

Australian perspective regarding recommendations for physical activity and exercise rehabilitation in pulmonary arterial hypertension

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Aim: To determine the opinion of health care professionals within Australia, regarding acceptable levels of exertion and symptoms, and referral for exercise rehabilitation in patients with pulmonary arterial hypertension (PAH).

Method: In 2010, 76 health care professionals at a specialist pulmonary hypertension meeting in Australia were surveyed using a self-administered questionnaire. The questionnaire included case studies of patients with PAH in World Health Organization (WHO) functional classes II–IV. For each case study, respondents were asked to report their opinion regarding the acceptable level of exertion and symptoms during daily activities, and whether they would refer the patient for exercise rehabilitation. Three additional questions asked about advice in relation to four specific physical activities.

Results: The response rate was 70% (n = 53). Overall, 58% of respondents recommended patients undertake daily activities 'as tolerated'. There was no consensus regarding acceptable levels of breathlessness or fatigue, but the majority of respondents considered patients should have no chest pain (73%) and no more than mild light-headedness (92%) during daily activities. Overall, 63% of respondents would have referred patients for exercise rehabilitation. There was little difference in opinion regarding the acceptable level of exertion or symptoms, or referral for exercise rehabilitation, according to functional class. However, the patients' functional class did influence the advice given regarding the specific physical activities.

Conclusion: In 2010, there were inconsistencies between individual health care professionals within Australia regarding appropriate levels of physical exertion and acceptable symptoms during daily activities. Almost two-thirds of the respondents reported they would refer patients for exercise rehabilitation.

Keywords: exercise rehabilitation, physical activity, pulmonary arterial hypertension

Introduction

Pulmonary arterial hypertension (PAH) is characterized by poor exercise tolerance and impaired functional capacity. In patients with left heart failure (LHF) and chronic obstructive pulmonary disease, similar impairments respond well to exercise rehabilitation.¹⁻³ Until recently, however, exercise rehabilitation was discouraged for patients with PAH due to a perceived high risk of adverse outcomes associated with physical exertion.⁴ Historically, symptoms were used to guide physical activity and patients were advised to avoid exertion associated with light-headedness, chest pain, or severe dyspnea.⁵ Otherwise, little consideration was given

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regarding recommendations for physical activity or exercise rehabilitation for individuals with PAH.

The recent introduction of PAH specific therapy has led to improved central hemodynamics and prognosis for patients with PAH.⁶ Despite therapy, however, many patients with PAH continue to experience significant impairment in physical function and quality of life.⁷ The benefits seen following exercise rehabilitation in patients with LHF and chronic obstructive pulmonary disease have been achieved in the absence of adverse events or clinical deterioration and exercise rehabilitation now constitutes an important adjunct to medical therapy in these populations.^{8,9} These findings have stimulated an interest in the effects of exercise rehabilitation in patients with PAH, but, prior to a feasibility study published in Japanese in 2005,¹⁰ there were no reports of exercise rehabilitation in this population. Between 2006 and June 2010, a further three studies, with a total of 51 subjects with PAH who had undergone rehabilitation, were published.^{11–13} Improvements in exercise capacity and quality of life were observed following rehabilitation, without any adverse events or clinical deterioration.^{10–13} Since June 2010, two further studies of exercise rehabilitation in subjects with PAH (with a total of 27 subjects) have been published.^{14,15}

It is possible the paucity of data regarding the impact of physical activity and exercise on outcomes in PAH has made it difficult for clinicians to provide consistent advice regarding physical activity and to identify which patients are suitable for exercise rehabilitation. The aim of this study was to determine the consistency of approach by health care professionals who manage patients with PAH within Australia, regarding appropriate levels of exertion and acceptable symptoms during physical activity, and referral for exercise rehabilitation.

Methods

The study used a cross sectional design with data collected via a self-administered questionnaire. The questionnaire was developed by the investigators specifically for this study (see Appendix 1). Although responses were anonymous, respondents were asked to indicate the type of institution that they worked in, their professional position, and the number of new PAH cases seen each year.

