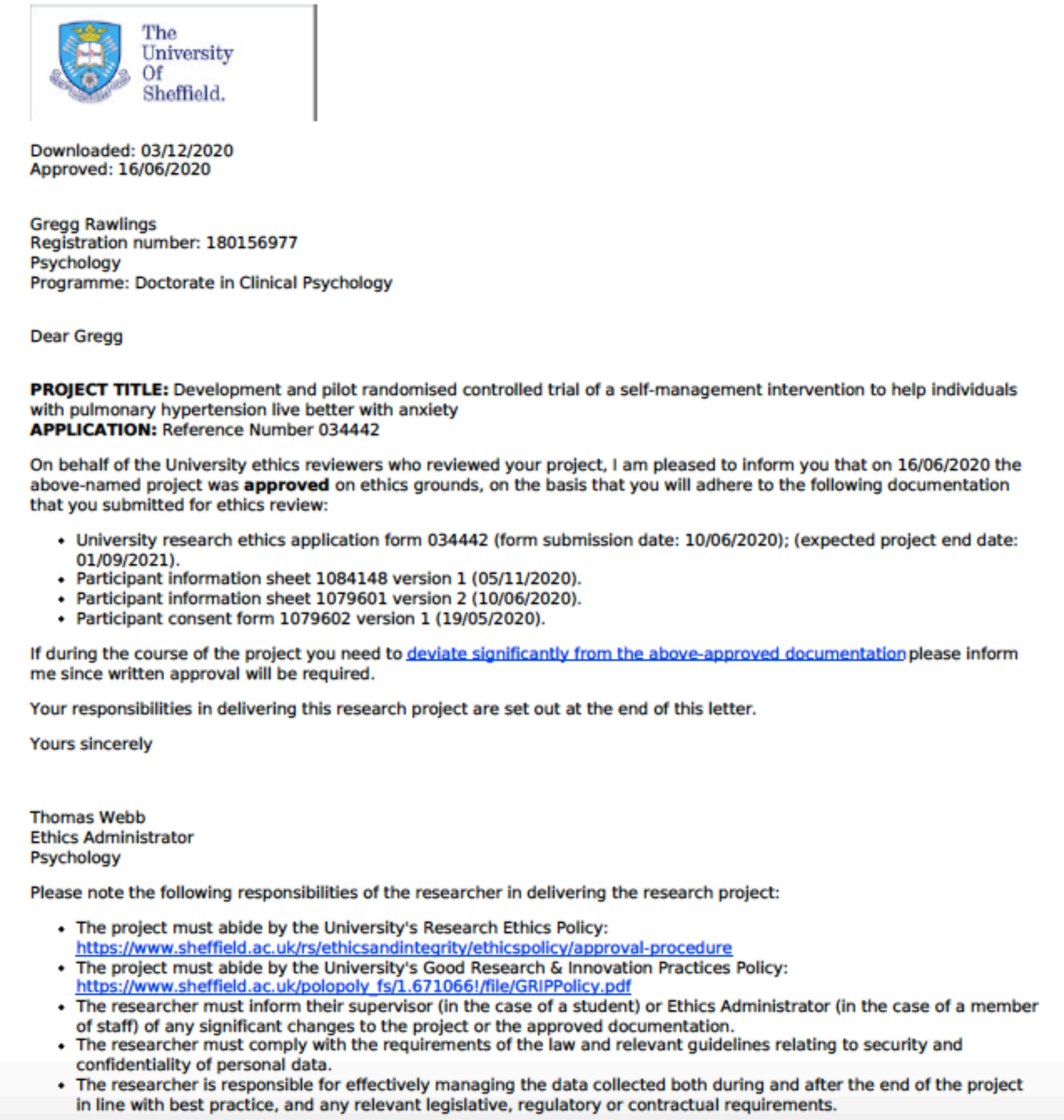
Section/Topic	Item No	Checklist item	Reported on page No
Title and abstract			
	1a	Identification as a pilot or feasibility randomised trial in the title	103
	1b	Structured summary of pilot trial design, methods, results, and conclusions (for specific guidance see CONSORT abstract extension for pilot trials)	104-105 (extension only used for journal)
Introduction			
Background and objectives	2a	Scientific background and explanation of rationale for future definitive trial, and reasons for randomised pilot trial	106-111
	2b	Specific objectives or research questions for pilot trial	109-111
Methods			
Trial design	3a	Description of pilot trial design (such as parallel, factorial) including allocation ratio	118
	3b	Important changes to methods after pilot trial commencement (such as eligibility criteria), with reasons	119-120
Participants	4a	Eligibility criteria for participants	119
	4b	Settings and locations where the data were collected	119-121
	4c	How participants were identified and consented	119-121
Interventions	5	The interventions for each group with sufficient details to allow replication, including how and when they were actually administered	112-117, 119-121 Appendix B
Outcomes	6a	Completely defined prespecified assessments or measurements to address each pilot trial objective specified in 2b, including how and when they were assessed	118, 121-123
	6b	Any changes to pilot trial assessments or measurements after the pilot trial commenced, with reasons	119-121
	6c	If applicable, prespecified criteria used to judge whether, or how, to proceed with future definitive trial	155

Sample size	7a	Rationale for numbers in the pilot trial	118-119
	7b	When applicable, explanation of any interim analyses and stopping guidelines	118
Randomisation:			
Sequence generation	8a	Method used to generate the random allocation sequence	118
	8b	Type of randomisation(s); details of any restriction (such as blocking and block size)	118
Allocation concealment mechanism	9	Mechanism used to implement the random allocation sequence (such as sequentially numbered containers), describing any steps taken to conceal the sequence until interventions were assigned	118
Implementation	10	Who generated the random allocation sequence, who enrolled participants, and who assigned participants to interventions	118 -121
Blinding	11a	If done, who was blinded after assignment to interventions (for example, participants, care providers, those assessing outcomes) and how	118
	11b	If relevant, description of the similarity of interventions	121
Statistical methods	12	Methods used to address each pilot trial objective whether qualitative or quantitative	124-126
Results			
Participant flow (a diagram is strongly recommended)	13a	For each group, the numbers of participants who were approached and/or assessed for eligibility, randomly assigned, received intended treatment, and were assessed for each objective	126-127
	13b	For each group, losses and exclusions after randomisation, together with reasons	126-127
Recruitment	14a	Dates defining the periods of recruitment and follow-up	118
	14b	Why the pilot trial ended or was stopped	118
Baseline data	15	A table showing baseline demographic and clinical characteristics for each group	128-134
Numbers analysed	16	For each objective, number of participants (denominator) included in each analysis. If relevant, these numbers should be by randomised group	134-147
Outcomes and estimation	17	For each objective, results including expressions of uncertainty (such as 95% confidence interval) for any estimates. If relevant, these results should be by randomised group	128-149
Ancillary analyses	18	Results of any other analyses performed that could be used to inform the future definitive trial	128-149
Harms	19	All important harms or unintended effects in each group (for specific guidance see CONSORT for harms)	118
	19a	If relevant, other important unintended consequences	118
Discussion			
Limitations	20	Pilot trial limitations, addressing sources of potential bias and remaining uncertainty about feasibility	153-154
Generalisability	21	Generalisability (applicability) of pilot trial methods and findings to future definitive trial and other studies	149-153

Interpretation	22	Interpretation consistent with pilot trial objectives and findings, balancing potential benefits and harms, and considering other relevant evidence	149-155
	22a	Implications for progression from pilot to future definitive trial, including any proposed amendments	149-155
Other information			
Registration	23	Registration number for pilot trial and name of trial registry	118
Protocol	24	Where the pilot trial protocol can be accessed, if available	118
Funding	25	Sources of funding and other support (such as supply of drugs), role of funders	4
	26	Ethical approval or approval by research review committee, confirmed with reference number	118

Eldridge, S.M., Chan, C.L., Campbell, M.J., Bond, C.M., Hopewell, S., Thabane, L. & Lancster, G. (2016). CONSORT 2010 statement: extension to randomised pilot and feasibility trials. *BMJ, 355*, Article i5239. https://doi.org/10.1136/bmj.i5239

Appendix D: Ethical Approval Letter



The
University
Of
Sheffield.

Approved: 16/06/2020

Gregg Rawlings
Registration number: 180156977
Psychology
Programme: Doctorate in Clinical Psychology

Dear Gregg

PROJECT TITLE: Development and pilot randomised controlled trial of a self-management intervention to help individuals with pulmonary hypertension live better with anxiety
APPLICATION: Reference Number 034442

On behalf of the University ethics reviewers who reviewed your project, I am pleased to inform you that on 16/06/2020 the above-named project was **approved** on ethics grounds, on the basis that you will adhere to the following documentation that you submitted for ethics review:

- University research ethics application form 034442 (form submission date: 10/06/2020); (expected project end date: 01/09/2021).
- Participant information sheet 1084148 version 1 (05/11/2020).
- Participant information sheet 1079601 version 2 (10/06/2020).
- Participant consent form 1079602 version 1 (19/05/2020).

If during the course of the project you need to deviate significantly from the above-approved documentation please inform me since written approval will be required.

Your responsibilities in delivering this research project are set out at the end of this letter.

