

**Exercise rehabilitation programmes for pulmonary hypertension – a systematic review of intervention components and reporting quality**

MCGREGOR, Gordon, POWELL, Richard, FINNEGAN, Susanne, NICHOLS, Simon <<http://orcid.org/0000-0003-0377-6982>> and UNDERWOOD, Martin

Available from Sheffield Hallam University Research Archive (SHURA) at:

<http://shura.shu.ac.uk/22697/>

---

This document is the author deposited version. You are advised to consult the publisher's version if you wish to cite from it.

**Published version**

MCGREGOR, Gordon, POWELL, Richard, FINNEGAN, Susanne, NICHOLS, Simon and UNDERWOOD, Martin (2018). Exercise rehabilitation programmes for pulmonary hypertension – a systematic review of intervention components and reporting quality. *BMJ Open Sport & Exercise Medicine*, 4 (1).

---

**Copyright and re-use policy**

See <http://shura.shu.ac.uk/information.html>

**Exercise rehabilitation programmes for pulmonary hypertension – a systematic review of intervention components and reporting quality.**

Gordon McGregor<sup>1,2\*</sup>, Richard Powell<sup>1</sup>, Susanne Finnegan<sup>3</sup>, Simon Nichols<sup>4</sup>, Martin Underwood<sup>3</sup>

\*Corresponding Author

Department of Cardiopulmonary Rehabilitation, Centre for Exercise & Health, University Hospitals Coventry & Warwickshire NHS Trust, Coventry, UK

gordon.mcgregor@uhcw.nhs.uk

Tel: 02476 234 570

<sup>1</sup> Department of Cardiopulmonary Rehabilitation, Centre for Exercise & Health, University Hospital, Coventry, UK

<sup>2</sup> School of Health & Life Sciences, Coventry University, Coventry, UK

<sup>3</sup> Clinical Trials Unit, Warwick Medical School, University of Warwick, Coventry, UK

<sup>4</sup> Sheffield Hallam University, Sheffield, UK

Key words: Consensus on Exercise Reporting Template (CERT), exercise rehabilitation, pulmonary hypertension

Abstract word count: 250

Word count: 3924

## **Abstract 250**

**Objectives:** To identify the components, and assess the reporting quality, of exercise training interventions for people living with pulmonary hypertension.

**Design:** Systematic review with analysis of intervention reporting quality using the Consensus on Exercise Reporting Template (CERT).

**Data sources:** Eligible studies in the Cochrane Systematic Review of exercise-based rehabilitation for pulmonary hypertension, updated with a new search of relevant databases from 1<sup>st</sup> August 2016 to 15<sup>th</sup> January 2018.

**Eligibility criteria:** Peer reviewed journal articles of randomised and non-randomised controlled trials, and non-controlled prospective observational studies, investigating dynamic exercise training interventions in adult humans with diagnosed pulmonary hypertension, reporting on at least one physiological and/or psychosocial outcome.

**Results:** Interventions typically involved cycle ergometry and walking. They were delivered as three-week in-patient, or out-patient and/or home-based programmes, lasting for four to 15 weeks. Components relating specifically to exercise prescription were described satisfactorily and in more detail than motivational/behavioural change strategies, adherence and fidelity. Mean Consensus on Exercise Reporting Template (CERT) score was 13.1 (range 8 to 17) out of a possible maximum score of 19. No studies fully reported every aspect of an exercise intervention to the standard recommended by CERT.

**Summary/conclusion:** Considerable variability was evident in the components and reporting quality of interventions for exercise rehabilitation studies in pulmonary hypertension. Interventional studies using exercise training should pay greater attention to describing motivational/ behavioural change strategies, adherence and fidelity. Detailed description of these parameters is essential for the safe and effective replication of exercise rehabilitation interventions for pulmonary hypertension in clinical practice.

**Trial registration number:** This systematic review is registered with PROSPERO 2018 CRD42018085558

## **SUMMARY**

- The reporting of exercise intervention components in clinical populations is often inadequate
- The Consensus on Exercise Reporting Template (CERT) is a useful tool with which exercise intervention reporting can be standardised
- Exercise prescription for people living with pulmonary hypertension is relatively well reported although no studies reported adequately on all CERT criteria.
- Motivational strategies, exercise adherence, and intervention fidelity are poorly reported in pulmonary hypertension studies.