Three scenarios reflective of clinical practice were presented as case studies. These case studies described adults with PAH, one in each of the WHO functional classes II, III and IV. The data presented for each case study included the hemodynamic status (ie, pulmonary artery pressure and

pulmonary vascular resistance index) obtained from right heart catheter assessment, right ventricular function measured at rest on an echocardiogram, 6-minute walk distance, and arterial blood gas data. For each case scenario, participants were instructed to respond to the same seven questions in accordance with their usual clinical management. The first question asked if they would perform any additional investigations. The remaining questions were designed to obtain data pertaining to the recommendations the respondent would provide regarding daily activity, acceptable symptoms on exertion, and whether they would refer the patient described in the case study for exercise rehabilitation.

Three additional questions, regarding four specific physical activities, were asked to determine the advice the respondents would give to patients with PAH, in each of the functional classes II–IV. These questions asked whether patients with PAH would be advised for or against lifting a 20 kg weight, exercising in a non-hospital gym, regularly using stairs and slopes, or adopting a sedentary lifestyle.

Piloting of the questionnaire was performed in a sample of seven health care professionals (medical practitioners, $n = 3$; nurses, $n = 2$; physiotherapists, $n = 2$) from the pulmonary hypertension clinic at a tertiary hospital (Royal Perth Hospital, Perth, Western Australia). These individuals were instructed to complete the questionnaire, and provide feedback about its ease of use, applicability, and length. No recommendations for change arose from this pilot study.

Participants

The survey was conducted at the Pulmonary Hypertension Perspectives Meeting (Sydney, Australia; Pfizer Pharmaceuticals Australia) in June 2010. Eighty-nine health care professionals who worked in pulmonary hypertension clinics within Australia attended the meeting. Seventy-six of these participants had a role in advising patients with PAH regarding physical activity and exercise. The remaining participants were pharmacists ($n = 12$) and a medical student ($n = 1$). Attendance was by invitation and the purpose of the meeting was to provide a forum for experts within the field of pulmonary hypertension to discuss best practice and future directions in the diagnosis and management of PAH.

This study was approved by the Human Research Ethics Committee of Curtin University. Participant consent was implied by return of the questionnaires.

Data analysis

Data analysis comprised descriptive statistics. Analysis was performed using SPSS software (v 18; SPSS Inc, Chicago, IL).

Results

Fifty-three completed questionnaires were returned from participants who have a role providing advice to patients regarding daily activities, or in referring patients for exercise rehabilitation. This represents a response rate of 70%. Some respondents did not answer all questions and therefore not all percentages presented in the results add to 100.

Demographics

Demographic data of the 53 respondents who completed the questionnaire are presented in Table 1.

Responses to the questions reflecting usual clinical practice, in relation to the case studies and four specific physical activities, are summarized in Table 2. Data pertaining to the responses to all of the questions are presented in Appendix 1.

Responses to questions relating to the case studies

Instructions for daily activity

The majority of respondents (67%) stated that they would have given the patients verbal advice and 33% would have given written advice.

Overall, 58% of respondents stated that they would advise the patients to undertake daily activities 'as tolerated', regardless of functional class (Table 2). The balance of respondents moderated their instructions according to functional class. For example, 25% advised moderate activity for patients in functional class II, 9% advised moderate activity for patients in functional class III, and only 4% advised moderate activity for patients in functional class IV (Table 2). Some respondents provided details on how their patients would have been guided regarding daily activities. These respondents reported that they would have: assessed the patient's exercise capacity, referred the patient for physiotherapy and/or pulmonary or cardiac rehabilitation, advised the patient to use oxygen therapy during activity, or

advised the patient to undertake symptom-limited exertion (Appendix 1).

Acceptable symptoms during daily activities

Overall, the majority (92%) of respondents considered that either no light-headedness (53%) or mild light-headedness (39%) was acceptable during daily activities. A minority (7%) considered that moderate light-headedness was acceptable. With regard to chest pain, 73% of respondents considered that no chest pain was acceptable. A further 20% advised that mild chest pain was acceptable. For both light-headedness and chest pain there was little variation according to functional class (Table 2).

In terms of the breathlessness experienced during physical activities, overall 20% of respondents considered minimal, 44% considered moderate and 31% considered breathlessness 'as tolerated' was acceptable during daily activities. However, more respondents considered moderate rather than minimal breathlessness (51% vs 11%, respectively) was acceptable for the patient in functional class IV. When considering fatigue levels during daily activities, overall 5% of respondents considered that no fatigue was acceptable. A further 27% considered minimal fatigue, 40% considered moderate fatigue, and 21% considered that fatigue 'as tolerated' was acceptable. With the exception of the different opinion pertaining to acceptable levels of breathlessness for patients in functional class IV compared to patients in classes II and III, there was little difference in the level of symptoms that was considered acceptable across the functional classes (Table 2).

