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Quality of Life

Program Description

The mission of *Advances in Pulmonary Hypertension* is to serve as the premiere forum for state-of-the-art information regarding diagnosis, pathophysiology, and treatment of pulmonary hypertension (PH). The 2018 Nice revision of the World Symposium on Pulmonary Hypertension (Simmonneu G, Montani D, Celermajer DS, et al. Haemodynamic definitions and updated clinical classification of pulmonary hypertension. Eur Respir J. 2019;53(1). DOI:10.1183/13993003.01913-2018) serves as a guide to categories of PH addressed in *Advances in Pulmonary Hypertension*. While focusing on Group 1 PH (PAH: pulmonary arterial hypertension), the other categories (Group 2, PH due to left heart disease; Group 3, PH due to lung diseases and/or hypoxia; Group 4, PH due to pulmonary artery obstructions; Group 5, PH with unclear and/or multifactorial mechanisms) are also addressed. This mission is achieved by a combination of invited review articles, roundtable discussions with panels consisting of international experts in PH, and original contributions.

Objectives

- Provide up-to-date information regarding diagnosis, pathophysiology, and treatment of PH.
- Serve as a forum for presentation and discussion of important issues in the field, including new paradigms of disease understanding and investigational trial design.

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Advances in Pulmonary Hypertension: Official Journal of the Pulmonary Hypertension Association is a quarterly publication directed by an editorial board of renowned pulmonary hypertension (PH) experts with oversight by PHA's Scientific Leadership Council. The mission of *Advances in PH* is to assist physicians in their clinical decision-making by informing them of important trends affecting their practice and providing an analysis of the impact of new findings and current information in peer-reviewed publications. Each article is reviewed and approved by members of the Editorial Board.

While most articles are invited by the Editorial Board, the following submissions will be considered for publication:

- Reviews that summarize and synthesize peer-reviewed literature to date on relevant topics
- Letters to the Editor
- Clinical case studies

Submitted manuscripts are reviewed by the Editorial Board and other experts in the field. Acceptance of manuscripts is determined by factors such as quality, relevance, and perceived value to clinical decision-making.

Advances in Pulmonary Hypertension's Web Platform

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The words of Dr. Francis Peabody still ring true almost 100 years after they were first uttered. "The secret of the care of the patient is in caring for the patient." As providers of patients with pulmonary hypertensive vascular disease (PVHD) we direct the pathway for a patient's medical care which has considerable impacts on their quality of life. It is imperative that our patients are viewed as people beyond their diagnosis. Pulmonary hypertension (PH) is a disease that affects patients across the lifespan. Each stage in the life of a PH patient presents its own unique challenges which require expert collaboration for holistic care and overall wellbeing. In this issue of *Advances in Pulmonary Hypertension* we have gathered patients, caregivers, physicians, nurses, social workers, and psychologists to address issues that affect all age groups as they navigate their way through life with PH. Our hope is that this will inspire you to think creatively to care your patients and families beyond their diagnosis with the goal of overall wellbeing and quality of life.

In the PHPN Corner, Allison D'Souza, RN offers a personal perspective of how living with PH has empowered her to be a better and more compassionate provider in "From Patient to Provider: How My Diagnosis Makes Me a Better Nurse." She has transitioned from a teenager adjusting to a new diagnosis to a pediatric cardiac ICU nurse guiding young patients to find hope.

Dr. Hilary DuBrock and her team at the Mayo Clinic provide an overview of how PH effects quality of life. They synthesize the evidence regarding palliative care in PH and other cardiopulmonary diseases. DuBrock and team make a strong argument to incorporate palliative care in the longitudinal management of patients with PH.

In "Perspectives on Mental Health Evaluation in Pediatric Pulmonary Hypertension: A Call to Action" an argument is made for more robust mental health screening practices integrated within care guidelines for pediatric PH. While providers recognize symptoms of anxiety and depression in their patients, few actually screen for anxiety and depression with intention to connect to mental health services. The authors from the AHP group of the Pediatric Pulmonary Hypertension Network (PPHNet) identify the existing barriers to accessing mental health services in pediatrics.