## INTRODUCTION

Pulmonary hypertension is a debilitating long-term condition characterised by severe exercise intolerance<sup>1</sup>. Pulmonary arterial pressure is abnormally raised due to dysfunctional endothelial cells and vascular smooth muscle, leading to maladaptive pulmonary vascular remodelling and increased right ventricular afterload<sup>2,3</sup>. Pulmonary and cardiovascular haemodynamics are progressively compromised, often during minimal physical exertion<sup>4,5</sup>. Consequently, exertional dyspnoea, fatigue and syncope are the most common symptoms, impacting profoundly on quality of life (QoL), morbidity and mortality<sup>6</sup>. Guidance from the World Symposium on Pulmonary Hypertension<sup>7</sup> identified five distinct subgroups: Group 1 - pulmonary arterial hypertension (PAH), Group 2 - pulmonary hypertension due to left heart disease, Group 3 – pulmonary hypertension due to lung diseases or hypoxia, or both, Group 4 - chronic thromboembolic pulmonary hypertension (CTEPH), Group 5 – pulmonary hypertension with unclear multifactorial mechanisms.

There are many similarities between pulmonary hypertension and conditions like chronic obstructive pulmonary disease and chronic heart failure; indeed, they often co-exist<sup>6</sup>. Exercise therapy improves exercise tolerance and QoL for people living with these conditions, therefore, it is plausible, that it will also help people living with pulmonary hypertension. Specific exercise therapy guidelines to maximise physiological and psychosocial gains, whilst ensuring safe participation, are available for some clinical conditions; for example, post-myocardial infarction<sup>8</sup> or chronic obstructive pulmonary disease<sup>9</sup>. Such robust recommendations are not available for pulmonary hypertension. Nevertheless, a 2017 Cochrane Review and meta-analysis found a meaningful short-term improvement in exercise capacity from participation in partly supervised, exercise-based, rehabilitation for people with moderately severe pulmonary hypertension; albeit from low

quality evidence <sup>10</sup>. Quality of life (SF-36) also improved, but the authors concluded that the magnitude of change may be insufficient to be clinically important.

To develop appropriate exercise training guidance for people with pulmonary hypertension, an understanding of exercise intervention components is essential. Systematic reviews commonly pool data from heterogeneous exercise training protocols, which may dilute the relevance and applicability of findings. Recent Cochrane Reviews of exercise-based rehabilitation in cardiac and pulmonary disease, whilst indicating the potential for benefit, were limited by the quality and diversity of exercise training interventions <sup>11 12</sup>. Replication of safe and effective clinical exercise therapy is reliant on access to detailed description of exercise intervention components. Information on important aspects of an exercise intervention; e.g. setting, facility, equipment, frequency, intensity, duration, modality, adherence, fidelity and safety, is required for subsequent implementation. Such reporting is often poor which may question the validity of published exercise protocols, and prevent safe and effective adoption of successful interventions. The Consensus on Exercise Reporting Template (CERT), recently developed and endorsed by an international panel of experts, allows standardised appraisal of published exercise rehabilitation interventions <sup>13 14</sup>. This document provides guidance on the minimum reporting standard considered necessary to allow replication of exercise interventions. Not only can this tool be used to retrospectively assess reporting quality, it can also help with protocol development. Whilst it is not intended to assess the scientific and methodological quality of studies *per se*, it should facilitate standardised reporting of exercise interventions and aid translation of research findings into everyday clinical practice

The aim of this review was to assess both the components, and reporting quality of studies investigating exercise-based rehabilitation in pulmonary hypertension. To ensure a standardised appraisal, we used the Consensus on Exercise Reporting Template (CERT). Collating the components of published exercise interventions also allowed us to make inferences as to future clinical research priorities in the field of exercise programming and prescription for people with pulmonary hypertension.

## **METHODS**

We first examined the included and excluded studies of the 2017 Cochrane Systematic Review of exercise-based rehabilitation in pulmonary hypertension<sup>10</sup>. We replicated this search, with some amendments, searching for original research published in academic journals as full-length, independently peer reviewed, randomised and non-randomised controlled trials, and non-controlled prospective observational studies investigating exercise training interventions in adults with pulmonary hypertension. Inspiratory/respiratory muscle training was only included where it formed part of an intervention which also employed dynamic exercise training. The included studies must have tested the effect of a dynamic exercise training (aerobic or strength) intervention on at least one physiological and/or psychosocial parameter. We did not place any restriction on publication language or study design, other than excluding case reports. We also excluded abstracts and conference proceedings as they do not qualify as full-length original articles, which the CERT checklist was designed to appraise. Shorter communications, such as abstracts, do not contain sufficient detail to enable scoring with CERT. The identified literature was hand-searched for additional publications. Subsequently, we updated the search to cover the period 1<sup>st</sup> August 2016 to 15<sup>th</sup> January 2018 using the following databases. (Appendix 1):

- The Cochrane Airways Register of Trials
- Cochrane Central Register of Controlled Trials (CENTRAL) via the Cochrane Register of Studies
- Medline (Ovid)
- Embase (Ovid)
- Physiotherapy Evidence Database (PEDro)

Two reviewers (GM and RP) determined eligibility by independently screening titles and abstracts. Where discrepancy relating to study inclusion existed, a consensus was reached through discussion and consultation with a third reviewer (SF). All eligible full-text articles were acquired to allow assessment with the Consensus on Exercise Reporting Template (CERT). Two reviewers (GM and RP) extracted all intervention component data to summarise each study in a standardised fashion ( 1).