Referral for exercise rehabilitation

Overall, 63% of respondents stated they would have referred patients for exercise rehabilitation and this did not vary depending on the patients' functional class (Table 2).

Responses to questions regarding specific physical activities

Respondents' advice regarding the recommendations for the four specific physical activities varied according to functional class. Fifty-seven percent of respondents would have advised for, and 36% against, patients in functional class II lifting a 20 kg weight. The majority of respondents would have advised patients in functional class II to use a non-hospital gym (70%) and to regularly use stairs and slopes (79%). The majority of respondents would also have advised patients in functional class II against adopting a sedentary lifestyle (91%).

Table 1 Demographics of the 53 respondents

Type of institution	Tertiary	41 (77)
	Secondary	8 (15)
	Private and public	3 (6)
	Other	1 (2)
Profession	Respiratory physician	19 (36)
	Cardiologist	8 (15)
	Rheumatologist	6 (11)
	Nurse	19 (36)
	Exercise physiologist	1 (2)

Note: Data are number of responses with percentages given in parentheses.

Table 2 Questionnaire responses from the 53 participants

		Class II	Class III	Class IV
Method to guide daily activity	Verbal	72	62	68
	Written	38	25	36
	Refer on for instruction	17	26	23
Refer for exercise rehabilitation	Yes	62	60	66
	No	38	32	34
Recommended level of exertion during daily activities	Mild	6	13	23
	Moderate	25	9	4
	As tolerated	57	57	60
Acceptable breathlessness during daily activities	Minimal	23	26	11
	Moderate	44	38	51
	As tolerated	30	28	34
Acceptable fatigue during daily activities	None	6	8	2
	Minimal	33	25	28
	Moderate	37	34	50
	As tolerated	22	25	17
Acceptable light-headedness during daily activities	None	57	51	51
	Mild	37	38	41
	Moderate	4	4	6
	To syncope	0	0	0
Acceptable chest pain during daily activities	None	78	74	68
	Mild	16	13	30
	Moderate	2	4	0
	As tolerated	4	2	2
Would you advise a patient to lift a 20 kg weight	Yes	36	9	0
	No	57	83	92
Would you advise exercise in a non-hospital gym	Yes	70	21	6
	No	23	72	87
Would you advise regularly using stairs and slopes	Yes	79	51	11
	No	13	42	81
Would you advise a sedentary lifestyle	Yes	2	8	17
	No	91	85	75

Notes: Data are percentage of responses. Some respondents did not answer all questions and therefore not all percentages add to 100. Raw data for the responses to all questions are given in Appendix I.

For patients in functional class III, 83% of respondents would have advised against lifting a 20 kg weight and against exercise in a non-hospital gym (72%). A similar number of respondents recommended that patients in functional class III did and did not regularly use stairs and slopes (51% and 42%, respectively). Eighty-five percent would have advised patients in functional class III against a sedentary lifestyle. The majority of respondents stated that they would have advised patients in functional class IV not to lift a 20 kg weight (92%), not to use a non-hospital gym (87%) and not to regularly use stairs or slopes (81%). The majority of respondents would have advised patients in functional class IV to adopt a sedentary lifestyle (75%).

Discussion

The main findings of this survey of health care professionals attending a pulmonary hypertension specific meeting in 2010 were: (1) there was inconsistency between individuals

regarding the acceptable level of exertion, breathlessness and fatigue during daily activities for patients with PAH; (2) there was a consensus that, for these patients, chest pain and anything more than mild light-headedness was not acceptable during daily activities; (3) approximately two-thirds of respondents stated they would refer patients for exercise rehabilitation; (4) the level of acceptable symptoms during daily activities and patterns of referral for exercise rehabilitation did not vary greatly according to the patients' functional class, but, (5) advice regarding specific physical activities varied according to functional class.