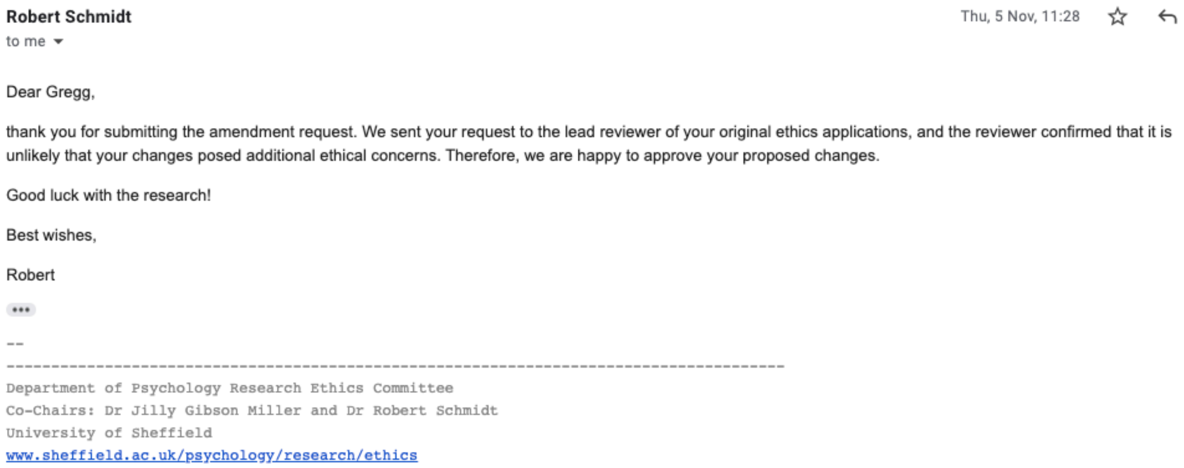
Yours sincerely

Thomas Webb
Ethics Administrator
Psychology

Please note the following responsibilities of the researcher in delivering the research project:

- The project must abide by the University's Research Ethics Policy: https://www.sheffield.ac.uk/rs/ethicsandintegrity/ethicspolicy/approval-procedure
- The project must abide by the University's Good Research & Innovation Practices Policy: https://www.sheffield.ac.uk/polopoly_fs/1.671066!/file/GRIPPolicy.pdf
- The researcher must inform their supervisor (in the case of a student) or Ethics Administrator (in the case of a member of staff) of any significant changes to the project or the approved documentation.
- The researcher must comply with the requirements of the law and relevant guidelines relating to security and confidentiality of personal data.
- The researcher is responsible for effectively managing the data collected both during and after the end of the project in line with best practice, and any relevant legislative, regulatory or contractual requirements.

Ethical approval for amendments regarding eligibility to include people who have received treatment for anxiety in the last twelve months, and to recruit people outside of the UK.



Robert Schmidt

to me

Thu, 5 Nov, 11:28

Dear Gregg,

thank you for submitting the amendment request. We sent your request to the lead reviewer of your original ethics applications, and the reviewer confirmed that it is unlikely that your changes posed additional ethical concerns. Therefore, we are happy to approve your proposed changes.

Good luck with the research!

Best wishes,

Robert

--

Department of Psychology Research Ethics Committee
Co-Chairs: Dr Jilly Gibson Miller and Dr Robert Schmidt
University of Sheffield
www.sheffield.ac.uk/psychology/research/ethics

Appendix E: Research Governance Sponsor Letter

Address: Gregg Rawlings
Clinical Psychologist
Department of Psychology
Cathedral Court

Clinical Psychology Unit
Department of Psychology
Cathedral Court
Sheffield

Date: 25.06.2020

Telephone: 0114 22 26650
Email: a.sinha@sheffield.ac.uk

Project title: Development and pilot randomised controlled trial of a self-management intervention to help individuals with pulmonary hypertension live better with anxiety

URMS number: **167568**

Dear Gregg,

LETTER TO CONFIRM THAT THE UNIVERSITY OF SHEFFIELD IS THE HUMAN-INTERVENTIONAL STUDY'S RESEARCH GOVERNANCE SPONSOR

As you are aware, as a University-sponsored human-interventional study, the study was subject to the University's risk assessment procedure, and your support in facilitating the procedure by completing the checklist is appreciated. The outcome of the risk assessment procedure is that the University has categorised the human-interventional study as being potentially low risk.

Therefore, I would be very grateful if you could **sign and date the enclosed self-certification statement at Annex 2 and arrange for your Head of School/Department to countersign it. The form should then be returned to [departmental research governance contact].**

The University expects that the human-interventional study will have in place present and effective systems and practices for a) safeguarding the dignity, rights, safety & well-being of participants recruited to the study and b) for ensuring the validity of the data collected, analysed, recorded and reported.

Your School/Department has reviewed the documents which confirm that the following is in place:

- Scientific approval;
- Ethical approval;
- Registration on a publicly accessible registry.

Accordingly, once the self-certification statement has been signed and returned, as the study's research governance sponsor the University authorises the study to commence. Please note that any aspects of the research requiring additional research governance approvals (e.g. site approval) must be obtained before the commencement of those aspects of the research.

You are expected to deliver the study in accordance with the University's policies and procedures, which includes the University's Good Research and Innovation Practices Policy: https://www.sheffield.ac.uk/rs/ethicsandintegrity/index.

As the Principal Investigator you are responsible for providing up-to-date study documentation to all relevant sites, and for monitoring the study on an ongoing basis. Your Head of School (and/or Head of Department in the case of the Medical School) is responsible for independently monitoring the study as appropriate. The monitoring responsibilities are listed at Annex 1**.** More details can be found on the University's research governance website: https://www.sheffield.ac.uk/rs/ethicsandintegrity/governance.

Yours sincerely,

Jaime Delgadillo

Dr Jaime Delgadillo

Director of Research Training, Clinical Psychology Unit

cc.
Head of Department

Glenn Waller

Glenn Waller

Appendix F: Clinical Trial Registration Form

ID: 034442 Anxiety Self-management Intervention for Pulmonary Hypertension NCT04450862

Record Status

In Progress → Entry Completed → Approved → Released → PRS Review → ***Public***

Record Owner: GRawlings
Last Update: 11/05/2020 14:51 by GRawlings
Initial Release: 06/19/2020
Last Release: 11/06/2020 Receipt (PDF)

Access List: [] Edit
Upload: Allowed Edit
PRS Review: Review History
Public Site: Last Public Release: 11/06/2020 View on ClinicalTrials.gov
FDAAA: Non-ACT (No FDA-regulated drug/device)

Protocol Section

Identifiers: NCT04450862 Unique Protocol ID: 034442
Brief Title: Anxiety Self-management Intervention for Pulmonary Hypertension
Module Status:
- Study Identification: ✓
- Study Status: ✓
- Sponsor/Collaborators: ✓
- Oversight: ✓
- Study Description: ✓
- Conditions: ✓
- Study Design: ✓
- Arms and Interventions: ✓
- Outcome Measures: ✓
- Eligibility: ✓
- Contacts/Locations: ✓
- IPD Sharing Statement: ✓
- References: ✓

Appendix G: Consent Form

The University Of Sheffield.

Department Of Psychology. Clinical Psychology Unit.

Doctor of Clinical Psychology (DClin Psy) Programme Clinical supervision training and NHS research training & consultancy

Clinical Psychology Unit
University of Sheffield
Cathedral Court,
Floor F,
1 Vicar Lane,
Sheffield,
S1 2LT

Telephone: 0114 222 2000
Fax: 0114 2226610
Email: g.rawlings@sheffield.ac.uk

Research Project: Development and pilot randomised controlled trial of a self-management intervention to help individuals with pulmonary hypertension live better with anxiety

Researcher: Gregg Rawlings

Participant identification number: …………………………..

Please read the information below and write your initials in the boxes if you agree to the statements.

1. I confirm that I have read the information sheet dated *[insert date]* for the above study. I have had the opportunity to consider the information, ask questions and have had these answered. ☐
2. I understand that my participation is voluntary and that I am free to withdraw at any time without giving any reason and without my medical care or involvement in Pulmonary Hypertension Association UK being affected. I can contact the lead researcher Gregg Rawlings on 0114 222 2000 or g.rawlings@sheffield.ac.uk if I wish to withdraw. ☐
3. I understand that the information I provide will be used to support other research in the future, and may be shared anonymously with other individuals, for example, for research publications. I give permission that the research team may use my data in publications, reports and other research outputs, only if they agree to preserve the confidentiality of my information. ☐
4. I understand that this project is a randomised controlled trial meaning that I may be randomised to the control group that do not receive the intervention immediately, but I will still be asked to complete a series of questionnaires. I understand that if I am in the control group and if the booklet is found to be helpful, I will receive the self-help intervention at a later date. ☐

5. I agree to provide my contact details for the purpose of this project and for Gregg Rawlings to contact me at the end of the intervention asking me to complete a series of questions. ☐

