



# Palliative care in pulmonary arterial hypertension

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## Purpose of review

Developments in the management of pulmonary arterial hypertension have significantly improved prognosis changing this from an acute to a chronic disease. Despite optimal treatment many patients still have a high-symptom burden both because of the disease and the side-effects of therapy, consequently there is an increasing need for a palliative care approach to improve the quality of life for this patient group. This review article will outline the need for palliative care support for patients with pulmonary arterial hypertension, discuss the barriers that currently exist and suggest how this may be improved.

## Recent findings

Studies have been conducted which explore the role of palliative care in pulmonary arterial hypertension including physicians attitudes and the current barriers that exist to prevent its implementation.

## Summary

Specialist palliative support is utilized in the minority of patients with pulmonary arterial hypertension despite a need for symptom control. Patients may benefit from the introduction of a palliative care approach as part of their standard care, but to achieve this there needs to be a greater understanding of the role of palliative care by both clinicians and patients and more research into the benefits for patients with pulmonary arterial hypertension.

## Keywords

palliative care approach, pulmonary arterial hypertension, quality of life

## INTRODUCTION

Pulmonary arterial hypertension (PAH) is a rare disease characterized by a progressive increase in pulmonary vascular resistance resulting in right heart failure and death. Over the past two decades there have been dramatic improvements in the management of the disease and the development of new drug therapies which have improved prognosis but life expectancy remains poor with an estimated 7-year survival of around 50% [1,2]. As there is currently no cure for PAH, therapy is targeted to improve symptoms, allowing patients to become more active and slowing disease progression [3].

## PALLIATIVE CARE IN PAH

Before the advent of disease modifying therapies patients with pulmonary hypertension had a mean life expectancy of approximately 2.8 years [4]. Recent improvements in treatment have improved survival but despite these patients continue to experience a constellation of symptoms [5] which negatively affect quality of life (QoL) [6] in what is now a chronic disease. The palliative care approach can improve the QoL of patients through early identification, assessment and treatment of pain

and other problems, including physical, psychosocial and spiritual [7]. This approach has mostly been studied and implemented in patients with cancer, but has also been found to be beneficial for patients suffering from chronic lung diseases, such as chronic obstructive pulmonary disease, interstitial lung disease and pulmonary hypertension [8,9], as well as in patients with chronic heart failure [10,11].

Nearly 70% of patients with PAH report moderate or severe dyspnoea and fatigue, the other most common symptoms are drowsiness (39.9%) and pain (34.9%) [12]. The physical symptoms may be further impacted by depression, which is severe in 15% of patients and mild to moderate in a further 40% [13], and by the patients' perceived lack of control over their condition [14].

The palliative care approach focuses on both the psychological and physical components of

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## KEY POINTS

- Patients with PAH may benefit from a palliative care approach as part of standard care.
- Palliative care can exist in parallel with aggressive PAH disease-targeted therapies.
- There is a need for more education of both clinicians and patients about the benefits of palliative care.
- The access to specialist palliative care provision needs to be improved to ensure that this is available to all patients with PAH when appropriate.

dyspnoea, patients may benefit from supplementary oxygen and by the careful use of opioids and anxiolytics to manage both the sensation of dyspnoea and anxiety, they are also encouraged to enrol in pulmonary rehabilitation programmes early in their disease course to learn how to control their breathlessness [15].

The guidelines for the management of PAH suggest referral to specialist palliative care services when appropriate [16]. It can be argued that as PAH is now a chronic disease and managed in reference centres by experienced multidisciplinary teams that palliation of symptoms should be part of the standard care for all patients. Referrals to specialist palliative care teams should be made when there are short-term acute problems beyond the expertise of the PAH team and for patients towards the end of life. Most physicians (91%) report utilizing palliative care consultations at least once in the previous year [17<sup>\*\*</sup>] but the patient perception of palliative care intervention is less with only 1.4% of patients reporting having a palliative care provider involved in their care [12]. The level of support has been found to increase towards the end of life but even at this late stage just 14% of patients had specialist palliative care providers involved in their care at time of death [18]. When questioned in this retrospective study many surrogates were unaware of specialist palliative care services available to the decedents, patients died with a high-symptom burden, especially dyspnoea and often in an Intensive Care setting [18]. There are parallels with the level of palliative care provision in other patient groups with noncancer end-stage diseases such as chronic obstructive pulmonary disease [19] as despite the recommendations of respiratory societies [20] the rates of involvement are also low. Noncancer patients also tend to be referred much later in the disease trajectory than cancer patients and have lower survival rates [21].