Two assessors, (GM and SF) independently reviewed each extracted manuscript using the 16-item CERT checklist (Appendix 2) <sup>13</sup>. Each item was scored ‘1’ (adequately reported) or ‘0’ (not adequately reported), and a score out of a total of 19 was calculated. Where studies cited methodology from previous work, information from the cited paper was also included in the score. We referred to the CERT examples of ‘good reporting’ to ensure scoring was standardised. Together, both assessors reviewed each of the 16 items for each of the extracted manuscripts to compare scores. Discrepancies were resolved through discussion between the two reviewers and further reference to the CERT Explanation and Elaboration Statement <sup>13</sup>. The review was conducted in accordance with PRISMA guidelines, and was registered with PROSPERO (2018 CRD42018085558).



**Figure 1.** PRISMA flow diagram.

## RESULTS

We included 19 studies, six of which were RCTs<sup>15-20</sup>, three non-randomised controlled trials<sup>21-23</sup>, and 10 non-controlled, prospective observational studies<sup>24-33</sup> (Supplementary file 1). Mean sample size was 35 (range 5 to 183) participants. Pulmonary hypertension aetiology classification was varied: seven studies exclusively enrolled group 1<sup>20 23 26 28 29 31 32</sup>, none groups two and three only, three group four only<sup>22 24 30</sup>, two group five only<sup>25 27</sup>, four groups one and four<sup>16 18 19 21</sup>, and three were mixed<sup>15 17 33</sup>. Intervention content was heterogeneous. Two studies (2/19; 11%) prescribed dynamic exercise training only<sup>15 20</sup>, five studies (5/19; 26%) dynamic exercise training in combination with resistance training<sup>21-23 25 27</sup>, and twelve studies (12/19; 63%) a multi-modality intervention consisting of dynamic exercise training, resistance training and respiratory muscle training<sup>16-19 24 26 28-33</sup>. All studies delivered an intervention that was supervised in an inpatient or outpatient setting, with 10 incorporating an inpatient phase. Eleven studies (11/19; 58%) combined unsupervised home-based exercise training with supervised sessions, or used home-based as a continuation beyond centre-based exercise<sup>16 18 19 22 24 28-33</sup>. Most studies (16/19; 84%) used cycle ergometry as the main modality of dynamic exercise, delivered alone, or in combination with walking and/or step climbing. Three studies used walking as the predominant modality of training<sup>15 20 24</sup>. There was considerable variation in the duration of the exercise intervention. Programmes lasted four<sup>26</sup>, eight<sup>17</sup>, ten<sup>15 20</sup>, twelve<sup>21-25 27</sup> or fifteen<sup>16 18 19 28-33</sup> weeks. Session duration, frequency and intensity were well described in all studies. Most studies detailed the methods of progression, and the use of heart rate, oxygen saturation and subjective measures (perceived dyspnoea and physical exertion) to adjust training intensity. All but four studies, which used exercise training as a stand-alone intervention<sup>17 19 25 27</sup>, adopted a multi-

disciplinary approach, combining exercise training with additional educational and psychosocial components (15/19; 79%).

CERT scores ranged from 8 to 17 (mean 13.1) out of a possible maximum score of 19 points (Figure 2). No studies adequately described all aspects of exercise interventions in accordance with CERT (figure 3). We detail the standard of reporting for each criteria below.

***Question 1: Detailed description of the type of exercise equipment***

Whilst it was common to describe the mode of activity e.g. cycle ergometry or walking, only one study included the make and model of the exercise equipment used<sup>17</sup>.

***Question 2: Detailed description of the qualifications, expertise, and/or training***

Fifteen studies (79%) identified the specific discipline delivering the exercise intervention e.g. physiotherapist, and the presence/nature of medical supervision e.g. respiratory physician. Other than stating, ‘experienced’ or ‘specialist’, only three studies described any additional post-graduate training or relevant continued professional development, undertaken by staff, or the extent of the experience required.

***Question 3: Describe whether exercises are performed individually or in a group***

This was poorly described, but in most studies the description of the intervention was sufficient to imply the format, hence 14 studies (74%) met this criterion.

***Question 4: Describe whether exercises are supervised or unsupervised; how they are delivered***

Eighteen studies (94%) reported the presence or absence of supervision but limited detail was provided. As these studies reported face-to-face supervision, there was less need to provide additional detail.

