

Use of eHealth in the management of pulmonary arterial hypertension: review of the literature

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ABSTRACT

Background Pulmonary arterial hypertension (PAH) is a severe chronic condition associated with poor quality of life and high risks of mortality and hospitalisation. The utilisation of novel diagnostic technologies has improved survival rates although the effectiveness of Electronic Health (eHealth) interventions in patients with a chronic cardiopulmonary disease remains controversial. As the effectiveness of eHealth can be established by specific evaluation for different chronic health conditions, the aim of this study was to explore and summarise the utilisation of eHealth in PAH.

Method We searched PubMed, CINAHL and Embase for all studies reporting clinical trials on eHealth solutions for the management of PAH. No limitations in terms of study design or date of publication were imposed.

Results 18 studies (6 peer-reviewed journal papers and 12 conference papers) were identified. Seven studies addressed the accuracy, safety or reliability of eHealth technologies such as intra-arterial haemodynamic monitoring of the pulmonary artery pressure, self-administered 6-Minute walk test App, computerised step-pulse oximeter and ambulatory impedance cardiography. Two studies evaluated eHealth as part of the medical management and showed a reduction in hospitalisation rate.

Conclusions The evidence of eHealth supporting the management of people with PAH is limited and only embraced through a few studies of small sample size and short-term duration. Given the proposed clinical benefits in heart failure, we postulate that the evaluation of eHealth for the clinical management of PAH is highly warranted.

INTRODUCTION

Pulmonary arterial hypertension (PAH) is an incurable and progressive disease caused by the narrowing of the blood vessels of the lungs. This disease is characterised by high blood pressure in the pulmonary artery that, if maintained over time, may lead to right heart failure and premature death.¹ PAH is a complex and multifactorial chronic disease² that pervades the holistic functioning of the individuals and their quality of life (QoL).³ Over the past few years, the efficacy of combination drug therapy and the utilisation of

diagnostic technologies (including imaging techniques and right heart catheterisation) have contributed to increasing the efficacy of therapeutic regimes, reducing morbidity and mortality in people with PAH.^{4,5}

The management of PAH is complex and requires intense and regular communication, in the first place between the patient and caregivers, but also among all members of the highly specialised centre-based multi-disciplinary care team⁶ and the external caregivers, including primary care centres, regional hospitals, supporting family and carers. In PAH care, all contributions to enhance communication can be of particular interest, for example, during the almost inevitable periods of drug modification or up-titration, or when drug side effects may potentially appear.⁷

Electronic Health (eHealth), which is referred to as a broad range of informatics applications for facilitating the management and delivery of healthcare,⁸ has been proposed to support the management of patients with chronic disease and as a possible tool to improve clinical outcomes in patients with heart failure (HF) and chronic obstructive pulmonary disease (COPD).^{9,10} The adoption of eHealth enables remote monitoring of patients and helps to identify early symptoms and respond promptly to exacerbations.¹¹ In addition, eHealth innovations enable self-management and contribute to shifting the balance of power and responsibility from healthcare professionals to patients,¹² which in turn is closely related to improvements in health-related QoL.¹³ In addition, mobile health (mHealth), which is a subset of eHealth and defined as the medical and public health practice supported by mobile devices, such as mobile phones, patient monitoring devices, personal digital assistants and other wireless devices,¹⁴ has been envisioned to play an important role for the management