In the current age, social media has been integrated into everyday life. Elise Whalen, NP and her co-authors from Texas Children's Hospital provide a unique discussion on social media participation as a support system among caregivers of children with PH.

Finally in the round table discussion, co-editors Claire Parker, NP and Nancy Bair, CNS gather Maddie Bonpin, a pediatric PH patient and her mother, Liza Bonpin, and Dr. Neal Chaisson to discuss transitions throughout the lifespan of a pulmonary hypertension patient.

Thank you to all the authors and participants who contributed to this issue. We are thankful for the PHA and Allen Press for their support and guidance to bring this discussion to publication. We hope this issue encourages you to actively engage in the improvement of quality of life and overall wellbeing of your patients.

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From Patient to Provider: How My Diagnosis Makes Me a Better Nurse

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As a nurse on an advanced lung disease and pulmonary transplant floor, I always brace myself when I have a patient with pulmonary hypertension (PH). Although they are my favorite patients, it always hits a little too close to home. One day, I was assigned a patient that had severe PH and was supposed to go home on hospice. As I attempted to get my tasks completed, we started talking about how she felt. Morbid jokes were sprinkled throughout our conversation, almost as an acceptance of our disease without a definitive cure. When I shared my diagnosis of idiopathic pulmonary arterial hypertension (IPAH), she looks at me and chuckles as she says "Well, I hope your outcome is better than mine." While our conversation continued, my PH specialist (who happened to be on PH service that week) walked into her room with a plan that did not necessarily include going home on hospice. After he left, she said, "I can't believe I found my two," as she heard one or two in every million are diagnosed with IPAH. Throughout that day, our bond grew immensely. At the end of my shift, she asked, "Where were you all of this time since my diagnosis?" Although our paths were very different, sharing my story gave her a bit of hope.

I was a *healthy* teenager prior to my diagnosis. My focus was school, horseback riding, and violin. However, slowly, I was losing the ability to ride my horse and walk to class in my small high school. Something in my body just told me to stop and take a break; I later I realized this was severe dyspnea on exertion and presyncope. One day, my biology teacher said people with heart murmurs have difficulty walking to their cars. This statement made me realize that something other than being lazy and subjectively overweight may be going on. That night, I told my dad we should schedule an appointment with my pediatrician to help figure out why I was so tired all the time.

The appointment was uneventful. With a shrug of the shoulders, my pediatrician recommended that I get more sleep and exercise. However, my CBC showed polycythemia, which led to a CT scan of my abdomen to check my kidneys. A radiologist found cardiomegaly, and I was sent to a cardiologist's office the next day. One of the first things he said to me was, "I think I know what you have, and if I am right, you will need to go to UCSF [University of California, San Francisco] immediately." After an echocardiogram with many concerned looks from both the echo tech and my cardiologist, I heard the words "pulmonary hypertension" for the first time. I had the choice of going via ambulance or driving with my parents. The stubborn student in me demanded that I go to my English presentation first, to which my cardiologist said ,"Well, considering you walked in here, I can't see why you wouldn't be able to go."

After getting through the rough afternoon traffic from Sacramento to San Francisco, I walked into the unit that would become my home for the next 9 days. I was taken for my first right heart catheterization, where I gained the title of "the worst pressures" these experienced PH providers had ever seen. My pressures were suprasystemic, and I had poor right ventricular function, which I later learned stratified me to a very poor prognosis. I was started on triple therapy, including subcutaneous Remodulin, all of which I am still on today. A few days later, my PH specialist, Jeff Fineman, came in and said, "there's a PH 5K in a month, and you are going to walk it." I, along with everyone else in the room, looked at him very confused. How could I walk a 5K when I could barely walk 238 meters on my 6-minute walk test? Jeff had a lot of hope during a time where it was difficult for me to comprehend what my future may entail. I was able to walk that 5K, with my PH and Cardiac Intensive Care Unit team behind me.