***Question 5: Detailed description of how adherence to exercise is measured and reported***

The methodology for measuring adherence was not well described, as was the case for the level of adherence achieved in the studies. Only six studies (32%) reported this adequately, by first describing *a-priori* how adherence would be assessed, and second, confirming the level of participant adherence in absolute or relative terms.

***Question 6: Detailed description of motivation strategies***

Only one study provided adequate information relating to any motivational strategies and behavioural change interventions used to complement exercise training<sup>26</sup>. In this study, whilst not exhaustive, sufficient detail was included to allow informed assessment of effective motivational strategies in this population.

***Question 7a: Detailed description of the decision rule(s) for determining exercise progression, 7b: Detailed description of how the exercise program was progressed***

The decision rule(s) for progression were defined in 17 studies (90%). Nevertheless, six studies (32%) did not provide sufficient information regarding how exercise programmes were actually progressed in practice.

***Question 8: Detailed description of each exercise to enable replication***

All programmes consisted of walking and/or cycling, requiring very little detailed instruction. Where additional exercise such as resistance training, respiratory muscle training or yoga was prescribed, this was generally well defined. Information regarding dumbbell weight (kg), lifting technique and/or a specific respiratory muscle training protocol were often thorough and detailed.

***Question 9: Detailed description of any home programme component***

Home exercise programmes were secondary components of centre-based interventions in all but one study which primarily tested home-based rehabilitation in pulmonary hypertension, with a secondary centre-based component<sup>24</sup>. As such, reporting was generally of a low standard.

***Question 10: Describe whether there are any non-exercise components***

Overall, a clear description of non-exercise components was provided. Sixteen studies (84%) contained a structured educational, physical and psychological disease self-management programme alongside the exercise intervention.

***Question 11: Describe the type and number of adverse events that occurred during Exercise***

Sixteen studies (84%) described this aspect well, using pre-defined criteria, and relating adverse events to clinical outcome and the impact on participant completion of the exercise intervention. Most also made reference to whether or not adverse events were related to the intervention itself.

***Question 12: Describe the setting in which the exercises are performed***

Clearly and concisely reported in 18 studies (95%).

***Question 13: Detailed description of the exercise intervention, 14a: Describe whether the exercises are generic (one size fits all) or tailored, 14b: Detailed description of how exercises are tailored to the individual***

All studies provided adequate detail of the exercise intervention. Within this criterion there was considerable variability in the level of detail, but all were deemed adequate in line with CERT recommendations. The intended method of exercise tailoring (individualisation), was also well described in all studies.

***Question 15: Describe the decision rule for determining the starting level***

Primarily from cardiopulmonary exercise testing (a common and informative test in the pulmonary hypertension population), 18 studies (95%) indicated methods for initial exercise prescription.

***Question 16a: Describe how adherence or fidelity is assessed/measured, 16b: Describe the extent to which the intervention was delivered as planned.***

No studies adequately reported how therapist adherence to appropriate protocol delivery would be measured, however, 12 studies (63%) referred to issues experienced by therapists which in practice could have led to sub-optimal intervention deliver

**Supplementary file 1.** Summary of study design and exercise intervention components

**Figure 2.** Consensus on Exercise Reporting Template (CERT) score by paper

**Figure 3.** Quality of exercise intervention reporting as assessed with the Consensus on Exercise Reporting Template (CERT).

## **DISCUSSION**

Using the Consensus on Exercise Reporting Template (CERT), we assessed the quality of exercise intervention reporting in 19 studies investigating the effect of exercise-based rehabilitation in people with pulmonary hypertension. Our main findings were 1) exercise prescription components in pulmonary hypertension rehabilitation were, overall, relatively well defined, and 2) motivational and behavioural change strategies, adherence and intervention fidelity were inadequately described. Formal analysis of data relating to efficacy, safety or clinical outcomes was not within the scope of this review. However, our appraisal of the literature did allow us to make broad recommendations for future clinical research priorities investigating exercise-based rehabilitation for people with pulmonary hypertension.

A striking observation was that 10/19 studies reported a one to three-week inpatient rehabilitation period prior to an outpatient and/or home based programme. Due to differences in health care provision, this is unlikely to be a plausible treatment option for most people with pulmonary hypertension. As an alternative, outpatient programmes provided at local facilities, offering an hour of exercise, two to three times weekly, are likely to be more feasible. However, the efficacy of this approach is yet to be confirmed. It is reassuring that no studies reported serious adverse events such as death.

The complexity of pulmonary hypertension and its management, and the potential for harm to result from exercise training, means that researchers and clinicians wishing to replicate published pulmonary hypertension rehabilitation programs, require a detailed description of every aspect of intervention delivery. In the translation of robust research findings to clinical practice, practitioners and healthcare commissioners need assurance that comprehensive replication of trial protocols is possible. For this to happen, there is a need for standardised and detailed reporting of exercise intervention components. Currently, throughout the clinical exercise literature, inadequate reporting is the norm.