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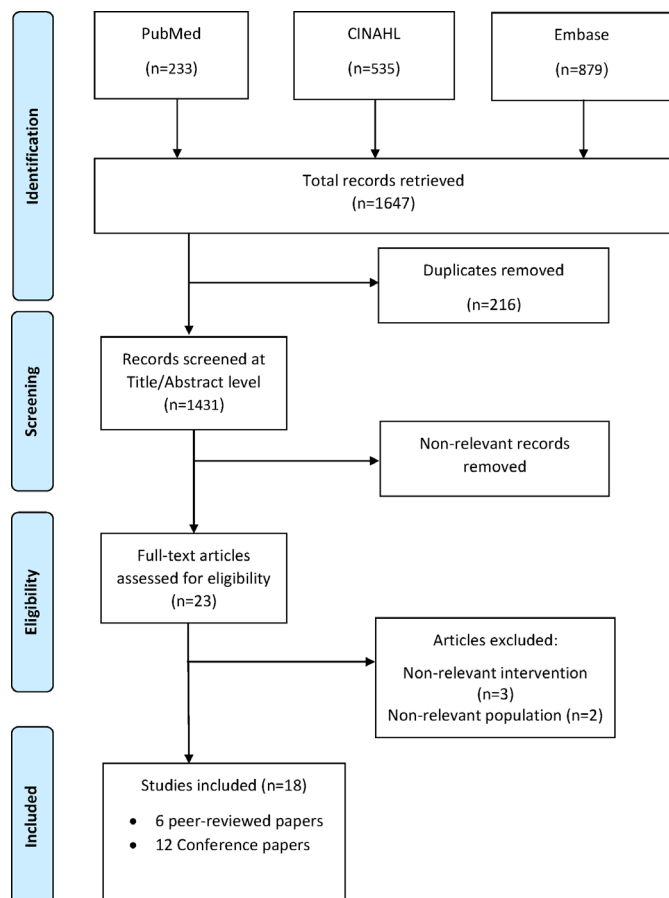


Figure 1 PRISMA flow diagram of the screening and selection of the studies.

of chronic conditions.¹⁵ As the impact of eHealth needs to be established specifically for different targeted populations,¹⁶ the objective of this study was reviewing the existing literature in an attempt to better understand what types of eHealth solutions have been tested, and the possible benefits that eHealth has brought about for the management of people with PAH.

METHODS

Search strategy

We searched PubMed, CINAHL and Embase for studies on eHealth interventions for people with PAH. A combination of MeSH terms and free-text keywords pertaining to the two main concepts of interest (ie, eHealth and pulmonary arterial hypertension) was used to develop a sensitive search query following the recent guides on PubMed searches.^{17–19} The detailed search strategy for PubMed is available (online supplementary appendix 1). The search query was modified for CINAHL and Embase according to their user guide. The results of electronic searches were exported to an EndNote library, and duplicate records were removed.

Study selection

Two independent reviewers screened the records at the title/abstract level to identify potentially relevant studies.

The inclusion criteria were journal papers as well as conference proceedings in English language in which eHealth had been used for improving the care of people with PAH, with no limitations on date of publication. The exclusion criteria were non-clinical studies (either pharmacological or technology-focused publications related to the development of technology devices), clinical trials published in a language other than English, expert opinions and letters to the Editor. Research articles on imaging and echocardiography, as well as reported surgical procedures, were excluded as considered out of the scope of this review.

Data extraction

The full text of potentially relevant papers was obtained for a further check against the study selection criteria and determining the final set of included papers for review. We extracted the following data: author and country, characteristics of the study (retrospective or prospective cohort study or case-series), trial quality (level of evidence, according to the Centre for Evidence-Based Medicine [CEBM], Oxford University),²⁰ the year and the type of publication (conference paper or journal paper), the study population, brief descriptions of the eHealth solution and the results of the study. Some of the authors had conducted more than one study on the same eHealth solution. For simplicity reasons, all studies presented by the same group of researchers are presented in a single row of the table, indicating their correspondent author's name, type and year of publication.

RESULTS

The electronic search resulted in 1431 unique records. Of these, 1406 non-relevant records were filtered out at title/abstract screening, based on the exclusion criteria. The full text of 23 studies were inspected for eligibility. Of these, five papers were excluded either due to non-relevant study setting (eg, the purpose of the trial was not assessing eHealth) or non-relevant patient population (eg, patients with HF without PAH). Eighteen studies were considered eligible for inclusion in this review: 6 peer-reviewed journal papers and 12 conference papers published in scientific journals. The PRISMA flow diagram of study selection is shown in figure 1. To facilitate the analysis and the discussion of the findings of this review, we split the studies into two groups: one group includes the studies that aimed to test the validity and/or the reliability of a specific eHealth solution (table 1), and the other group includes the studies that evaluated eHealth as part of the medical management of PAH, with the possibility of reporting clinical endpoints such as hospitalisation and QoL (table 2).