After my diagnosis, I had one main goal – to live as normal of a life as possible. I begged my parents to let me move away from home for college. I had learned how to manage my medications and doctors' appointments and wanted to experience a true freshman year. I attended Cal Poly SLO, and I truly thrived. During my first year, I started working as a medical assistant for a family practice and achieved the President's List every quarter. That summer, I started shadowing my PH team's nurse practitioners, who inspired me to pursue nursing. I transferred to UCLA's School of Nursing, where I began my journey to be a pediatric Cardiac Intensive Care Unit nurse. It was a journey full of ups and downs. I had people tell me they were not sure that I was cut out to be a nurse because

Key Words—nurse, patient, pulmonary hypertension
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of their preconceived notions of people with PAH. However, I proved them all wrong as I graduated at the top of my class in 2020.

I moved to Pittsburgh, Pennsylvania, to pursue a job as a nurse in an advanced lung disease and pulmonary transplant floor. I had to go through a bit of an adjustment, learning how to take care of myself while taking care of very ill patients. This included taking diuretics and dealing with subcutaneous Remodulin site changes while working a physically demanding job. I quickly learned that I must take care of myself first in order to provide the best care for my patients. Working with this population has been incredibly rewarding, but also very difficult to see patients severely ill after their life-saving lung transplants, as I know it is likely a part of my future.

My patients with PH always have a special place in my heart. Despite the large age differences, we have a shared experience that brings us closer together. Although I have shed many tears for these patients, I am truly thankful for the days that I get to spend with them. When I care for adult PH patients, I have a difficult time not seeing myself in their shoes one day, struggling for every breath and having to make some of the most difficult decisions of their life. A few months after our initial meeting, I saw my would be hospice patient was a patient on my unit again. When I went to see her, the first thing she said was, "Aren't you surprised I'm not dead yet?" She passed away a few nights later, but our relationship taught me how to find the humor in every situation. Although these relationships with patients have been incredibly difficult because of the often harsh reality of PAH, it makes each grueling day at work worth it knowing that I made a difference.

As I transition to my new job in the pediatric Cardiac Intensive Care Unit, I hope that I will show children and their families some hope in their diagnosis. I want them to know that they can pursue their dreams despite many of the challenges that their diagnosis may throw at them. Without PAH, I would not be the nurse I am today, and for that, I am forever thankful for my diagnosis and for being a part of this amazing PH community.

REGULAR ARTICLE

Palliative Care and Pulmonary Arterial Hypertension

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Background: Pulmonary arterial hypertension (PAH) is a chronic and progressive disease associated with impaired health-related quality of life and survival. Palliative care (PC) is patient- and family-centered care provided by an interdisciplinary team with an overarching goal of alleviating suffering and improving quality of life for patients with advanced illness. PC in other chronic diseases is associated with improved quality of life, symptom management, illness understanding, and reduced caregiver burden, but there is limited data regarding PC in PAH. Despite limited evidence, there is strong rationale for involving PC specialists in the longitudinal management of PAH.

Implications for Clinicians: There are currently no guidelines to help clinicians determine the most appropriate timing for referral of PAH patients to PC specialty teams. Consequently, referrals are limited and often delayed. Adoption of a standardized approach to PC referrals based on clinical or patient triggers could facilitate earlier involvement of PC as an adjunct to ongoing PAH disease-directed care.

Conclusions: Incorporation of PC in the longitudinal management of PAH may be beneficial to address the multidimensional aspects of living with a chronic and life-limiting illness.

INTRODUCTION

Pulmonary arterial hypertension (PAH) is a chronic and progressive pulmonary vascular disease associated with significant morbidity and mortality. PAH is characterized by pulmonary vasoconstriction and vascular remodeling which can ultimately lead to right heart failure and death. PAH is also associated with significant symptoms as well as impaired health-related quality of life (HRQOL)[1] across varied physical, social, and emotional domains.[1,2] PAH therapeutics delay disease progression and improve symptoms, hemodynamics, exercise capacity, and survival but unfortunately are also commonly associated with adverse effects which may further impair HRQOL.[3] Despite the importance of HRQOL in the overall patient experience of living with PAH, few therapies are available to specifically address and improve HRQOL in PAH. Palliative care (PC) provides patient- and family-centered care with an overarching goal of alleviating suffering and improving QOL for patients with advanced illness. Despite its potential benefits, PC is underutilized in PAH, and there is little data to guide clinicians regarding how to incorporate PC into their clinical practice. This review provides an overview of PC and its utility in other cardiopulmonary diseases, summarizes available evidence regarding PC in PAH, and suggests potential roles for PC in the longitudinal management of patients with PAH.