All studies adequately described the intended protocol for frequency, intensity, time, type (FITT), progression, individualisation and starting level. This is a surprising finding given the results of other reviews. A quality assessment of exercise training principles in breast cancer survivors, whilst not using CERT, identified that only 29% of studies appropriately applied and explained exercise progression, 67% starting values, and 38% physiological overload<sup>34</sup>. Likewise, an appraisal of exercise intervention quality in sub-acute and chronic stroke survivors identified only one of 37 RCTs which adequately described all principles of physical exercise training<sup>35</sup>. Both reviews concluded that incomplete and inconsistent

reporting of exercise prescription components would prevent the utility and reproducibility of exercise interventions in these populations. The conflicting finding of adequate exercise intervention description in pulmonary hypertension studies may well reflect the explicit need for highly individualised prescription and clinical safety in pulmonary hypertension patients with a potentially high risk of exercise induced complications.

Whilst exercise prescription reporting was relatively well defined in the included studies, other components, also considered important, were not. For example, psychological and motivational strategies were poorly described in all but one of 19 studies. Most studies provided a brief overview of patient educational programmes covering disease specific pathophysiology and management by way of generalised group information sessions. Good compliance and adherence to exercise is primarily achieved through effective behavioural change interventions. Regular goal setting combined with motivational interviewing, patient activation, exercise diaries, positive feedback and reinforcement, 'patient contracts', and theoretical models, are known to enhance short and long-term exercise adherence<sup>36-38</sup>. These strategies are perhaps most important in clinical populations in which disease specific symptoms and psychological burden commonly prevent participation in physical activity.

In people living with pulmonary hypertension, exercise can be a very uncomfortable experience, which reinforces negative perceptions and fear of harm. Breathlessness on exertion naturally leads to short and longer-term anxiety and depression, resulting in exercise avoidance and an inability to undertake even the simplest of physical tasks. Most of the reviewed studies incorporated management of breathlessness, but there was no detail as to how this would form part of a motivational, behavioural change strategy to enhance

compliance and adherence. Clear and detailed reporting of this aspect of exercise programming is essential, and is a major weakness of the identified literature.

The methodology for recording and reporting adherence was identified as inadequate in this review. In approximately two-thirds of studies, it was not possible to determine the strategy for measuring adherence, and/or the actual level of adherence achieved. As such, a sufficient understanding of the volume or minimum dose of exercise which may lead to benefit in pulmonary hypertension was not attainable. This is a critical flaw in these studies. Equally as important as the demonstration of physiological and psychosocial efficacy, is the ability to determine the appropriate dose of exercise required to facilitate meaningful change. This is essential knowledge for practitioners wishing to assess the feasibility of exercise training for people with pulmonary hypertension in clinical practice. Adherence, which has numerous confounding factors including patient motivation, can be recorded accurately as absolute or relative attendance, frequency, intensity and duration. It is unclear why this would not be well defined, as it is simple to monitor, particularly in supervised programs. To ensure adequate information in this area, transparency of reporting should become the norm, regardless of the methodological shortcomings it may expose.

Only one study reported any fidelity strategies. Twelve (63%) reported how accurately and successfully interventions were delivered in line with their protocols. The strategies employed to ensure that practitioners deliver interventions as per the protocol, and the level of precision with which they were delivered, are both critical factors to confirm adequate training stimulus and individualisation. Fidelity is an important component of protocol development and implementation, and should be considered a reporting priority in future studies.



Seventeen studies (90%) were concise and accurate in reporting the rule(s) for determining exercise progression. Methods for monitoring the intensity of exercise, and participant response, were presented in considerable detail, in addition to providing an understanding of how these responses would translate to the progression of exercise prescription. However, implementation of intervention progression in practice was incompletely defined in a third of studies, consistent with the literature in other clinical conditions<sup>34 39 40</sup>. This practice may relate to poor recording, reluctance to report inefficiencies, or a lack of understanding of the importance of this parameter. Regardless, as with adherence and fidelity, this information supports the successful replication of interventions. Lack of exercise progression can lead to sub-optimal exercise prescription and may negatively impact outcomes. Awareness of participant tolerance, and the best strategies by which exercise can be progressed, are vital for safe and effective delivery.

As a general observation, even those parameters deemed to be adequately reported as per CERT, may benefit from additional detail. Whilst qualifications/expertise, and supervision were adequately described in 79% and 94% of studies respectively, reporting in both categories could be strengthened. Specific reference to qualifications, expertise and experience, rather than simply detailing staff specialty (with implied knowledge and competence) would be advantageous. Likewise, more thorough explanation of the nature and level of supervision would be beneficial, particularly in cases where face-to-face supervision was limited i.e. home programmes. Three areas in which reporting was considered extensive and appropriate were 1) additional exercise components e.g. respiratory muscle training, resistance training, yoga, 2) non-exercise components such as disease specific education, and 3) adverse events.