In the reviewed studies, four different eHealth solutions have been tested: (1) wireless implantable haemodynamic monitor (IHM) to measure pulmonary artery pressure, (2) wireless pedometers and smartphone apps for self-administered 6-minute walk test (6MWT), (3)

Table 1 Summary of studies evaluating the validity/reliability of eHealth for PAH

Author Country (year of publication)	Type of study/level of evidence (CEBM)/type of publication	Study population	Intervention	Summary of results	Significance level
Arelli <i>et al</i> USA (2012) ³⁷	Case-series/level 4/CP	18 patients with right heart catheterisation-confirmed PH	A portable IC device with real-time wireless monitoring via a Bluetooth adapter to determine HR, CO, CI and SV during the 6MWT	The IC increased the value of 6MWT and provided insight into the haemodynamic changes during exercise in PH	All the comparisons at rest vs activity were statistically significant (p<0.001)
Biederman <i>et al</i> USA (2015) ²⁸	Case-series study/level 4/CP	10 patients with PAH	Longitudinal CardioMEMS measuring of PAP, HR and CO and CMR at baseline and during pharmacological stress were performed	Cardiomems may contribute to providing an accurate calculation of non-invasive CO in PAH	NR
Brooks <i>et al</i> USA (2014, 2015) ^{31 32}	1. Case-series study/level 4/CP 2. Prospective study/level 1b/JP	CP: 52 participants half of whom had CHF and PH JP: 103 participants with PH and CHF	SA-6MWT app for independent use at home	1. 6MWTApp accurately predicted the distance walked during a 6MWT and 2. It was easy to use and yielded accurate repeatable measurements in the clinic and at home	1. Good correlation with measured distance, r=0.89 (95% CI 0.82 to 0.95). For those with PAH, the r=0.75 (95% CI, 0.54 to 0.96) 2. Home-based walked distance (SA-6MWTApp) were highly repeatable (CV=4.6%) and correlated with in-clinic-measured distance (r=0.88; 95% CI 0.87 to 0.89)
Fox <i>et al</i> Israel (2011, 2013) ^{35 36}	1. Case-series study/level 4/JP 2. Prospective cohort study with good follow-up (>80%)/level 1b/JP	1. 44 patients with PAH 2. 86 subjects (52 with PH)	Step oximetry system linked to a computer	1. The step-oximetry test was an informative test of functional capacity among patients with PH. 2. Patients with PH showed significant limitation in step climbing ability that correlated with functional class and 6MWT	1. Correlation between 6MWT and step climbing velocity r=0.66 (p<0.0001) and O ₂ desaturation during step climbing correlated with DLCO (r=-0.65, p=0.003) 2. Correlation between exercise performance on the step and 6MWT-climb index (r=-0.77, p<0.0001); saturation deviation on the step test correlated with diffusion capacity of the lung (r=-0.49, p=0.001)
Fruhwald <i>et al</i> Austria (2003) ²⁹	Case-series study/level 4/JP	5 patients with PAH who had received long-term treatment with aerosolized Iloprost	IHM in patients who had received long-term treatment with Iloprost	The IHM enabled reproducible measuring of drug-induced variations in PAP	Mean PAP from 68±13 to 49±11 mm Hg (mean time of 49±8 min). Thereafter, PAP returned to pre-inhalation levels

Continued

Table 1 Continued

Author Country (year of publication)	Type of study/level of evidence (CEBM)/type of publication	Study population	Intervention	Summary of results	Significance level
Gregoriotti <i>et al</i> Argentina (2015 and 2016) ^{33 34}	Case-series study/level 4/CP	162 and 118 patients, respectively, with PH	Utilisation of a DP during the 6MWT, and its possible association with other parameters of clinical and prognostic relevance in PH	The addition of a DP to the 6MWT provided valuable data for evaluation and follow-up of patients with PH The number of footsteps was inversely correlated to clinical and laboratory parameters of prognostic relevance in PAH	In both studies, there were significant associations between pro-BNP levels and the number of footsteps (p=0.001) and WHO functional class (p=0.003 and p=0.009, respectively)
Tonelli <i>et al</i> USA (2013, 2014) ^{38 39}	Prospective cohort study with good follow- up (>80%)/level 1b/ 1. CP and 2. JP	1. 43 subjects (28 with PH and 15 healthy controls) 2. 50 subjects, 20 of them with PH	A portable impedance cardiography with wireless monitoring via a Bluetooth to determine HR and CO during a 6MWT and HR acceleration and decay slopes during the 6MWT	1. Wireless impedance cardiography showed lower distance walked, heart rate recovery (1 min) and CO change in PH under treatment than in healthy controls 2. The HR acceleration and decay slopes during the 6MWT were different in PH, compared with other lung diseases and healthy controls	All the comparisons in HR curves and acceleration rates were statistically significant (p<0.001)