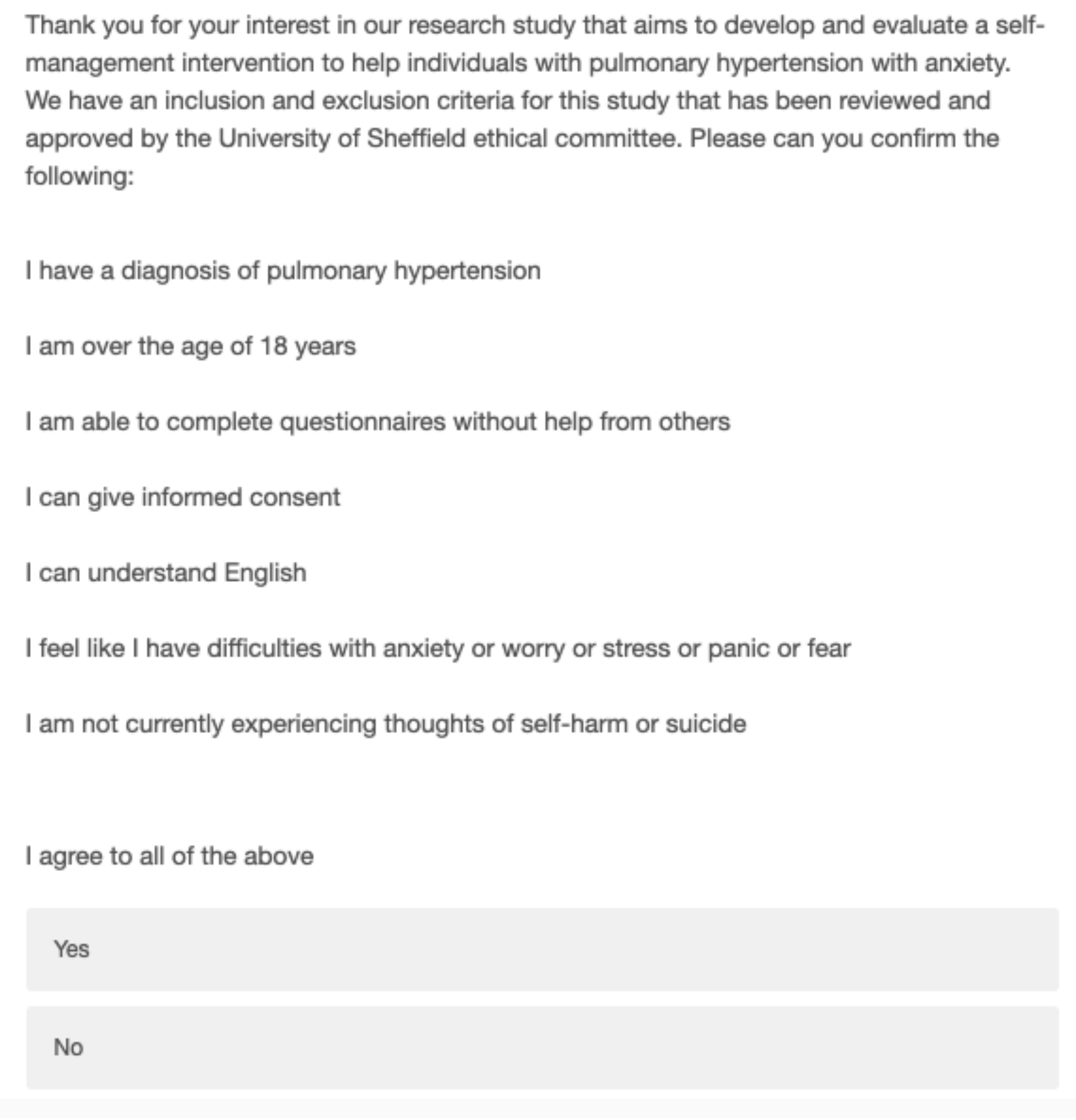
6. I agree to be contacted by Gregg Rawlings partway through the intervention asking me questions about my experience of the self-help intervention. ☐

7. I agree to be contacted by Gregg Rawlings after my involvement in the study has finished to ask me questions about my experience of the intervention. ☐

8. I agree to take part in this study and understand that the data will be used as part of a doctoral degree. ☐

________________________	_______________	____________________
Name of Participant	Date	Signature
________________________	_______________	____________________
Name of Researcher	Date	Signature

Appendix H: Screening Questionnaire



Thank you for your interest in our research study that aims to develop and evaluate a self-management intervention to help individuals with pulmonary hypertension with anxiety. We have an inclusion and exclusion criteria for this study that has been reviewed and approved by the University of Sheffield ethical committee. Please can you confirm the following:

I have a diagnosis of pulmonary hypertension

I am over the age of 18 years

I am able to complete questionnaires without help from others

I can give informed consent

I can understand English

I feel like I have difficulties with anxiety or worry or stress or panic or fear

I am not currently experiencing thoughts of self-harm or suicide

I agree to all of the above

Appendix I: Study Advert Example

PHA UK
@PHA_UK

Experiencing some anxiety and worry is a normal part of life, but it's important to access help when it becomes too much.
Our partnership study with University of Sheffield psychologists is now available to anyone with #PulmonaryHypertension: bit.ly/37OCIGc

11:53 am · 8 Dec 2020 · Twitter Web App

Appendix J: Participant Information Sheet

The
University
Of
Sheffield.

Department Of Psychology. Clinical Psychology Unit.

Doctor of Clinical Psychology (DClin Psy) Programme Clinical supervision training and NHS research training & consultancy

Clinical Psychology Unit
University of Sheffield
Cathedral Court,
Floor F,
1 Vicar Lane,
Sheffield,
S1 2LT

Telephone: 0114 222 2000
Fax: 0114 2226610
Email: g.rawlings@sheffield.ac.uk

1. Research Project title:

Development and pilot randomised controlled trial of a self-management intervention to help individuals with pulmonary hypertension live better with anxiety

2. Researchers involved:

Dr Gregg Rawlings, Professor Nigel Beail, Dr Iain Armstrong, Dr Andrew Thompson

3. Invitation:

You are being invited to take part in a research project. Before you decide whether or not to participate, it is important for you to understand why the research is being done and what it will involve. Please take time to read the following information carefully and discuss it with others if you wish. Ask us if there is anything that is not clear or if you would like more information. Take time to decide whether or not you wish to take part. Thank you for reading this

4. What is the project's purpose?

Up to 50% of individuals with pulmonary hypertension experience symptoms of anxiety to the point where it has a considerable negative impact on daily life. Symptoms of anxiety may include fear, panic, excessive worry and stress. Cognitive Behavioural therapy is a type of talking treatment that works by helping people to alter their attitudes and behaviours by exploring their cognitions, emotions, physiology and behaviours. This project looks to develop and investigate a self-help intervention based on principles of Cognitive Behavioural therapy to help individuals with pulmonary hypertension manage anxiety.

This research is being conducted by Gregg Rawlings, a Trainee Clinical Psychologist at the University of Sheffield. This study is being carried out as part of his doctoral research project

5. Why have I been chosen to take part?

You have been invited to take part in this research study as you have a diagnosis of pulmonary hypertension and are a service user of a Pulmonary Hypertension Association UK.

6. Do I have to take part?

No. It is up to you to decide whether or not to take part. Your decision will not affect the level of care you receive or involvement with the Pulmonary Hypertension Association. Participation in this research study is voluntary. You have the right to withdraw from the study at any time without providing a reason. If you wish to withdraw from the research, please contact Gregg Rawlings.

7. What will happen to me if I take part?

If you wish to take part in this study, we will ask you to complete a series of questionnaires asking about some difficulties that you may be experiencing associated with anxiety. More specifically, anxiety, depression, quality of life, breathing difficutlies, your perception of self-control and, behaviours and thoughts towards coping. You will then be put into one of two groups at random. One group will receive the self-help intervention within a few days. The other group will receive the intervention once the study is completed in September 2021, but only if the intervention was found to be helpful.

This intervention is based on a type of therapy called Cognitive Behavioural Thearpy, which has been shown to help people with pulmonary hypertension manage anxiety. You will be asked to work through the intervention over the next four weeks alone, in your own time and at home. The intervention includes information about pulmonary hypertension and anxiety and how the two can interact, and a range of techniques and skills to help you to better understand and cope with symptoms of anxiety, such as excessive worry, fear, panic, nervousness and stress.

After four weeks we will contact you again asking you to complete the same questions as at the start. We will also ask you permission to contact you at week two and after you have completed the questionnaires for the second time to ask you about your experiences of the intervention and participation in the study.

8. What are the possible risks of taking part in this study?

There are no significant risks associated with taking part in the study. Some of the information in the intervention may be difficult to read, as it will be asking you to think about your anxiety and emotional difficulties. Some of the tasks may also cause some initial distress or uncomfortable. The intervention aims to help you to develop a range of coping skills to help manage with anxiety and distress. You will also be provided the details of services and organisations you can contact for further support. This information can be found at the end of the intervention materials. Alternatively, please contact Gregg Rawlings, his supervisors or PHA UK who can provide you with this information. If you feel too distressed or the intervention is having a negative impact on you, you can stop at any time.

9. What are the possible benefits of taking part?

Self-help intervention s have helped people with other medical conditions in other research trials to manage their anxiety better. This can have a positive effect on other areas of your life, for example it may help to improve your mood and quality of life. The information you provide will also help to further develop the self-help intervention hopefully making it more useful for other people with pulmonary hypertension. The information you provide us with may also help us to better understand the impact of anxiety in pulmonary hypertension, and how people with the condition can be supported better.

10. Will my taking part in this project be kept confidential?
All the information that we collect about you during the course of the study will be kept strictly confidential. You will not be able to be identified in any reports or publications.