The lack of evidence regarding the safety and impact of physical exertion on hemodynamics in patients with PAH has made it difficult for clinicians to provide recommendations regarding physical activity. Until recently, physical activity guidelines for patients with PAH were only available from sources such as the Pulmonary Hypertension Association⁵ and were based upon studies in subjects with LHF. Patients

with PAH were advised to undertake light to moderate aerobic exercise and to avoid exercise to the point of light-headedness, chest pain, or severe dyspnea. Patients with severe functional limitation, and those with a history of dizziness and/or fainting, were advised against undertaking an exercise program.⁵

The inconsistency in response in our study, regarding acceptable symptoms of breathlessness and fatigue during physical activity, is likely to reflect uncertainty about the significance of these symptoms. In contrast, the consensus regarding the avoidance of light-headedness, a symptom suggestive of impaired cardiac output,¹⁶ and the majority view recommending avoidance of chest pain, a symptom suggestive of myocardial ischemia,¹⁷ is likely to reflect the understanding that these symptoms can represent life-threatening events. The inconsistencies in opinion regarding the appropriate level of exertion and advice for common daily activities, such as lifting a 20 kg weight and the regular use of stairs and slopes, suggests uncertainty about the impact of general physical activity on patient outcome.

Recently, one randomized controlled trial,¹¹ one non-randomized controlled trial,¹⁵ three intervention trials^{10,12,14} and one case report on exercise rehabilitation¹³ have been published. The subjects in these studies had idiopathic PAH (n = 83), chronic thrombo-embolic pulmonary hypertension (n = 8), or PAH associated with connective tissue disease (n = 10), or congenital heart disease (n = 1). These publications reported positive outcomes associated with exercise rehabilitation, achieved in the absence of adverse events. To the authors' knowledge, only one published position statement on pulmonary rehabilitation⁹ and no position statements or guidelines on cardiac rehabilitation include specific guidelines for exercise rehabilitation in PAH. In 2008, one publication reviewed exercise responses in PAH and discussed exercise prescription for this population.⁴

In the current study, the finding that only two-thirds of respondents recommended exercise rehabilitation for patients in functional classes II and III suggests that, in 2010, there was uncertainty regarding this intervention, despite publications describing benefits of exercise training for this population.¹⁰⁻¹³ This uncertainty may be related to the low number of subjects in the published reports and the publication of only one randomized controlled trial of exercise rehabilitation.

The only reported study of exercise rehabilitation that included a substantial number of patients in functional class IV was published in Japanese.¹⁰ However, in our study it was notable that the majority of respondents stated that they would refer patients in functional class IV for exercise

rehabilitation, despite the very limited evidence for exercise rehabilitation in this subgroup. It is possible this finding relates to a perception that exercise rehabilitation for patients in functional class IV will improve functional capacity and minimize symptoms during activities of daily living. It may also relate to the perceived need for patients with severe disease to be educated about the risks of overexertion and to be supervised during exercise. The advice for patients in functional class IV, regarding the specific physical activities presented in this study, demonstrated a conservative approach towards physical activity that was not supervised by a health care professional.

The point prevalence of idiopathic PAH in Australia is estimated at 40 per million¹⁸ and therefore, based upon Australian population statistics,¹⁹ the estimated number of patients with idiopathic PAH in Australia is approximately 900. In 2010, medical practitioners in 47 pulmonary hypertension clinics were licensed to prescribe PAH specific pharmaceutical therapies.²⁰ The rarity and complex nature of PAH, and strict licensing conditions associated with the prescription of PAH medications, means a limited number of health care professionals are involved in the management of patients with PAH. The relatively small number of participants in this study reflects the limited number of health care professionals, within Australia, who work with patients who have PAH.

Limitations

Attendance at the meeting at which the survey was performed was by invitation, resulting in likely selection bias. However, it also ensured the respondents were directly involved in the management of patients with PAH.

It was not possible to identify any overlap between respondents, in terms of the institutions that were represented, due to a deliberate decision to maintain the respondents' anonymity. However, the preservation of anonymity is likely to have had a positive influence over the response rate and encouraged open responses.

Conclusion

In 2010, health care professionals within Australia were inconsistent in opinion regarding appropriate levels of exertion and acceptable symptoms during daily activities. There appeared to be some uncertainty regarding the role of exercise rehabilitation in PAH. The findings of this study identify a need for further research to support the development of guidelines on physical activity and exercise rehabilitation for the PAH population in Australia.

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Author contributions

Robin Fowler was responsible for the design of this study, collection of the data, analysis and interpretation of the data and preparation of the manuscript. Sue Jenkins was responsible for the design of this study, interpretation of the data and preparation of the manuscript. Andrew Maiorana was responsible for the design of this study and preparation of the manuscript. Kevin Gain was responsible for analysis and interpretation of the data and preparation of the manuscript. Gerry O'Driscoll was responsible for the design of this study and preparation of the manuscript. Eli Gabbay was responsible for the design of this study, interpretation of the data and preparation of the manuscript.

