

# Serious illness conversations in pulmonary hypertension

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## Abstract

Pulmonary arterial hypertension has evolved from a fatal disease with few treatment options to a chronic condition with improved survival. This improvement is possible through development of effective therapies as well as the expansion of risk stratification scores to assist clinical decision making. Despite improved disease control, quality of life, and overall prognosis, many challenges remain. The treatment itself is burdensome, with significant impact on quality of life. Many patients with pulmonary arterial hypertension still present with advanced, often end-stage disease. Increased use of mechanical circulatory support and catheter-based interventions have expanded use of extracorporeal life support and right ventricle assist devices. For these reasons as well as the long-term relationships pulmonary hypertension physicians have with patients and their families, navigating the course of the illness in a considered, proactive way is essential. Understanding individual goals and revisiting them as they change over time requires comfort with the conversation itself. There are many barriers and challenges to having effective, compassionate conversations in the clinical setting with time constraints being the most often cited. Compressed visits are necessarily focused on the clinical aspects, therapy and medication adherence and tolerance. Clinicians are sometimes wary of diminishing hope in the face of ongoing treatment. Having sufficient experience and comfort with these discussions can be empowering. In this paper, we discuss the challenges involved and propose a framework to assist in incorporating these discussions into clinical care.

## Keywords

communication, serious news, goals of care, quality of life, values-concordant care

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## Introduction

Pulmonary hypertension (PH) is a serious and progressive disease of the pulmonary vasculature with multifactorial pathogenesis. It can be divided into five distinct categories including: pulmonary arterial hypertension (PAH) (Group 1), PH due to left heart disease (Group 2), PH due to lung diseases and/or hypoxemia (Group 3), PH due to pulmonary artery obstruction including chronic thromboembolism (Group 4), and PH related to multifactorial causes (Group 5).<sup>1</sup> Regardless of etiology, PAH denotes the presence of pulmonary vasculopathy with attendant intimal proliferation, medial hypertrophy, and plexiform lesions. The obstructive remodeling of pulmonary vascular bed, in addition to vasoconstriction leads to a progressive increase in pulmonary vascular resistance and increased afterload on the right ventricle (RV). Whether the RV adapts or

maladapts in response is the key determinant of symptoms and survival as death is most often the result of RV failure.<sup>2</sup> Assessing RV function – including measurements of mean right atrial pressure, cardiac output, and cardiac index – is important as it has been shown to be a significant predictor of survival in PAH.<sup>3</sup>

The diagnosis of PAH can be challenging and is often delayed due to under recognition, with subsequent implications on survival, as delayed diagnosis is associated with higher mortality.<sup>4</sup> Prior to the availability of PAH-specific therapy, patients captured in the original National Institutes of Health registry had a median survival from diagnosis of

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only 2.8 years, and 5-year survival of 34%.<sup>5</sup> By contrast, the more recent registry to evaluate early and long-term PAH disease management (REVEAL Registry), demonstrated significant improvements in 5-year survival estimated at 57%.<sup>6</sup> Similar survival rates have been reported in contemporary registries of European patients with PAH.<sup>7</sup> The availability of a wider assortment of medications over the past three decades has resulted in improved patient survival and quality of life. Despite improvements in survival, patients with PAH face multiple hospitalizations over time and challenges with functional status and in the event of circulatory arrest, long-term outcomes from resuscitation remain poor.<sup>8</sup>

Patients who progress to severe right-sided heart failure require comprehensive care and whenever possible, should be treated at centers capable of providing a range of advanced treatment options including medical therapy, extracorporeal life support, and lung transplantation. These centers have expertise in knowing both when to apply and when to not recommend specific advanced therapy. Advanced therapies such as extracorporeal membrane oxygenation and right ventricular assist devices may be considered in very select patients.<sup>9</sup> Conversations framing the risks and benefits of such interventions, in the face of admittedly uncertain outcomes, require deliberate shared decision making.<sup>10</sup>

Given the complexity of care required of these patients, there has been a shift towards accrediting centers who have demonstrated excellence in caring for these patients. In rare disease states, it is not only the diagnostic testing that requires a high standard but also the therapeutic management decisions, multidisciplinary healthcare team members familiar with the care of rare disease patients, and discussions of ongoing treatment in the context of prognosis, quality of life, and mortality. Inpatient pharmacotherapy decisions are improved by clinical pharmacists embedded in the PH team, and help to ensure patients have proper training and access to advanced therapies to ensure safe transitions of care.<sup>11,12</sup> Clinician-led conversations about serious illness that are intended to seek to elicit goals, values, and preferences is the first step in allowing for shared decision making. Importantly, such conversations are associated with less aggressive care at end of life, positive family outcomes, decreased anxiety, and distress at end of life as well as enhanced goal-consistent care.<sup>13,14</sup>

