

Organization and Delivery of Palliative Care for Pulmonary Arterial Hypertension: Case-based Discussion

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Our patient, as seen through the eyes of a palliative medicine team

This 43-year-old woman presents a history that serves as a valuable foundation for discussion of need for a structured platform for delivery of palliative care for persons with pulmonary arterial hypertension and associated right heart failure; as such, her case vignette is succinctly reviewed, with emphasis placed on features relevant to this topic.

Pre-existing and lifelong disease was present prior to diagnosis of pulmonary arterial hypertension (PAH), with major effects of additional non-PAH disease on functional capacity, quality of life and quantity of life, as contrasted to other age- and gender-matched persons. In-office outpatient therapy was intense and frequent, and allowed home life to be active and independent, albeit with likely adjustment of expectations to allow for such. With diagnosis of PAH there came a new uncertainty of outcomes, with high likelihood of development of additional multiple organ system life-altering disease. Effects of disease, chronic maintenance therapies, crisis-based salvage treatments, and heroic alternatives on symptoms, quality, and goals of life appeared not to have been fully discussed pre-emptively, reviewed in iterative fashion, nor periodically screened. Unexpected absence from disease-specific care was followed by development of new organ system diseases that multiplied symptomatology, altered prognosis, and increased intensity and burden of both hospital- and home-based care. Over an ensuing 7 years, the patient weathered multi-

ple, frequent, prolonged, and at times life-threatening intensive hospital stays, often with deep prognostic uncertainty. Each time she rebounded with lesser effect of medical therapies, lesser return to function and with greater chronic symptomatology and physical wasting, but to a life that she felt was meaningful and fulfilling at home, with family and purpose. Goals of care and end-of-life discussions appeared to occur first with surrogates in crisis-related fashion and timing, with more in-depth conversations about tradeoffs and prioritizing quality of life with the patient and family over time. Ultimately and especially over the past year, palliative care consultation and continuous palliative therapies were discussed with the patient, and were offered and extended to maintenance care, concomitant with disease-targeted treatments, allowing for optimal management of symptoms and maintenance of perceived quality and goals of care. Her goals of care are shifting toward fewer interventions at the end of life (now DNR/DNI) and allowing tradeoffs for function and quality over time (escalating diuretic use despite risk of renal failure; catheter

placement to manage recurrent ascites at home despite risk of life-threatening infection). Given her cachexia, signs of progressive multiple organ failure, and diminishing functional status, her prognosis may be limited to months (or conceivably shorter in the event of acute exacerbation or complication), but both time and functional prognosis remain uncertain, her prognostic awareness and informational preferences about prognosis are unknown, and we are not aware whether her advance care planning and wishes are documented.

EMERGING DATA RELEVANT TO ORGANIZATION AND DELIVERY OF PALLIATIVE MEDICINE IN PAH

High symptom burden underlies a life with PAH and intensifies as disease progresses, and ideal therapy incorporates interdisciplinary collaboration and continuity of care across settings to optimize and coordinate disease-directed and adjuvant therapies.¹⁻³ This is compounded by imperfect predictability of clinical trajectory, pointing to need for access to primary and specialty palliative care along the entirety of illness. Early diagnostic uncertainty exists for patient, family, and primary clinician, and typically sets a stage for future encounters and care. Symptoms (particularly shortness of breath and fatigue) predating diagnosis are nonspecific. Time to diagno-

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sis from onset of symptoms is prolonged (1 year on average), leaving patient and family with potential for distrust in the relationship with their primary medical team.^{4,5} Given that known disease-modifying therapies currently extend life for fewer than 5 to 10 years for most PAH patients, guilt and anger regarding delay in diagnosis may exist, and may intensify concerns that delayed diagnosis could mitigate benefits of disease-modifying interventions.

Early clinical course is often seen as one of marked improvement, but, retrospectively, patients and clinicians come to realize that this regaining of a modicum of function is better classified as freedom from greatest disability (analogous to pleasure being defined as the absence of pain). The phenomenon of “awakening” from the depths of severe heart failure (decreased mental acuity, depression, despair and anxiety, severe fatigue, anorexia, thirst, cachexia, bloating, edema, and ascites) is frequently accompanied by excessive hope by patients, families, and care providers that is typically less realistic than recognized outcomes from disease-modifying therapies (which carry their own uncertainties in effectiveness, adverse side effects, and complications). PAH specialty medical care teams can be misperceived as having full control over disease, creating an environment of idealistic but impractical outcomes from current and future disease-targeting therapies.