11. What is the legal basis for processing my personal data?
We are required to inform you that the legal basis we are applying in order to process your personal data is that 'processing is necessary for the performance of a task carried out in the public interest' (Article 6(1)(e)). Further information can be found in the University's Privacy Notice https://www.sheffield.ac.uk/govern/ data-protection/privacy/general. As we will be collecting some data that is defined in the legislation as more sensitive (i.e. information about your medical condition), we also need to let you know that we are applying the following condition in law: that the use of your data is 'necessary for scientific or historical research purposes.

12. What will happen to the data collected, and the results of the project?
All information that you provide will be stored on a password-protected computer. Only members of the research team will have access to this information. When the study is complete, all paper files will be disposed of securely and electronic files will be stored for a maximum of 6 years at the University of Sheffield. They will be securely disposed of after this time. We plan to publish the findings in an international scientific research journal. The findings will also be shared with medical professionals and patients at conferences and presentations. You will not be identified in any report or publication.

13. Who is organising and funding the research?
This research project is funded by the University of Sheffield and Pulmonary Hypertension Association UK. The study is sponsored by the University of Sheffield.

14. Who is the data controller?
The University of Sheffield will act as the Data Controller. This means that the university is responsible for looking after your information and using it properly.

15. Who has ethically reviewed the project?
The project has been reviewed by the Department of Psychology, University of Sheffield. The study has ethical approval gained from the University of Sheffield's Ethics Review Procedure, as administered by Department of Psychology.

16. Useful contact numbers should you need additional support
If you are concerned about anything about the project, please contact Gregg Rawlings in the first instance. You are also to contact his research supervisors:

Professor Nigel Beail
Clinical Psychology Unit
University of Sheffield
Cathedral Court,
Floor F,
1 Vicar Lane,
Sheffield,
S1 2LT

Dr Iain Armstrong
Thorncliffe Park,
Unit 1 Newton Business Centre,
Newton Chambers Rd,
Sheffield
S35 2PH

If you still feel your concerns have not been addressed satisfactory, please contact the Head of Department, Professor Glenn Waller, Head of Psychology Department, University of Sheffield, Cathedral Court, Floor F, 1 Vicar Lane, Sheffield, S1 2LT who will then escalate your complain through the appropriate channels.

If the complaint relates to how the participants' personal data has been handled, information about how to raise a complaint can be found in the University's Privacy Notice: https://www.sheffield.ac.uk/govern/data-protection/privacy/general.

Thank you for considering taking part in this research project

Appendix K: Adherence and Acceptability Check-In Questionnaire

Hello, my name is Gregg Rawlings, can I please speak to [participants name]. I am contacting you in regard to the study that you are taking part in looking to develop and evaluate a self-management intervention to help individuals with pulmonary hypertension to manage anxiety and panic. PHA UK and the University of Sheffield are doing this project.

You have provided me with permission to contact you partway through the study to discuss with you how you are getting on with the intervention.

*Is this a suitable time to speak?

*> If no arrange a suitable time to call if possible

*>If yes, this phone call will only be brief and should take no more than 10 minutes. I have a series of questions to ask you about your experiences of the intervention so far:

1. *Are you comfortable talking on the phone with me about your experiences of the intervention (i.e. do you have any concerns with confidentiality?) explore
2. *It is a good idea to take this call in a space that is private and quiet, is that possible? (i.e. are there any distractions and is the participant alone?)
3. Can I confirm that you have received the intervention?
    a. Yes – move to question 2
    b. *Confirm postal address and arrange for another intervention to be posted
4. Have you managed to have a look at the booklet? Yes/No
    a. No, explore reasons why
    b. Yes, how much have you looked at the intervention?
        i. 1 (not at all) – 5 (a great deal)
5. What section are you up to?
    a. Multi-choice
6. How much have you understood the information in the intervention so far?

        i. 1 (not at all) – 5 (a great deal)

7. How distressing have you found the intervention so far?
    i. 1 (not at all) – 5 (a great deal)
8. How difficult have you found using the intervention?
    i. 1 (not at all) – 5 (a great deal)
9. How often have you been using the skills you have developed so far from the intervention?
    i. 1 (not at all) – 5 (a great deal)
10. How much do you feel the intervention is helping you with your anxiety?
    i. 1 (not at all) – 5 (a great deal)
11. How much do you feel the intervention is helping you in other areas of your life?
    i. 1 (not at all) – 5 (a great deal)
12. How much do you think the intervention can help you with your anxiety?
    i. 1 (not at all) – 5 (a great deal)
13. How much do you think the intervention can help you in other areas of your life?
    i. 1 (not at all) – 5 (a great deal)
14. Do you intend to finish the intervention?
    i. 1 (no intention) – 5 (definitely intend to)
15. Do you have any other comments about the intervention at this stage? (open ended question)

Thank you for your time. Please remember we will contact you again in two weeks when you should have completed the intervention asking you to complete a series of questions.

Appendix L: Final Acceptability Questionnaire

Dear Participant,

Thank you for taking part in the research study that aimed to develop and evaluate a self-management intervention to help individuals with pulmonary hypertension manage difficulties associated with anxiety and panic.

To help us to better understand your experiences of using the self-help intervention and taking part in the study, we have devised a questionnaire asking you about your experiences that we would like you to answer. This questionnaire should take approximately 10-15 minutes to complete. We are interested in your honest opinion, whether they are positive or negative. Thank you very much, we appreciate your help:

1. What motivated you to take part in this research study? (open ended)
2. How much did the intervention help you to manage your anxiety?
   a. 1 (not at all) – 5 (a lot)
   b. Please expand on your answer:
3. How much did the intervention help you in other areas of your life?
   a. 1 (not at all) – 5 (a lot)
   b. Please expand on your answer:
4. How likely are you to recommend the intervention to another person with pulmonary hypertension?
   a. 1 (not at all) – 5 (a lot)

5. Please rate each of the following sections in terms of how helpful it was for managing your anxiety: 1 (not at all helpful) – 5 (extremely helpful)

Component	1-5
Booklet 1	
Booklets 2 & 3	
Booklet 4 – focusing on behaviours	
Booklet 4 – other information on anxiety	
Homework tasks	

6. The questionnaires asking about my anxiety, mood, quality of life and behaviours were relevant to my difficulties
    a. 1 (strongly disagree) – 5 (strongly agree)
    b. Please expand:
7. I feel more in control of my anxiety
    a. 1 (strongly disagree) – 5 (strongly agree)
8. I was given enough time to work through the intervention
    a. 1 (strongly disagree) – 5 (strongly agree)
9. I valued that the intervention was specific to pulmonary hypertension
    a. 1 (strongly disagree) – 5 (strongly agree)
10. I think when people are diagnosed with pulmonary hypertension they would benefit from being given this intervention
    a. 1 (strongly disagree) – 5 (strongly agree)
11. I think relatives and friends of those with pulmonary hypertension would benefit from using this intervention
    a. 1 (strongly disagree) – 5 (strongly agree)
12. I valued being contacted partway through the study by a member of the research team
    a. 1 (strongly disagree) – 5 (strongly agree)
13. The level of support I received from the research team was sufficient

   a. 1 (strongly disagree) – 5 (strongly agree)
   a. Please expand on your answer

14. How do you plan to continue using the skills you have developed by using the intervention (open ended):
15. Overall, how would you rate the self-help intervention?
   c. 1 (Poor) – 5 (Excellent)
   d. What was your experience of taking part in this research study?
16. What did you like the most about the intervention? (open ended)
17. What did you like the least about the intervention? (open ended)
18. Do you have any other comments about the intervention at this stage? (open ended question)

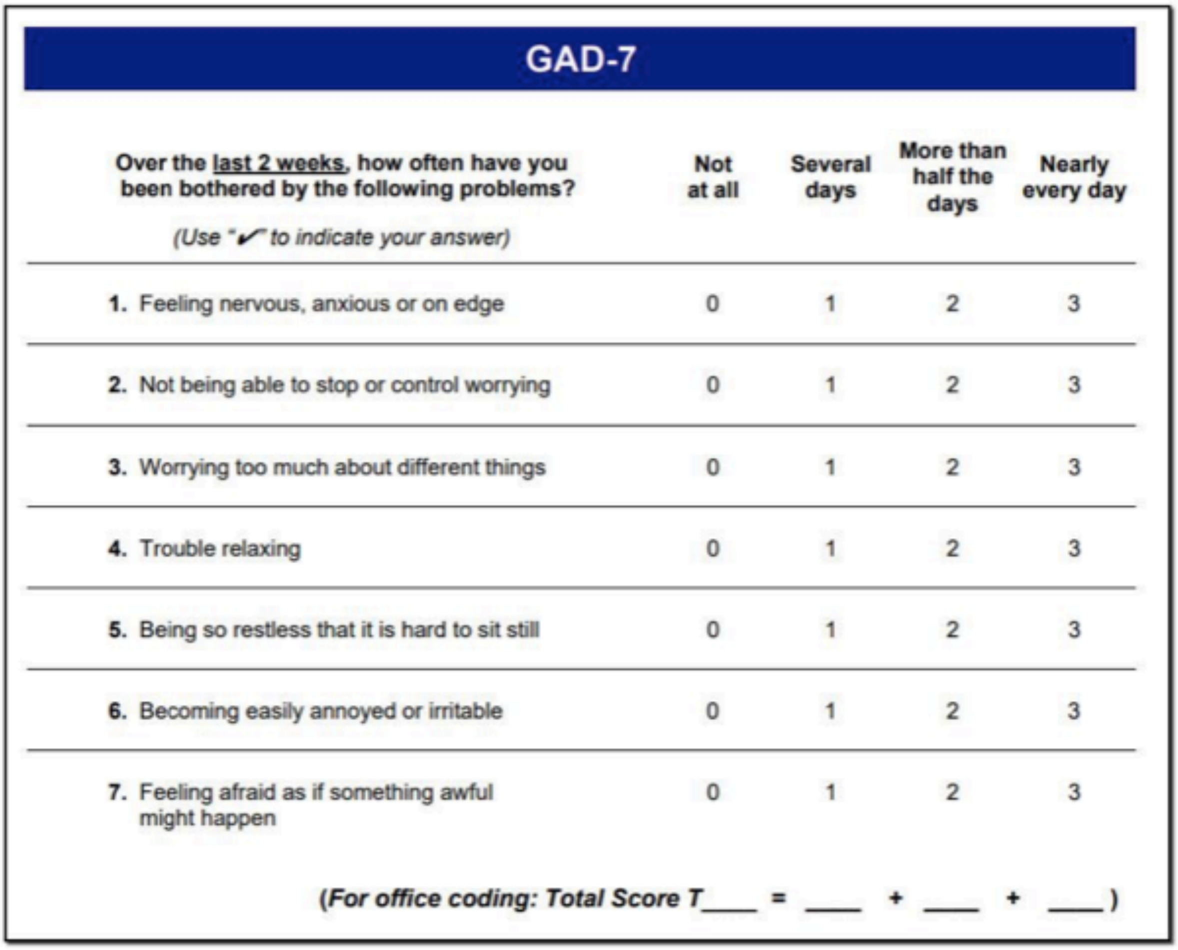
Thank you for your time and participation in the study.