Despite the obvious indication for these conversations, and the very real benefits that patients and families derive from them, they are not reliably embedded in clinical practice. Although many paradigms for structured communication around serious news exist, to our knowledge they have not been systematically applied to the population of PAH patients in a routine and standardized way. This paper will identify barriers to effective communication in PAH patients, review best practices, and offer solutions to overcoming the obstacles to apply best

practices to implement serious news conversations in the PAH population.

## **Barriers to effective serious illness conversations**

Unlike more common diseases like cancer, the rare nature of PAH makes it unlikely that patients and families will have provisional awareness of it as a “devastating diagnosis.” Patients often remain optimistic about their long-term survival and transplant candidacy. This complicated understanding of prognosis is compounded in situations of low health literacy. It falls to the clinician to ensure effective, compassionate disease-state education.

There are several factors that may result in clinicians avoiding or simply delaying conversations around serious illness. Uncertainty of prognosis can be difficult to communicate and some clinicians may harbor fears that such a conversation would serve to take away hope from their patients.<sup>15</sup> Clinicians may be uncertain of their skills at broaching the topic and may not have spent time cultivating a skillset that allows for emotion-laden, complex conversations. To do this work effectively and compassionately, it can be helpful to seek out professional development in the form of communication training. A variety of models share the goal of understanding a patient’s story before co-designing a plan of care that aligns with their values. An accessible communication skillset for conflict is similarly invaluable when intra-family dynamics or inter-care team disagreements create discord around decisions.

Identifying the right time for the conversation from the patient perspective with respect to both the disease trajectory and titration of therapy is challenging. Early clinical encounters in PH are often focused on building trust, outlining diagnostic strategies, and educating patients on treatment options. Patients generally derive benefit from the initiation of therapy, and it is natural to focus on the improvement in subsequent encounters. Later in the disease course, there may be need for expedited decisions or frequent admissions for RV failure. Finding the time for such conversations can be a logistical challenge. Outpatient clinics are tightly booked and discussions during acute hospitalizations present their own challenges.

When these conversations are forced to occur, it is too often in urgent situations, and necessarily focused on establishing resuscitation and intubation status. Conversations undertaken in extremis do not allow opportunity to address preferences or explore understanding. Conversations about patient centered goals such as quality of life, caregiver or family support, candidacy for advanced therapies, symptom relief and mitigation of emotional, spiritual and physical suffering require time and space to truly be generative.

### Which patients?

Because of the high morbidity and mortality associated with PAH, serious illness conversations should be conducted with all patients. PAH is unique in that withdrawal of prostacyclin medication can result in refractory RV failure, and as such, transparent discussion that goes beyond risks and benefits and considers the emotional toll of being dependent on continuous therapy is warranted. Transitions off these medications for hospice care confers additional layers of complexity that must be navigated skillfully, with nuanced discussion.<sup>16</sup> At all points in the treatment course, the trade-offs in terms of quality of life and symptom relief must consider patient lifestyle factors, support network and capacity to absorb and integrate difficult news.

These conversations of course are not singular. They should occur at regular intervals while on therapy to reassess whether goals are being achieved. Clinicians must strive to cultivate a safe space where patients feel comfortable disclosing the difficulties of disease and treatment as well as the trade-offs, regrets, and successes of decisions already made. Patient should be stable and able to consider goals, values, and preferences and not under duress of acute illness.

### When?

Although the ideal is to have these conversations building trust and facilitating an ongoing relationship longitudinally, sometimes this is not possible, and it can be helpful to identify clinical triggers that would prompt a conversation, such as clinical deterioration over time, multiple inpatient admissions, or in the event of need for transplant evaluation. For clinicians who utilize risk stratification, high-risk patients can be identified, and conversations approached with the context of prognosis. Possible other triggers include an event that harbors prognostic significance for the patient, at the request of patient/family, changes in function or dependence, and recurrent or prolonged hospitalizations.