Relief from severe heart failure can place the PAH patient in a middle ground of improved capacity that appears normal and is perceived as free of goal-altering symptoms, but, in fact, is a medical state marked by little, if any, medical and physical reserve. Symptoms and functional incapacity remain highly prevalent in waxing, waning, and often spiraling fashion despite optimal disease-targeting therapies. (Of import, symptom presence and effects are typically under-recognized, under-discussed, and under-treated by patients and PAH specialty clinicians.^{2,3,6})

As illness progresses, patients' goals of care are often not iteratively reviewed, and nature and timing of end of life are frequently misperceived and left undiscussed. Patient-family-clinician

mismatch in expectations and understanding of goals of care at end of life for PAH patients can lead to high resource utilization with frequent death in hospital and in the intensive care unit (ICU). Patient choice to attempt acute salvage therapy is common, but often accompanied by choice not to implement intense chronic maintenance disease-targeting therapy (such as intravenous [IV] prostaglandin therapy).^{3,4,6}

PRIORITIES FOR PALLIATIVE CARE IN PAH

These findings exemplified by our patient case, as well as by existing literature, underscore the need for comprehensive, structured, systematic, and longitudinal delivery of palliative care, over the entire course of PAH.⁷

Our patient vignette highlights marked unmet symptom recognition, definition, attention, and care over her life experience with PAH, in many ways threatening her outcomes until identified and mitigated. Comprehensive symptom management remains a cornerstone of palliative medicine for patients and families affected by PAH.⁸ Such would include systematic screen, assessment and management of depression, anxiety, insomnia, anorexia, and pain. In particular, aggressive management of dyspnea is highlighted, especially toward the end of life (including opioids and anxiolytics as needed).

Serious illness conversations could have aided the patient, family, and care providers at multiple levels.⁹ Standardized conversations allow for assessment of patients' understanding of illness, prognostic awareness, informational preferences, goals, and values—all in increasingly normalized fashion for patients, family, and medical care teams. Recognition of higher-risk features could trigger a systematic approach to identify, embark upon, and document advance care planning conversations, and presents opportunities for explicit exploration of wishes about end-of-life care, and recommendation for hospice when it supports patient and family goals. Sharing and coordinating status and knowledge of these processes permits greater alignment between care teams, creates opportunities for improvements

in shared decision-making, and underscores the need for training in serious illness communication for involved teams.

The depth and extent of interventions necessary to accomplish such coordination and practice for patients with PAH and for their extended family and support units suggest a need for an interdisciplinary team approach in iterative fashion across the palliative care process and patient lifetime, assisting in coping, quality of life, complex decision-making, and end-of-life planning.¹⁰⁻¹³ This includes development, establishment, and implementation of a system for bereavement follow-up and care, as the high incidence of in-hospital (and in-ICU) death carries risk of complicated or prolonged grief reaction for involved families.

OPPORTUNITIES TO IMPLEMENT PALLIATIVE CARE ALONG THE TRAJECTORY OF PAH

Interdisciplinary Crisis Intervention

Crises can be characterized as abrupt life-altering occurrences due to disease-specific or external influences. Sentinel contributors to crisis can be characterized as those involving progression of disease, complications or new concomitants of illness, and limits of prognosis.⁷

Initial diagnosis, as well as each worsening of medical functional status or complicating event (arrhythmia, worsened dyspnea or edema or heart failure classification, cough, bloating, weight loss, hemoptysis, sepsis, gastrointestinal bleeding), hospitalization or transition of care level (mechanical ventilation, intensive care), addition or titration of medical disease-targeting therapy or IV inotropes (regardless of directional change, whether due to disease worsening or drug intolerance), or implementation or denial of catheter-based (shunt creation, balloon pulmonary angioplasty) or surgical mechanical therapy (right heart or lung support, transplantation) carries potential to influence and limit prognosis and psychosocial, spiritual, and life goals. An aide to recognition of the potent influence of such occurrences is incorporation of the reflection, “would

you be surprised if the patient died in the next year?" within standardized clinical assessment.⁹ Likewise, newly present or recognized tensions within the network of community, family, religious, and financial supports contribute to instability, uncertainty, alterations in clarity, and change in life perception and quality.