PAH IS ASSOCIATED WITH SIGNIFICANT MORBIDITY AND MORTALITY

HRQOL is a multidimensional patient-reported outcome (PRO) that refers to the general wellbeing of an individual. PAH negatively impacts HRQOL across the spectrum of physical, social, and emotional domains.[1,2] PAH is associated with symptoms, such as dyspnea, fatigue, impaired exercise capacity, lower extremity edema, lightheadedness, syncope, and depression.[4,5] Although there have been significant recent advances in treatment of PAH, PAH-targeted therapies are commonly associated with side effects which carry an additional symptom burden, and can include headache, heartburn, nausea, flushing, dizziness, edema, and pain, among others. Despite treatment, most patients continue to experience symptoms that impact their HRQOL as well as other aspects of their lives, such as employment and social interactions.[6] PAH can also be associated with emotional and psychological distress, manifested as feelings of frustration, anger, and sadness.

HRQOL is a significant predictor of outcomes in PAH.[2] Patients with impaired HRQOL at diagnosis have increased hospitalizations and worse survival, even after adjusting for disease severity.[1,2] Despite its importance as a prognostic marker and its importance to an individual's overall wellbeing and daily life, HRQOL is not routinely assessed in clinical practice and is rarely con-

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sidered a primary endpoint for clinical trials or other interventions. Although current PAH therapeutics can improve variables, such as functional class, pulmonary hemodynamics, and 6-minute walk distance, they often do not address the multidimensional aspects of PAH such as the cognitive, emotional, social, and financial impacts.

In addition to its impact on HRQOL, PAH is a progressive disease associated with impaired long-term survival.[7] Despite the adverse prognosis associated with PAH, studies have found that completion of advance health care directives and utilization of PC services are low. According to one single-center study, less than half of PAH patients had completed advance health care directives at the time of death.[8] Despite the poor prognosis associated with interventions such as cardiopulmonary resuscitation (CPR) and mechanical ventilation in PAH, 31% and 40% of patients, respectively, received CPR and mechanical ventilation at the end of life (EOL)[8,9] and 80% of patients died in a health care setting.[8] More than half of patients died in the intensive care unit with only 8.6% of patients receiving PC before death[8]. Overall in-hospital mortality for patients with PAH and right heart failure is high, with estimates ranging from 14% to as high as 30% to 48% for patients admitted to the intensive care unit.[9,10] Despite the poor prognosis associated with hospitalizations, PC was consulted in only 2.2% of PAH hospitalizations according to a national study.[11] Overall, these studies suggest that there is an unmet need in PAH to improve prognostic awareness and communication with both patients and caregivers regarding goals of care to ensure the PAH care team is providing goal-consistent care throughout the continuum of disease.

PC: AN OVERVIEW

Patients with serious illness encounter numerous issues that can detrimentally affect their perceived QOL. PC is an interdisciplinary model that provides patient- and family-centered care, with an overarching goal to alleviate suffering and to improve QOL for patients with advanced illness. It is a common misconception that PC is synonymous with EOL care and hospice; the two are on the same spectrum but distinct entities. For patients to receive the full benefits of PC, early integration, in conjunction with disease-directed therapy, is likely the best plan of care (Figure 1). Incorporating palliative principles early into a patient's disease course can result in improved QOL and symptom management, better illness understanding, completion of advance directives, enhancement in coping skills, and reduction in caregiver burden.[12–14] Contrary to many patient and medical provider fears, PC is not associated with reduced survival.[15,16] Therefore, the World Health Organization recommends inclusion of PC early in the course of illness.[17]