Future research should prioritise the assessment of pulmonary hypertension exercise training interventions in the context of clinical service models currently known to be physiologically/psychosocially beneficial and cost-effective in other diseases. It may be possible to seamlessly integrate patients with all sub-groups of pulmonary hypertension into existing cardiovascular and pulmonary rehabilitation programmes. Trials should ensure provision of additional expert supervision to manage the potential risk of exercise induced complications in this population<sup>41 42</sup>.

## **STRENGTHS AND LIMITATIONS**

We conducted a systematic literature search, and objectively summarised results using the Consensus on Exercise Reporting Template (CERT). Our search strategy was inclusive, incorporating not only RCTs, but also non-randomised controlled studies and prospective observational studies. We believe the findings have relevance to the clinical exercise community, both academics and clinicians. They also advance the knowledge of exercise prescription and intervention reporting. However, some limitations should be acknowledged. Whilst it was possible to assess the quality of intervention reporting, and recommend future research priorities, we did not formally assess the efficacy of exercise interventions in pulmonary hypertension, nor the methodological quality of the studies included. Further, our conclusions are primarily based on group 1 pulmonary arterial hypertension, and mostly reflect exercise provision with an in-patient component.

## **CONCLUSION**

We did not find any studies that adequately reported all aspects of exercise training interventions in pulmonary hypertension. We did, however, identify some areas in which

sufficient information was provided. Exercise prescription was often detailed and well reported, as were non-exercise components and adverse events. Conversely, adherence, fidelity and motivational strategies were poorly described. We encourage the detailed reporting of interventions in future studies, with particular attention to these parameters. The adoption of exercise rehabilitation for pulmonary hypertension may be unnecessarily restricted due to poor reporting. To allow fuller descriptions of interventions, we recommend the publication of trial protocols or intervention development papers, and the use of supplementary online material. Much of the current data are based on a three-week residential programme in relatively fit people with an average baseline six-minute walking distance of over 400 m. Thus, whilst it appears that exercise training may be beneficial, there are presently insufficient data to allow practitioners to universally replicate effective exercise based interventions for people living with pulmonary hypertension.

**Competing interests:**

MU has received multiple research grants as chief investigator or co-investigator from the National Institute of Health Research and Arthritis Research UK. He is a shareholder and director in Clinvivo Ltd. He is co-investigator on an NIHR funded trial in which ORTHOSPACE Ltd is providing training and surgical implants. He is part of an academic partnership involving Serco Ltd. GM is a director of Atrium Health Ltd.

**Contributorship:** MU and GM were jointly responsible for the concept of the review. All authors jointly developed the protocol. RP and GM conducted the literature search, and screened and assessed the articles. GM and SF undertook intervention reporting assessment using CERT. GM drafted the manuscript. All authors approved the final version of the manuscript.

**Acknowledgements:** none

**Funding:** This research received no specific grant from any funding agency in the public, commercial or not-for-profit sectors