Disclosures

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The meeting organisers had no input or influence over the content of the questionnaire, the data analysis or this manuscript. They supported the project by photocopying, distributing, and collecting the questionnaires.

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Appendix I

Questionnaire used in the study together with the responses to each questions.

Note: Data given in the responses are number of responses with percentages given in parentheses.

Activity and exercise guidelines for patients with pulmonary arterial hypertension (PAH)

Background

There is very little in the literature to guide exercise and physical activity recommendations for patients with PAH. The

Royal Perth Hospital Advanced Lung Disease/Pulmonary Hypertension Research Group led by Associate Professor Eli Gabbay, in conjunction with the Curtin University School of Physiotherapy, is seeking to determine expert opinion on exercise and physical activity guidelines for patients with PAH. We therefore request that you complete and return the following questionnaire.

Participation is purely voluntary but we strongly encourage your contribution. There is no individual identifying information on this questionnaire and therefore anonymity will be protected. Information gained from this questionnaire may be used in future presentations and publications.

Thank you for taking the time to complete this questionnaire.

For each of the following questions please circle your chosen answer.

Total number of respondents

Type of institution:

- a) Tertiary/major hospital
- b) Secondary/regional hospital
- c) Private practice
- d) General practice
- e) Other – please specify _____

Position:

- a) Respiratory (thoracic) physician
- b) Cardiologist
- c) Rheumatologist
- d) Nurse specialist
- e) Exercise physiologist
- f) Other – please specify _____

Number of new PAH patients seen per year:

- a) 0–10
- b) 11–50
- c) 51–100
- d) 101–150
- e) 151–200
- f) 201–300
- g) >300

Results

(n = 53)

n (%)

41 (77)

8 (15)

3 (6)

0

1 (2)

19 (36)

8 (15)

6 (11)

19 (36)

1 (2)

18 (34)

24 (45)

4 (8)

4 (8)

0

2 (4)

0

Two case studies are presented. Please circle the answer(s) that best represents your usual response.

Case 1

44-year-old female

WHO functional Class IV

Six-minute walk distance 85 m on room air.

ABGs on room air PaO₂ 51 mmHg (6.8 kPa), PaCO₂ 42 mmHg (5.6 kPa)

PAP 92/55, mean PAP 67 mmHg on resting RHC study

PVRI 2291 dyne/s/m²/cm⁵

Severely dilated right ventricle with severe global impairment of RV systolic function
No evidence of thromboembolic disease

All usual tertiary hospital-based assessment related to diagnosis and medical management has been performed

In relation to exercise and activity prescription:

1. What further investigations would you perform?

Results

n (%)

- | | |
|----------------------------------|---------|
| a) None | 28 (53) |
| b) Exercise right heart catheter | 8 (15) |
| c) Exercise echo | 5 |
| d) Other – please explain _____ | |

Results: lung function tests (n = 2), cardiopulmonary exercise test (CPET) (n = 1), CT angiogram (n = 1), overnight oximetry (n = 1)

In the absence of these results:

2. What instructions for daily activities would you give?

- | | |
|---|---------|
| a) None | 0 |
| b) Recommend only mild exertion | 12 (23) |
| c) Recommend moderate exertion | 2 (4) |
| d) Instruct the patient to continue activity as tolerated | 32 (60) |
| e) Other – please explain _____ | |

Results: Refer to physio/pulmonary rehab (n = 6), oxygen therapy (n = 4), symptom limited exercise (n = 2), overnight oximetry (n = 1)

For this question, more than one answer may be selected

3. How would you guide the patient with respect to their daily activities? Would you:

- | | |
|--|---------|
| a) Give verbal guidelines | 36 (68) |
| b) Give written guidelines | 19 (36) |
| c) Refer the patient for instruction | 12 (23) |
| d) Refer the patient for exercise rehabilitation | 35 (66) |
| e) Other – please explain _____ | |

For this patient, which of the following symptoms would you consider to be acceptable during daily activities?