Appendix M: Demographic Questionnaire

Please answer the following questions:

1. Full name:
2. Date of birth:
3. Gender:
4. Address including postcode and country:
5. Telephone number:
6. Email address:
7. Employment status:
    a. Full time, Part time, not employed, retired
8. Years of education:
9. Ethnicity: (open ended)
10. PH diagnosis
    a. Idiopathic PH, connective tissue disease, chronic thromboembolic PH, familial PH, congenital PH, other, not sure
11. PH functional class
    a. Class I, Class II, Class III, Class IV
12. How long have you had pulmonary hypertension?
13. Have you been prescribed medicaton to help you with anxiety in the last 12 months?
    a. Yes, No, Not sure
14. Have you received psychologu therapy to help with anxiety in the last 12 months?
    a. Yes, No, Not sure

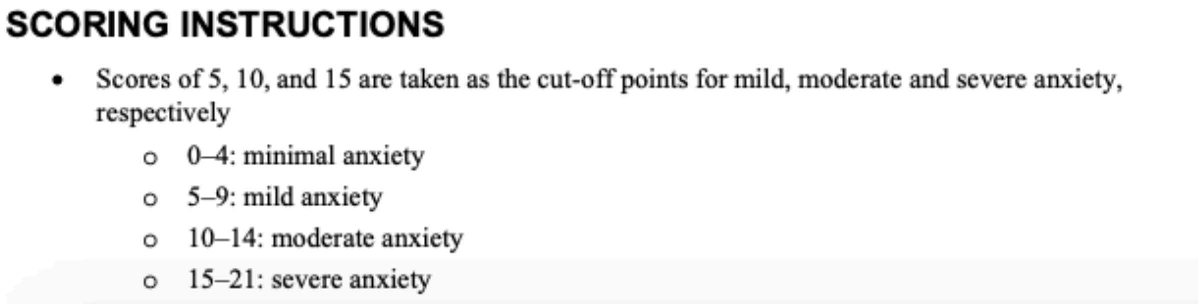
Appendix N: Generalised Anxiety Disorder 7 Questionnaire (GAD-7)



GAD-7

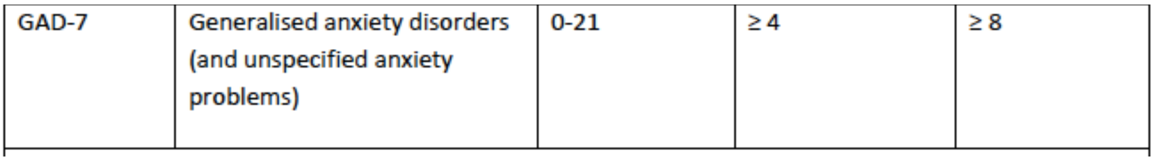
Over the last 2 weeks, how often have you been bothered by the following problems? (Use "✔" to indicate your answer)	Not at all	Several days	More than half the days	Nearly every day
1. Feeling nervous, anxious or on edge	0	1	2	3
2. Not being able to stop or control worrying	0	1	2	3
3. Worrying too much about different things	0	1	2	3
4. Trouble relaxing	0	1	2	3
5. Being so restless that it is hard to sit still	0	1	2	3
6. Becoming easily annoyed or irritable	0	1	2	3
7. Feeling afraid as if something awful might happen	0	1	2	3

(For office coding: Total Score T____ = ____ + ____ + ____)



SCORING INSTRUCTIONS

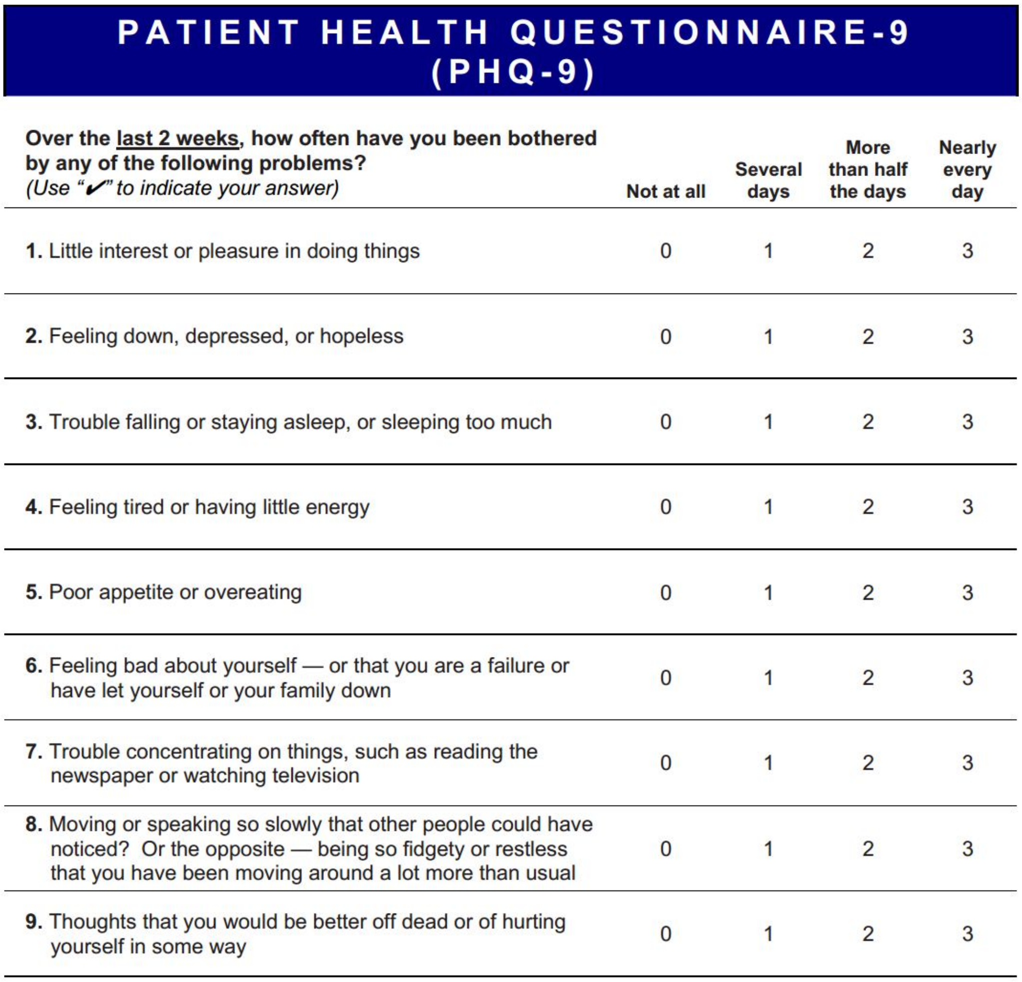
- Scores of 5, 10, and 15 are taken as the cut-off points for mild, moderate and severe anxiety, respectively
    - 0–4: minimal anxiety
    - 5–9: mild anxiety
    - 10–14: moderate anxiety
    - 15–21: severe anxiety



GAD-7	Generalised anxiety disorders (and unspecified anxiety problems)	0-21	≥ 4	≥ 8