The involvement of palliative care in patients with pulmonary hypertension seeks to increase

comfort and dignity for as long as the patients have to live with the emphasis on identifying and treating symptoms and attempting to understand the patient's goals of care [6]. This can be delivered in parallel to aggressive medical therapy when patients remain symptomatic because of the underlying disease or from the side-effects of treatment. The facilitation of advanced care planning early in the disease can also allow patients the peace of mind to know that their priorities will be honoured if they come to a stage when they can no longer speak for themselves and can help reduce some of the burden of end-of-life decisions for next of kin [22].

## TIMING OF PALLIATIVE CARE REFERRAL

Palliative care can be introduced at any time in the disease process to improve symptoms, the World Health Organisation's definition of palliative care mentions that it should be applicable early in the course of an illness, in conjunction with other therapies that are intended to prolong life [7]. Palliation of symptoms should be considered in all patients with PAH and, as there are no restrictions regarding specific prognosis, specialist palliative care referrals can be made at any time in the disease trajectory although end of life and hospice care in some countries requires a terminal diagnosis with a life expectancy of less than 6 months. As adult patients usually have decision-making capacity early in the chronic disease process it is important to determine their immediate and long-term goals of care. It is suggested that the discussions about palliative care should be guided by their articulated goals, expressed preferences and questions [15].

PAH is a chronic disease with long-term limitations and intermittent serious episodes and in comparison to cancer narratives, which have a well-rehearsed beginning, middle and anticipated end, people with diseases which involve organ failure struggle to tell their stories being unclear when the illness began, or how one event linked to another [23]. It has been shown that ambulatory patients with heart failure tend to substantially overestimate their life expectancy compared with model-based predictions, this discordant prediction of survival may affect decision making regarding advanced therapies and end-of-life planning [24] as patients with a noncancer disease speak less about death, hoping instead to avoid further deterioration and often do not have a clue about what will happen at the end of life [23].

In PAH acute deteriorations in right heart failure are generally associated with admission to hospital and each exacerbation maybe life-threatening. If the patient survives there is often a gradual

deterioration in health and functional status. The timing of death, however, remains uncertain and in patients with advanced heart failure – more than 60% died when expected to live for at least a further 6 months [25]. It has been shown that clinicians are often over optimistic about the patient's prognosis and the stronger the doctor–patient relationship is the less accurate the prognosis becomes [26], this can potentially lead to more invasive and potentially inappropriate intervention at the end of life.

This prognostic uncertainty should not prevent us talking with our patients about the issue of palliative care as it is easy to fall into the trap of prognostic paralysis [27]. With appropriate training and experience all PAH teams should be able to manage the basic palliative care needs of their patients. It has been suggested that palliative care becomes an essential component of comprehensive care for all patients with chronic critical illness as the disease enters the chronic phase and that close attention to palliative care principles is a paramount responsibility for all clinicians on the treatment team [28]. Normalizing the discussions about palliative care support and end-of-life issues and introducing them earlier allows patients to learn about their right to high-quality symptom management. This could help patients to make more informed choices, achieve better palliation of symptoms and have more opportunity to work on issues of life closure [29].

The optimal time to involve a palliative care specialist will be different for each patient; consults should be considered when patients present with:

- (1) challenging psychological, social or spiritual symptom management issues;
- (2) a deterioration in health status;
- (3) a significant symptom burden despite optimal therapy;
- (4) recurrent admissions with right heart failure;
- (5) high scores in PAH QoL questionnaires;
- (6) a need for family support and planning.

## **BARRIERS TO ACHIEVING OPTIMAL PALLIATIVE CARE**

The palliative care approach can be used by all clinicians to manage symptoms at all stages of the disease, this concept is often misunderstood and confused with end of life or hospice care by both by patients and their physicians [12] although the two are distinct. There is a common misconception that palliative care involvement may prevent concurrent treatment with PAH targeted therapies and whilst this may be the case in some countries, for patients entering hospice care at the end of life it is

possible to aggressively treat the underlying disease whilst also treating the persistent symptoms. Despite this 20% of physicians felt that palliative care involvement resulted in a difficulty in treating patients as aggressively as necessary as there was a restriction on medications that can be used in the palliative care setting [17<sup>\*\*\*</sup>].