There are many different topics clinicians need to be able to address. First will be delivering the news of this serious diagnosis, along with discussion of prognosis based on validated risk scores. In cases of heritable disease, there may be an additional need for genetic counseling. Quickly following are discussions regarding treatment options, which depending on the severity of disease may include recommendations for fairly rapid initiation of multiple oral drugs that are quite costly, or in the most severe cases, a regimen which includes parenteral prostacyclin therapy, all of which have significant side effects. Discussing anticipated disease trajectory and impact on functional capacity and quality of life is necessary, though admittedly difficult to predict given variability in underlying driving factors and individual response to therapy. In the event of disease progression, discussions around potential need for lung transplantation may become necessary. End-of-life discussions may have the added challenge that discontinuation of parenteral therapy will hasten death.

There are several well-validated approaches to engaging patients with serious illness in shared decision making. Most share a common set of goals: contextualize the situation for the patient, elucidate patients' goals and priorities as well as their values, attend to emotion, describe treatment options and include palliative approaches that most align with patient goals, affirm continued commitment to support patient and family even as goals and treatment may change.<sup>17-19</sup> Here, we will focus on methodology that is utilized in VitalTalk curriculum, as well as the CLEAR (Connect, Listen, Empathize, Align, Respect) curriculum at Henry Ford Health System, which has its foundation in VitalTalk methodology.<sup>20,21</sup> Both programs provide evidence-based communication skills training utilizing improvisational actors to provide feedback in real time to clinicians.

### REMAP

Matching patient values with treatment recommendations requires a structured conversation, and REMAP (REframe, Map, Align, Plan) is a roadmap that allows for co-creation of a treatment plan with emphasis on patient values and goals. Preferences for content and delivery vary between patients, as does health literacy, degree of acceptance, and openness to difficult conversations. The sequence and cadence of this template allows clinicians to elicit the patient's preferences about how much information they feel suited to receive.

### REframe

Opening the conversation by exploring the patient's understanding allows the clinician to appreciate the degree of acceptance and understanding of illness. When clinicians "Reframe," they assess the patient's understanding of their illness trajectory and, if necessary, may provide new information. When assessing understanding, clinicians should be attuned to knowledge gaps that may need to be filled, including burden of disease, functional expectations, and trajectory.

*Ask-Tell-Ask:* With this micro-skill the clinician engages the patient by asking first what s/he knows, allows for delivering news in a larger context, and then waits for the patient's reaction, where emotion is expected. Once emotion has been tended to, the clinician may follow with a second ask to ensure the patient understood the news delivered. This is often referred to as a teach back.

Example:

PH clinician: What have you heard so far about what's going on with your heart?

Patient: When I was in the hospital, they told me my right heart was failing, but I'm not sure what that means.

PH clinician: You're right that your heart is failing. Would it be ok if we talk more about what may be causing it to fail?

Clinicians should expect that patients will respond with emotion when hearing serious news and must be ready to tend to those emotions concurrently. The E in RE is a reminder to deploy Empathy. Done effectively, this allows the emotional intensity to decrease enough that discussion can continue. This can be accomplished through the NURSE statements (Table 1).<sup>22</sup>

**Map: Mapping patient values**

Clinicians should intentionally explore the patient’s values with genuine curiosity and without predetermined agenda before presenting treatment options. Most people have complex sets of values that can sometimes be in opposition to each other. They may wish to focus on maximizing quality of life over quantity while still citing a desire to participate in future milestone events. Helping the patient articulate these goals helps maintain a sense of personhood and mitigates feelings of hopelessness. This is also a time when fears such as symptom burden, suffering, and family dynamics can be explored. Articulating fears helps the patient to feel seen/understood and enhances therapeutic alliance. Asking open-ended questions aids in facilitating exploration of additional components of a patient’s value system, such as sources of strength or what functional changes and/or circumstances might lead the patient to reconsider or alter the focus of their plan of care. This will provide important information necessary to formulate an appropriate management plan that honors the patient’s stated values. When using this technique, it is important to use open-ended questions and is essential to continue exploring as the patient provides information.

Examples:

In order to figure out the best plan for you, it would help me if we spend some time talking about the things that are most important to you.

Would you be open to sharing a bit about what your life is like outside of the hospital?

If you were to get sicker, are there things you know you would want to avoid?

As you think about the future, what concerns you?

Follow-up:

I hear you saying that you want to avoid machines if possible, could you tell me more about what you mean by that?