## References

1. Kiely DG, Elliot CA, Sabroe I, et al. Pulmonary hypertension: diagnosis and management. *BMJ* 2013;346:f2028. doi: 10.1136/bmj.f2028
2. Howard LS. Prognostic factors in pulmonary arterial hypertension: assessing the course of the disease. *Eur Respir Rev* 2011;20(122):236-42. doi: 10.1183/09059180.00006711
3. Babu AS, Arena R, Morris NR. Evidence on Exercise Training in Pulmonary Hypertension. *Adv Exp Med Biol* 2017;1000:153-72. doi: 10.1007/978-981-10-4304-8\_10
4. Arena R, Lavie CJ, Borghi-Silva A, et al. Exercise Training in Group 2 Pulmonary Hypertension: Which Intensity and What Modality. *Prog Cardiovasc Dis* 2016;59(1):87-94. doi: 10.1016/j.pcad.2015.11.005
5. Tran DL, Lau EMT, Celermajer DS, et al. Pathophysiology of exercise intolerance in pulmonary arterial hypertension. *Respirology* 2018;23(2):148-59. doi: 10.1111/resp.13141
6. Guazzi M, Naeije R. Pulmonary Hypertension in Heart Failure: Pathophysiology, Pathobiology, and Emerging Clinical Perspectives. *J Am Coll Cardiol* 2017;69(13):1718-34. doi: 10.1016/j.jacc.2017.01.051
7. Simonneau G, Gatzoulis MA, Adatia I, et al. Updated clinical classification of pulmonary hypertension. *J Am Coll Cardiol* 2013;62(25 Suppl):D34-41. doi: 10.1016/j.jacc.2013.10.029
8. ACPICR. Standards for physical activity and exercise in the cardiac population. [http://acpicr.com/sites/default/files/ACPICR\\_Standards\\_2015\\_0.pdf](http://acpicr.com/sites/default/files/ACPICR_Standards_2015_0.pdf),. 2015
9. Bolton CE, Bevan-Smith EF, Blakey JD, et al. British Thoracic Society guideline on pulmonary rehabilitation in adults. *Thorax* 2013;68 Suppl 2:ii1-30. doi: 10.1136/thoraxjnl-2013-203808
10. Morris NR, Kermeen FD, Holland AE. Exercise-based rehabilitation programmes for pulmonary hypertension. *Cochrane Database Syst Rev* 2017;1:CD011285. doi: 10.1002/14651858.CD011285.pub2
11. Anderson L, Thompson DR, Oldridge N, et al. Exercise-based cardiac rehabilitation for coronary heart disease. *Cochrane Database Syst Rev* 2016(1):CD001800. doi: 10.1002/14651858.CD001800.pub3
12. McCarthy B, Casey D, Devane D, et al. Pulmonary rehabilitation for chronic obstructive pulmonary disease. *Cochrane Database Syst Rev* 2015(2):CD003793. doi: 10.1002/14651858.CD003793.pub3
13. Slade SC, Dionne CE, Underwood M, et al. Consensus on Exercise Reporting Template (CERT): Explanation and Elaboration Statement. *Br J Sports Med* 2016 doi: 10.1136/bjsports-2016-096651
14. Slade SC, Dionne CE, Underwood M, et al. Consensus on Exercise Reporting Template (CERT): Modified Delphi Study. *Phys Ther* 2016;96(10):1514-24. doi: 10.2522/ptj.20150668
15. Chan L, Chin LMK, Kennedy M, et al. Benefits of intensive treadmill exercise training on cardiorespiratory function and quality of life in patients with pulmonary hypertension. *Chest* 2013;143(2):333-43. doi: 10.1378/chest.12-0993
16. Ehlken N, Lichtblau M, Klose H, et al. Exercise training improves peak oxygen consumption and haemodynamics in patients with severe pulmonary arterial hypertension and inoperable chronic thrombo-embolic pulmonary hypertension: a prospective, randomized, controlled trial. *Eur Heart J* 2016;37(1):35-44. doi: 10.1093/eurheartj/ehv337