Acute referral for, availability of, and incorporation of both specialty-level and primary palliative care practice in such settings has the potential to impact outcomes in a patient-centered fashion. This includes the ability to improve and align patient and clinician understanding of patient-specific prognosis, as well as to better ensure communication within an often-complex multidisciplinary support team. Systematic palliative care allows for earlier identification, assessment, and treatment of important symptoms, while ensuring patient awareness of presence and import of high-quality adjunctive therapy of, and support for, psychosocial and spiritual tensions. At the same time, standardized and normalized discussions regarding goals of care (allowing for most appropriate therapeutic choices) can be initiated and extended, including awareness of nature, therapies available for, and imminence of end of life when deemed appropriate.

Patients and families tend to respond to direct face-to-face clinician encounters during crises. Direct palliative care team involvement at these inpatient and outpatient junctures appears appropriate and should be built into multidisciplinary PAH protocols to handle such crises. Extension of checklists developed during the healthier interstage between crises (see below) may serve as a stabilizing reference point and foundation for discussion during these highly emotion-filled crises.

Longitudinal Outpatient Care

The interval between crises can potentially be best characterized by the lyrics, "the space between the tears we cry is the laughter that keeps us coming back for more."¹⁴ Deeply imbedded within this phase of outpatient care is a clarity of thought that allows for greater alignment of realism and hope, for improved

clinician-patient-family alignment in education and perceptions, for development of coping strategies, and for implementation of plans to achieve greatest closure in personal, family, social, career, community, and spiritual goals.¹¹⁻¹³

Use of structured group sessions and educational forums may allow for improved symptom recognition and utilization of available resources by patients and families to optimally control symptoms; understanding of typical disease course and social, psychological, financial, and spiritual influences, as well as nature and availability of disease-specific and palliative supports; and definition and communication of medical, personal, psychosocial, and spiritual goals. Such sessions may be directed toward patients, families, PAH experts and associated care providers, and led by palliative care team members or advocate/champions within the PAH specialty team.

Creation of a patient-family-clinician checklist—inclusive of disease and current prognosis understanding, symptom awareness, development and expression of goals of care (and specific choices within goals that are personally relevant to identify), development and expression of goals of life/career/family/community/spirituality, and development and implementation of strategies designed to best accomplish these—can be implemented by members of the PAH specialty team and iteratively reviewed at each patient visit. This type of checklist offers potential to decrease anxiety and depression, align awareness and goals of each member in the PAH therapeutic relationship, decrease misperceptions and errors, allows a sense of greater patient dignity in having their wishes recognized and imbedded in care planning, and extends a greater sense of control over the unknown and oftentimes inexorably deteriorating PAH-specific medical state. This checklist can be reviewed with palliative care clinicians either on referral for face-to-face palliative care patient consultation, or during multidisciplinary meeting or review. Further, checklists can serve as a foundation for crisis-specific review (see above).

As clinical guidelines develop to further support incorporation of palliative care within management of patients with PAH, standardized systems of education and accrual of competencies in key aspects of palliative medicine must be developed, implemented, and assessed for trainees in cardiovascular and pulmonary vascular disease. Advocacy for such training in similar states of advanced heart failure has highlighted the need for education regarding symptom management along disease trajectory, as well as skill development in serious illness communication (advanced care planning, discussions regarding bad news or prognosis, conversations on goals of care as disease progresses and towards end of life, recommendations for palliative care and hospice).¹⁵

NEXT STEPS

It is highly likely that until effects of palliative care practice and intervention (within varied models of inpatient and outpatient specialty-level and primary palliative care) for patients with PAH are further studied and assessed; systematic primary palliative care education is implemented; and regional and national patient, clinician, and health-insurance organizations (Pulmonary Hypertension Association Comprehensive and Regional Centers of Care Guidelines, American Heart Association/American College of Cardiology and European Society of Cardiology PAH Care Guidelines) create consensus and guidelines documents that are inclusive of integration of palliative care practices throughout the course of PAH and associated right heart failure, organization and structure of palliative care within PAH care environments will be varied in structure, performance, and outcomes.^{7,16-20} It is our hope that this vignette-associated document will serve as a call to arms for further study, as well as creation of consensus guidelines for consistent integration of palliative care practice beginning at diagnosis and extending through the entirety of life for patients with PAH.

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