**Align: Aligning with the patient’s stated values**

This step involves reflecting and summarizing the values the patient has expressed. By summarizing, clinicians are deliberately demonstrating their understanding of the patient’s values and priorities. The summary may lead to further mapping as the patient clarifies, expands, or qualifies what is important.

Examples:

It sounds as if your independence is very important to you, and you don’t want to feel as if you are a burden to your family.

It seems you are open to trying therapy that may have significant side effects, so long as it has a good chance of helping you to be more active.

The patient may either clarify a summary of what they have shared, which would afford the opportunity to continue mapping, or they could say, “Yes, that’s exactly how I feel,” which would prompt the clinician to continue to the next step.

*Asking permission:* Prior to moving on to the next step, it is important to ask for permission to move forward in the

**Table 1.** Statements to help deal with and support emotion.

NURSE statements for articulating empathy		
	Example	Notes
Naming	It sounds like you are frustrated.	In general, turn down the intensity a notch when you name the emotion.
Understanding	This helps me understand what you are thinking.	Think of this as another kind of acknowledgment but stop short of suggesting you understand everything (you do not).
Respecting	I can see you have really been trying to follow our instructions.	Remember that praise also fits in here (e.g. I think you have done a great job with this).
Supporting	I will do my best to make sure you have what you need.	Making this kind of commitment is a powerful statement.
Exploring	Could you say more about what you mean when you say that. . .	Asking a focused question prevents this from seeming too obvious.

From VitalTalk.<sup>23</sup>



conversation in order to respect the significance of the conversation and allow the patient to control the amount of information they feel they can receive in one encounter. Consider something like, “Would it be ok if I propose a plan that takes what you’ve told me into account?”. This provides the patient with an opportunity to decline if they are not in an emotional space where they are prepared to discuss the matter further.

### **Plan: Propose a plan**

At this point, the clinician proposes a treatment plan that has the best chance of achieving the patient’s values and goals by coupling the patient’s stated values with the clinician’s knowledge of medical treatments. The proposal is followed by an open-ended question that is intended to assess the patient’s reaction to the plan and identify need for follow-up information. An example may be, “What do you think about what I’ve said so far?”. Approaching the discussion with genuine curiosity helps promote honesty and diminishes hierarchy.

Example:

Given what you’ve told me about your goal to maximize your time as much as possible, I would recommend that you consider the intravenous medication we discussed.

Since you shared that avoiding repeat admissions to the hospital is the most important thing, let’s spend some time talking about disease self-management.

*Closing the discussion:* This provides another opportunity to assess the patient’s understanding of the proposed plan, an opportunity for the patient to ask questions, and a space for the clinician to affirm intention and commitment to guide the patient through their serious illness, thus further creating trust.

Examples:

What questions do you have for me about what we’ve discussed today?

I want you to know our team will be with you every step of the way.

Though I wish there was a cure for this condition, I’m confident that the path you’ve chosen is the best plan for you.

### **Sharing of prognosis**

Of course, illness may sometimes progress despite treatment and clinicians must give honest appraisals of prognosis and treatment effectiveness. Clinician disclosure of prognostic information, expressed in concrete and specific detail, is ethically responsible as it allows the patient to have

agency over how they spend their remaining time. This can be accomplished in many ways, including time-based projection or a more general discussion of the anticipated functional trajectory. Clinicians can chart the path of what is ahead with an honest discussion of what to anticipate in terms of further medical problems, hospitalizations and procedures, and how this might affect quality of life. It is important to acknowledge uncertainty of prognosis and align with hopefulness, “While I hope your experience is different than the majority of the patients, I want you to have a sense of what you can expect.”

Examples:

When we look at similar patients to you, with similar risk factors, we find that most are alive at 5 years.

I’m worried that you are going to have more frequent and longer admissions to the ICU for heart failure.

We’re in a different place now, and I’d like to ask your permission to talk about what I fear is ahead.

### **Conclusion**

As patients and their families affected by incurable diseases navigate the complex road of treatment and caregiving, their needs and wishes will evolve. Clinicians play a crucial role that involves knowing when to broach difficult conversations, give guidance, when to prognosticate and when to align with hope. Clinicians can learn to sense opportunities in reports of patients’ symptoms, clinical deterioration, or need for additional therapy. These conversations, done skillfully, help foster both trust and healing. They can empower patients to take an active role in disease self-management, share their fears honestly and when appropriate transition their goals of care.

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### **Authors’ contribution**

Each author contributed to the conception, design of the work, drafting as well as critical revision of the article and final approval.

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