Over half of physicians have concerns that a palliative care approach would not be approved by patients and their families. The most common reason to discuss palliative care was at the end of life when actively dying (59%) or when a hospice referral was made (40%). Physicians were concerned that a palliative care approach would be perceived as giving up hope and 43% were concerned about demoralizing the patient and their family [17<sup>\*\*\*</sup>]. Although, clinicians may often be concerned about the consequences of discussions about prognosis, in the appropriate context and when conducted with compassion such conversations do not take away hope [30]. It has been shown that many patients with advanced illness often would like to have these conversations although they are often lacking [31]. By introducing these discussions as part of standard care all patients would be offered the opportunity to consider their options and to make their wishes known to both healthcare professionals and their family.

Although 88% of physicians reported a high degree of confidence regarding discussing end-of-life care plans with patients and relatives only 43% of these physicians felt comfortable in assessing end-of-life, whereas 33% felt happy managing it [17<sup>\*\*\*</sup>]. Despite reporting a confidence in discussing palliative care it is not always considered as a treatment option when managing the care patients with PAH. When faced with the care of patient who has only 6 months to live only 40% of PAH physicians considered a specialist palliative care consult and only 12% considered referral to a hospice [17<sup>\*\*\*</sup>].

Patients on continuous intravenous prostanoid therapies to treat PAH face unique challenges with respect to palliative and end-of-life care. The role of prostanoids as life-prolonging therapies may inhibit initial specialist palliative care consultation and the complexity of use may constitute an impediment to patients with PAH benefiting from palliative expertise in the hospice care setting [32]. Some healthcare systems will not fund prostanoid therapy for patients that need hospice care which can result in patients being reluctant to enter hospice as they have a personal moral objection to the discontinuation or downward titration of prostacyclin therapy, viewing it as suicide [33].

There are concerns that palliating symptoms of breathlessness with opioids may hasten death

through respiratory depression although several observational studies have found no evidence to support this theory [34–36]. There is, however, evidence to support that adding palliative care to disease-targeted therapy in other conditions can result in less depression and improved survival [37].

## LOOKING FORWARD

It has been suggested that one way to better integrate palliative care is to adopt a model in which clinicians treat the disease whereas at the same time the palliative care team treats the symptoms, provides emotional support and facilitates discussions about goals of care with all clinicians working towards a shared goal of sustaining life [38]. Despite this there are only limited specialist palliative care resources available and as it is, therefore, essential the PAH team address the palliative care needs of the patient whilst also aggressively treating the underlying disease. To ensure that this is possible there needs to be education and training of the PAH teams and ongoing evaluation of the perception of information received and collaboration across boundaries and disciplines. To improve communication with all concerned the trajectory of PAH also needs to be better defined and better understood. Information that provides an accurate depiction of the palliative care approach and potential benefits for patients may encourage more open dialogue and willingness to consider earlier in disease and achieve best possible QoL.

Although, increasing numbers of patients suffering from nonmalignant diseases are equally in need of specialist palliative care they are only slowly getting access to this and hospice services, mainly because of inadequate financing models and more political developments are needed to fill this gap [39]. We could argue that specialist palliative care should aspire to treat all patients and should fight for more resources but, depending on the robustness of local health economies, the palliative care approach may need to develop increasing flexibility and adaptability to enable the successful management of patients with PAH.

## CONCLUSION

Future PAH guidelines should consider recommending the need for a palliative care approach to be part of the routine management of the patient who has symptoms despite optimal treatment of the underlying disease [15]. The normalization of the role of palliative care in PAH management and the focus on symptom relief may alleviate some of the stigmas around palliative care with the emphasis moving

from the traditional association with dying to living well with a chronic disease. There is, however, a need for education to ensure that PAH patients and clinicians have an accurate depiction of palliative care and are aware of the potential benefits. Although the palliative care approach has the potential to play an important role in the care of PAH, it is essential that more information is gathered through research examining the impact of interventions and the best practice for integration into patient care.

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- of special interest
- of outstanding interest

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