17. Gonzalez-Saiz L, Fiuza-Luces C, Sanchis-Gomar F, et al. Benefits of skeletal-muscle exercise training in pulmonary arterial hypertension: The WHOLEi+12 trial. *Int J Cardiol* 2017;231:277-83. doi: 10.1016/j.ijcard.2016.12.026
18. Mereles D, Ehlken N, Kreuzer S, et al. Exercise and respiratory training improve exercise capacity and quality of life in patients with severe chronic pulmonary hypertension. *Circulation* 2006;114(14):1482-9. doi: 10.1161/CIRCULATIONAHA.106.618397
19. Ley S, Fink C, Risse F, et al. Magnetic resonance imaging to assess the effect of exercise training on pulmonary perfusion and blood flow in patients with pulmonary hypertension. *Eur Radiol* 2013;23(2):324-31. doi: 10.1007/s00330-012-2606-z
20. Weinstein AA, Chin LM, Keyser RE, et al. Effect of aerobic exercise training on fatigue and physical activity in patients with pulmonary arterial hypertension. *Respir Med* 2013;107(5):778-84. doi: 10.1016/j.rmed.2013.02.006
21. Fox BD, Kassirer M, Weiss I, et al. Ambulatory rehabilitation improves exercise capacity in patients with pulmonary hypertension. *J Card Fail* 2011;17(3):196-200. doi: 10.1016/j.cardfail.2010.10.004
22. Fukui S, Ogo T, Takaki H, et al. Efficacy of cardiac rehabilitation after balloon pulmonary angioplasty for chronic thromboembolic pulmonary hypertension. *Heart* 2016;102(17):1403-9. doi: 10.1136/heartjnl-2015-309230
23. Martinez-Quintana E, Miranda-Calderin G, Ugarte-Lopetegui A, et al. Rehabilitation program in adult congenital heart disease patients with pulmonary hypertension. *Congenit Heart Dis* 2010;5(1):44-50. doi: 10.1111/j.1747-0803.2009.00370.x
24. Inagaki T, Terada J, Tanabe N, et al. Home-based pulmonary rehabilitation in patients with inoperable or residual chronic thromboembolic pulmonary hypertension: a preliminary study. *Respir Investig* 2014;52(6):357-64. doi: 10.1016/j.resinv.2014.07.002
25. Mainguy V, Maltais F, Saey D, et al. Effects of a rehabilitation program on skeletal muscle function in idiopathic pulmonary arterial hypertension. *J Cardiopulm Rehabil Prev* 2010;30(5):319-23. doi: 10.1097/HCR.0b013e3181d6f962
26. Bussotti MG, P. Pedretti, R. Kransinska, R. Di Marco, S. Corbo, P. Marchese, G. Totaro, P. Sommaruga, M. Effects of an Outpatient Service Rehabilitation Programme in Patients Affected by Pulmonary Arterial Hypertension: An Observational Study. *Cardiovascular & Haematological Disorders-Drug Targets* 2016;16(1)
27. de Man FS, Handoko ML, Groepenhoff H, et al. Effects of exercise training in patients with idiopathic pulmonary arterial hypertension. *Eur Respir J* 2009;34(3):669-75. doi: 10.1183/09031936.00027909
28. Grunig E, Ehlken N, Ghofrani A, et al. Effect of exercise and respiratory training on clinical progression and survival in patients with severe chronic pulmonary hypertension. *Respiration* 2011;81(5):394-401. doi: 10.1159/000322475
29. Grunig E, Maier F, Ehlken N, et al. Exercise training in pulmonary arterial hypertension associated with connective tissue diseases. *Arthritis Res Ther* 2012;14(3):R148. doi: 10.1186/ar3883
30. Nagel C, Prange F, Guth S, et al. Exercise training improves exercise capacity and quality of life in patients with inoperable or residual chronic thromboembolic pulmonary hypertension. *PLoS One* 2012;7(7):e41603. doi: 10.1371/journal.pone.0041603
31. Becker-Grunig T, Klose H, Ehlken N, et al. Efficacy of exercise training in pulmonary arterial hypertension associated with congenital heart disease. *Int J Cardiol* 2013;168(1):375-81. doi: 10.1016/j.ijcard.2012.09.036
32. Kabitz HJ, Bremer HC, Schwoerer A, et al. The combination of exercise and respiratory training improves respiratory muscle function in pulmonary hypertension. *Lung* 2014;192(2):321-8. doi: 10.1007/s00408-013-9542-9
33. Grunig E, Lichtblau M, Ehlken N, et al. Safety and efficacy of exercise training in various forms of pulmonary hypertension. *Eur Respir J* 2012;40(1):84-92. doi: 10.1183/09031936.00123711

34. Neil-Sztramko SE, Winters-Stone KM, Bland KA, et al. Updated systematic review of exercise studies in breast cancer survivors: attention to the principles of exercise training. *Br J Sports Med* 2017 doi: 10.1136/bjsports-2017-098389
35. Ammann BC, Knols RH, Baschung P, et al. Application of principles of exercise training in sub-acute and chronic stroke survivors: a systematic review. *BMC Neurol* 2014;14:167. doi: 10.1186/s12883-014-0167-2
36. Nicolson PJA, Bennell KL, Dobson FL, et al. Interventions to increase adherence to therapeutic exercise in older adults with low back pain and/or hip/knee osteoarthritis: a systematic review and meta-analysis. *Br J Sports Med* 2017;51(10):791-99. doi: 10.1136/bjsports-2016-096458
37. Roter DL, Hall JA, Merisca R, et al. Effectiveness of interventions to improve patient compliance: a meta-analysis. *Med Care* 1998;36(8):1138-61.
38. Michie S, Ashford S, Sniehotta FF, et al. A refined taxonomy of behaviour change techniques to help people change their physical activity and healthy eating behaviours: the CALO-RE taxonomy. *Psychol Health* 2011;26(11):1479-98. doi: 10.1080/08870446.2010.540664
39. Fairman CM, Hyde PN, Focht BC. Resistance training interventions across the cancer control continuum: a systematic review of the implementation of resistance training principles. *Br J Sports Med* 2017;51(8):677-85. doi: 10.1136/bjsports-2016-096537
40. Mack DE, Wilson PM, Santos E, et al. Standards of reporting: the use of CONSORT PRO and CERT in individuals living with osteoporosis. *Osteoporos Int* 2017 doi: 10.1007/s00198-017-4249-z
41. Galie N, Humbert M, Vachiery JL, et al. 2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension: The Joint Task Force for the Diagnosis and Treatment of Pulmonary Hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS): Endorsed by: Association for European Paediatric and Congenital Cardiology (AEPC), International Society for Heart and Lung Transplantation (ISHLT). *Eur Heart J* 2016;37(1):67-119. doi: 10.1093/eurheartj/ehv317
42. Alison JA, McKeough ZJ, Johnston K, et al. Australian and New Zealand Pulmonary Rehabilitation Guidelines. *Respirology* 2017;22(4):800-19. doi: 10.1111/resp.13025