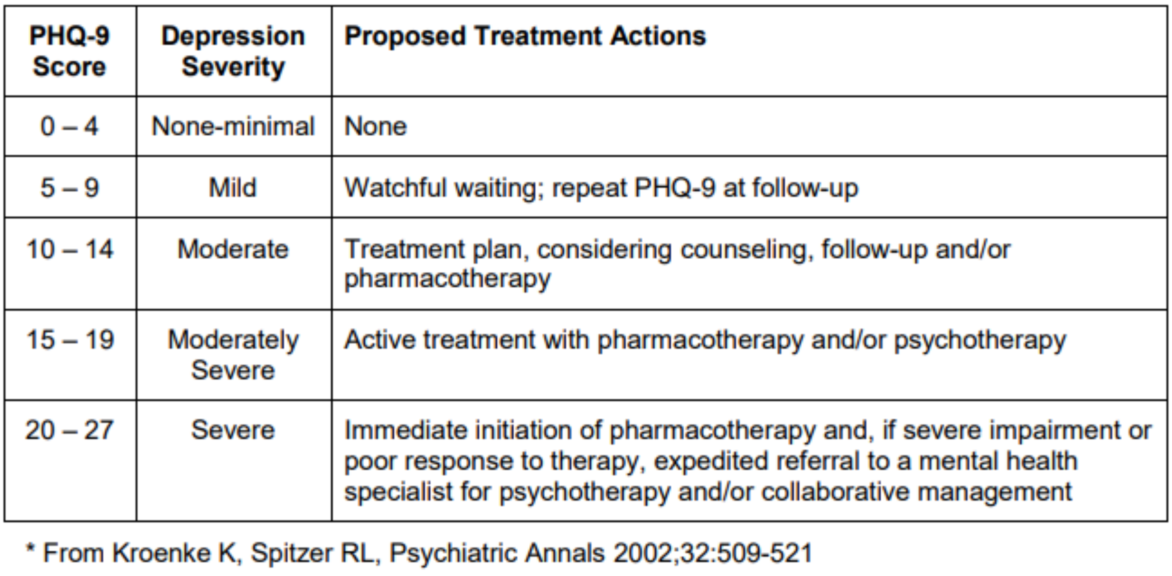
Clark, D., & Oates, M. (2014). *Improving Access to Psychological Therapies: Measuring improvement and recovery adult services (Version 2)*. http://www.oxfordahsn.org/wp-content/uploads/2015/11/measuring-recovery-2014.pdf

Appendix O: Patient Health Questionnaire 9 (PHQ-9)



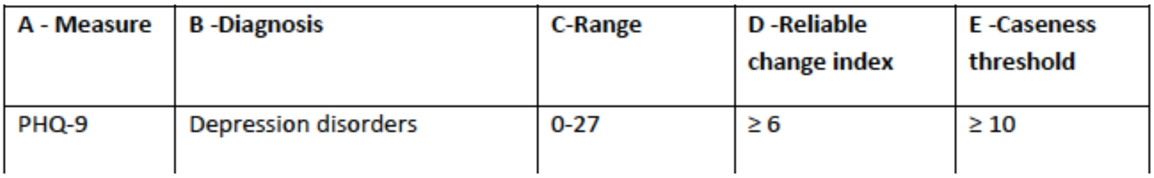
PATIENT HEALTH QUESTIONNAIRE-9 (PHQ-9)

Over the last 2 weeks, how often have you been bothered by any of the following problems? *(Use "✔" to indicate your answer)*	**Not at all**	**Several days**	**More than half the days**	**Nearly every day**
1. Little interest or pleasure in doing things	0	1	2	3
2. Feeling down, depressed, or hopeless	0	1	2	3
3. Trouble falling or staying asleep, or sleeping too much	0	1	2	3
4. Feeling tired or having little energy	0	1	2	3
5. Poor appetite or overeating	0	1	2	3
6. Feeling bad about yourself — or that you are a failure or have let yourself or your family down	0	1	2	3
7. Trouble concentrating on things, such as reading the newspaper or watching television	0	1	2	3
8. Moving or speaking so slowly that other people could have noticed? Or the opposite — being so fidgety or restless that you have been moving around a lot more than usual	0	1	2	3
9. Thoughts that you would be better off dead or of hurting yourself in some way	0	1	2	3



PHQ-9 Score	**Depression Severity**	**Proposed Treatment Actions**
0 – 4	None-minimal	None
5 – 9	Mild	Watchful waiting; repeat PHQ-9 at follow-up
10 – 14	Moderate	Treatment plan, considering counseling, follow-up and/or pharmacotherapy
15 – 19	Moderately Severe	Active treatment with pharmacotherapy and/or psychotherapy
20 – 27	Severe	Immediate initiation of pharmacotherapy and, if severe impairment or poor response to therapy, expedited referral to a mental health specialist for psychotherapy and/or collaborative management

* From Kroenke K, Spitzer RL, Psychiatric Annals 2002;32:509-521



A - Measure	B -Diagnosis	C-Range	D -Reliable change index	E -Caseness threshold
PHQ-9	Depression disorders	0-27	≥ 6	≥ 10

Clark, D., & Oates, M. (2014). *Improving Access to Psychological Therapies: Measuring improvement and recovery adult services (Version 2)*. http://www.oxfordahsn.org/wp-content/uploads/2015/11/measuring-recovery-2014.pdf

Appendix P: emPHasis10 – Health-Related Quality of Life Measure

emPHasis10

NHS/Hospital number:

Name:

Date of birth:

This questionnaire is designed to determine how pulmonary hypertension (PH) affects your life. Please answer every question by placing a tick over the ONE NUMBER that best describes your recent experience of living with PH.

For each item below, place a tick (✓) in the box that best describes your experience.

I am not frustrated by my breathlessness	0	1	2	3	4	5	I am very frustrated by my breathlessness
Being breathless never interrupts my conversations	0	1	2	3	4	5	Being breathless always interrupts my conversations
I do not need to rest during the day	0	1	2	3	4	5	I always need to rest during the day
I do not feel exhausted	0	1	2	3	4	5	I always feel exhausted
I have lots of energy	0	1	2	3	4	5	I have no energy at all
When I walk up one flight of stairs I am not breathless	0	1	2	3	4	5	When I walk up one flight of stairs I am very breathless
I am confident out in public places/crowds despite my PH	0	1	2	3	4	5	I am not confident at all in public places/crowds because of my PH
PH does not control my life	0	1	2	3	4	5	PH completely controls my life
I am independent	0	1	2	3	4	5	I am completely dependent
I never feel like a burden	0	1	2	3	4	5	I always feel like a burden

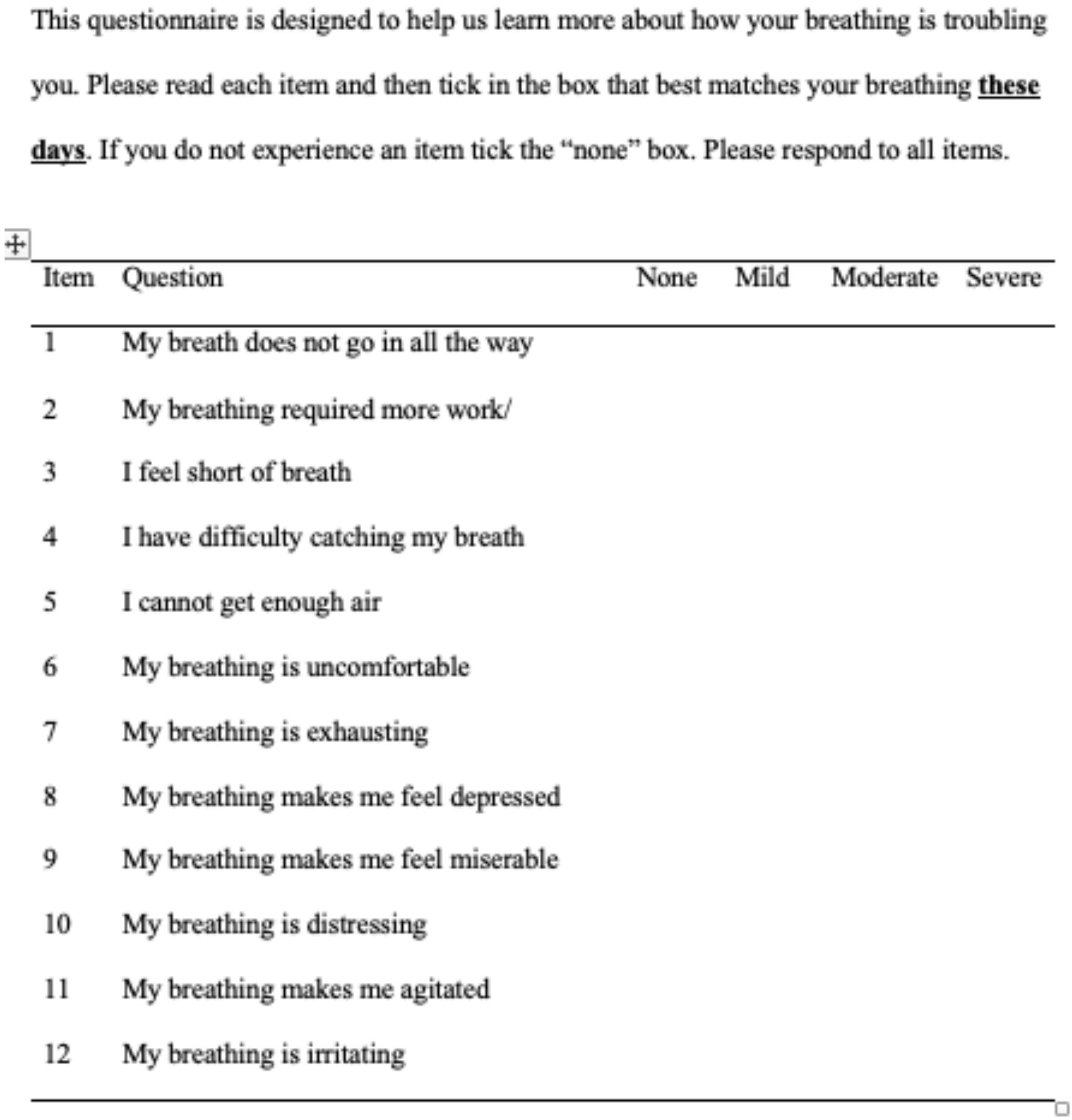
Total:

Date:

pha UK
pulmonary hypertension association

MANCHESTER
1824
The University of Manchester

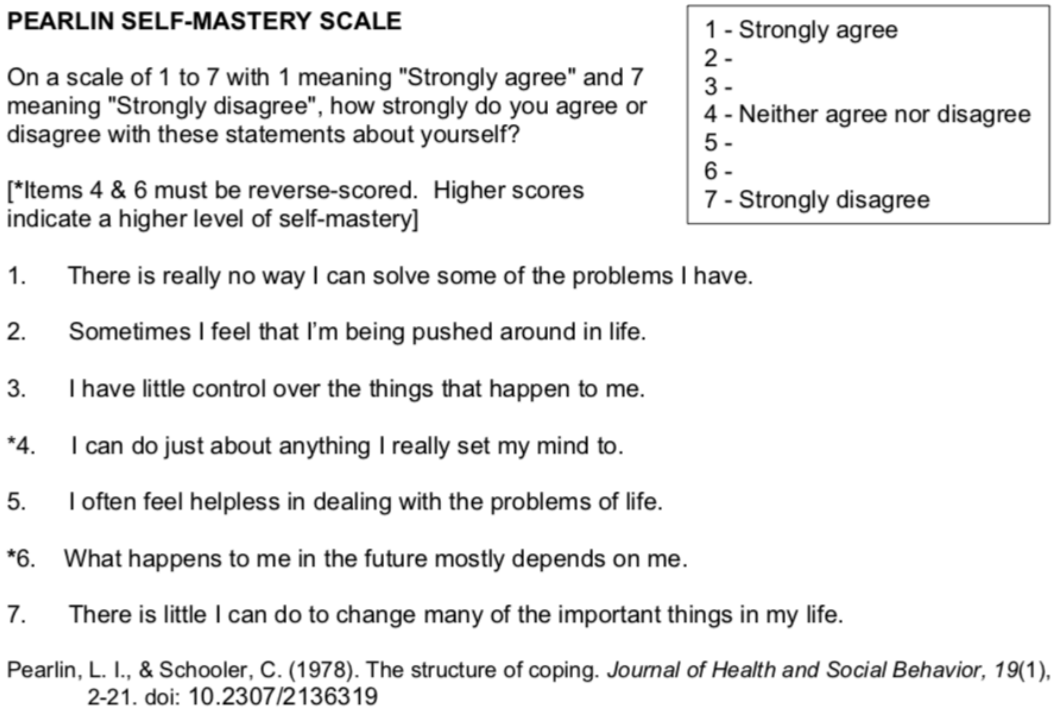
Appendix Q: Dyspnoea 12 (D12) questionnaire



This questionnaire is designed to help us learn more about how your breathing is troubling you. Please read each item and then tick in the box that best matches your breathing **<u>these days</u>**. If you do not experience an item tick the “none” box. Please respond to all items.

Item	Question	None	Mild	Moderate	Severe
1	My breath does not go in all the way				
2	My breathing required more work/				
3	I feel short of breath				
4	I have difficulty catching my breath				
5	I cannot get enough air				
6	My breathing is uncomfortable				
7	My breathing is exhausting				
8	My breathing makes me feel depressed				
9	My breathing makes me feel miserable				
10	My breathing is distressing				
11	My breathing makes me agitated				
12	My breathing is irritating				

Appendix R: Self-Mastery Scale



PEARLIN SELF-MASTERY SCALE

On a scale of 1 to 7 with 1 meaning "Strongly agree" and 7 meaning "Strongly disagree", how strongly do you agree or disagree with these statements about yourself?

[*Items 4 & 6 must be reverse-scored. Higher scores indicate a higher level of self-mastery]

1 - Strongly agree
2 -
3 -
4 - Neither agree nor disagree
5 -
6 -
7 - Strongly disagree

1. There is really no way I can solve some of the problems I have.

2. Sometimes I feel that I’m being pushed around in life.

3. I have little control over the things that happen to me.

*4. I can do just about anything I really set my mind to.

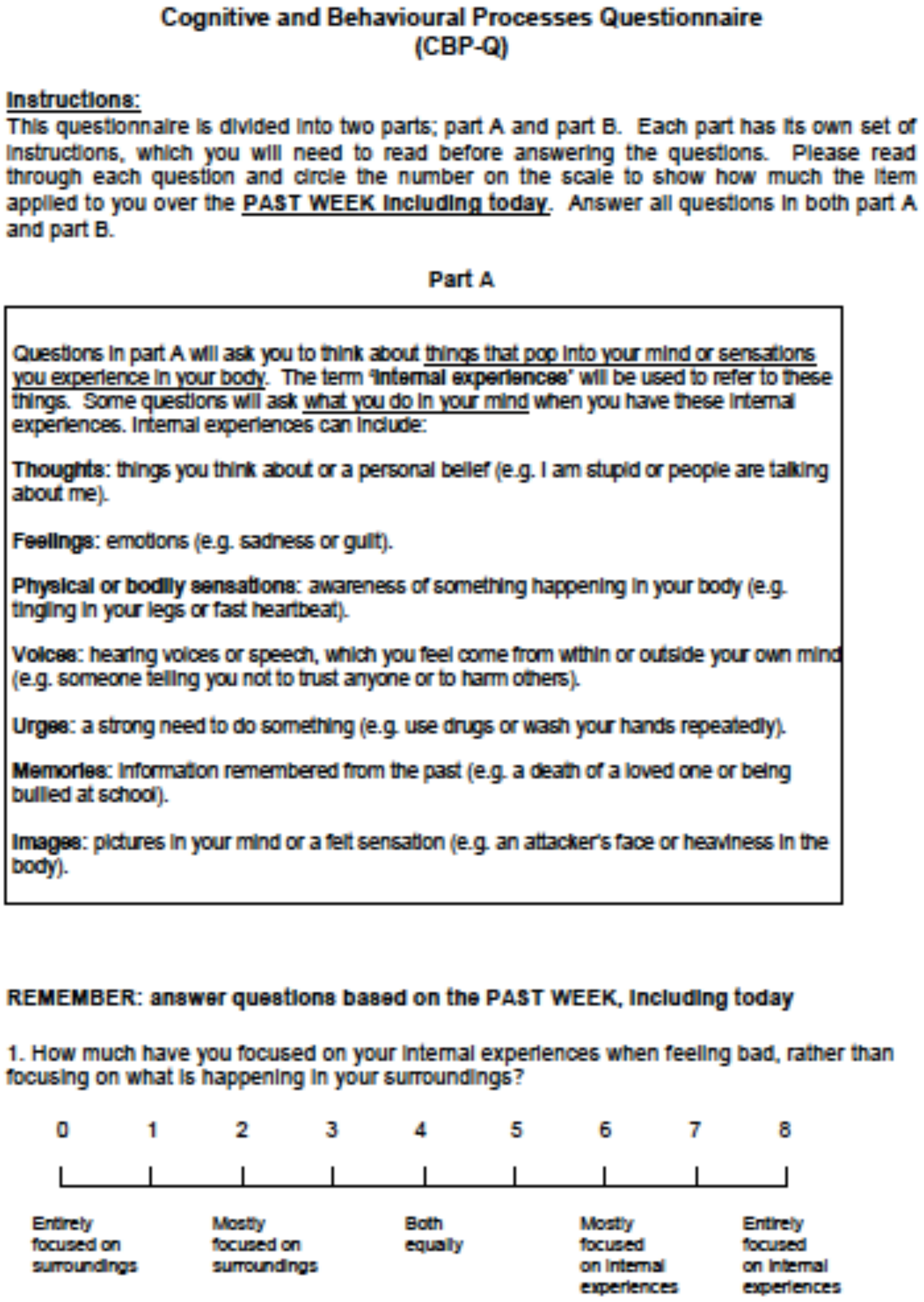
5. I often feel helpless in dealing with the problems of life.

*6. What happens to me in the future mostly depends on me.

7. There is little I can do to change many of the important things in my life.

Pearlin, L. I., & Schooler, C. (1978). The structure of coping. *Journal of Health and Social Behavior, 19*(1), 2-21. doi: 10.2307/2136319

Appendix S: Cognitive Behavioural Processes Questionnaire



Cognitive and Behavioural Processes Questionnaire
(CBP-Q)

Instructions:

This questionnaire is divided into two parts; part A and part B. Each part has its own set of instructions, which you will need to read before answering the questions. Please read through each question and circle the number on the scale to show how much the item applied to you over the **PAST WEEK including today**. Answer all questions in both part A and part B.