4. Breathlessness

- | | |
|--|---------|
| a) Minimal | 6 (11) |
| b) No more than moderate | 27 (51) |
| c) As much as the patient can tolerate | 18 (34) |
| d) Other – please explain _____ | |

5. Light-headedness

- | | |
|---------------------------------|---------|
| a) None | 27 (51) |
| b) Mild | 22 (41) |
| c) Moderate | 3 (6) |
| d) To the point of syncope | 0 |
| e) Other – please explain _____ | |

6. Fatigue

- | | |
|------------|---------|
| a) None | 1 (2) |
| b) Minimal | 15 (28) |

- | | |
|--|---------|
| c) Moderate fatigue | 26 (50) |
| d) As much as the patient can tolerate | 9 (17) |
| e) Other – please explain _____ | |

7. Chest pain

- | | |
|--|---------|
| a) None | 36 (68) |
| b) Mild | 16 (30) |
| c) Moderate | 0 |
| d) As much as the patient can tolerate | 1 (2) |
| e) Other – please explain _____ | |

Following 6 months on a maximal dose of Epoprostenol this same patient has the following results:

WHO functional class II

6MWD 405 m on room air

Echo: mild RV dysfunction and RV dilatation

Mean PAP 45 mmHg

PVRI 815 dynes/s/m²/cm⁵

8. What further investigations would you perform?**Results**

- | | n (%) |
|----------------------------------|--------------|
| a) None | 36 (68) |
| b) Exercise right heart catheter | 5 (9) |
| c) Exercise echo | 3 (6) |
| d) Other – please explain _____ | |

Results: Six-minute walk test (6MWT) (n = 1), CPET (n = 2)

In the absence of any other results:**9. What instructions for daily activities would you give?**

- | | |
|---|---------|
| a) None | 0 |
| b) Recommend only mild exertion | 3 (6) |
| c) Recommend moderate exertion | 13 (25) |
| d) Instruct the patient to continue activity as tolerated | 30 (57) |
| e) Other – please explain _____ | 6 (11) |

Results: Assess exercise capacity/6MWT (n = 3), review by physio/continue pulmonary rehab (n = 2), oxygen therapy (n = 2), symptom limited exercise (n = 3)

For this question, more than one answer may be selected**10. How would you guide the patient with respect to their daily activities? Would you:**

- | | |
|--|---------|
| a) Give verbal guidelines | 38 (72) |
| b) Give written guidelines | 20 (38) |
| c) Refer the patient for instruction | 9 (17) |
| d) Refer the patient for exercise rehabilitation | 33 (62) |
| e) Other – please explain _____ | |

For this patient, which of the following symptoms would you consider to be acceptable during daily activities?

11. Breathlessness

- | | |
|--|---------|
| a) Minimal | 12 (23) |
| b) Moderate | 23 (44) |
| c) As much as the patient can tolerate | 16 (30) |
| d) Other – please explain _____ | |

12. Light-headedness

a) None	30 (57)
b) Mild	20 (37)
c) Moderate	2 (4)
d) To the point of syncope	0
e) Other – please explain _____	

13. Fatigue

a) None	3 (6)
b) Minimal	18 (33)
c) Moderate	20 (37)
d) As much as the patient can tolerate	12 (22)
e) Other – please explain _____	

14. Chest pain

a) None	41 (78)
b) Mild	8 (16)
c) Moderate	1 (2)
d) As much as the patient can tolerate	2 (4)
e) Other – please explain _____	

Case 2

36-year-old female

WHO functional Class III

Six-minute walk distance 305 m on room air

ABGs on room air PaO₂ 88 mmHg (11.7 kPa), PaCO₂ 38 mmHg (5.1 kPa)

PAP 62/28, **mean PAP** 38 mmHg on resting RHC study

PVRI 671 dyne/s/m²/cm⁵

Moderately dilated right ventricle with moderate global impairment of RV systolic function

No evidence of thromboembolic disease

All usual tertiary hospital based assessment related to diagnosis and medical management has been performed.