CEBM, Centre for Evidence-Based Medicine; CHF, congestive heart failure; CI, cardiac index; CMR, cardiovascular magnetic resonance; CO, cardiac output; CP, conference paper; CV, coefficient of variation; DLCO, diffusing capacity of the lungs for carbon monoxide; DP, digital pedometer; HR, heart rate; IC, impedance cardiography; IHM, implantable haemodynamic monitor; JP, journal paper; 6MWT, 6-minute walk test; 6MWTApp, 6-minute walk test app; NR, not reported; PAH, pulmonary arterial hypertension; PAP, positive airway pressure; PH, pulmonary hypertension; PVR, pulmonary vascular resistance; SV, stroke volume.

Table 2 Summary of studies evaluating eHealth as part of the medical management in PAH

Author/ country	Type of study/level of evidence (CEBM)/type (and year) of publication	Study population	Intervention	Results	Significance level
Benza <i>et al</i> USA ²¹⁻²⁶	1. Case-series studies/level 4/CP (2012, 2015 and 2016) 2. Retrospective cohort study/level 2b/JP (2015)	11 patients with advanced PAH, 6 of them with a recent hospitalisation for RHF CHAMPION study population	Haemodynamically guided management of patients through Cardiomems	1. CardioMEMS monitoring was safe at rest and during a 6MWT in PAH, and reduced hospitalisation rates in HF patients with PH 2. CardioMEMS associated to a greater PAP reduction through medication changes in the ambulatory setting. Among patients with PH, there was a reduction in the composite endpoint of death and HF hospitalisation with ongoing knowledge of haemodynamics, but no difference in survival	1. 36% reduction in HF-related hospitalisation rates (0.60 vs 0.94, HR 0.64, 95% CI 0.51 to 0.81, p=0.0002) 2. Composite endpoint: HR 0.74, 95% CI 0.55 to 0.99, p=0.04 Survival: HR 0.78, 95% CI 0.50 to 1.22, p=0.28
Raina <i>et al</i> USA ³⁰	Retrospective cohort study/ level 2b/CP (2014)	314 WHO Group 2 patients with PH from the CHAMPION study	Retrospective analysis of Cardiomems patients	Cardiomems associated with hospitalisation reductions in Group 2 patients with PAH	HR 0.64, 95% CI 0.51 to 0.81

CEBM: Centre for Evidence-Based Medicine, Oxford University(<https://www.cebm.net/2009/06/oxford-centre-evidence-based-medicine-levels-evidence-march-2009/>). CHAMPION study population: 550 patients with heart failure with Cardiomems; 314 of them with WHO Group 2 PAH. CEBM, Centre for Evidence-Based Medicine; CP, conference paper; JP, journal paper; PAH, pulmonary arterial hypertension; PAP, pulmonary arterial pressure; PH, pulmonary hypertension; RHF, right heart failure.

step-oximetry devices to capture wireless information on exercise capacity and (4) ambulatory impedance cardiography to evaluate haemodynamic variables during all daytime activity. The findings are summarised as follows:

1. Benza *et al* explored the feasibility and safety of CardioMEMS® (Abbott Laboratories; Atlanta, USA) demonstrating that its use was safe at rest, during exercise^{21–23} and in combination with the use of cardiac MRI,²⁴ and was associated with a greater pulmonary artery pressure reduction through medication changes in the ambulatory setting.²⁵ The use of CardioMEMS was associated with a 36% reduction in heart failure-related hospitalisation rates (0.60 vs 0.94, HR 0.64, 95% CI 0.51 to 0.81, $p=0.00020$).²⁶ In a retrospective analysis of the CHAMPION trial, 314 patients with HF who met the definition of pulmonary hypertension (PH) (151 with CardioMEMS vs 163 control) were followed for an average of 15 months. The CardioMEMS group experienced a reduction in a composite endpoint of death and HF hospitalisation (HR 0.74, 95% CI 0.55 to 0.99, $p=0.04$).²⁷ Biederman *et al* showed that the use of CardioMEMS allowed reliable measurements of right ventricle cardiac output.²⁸ Fruhwald *et al* used an early model of IHM (Chronicle, model 9520; Medtronic, USA) to demonstrate the need for improvement of treatment models with long-term aerosolised Iloprost.²⁹ Raina *et al* conducted a retrospective analysis of the CHAMPION trial concluding that in patients with HF with marked limitation of their physical activity, those with pulmonary hypertension (Group 2 of the WHO classification) had higher hospitalisation rates than those without pulmonary hypertension (0.77/year vs 0.37/year, HR 0.49, 95% CI 0.39 to 0.61), and that in patients with and without PH, ongoing knowledge of CardioMEMS data resulted in a significant reduction in HF hospitalisation (HR 0.64, 95% CI 0.51 to 0.81 for patients with PH and HR 0.60, 95% CI 0.41 to 0.89 for non-PH). The relative risk reduction (RRR) was lowest in the patients with transpulmonary pressure gradient >15 mm Hg (RRR 30%) and pulmonary vascular resistance >3 Woods Units (RRR 33%). In addition, there was a non-significant trend towards improved survival with knowledge of CardioMEMS parameters (HR 0.78, 95% CI 0.50 to 1.22).³⁰
2. Brooks *et al* and Gregoriotti *et al* showed the feasibility and accuracy of a smartphone-based self-administered 6-MWT (6MWTApp) and a digital pedometer in measuring physical activity level and exercise capacity. The number of footsteps during the 6MWT correlated with clinical and laboratory parameters of prognostic relevance in patients with PH such as pro-BNP levels and WHO functional class and provided valuable data for the evaluation and the follow-up of people with PH.^{31–34}
3. Fox *et al* demonstrated that a step oximetry system equipped with pressure sensors and pulse oximeter linked to a computer provided useful information on the functional capacity of patients with WHO group

1 PAH (idiopathic form of PAH) and group 4 PAH (chronic thromboembolic PH), strongly correlating with functional class and 6MWT walk distance.^{35 36}

4. Arelli *et al* showed a real-time wireless impedance cardiography (Physioflow Enduro; Manatec Biomedical, Paris, France) for non-invasive determination of haemodynamic parameters in pulmonary hypertension patients during their 6MWT. Stroke volume, cardiac output and cardiac index increased with activity in direct relation to the distance walked, providing valuable insights into the haemodynamic changes during exercise.³⁷ Other authors (Tonelli *et al*) used this portable device to enable continuous non-invasive evaluation of haemodynamic variables on a beat-to-beat basis during all daytime activity. In another study, the same researchers demonstrated that the acceleration and decay of the heart rate slopes during a 6MWT exercise in patients with PAH were different compared with lung airway diseases and healthy controls.^{38 39}

DISCUSSION

To our knowledge, this is the first literature review investigating the use of eHealth in PAH. In general, the reported eHealth solutions have demonstrated to be safe, accurate and reliable, and could contribute to the clinical management of people with pulmonary hypertension. In particular, the reported association between CardioMEMS and a reduction in hospitalisations rates in PAH Group 2 (patients with left-sided HF and concomitant PH) appears promising. In another study (based on the same HF population), the use of this device was associated with a reduction in a composite endpoint of death and HF hospitalisation, although without any difference in survival. CardioMEMS combined with CMR could also contribute to providing an accurate calculation of non-invasive cardiac output, necessary to estimate non-invasive pulmonary vascular resistance, a key feature in the diagnosis of patients with PAH.

The smartphone-based self-administered 6MWT app (SA-6MWT App) demonstrated to be accurate and reliable at clinics and at home. A digital footstep counting and a computerised step-pulse oximeter during the 6MWT added valuable data for evaluation and follow-up of these patients, showing a strong correlation to clinical and laboratory parameters of prognostic relevance in PAH. The ambulatory impedance cardiography increased the value of 6MWT providing valuable insights into the haemodynamic changes that occur during exercise in patients with PAH, under treatment and in comparison, with other lung diseases and healthy controls.

In general, this review highlights a lack of studies assessing the impact of eHealth on hard clinical endpoints (such as mortality and hospitalisations) and on QoL. This makes it difficult to draw further conclusions on the effects that the introduction of eHealth may have for the people living with PAH, their health professionals and their families. However, based on promising reports in the HF domain,^{40 41} possible benefits of using eHealth in the management of PAH can be postulated.