Part A

Questions in part A will ask you to think about things that pop into your mind or sensations you experience in your body. The term **'internal experiences'** will be used to refer to these things. Some questions will ask what you do in your mind when you have these internal experiences. Internal experiences can include:

Thoughts: things you think about or a personal belief (e.g. I am stupid or people are talking about me).

Feelings: emotions (e.g. sadness or guilt).

Physical or bodily sensations: awareness of something happening in your body (e.g. tingling in your legs or fast heartbeat).

Voices: hearing voices or speech, which you feel come from within or outside your own mind (e.g. someone telling you not to trust anyone or to harm others).

Urges: a strong need to do something (e.g. use drugs or wash your hands repeatedly).

Memories: information remembered from the past (e.g. a death of a loved one or being bullied at school).

Images: pictures in your mind or a felt sensation (e.g. an attacker's face or heaviness in the body).

REMEMBER: answer questions based on the PAST WEEK, including today

1. How much have you focused on your internal experiences when feeling bad, rather than focusing on what is happening in your surroundings?

0	1	2	3	4	5	6	7	8
Entirely focused on surroundings		Mostly focused on surroundings		Both equally		Mostly focused on internal experiences		Entirely focused on internal experiences

REMEMBER: answer questions based on the PAST WEEK, including today

2. How much have you tried to mentally avoid or get rid of unpleasant internal experiences, rather than just noticing them and letting them pass?

0	1	2	3	4	5	6	7	8
Always just noticed & let them pass		Mostly just noticed & let them pass		Both equally		Mostly avoided or tried to get rid of internal experiences		Always avoided or tried to get rid of internal experiences

3. How much have you tried to change or mentally control your internal experiences when feeling bad, rather than just noticing them and letting them pass?

0	1	2	3	4	5	6	7	8
Always just noticed & let them pass		Mostly just noticed & let them pass		Both equally		Mostly changed or mentally controlled		Always changed or mentally controlled

4. How much have you gone over and over past experiences when feeling bad, rather than doing the things that are important to you?

0	1	2	3	4	5	6	7	8
Always did what was important to me		Mostly did what was important to me		Both equally		Mostly gone over & over past experiences		Always gone over & over past experiences

5. How much have you worried about bad things that might happen in the future, rather than doing the things that are important to you?

0	1	2	3	4	5	6	7	8
Always did what was important to me		Mostly did what was important to me		Both equally		Mostly worried about bad things happening		Always worried about bad things happening

REMEMBER: answer questions based on the PAST WEEK, including today

6. How much have you judged yourself or your appearance to other people when feeling bad, rather than just noticing people around you?

0	1	2	3	4	5	6	7	8
Always just noticed people around me		Mostly just noticed people around me		Both equally		Mostly judged self or appearance to others		Always judged self or appearance to others

7. How much have you let your internal experiences rather than what you see and hear in the moment, guide what you do?

0	1	2	3	4	5	6	7	8
Entirely guided by that seen & heard in the moment		Mostly guided by that seen & heard in the moment		Both equally		Mostly guided by internal experiences		Entirely guided by internal experiences

8. How much have you analysed past events for answers when feeling bad, rather than doing the things that are important to you?

0	1	2	3	4	5	6	7	8
Always did what was important to me		Mostly did what was important to me		Both equally		Mostly analysed past event for answers		Always analysed past events for answers

Part B

Questions in part B will ask you to think about how you act when **'feeling bad'**.
For example:

You may pay more attention to what you think might be harmful or threatening in your surroundings (e.g. dirt or spiders).

You may avoid things that distress you (e.g. people or situations).

You may avoid dealing with actual problems (e.g. debt or conflict in a relationship).

You may start doing an activity too often to distract yourself (e.g. tidying or exercise).

You may do something to try to stop negative feelings (e.g. harm yourself or leave a situation).

REMEMBER: answer questions based on the PAST WEEK, including today

1. How much have you looked for possible harm or threats in your surroundings when feeling bad, rather than just noticing things around you?

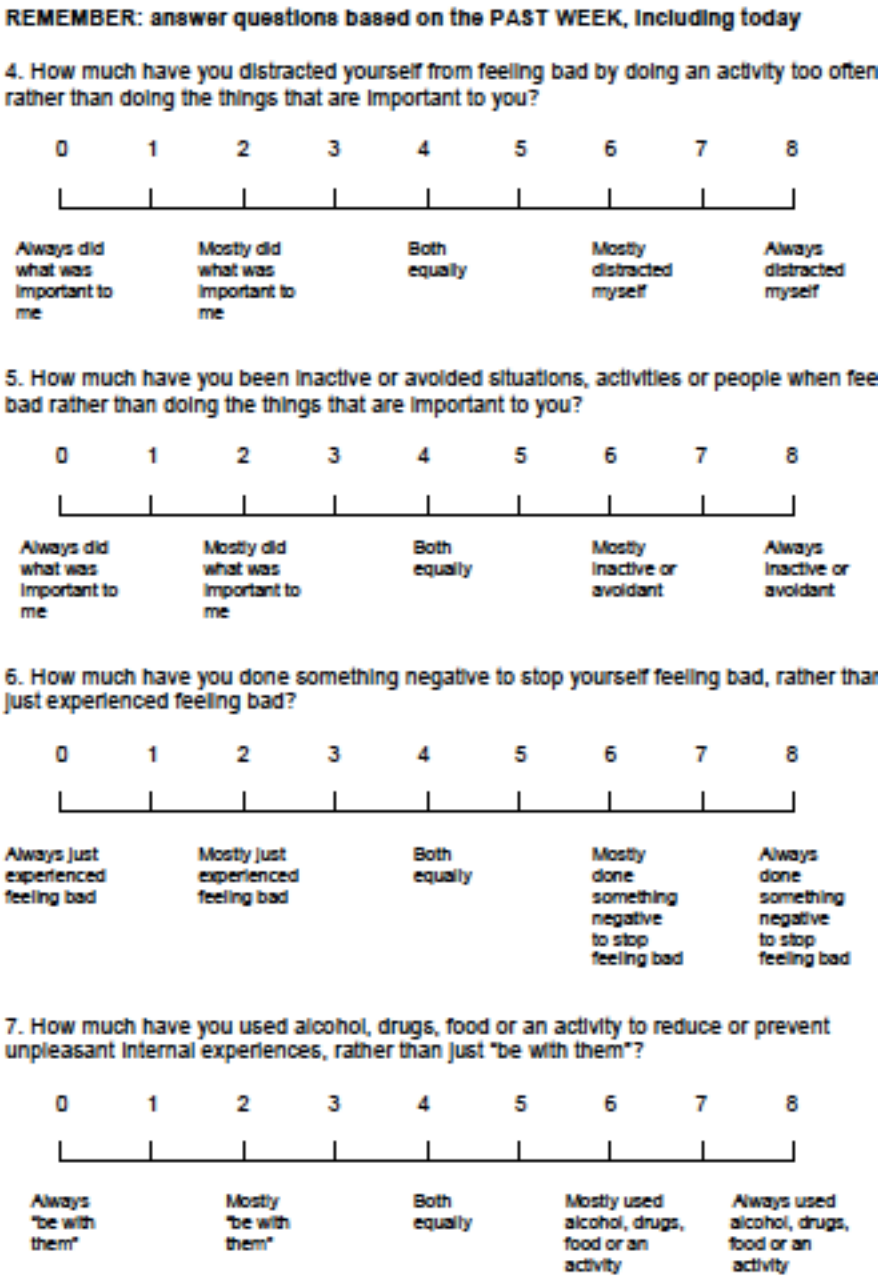
0	1	2	3	4	5	6	7	8
Always just noticed things around me		Mostly just noticed things around me		Both equally		Mostly looked for harm or threats		Always looked for harm or threats

2. How much have you looked for things in your surroundings to make you feel safe when feeling bad, rather than just noticing things around you?

0	1	2	3	4	5	6	7	8
Always just noticed things around me		Mostly just noticed things around me		Both equally		Mostly looked for things to make self feel safe		Always looked for things to make self feel safe

3. How much have you avoided dealing with an actual problem when feeling bad, rather than doing something to solve the problem?

0	1	2	3	4	5	6	7	8
Always tried to solve problem		Mostly tried to solve problem		Both equally		Mostly avoided dealing with problem		Always avoided dealing with problem



REMEMBER: answer questions based on the PAST WEEK, including today

4. How much have you distracted yourself from feeling bad by doing an activity too often rather than doing the things that are important to you?

0	1	2	3	4	5	6	7	8
Always did what was important to me		Mostly did what was important to me		Both equally		Mostly distracted myself		Always distracted myself

5. How much have you been inactive or avoided situations, activities or people when fee bad rather than doing the things that are important to you?

0	1	2	3	4	5	6	7	8
Always did what was important to me		Mostly did what was important to me		Both equally		Mostly inactive or avoidant		Always inactive or avoidant

6. How much have you done something negative to stop yourself feeling bad, rather tha just experienced feeling bad?

0	1	2	3	4	5	6	7	8
Always just experienced feeling bad		Mostly just experienced feeling bad		Both equally		Mostly done something negative to stop feeling bad		Always done something negative to stop feeling bad

7. How much have you used alcohol, drugs, food or an activity to reduce or prevent unpleasant internal experiences, rather than just "be with them"?

0	1	2	3	4	5	6	7	8
Always "be with them"		Mostly "be with them"		Both equally		Mostly used alcohol, drugs, food or an activity		Always used alcohol, drugs, food or an activity