In relation to exercise and activity prescription:**Results****15. What further investigations would you perform?****n (%)**

a) None	27 (51)
b) Exercise right heart catheter	8 (15)
c) Exercise echo	8 (15)
d) Other – please explain _____	

Results: Lung function tests (n = 1), CT pulmonary angiogram (n = 1), 6MWT (n = 1), overnight oximetry (n = 1)

In the absence of these results:**16. What instructions for daily activities would you give?**

a) None	0
b) Recommend only mild exertion	7 (13)
c) Recommend moderate exertion	5 (9)
d) Instruct the patient to continue activity as tolerated	30 (57)
e) Other – please explain _____	7 (13)

Results: Assess exercise capacity (n = 1)/6MWT (n = 2)/exercise right heart catheter (n = 1), cardiac or pulmonary rehab (n = 2), oxygen therapy (n = 1), symptom limited exercise (n = 3)/no strenuous exercise/Borg <6

For this question, more than one answer may be selected

17. How would you guide the patient with respect to their daily activities? Would you:

- | | |
|--|---------|
| a) Give verbal guidelines | 33 (62) |
| b) Give written guidelines | 13 (25) |
| c) Refer the patient for instruction | 14 (26) |
| d) Refer the patient for exercise rehabilitation | 32 (60) |
| e) Other – please explain _____ | |

For this patient, which of the following symptoms would you consider to be acceptable during daily activities?

18. Breathlessness

- | | |
|--|---------|
| a) Minimal | 14 (26) |
| b) Moderate | 20 (38) |
| c) As much as the patient can tolerate | 15 (28) |
| d) Other – please explain _____ | |

19. Light-headedness

- | | |
|---------------------------------|---------|
| a) None | 27 (51) |
| b) Mild | 20 (38) |
| c) Moderate | 2 (4) |
| d) To the point of syncope | 0 |
| e) Other – please explain _____ | |

20. Fatigue

- | | |
|--|---------|
| a) None | 4 (8) |
| b) Minimal | 13 (25) |
| c) Moderate | 18 (34) |
| d) As much as the patient can tolerate | 13 (25) |
| e) Other – please explain _____ | |

21. Chest pain

- | | |
|--|---------|
| a) None | 39 (74) |
| b) Mild | 7 (13) |
| c) Moderate | 2 (4) |
| d) As much as the patient can tolerate | 1 (2) |
| e) Other – please explain _____ | |

Please circle either the yes or no response to each of the questions below:

22. For a patient with established PAH in WHO functional class II would you advise:

- | | Results (n, %) (n, %) | |
|--------------------------------------|-----------------------|-------------|
| a) Lifting 20 kg in weight | Yes (19, 36) | No (30, 57) |
| b) Exercising in a non-hospital gym | Yes (37, 70) | No (12, 23) |
| c) Regularly using stairs and slopes | Yes (42, 79) | No (7, 13) |
| d) A sedentary lifestyle | Yes (1, 2) | No (48, 91) |

23. For a patient with established PAH in WHO functional class III would you advise:

- | | | |
|--------------------------------------|--------------|-------------|
| a) Lifting 20 kg in weight | Yes (5, 9) | No (44, 83) |
| b) Exercising in a non-hospital gym | Yes (11, 21) | No (38, 72) |
| c) Regularly using stairs and slopes | Yes (27, 51) | No (22, 42) |
| d) A sedentary lifestyle | Yes (4, 8) | No (45, 85) |

24. For a patient with established PAH in WHO functional class IV would you advise:

- | | | |
|-------------------------------------|------------|-------------|
| a) Lifting 20 kg in weight | Yes (0) | No (49, 92) |
| b) Exercising in a non-hospital gym | Yes (3, 6) | No (46, 87) |

- | | | |
|--------------------------------------|-------------|-------------|
| c) Regularly using stairs and slopes | Yes (6, 11) | No (43, 81) |
| d) A sedentary lifestyle | Yes (9, 17) | No (40, 75) |

Further comments about exercise/activity instructions, or the questionnaire:

Results:

- 1) Important all patients given an exercise prescription based on professional, experienced assessment (eg, physio); avoid heavy lifting and anaerobic activity; avoid breath holding (eg, during swimming)
- 2) Exercise to tolerance after supervised exercise in hospital gym for all class II/III patients
- 3) As good as a drug. Should be used in all cases
- 4) Most patients in WHO II and III could do a rehab program but it would be PH related and most would benefit from one-on-one supervised training with a specialist
- 5) For majority of patients exercise is advisable in consultation with treating physician and physiotherapist/pulmonary rehab co-ordinator in a monitored environment. Activity is always recommended however should be adjusted to patients' condition. Isometric exercises, gentle walking, and use of oxygen should be discussed individually with patients and carers.

Thank you again for your participation.

A/Prof Sue Jenkins is a lecturer and Robin Fowler is a PhD student at Curtin University.

If you wish to be informed of the collective results of this survey, please contact Robin Fowler via email at the following address:

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