In particular, some additional observations may be considered. First, looking at the characteristics of the study participants, most participants pertained to Group 2 of the WHO classification, haemodynamically referred to as post-capillary PAH. This is a crucial observation as the patients with left-sided HF have a completely different management (and treatment) compared with the WHO Group 1 of patients with PAH (haemodynamically referred to as pre-capillary PAH). This type of PAH includes, along with the idiopathic form of the disease, the connective tissue disease-associated PAH. Both conditions are considered rare diseases⁴² (or perhaps underdiagnosed).⁴³ These entities present a management challenge for PAH services and health organisations derived from a high dependency of frequent visits at tertiary centres and highly specialised care, with close supervision of health measures, as well as symptoms and treatment monitoring. We propose that any future attempt to trial the impact of eHealth in PAH should only recruit patients with precapillary PAH, excluding participants with PH associated with HF or severe chronic obstructive lung disease.

Second, one of the most important factors determining the success versus failure of an eHealth intervention may be dependent on delivery of an effective medical intervention and the presence or absence of required medical action. Thus, it is important to examine the whole workflow that the eHealth intervention will support and the impact on the clinical management.⁴⁴ In the present review, only two studies^{25 30} considered the use of eHealth as part of the medical management (table 2). In both cases, most of the patients pertained to the post-capillary form of PAH, which represents a serious limitation in terms of validating the usefulness of these eHealth interventions in patients living with precapillary forms of PAH.

Third, in the two studies referred to in table 2, the use of eHealth showed reductions in hospitalisation and, in one of them, a non-significant trend towards improved survival.^{26 30} Again, it was not possible to extrapolate this positive outcome to the patients with the precapillary form of the disease, as most of the participants included in these studies were patients with post-capillary PH.

Lastly, none of these studies assessed the impact that eHealth has on the well-being of the patients, usually represented by the assessment of QoL. Hence, the assessment of QoL is of great interest in PAH,⁴⁴ as in any other incurable disease where new treatments with small survival benefits may be offset by QoL deterioration.⁴⁵ Thus, specific tools have been developed to measure QoL in people with PAH.⁴⁶

Opportunities and future directions

Since most of the patients with PAH live geographically away from highly specialised PAH centres (especially in sparsely populated areas), these patients generally need to travel long distances for their regular clinical appointments. The remote monitoring of health data and vital signs (such as blood pressure, heart rate and oxygen saturation [at rest or during exercise]) could bring some benefits.^{41 47} This possibility becomes more relevant in such cases where urgent consultations and first-line visits can only be attended by local non-PAH

specialised health services.⁴⁸ The monitoring of clinical symptoms such as weight gain, light-headedness, hypotension, fainting episodes (syncope), chest pain and palpitations or increasing shortness of breath, either at rest, during daily exercise or during home-based pulmonary rehabilitation sessions,⁴⁹ could contribute to the prompt detection of clinical deterioration and early intervention.

Considering the effects of eHealth in empowering people with chronic disease by improving their self-management skills⁵⁰ and their QoL,⁵¹ a similar positive impact in patients with PAH could be proposed. Unfortunately, based on the current evidence, this hypothesis cannot be corroborated. For this reason, QoL should be considered as the main endpoint in future studies. Some learnings can be taken from eHealth trials in patients with HF. For example, the indistinct inclusion in HF trials of patients with different aetiology (preserved ejection fraction vs reduced ejection fraction) has led to uncertainties in the study outcomes.⁵² In a similar way, PAH studies should include only patients with pre-capillary forms of PAH, differentiated from left-sided-associated disease, as they are different entities with a dissimilar management and treatment.

Finally, as the success (or failure) of eHealth may rely on delivery of an effective medical intervention, the impact of eHealth on patients with PAH should be evaluated with the technology integrated into their medical management. In addition, the use and grade of satisfaction from patients and caregivers could be a relevant domain to evaluate.

CONCLUSION

The evidence on the use of eHealth in patients with PAH is scant and mainly envisioned to validate the accuracy and reliability of specific technology tools. However, the potentials of using eHealth to improve the management of patients with PAH needs to be considered. A comprehensive analysis of the impact that the utilisation of eHealth may have on hard clinical outcomes and QoL needs to be addressed. We see this study as a general call for researchers to investigate the impact that eHealth may have on the holistic management of people with PAH.

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