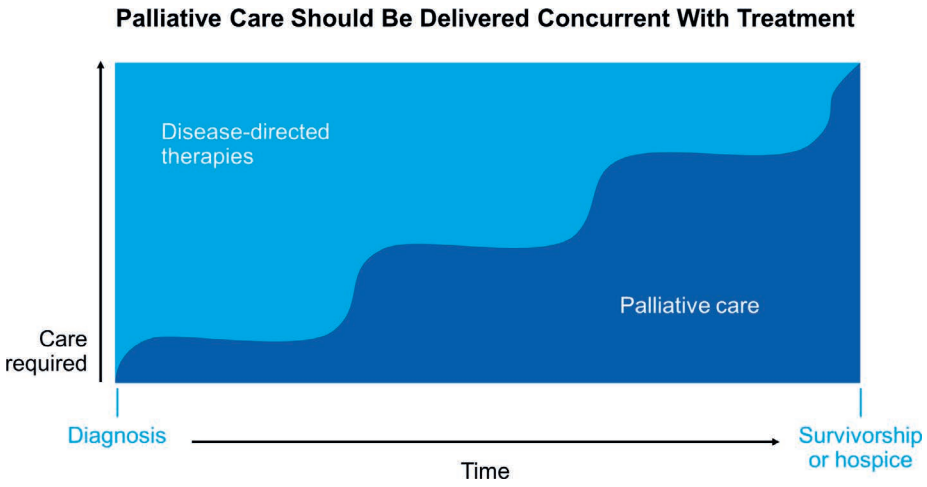

Figure 1: A model of palliative care delivered concurrently with disease-directed therapies.[41]

PC IN OTHER CARDIOPULMONARY CONDITIONS

PC has traditionally been rooted in the oncologic setting, but authors of studies have shown that PC can have a dramatic impact on patients with advanced nononcologic disease, including cardiopulmonary disease.[15,16,18–24] Like PAH, patients with advanced congestive heart failure (CHF) often experience progressive and debilitating physical, psychosocial, and spiritual concerns. In addition to the predicted symptoms of dyspnea and fatigue, many patients with CHF also suffer with pain and depression.[25,26] Spiritual distress has also been identified as a concern in patients with CHF.[27] Compelling evidence of benefit for integration of PC for patients with advanced heart failure has been demonstrated in numerous trials, showing improved QOL and patient satisfaction, improved physical and mental health and spiritual wellbeing, and completion of advance directives.[16,18–20] Most major cardiology societies, the Joint Commission, and the Centers for Medicare and Medicaid Services have developed guidelines for incorporation of PC in patients with advanced cardiac disease.[24]

Likewise, patients with advanced pulmonary disease, such as chronic obstructive pulmonary disease (COPD) and interstitial lung disease, have reported poor QOL, high symptom burden, involving both physical and mental health, and high rates of caregiver distress.[21,22] Breathlessness is an exceedingly common manifestation of advanced pulmonary disease despite optimal medical therapy for the underlying illness. Breathlessness can be substantially improved in the setting of an interdisciplinary care model incorporating PC into usual care.[15,21] Mood disorders and existential distress are often unrecognized and undertreated in patients with advanced pulmonary disease.[23] Furthermore, it is well established that untreated psychosocial and spiritual distress may influence a patient's treatment preferences.[23] Given the palliative approach to care, which includes close attention to emotional, social, and spiritual health domains, an opportunity exists to significantly impact outcome measures, both in terms of symptom

management and perceived qualify of life, and patients' treatment preferences.

Given the severe symptoms with advanced cardiopulmonary disease, advance care planning is very important, yet authors of one study demonstrated that less than one-third of patients with oxygen-dependent COPD had completed an advance directive.[28] Patients with advanced cardiopulmonary disease demonstrate a high rate of health care utilization in the last year of life, often in dissonance with their goals. The average median annual cost for a patient with advanced CHF is $24383, with the majority stemming from hospitalizations.[29] Patients with advanced COPD incur additional costs with mean estimates of $34911 in the last 6 months of life, again, largely related to inpatient care with substantial intensive care utilization.[30] Involvement of PC has been shown to reduce health care expenditures in the last year of life by helping patients and families navigate difficult decision making, provision of care that is in line with patient-specific goals and preferences, and timely referral to hospice.[16] Historically, most patients with advanced cardiopulmonary disease died in the hospital; however, palliative involvement is associated with increased likelihood of dying at home, which aligns with most patients' preferences.[31]

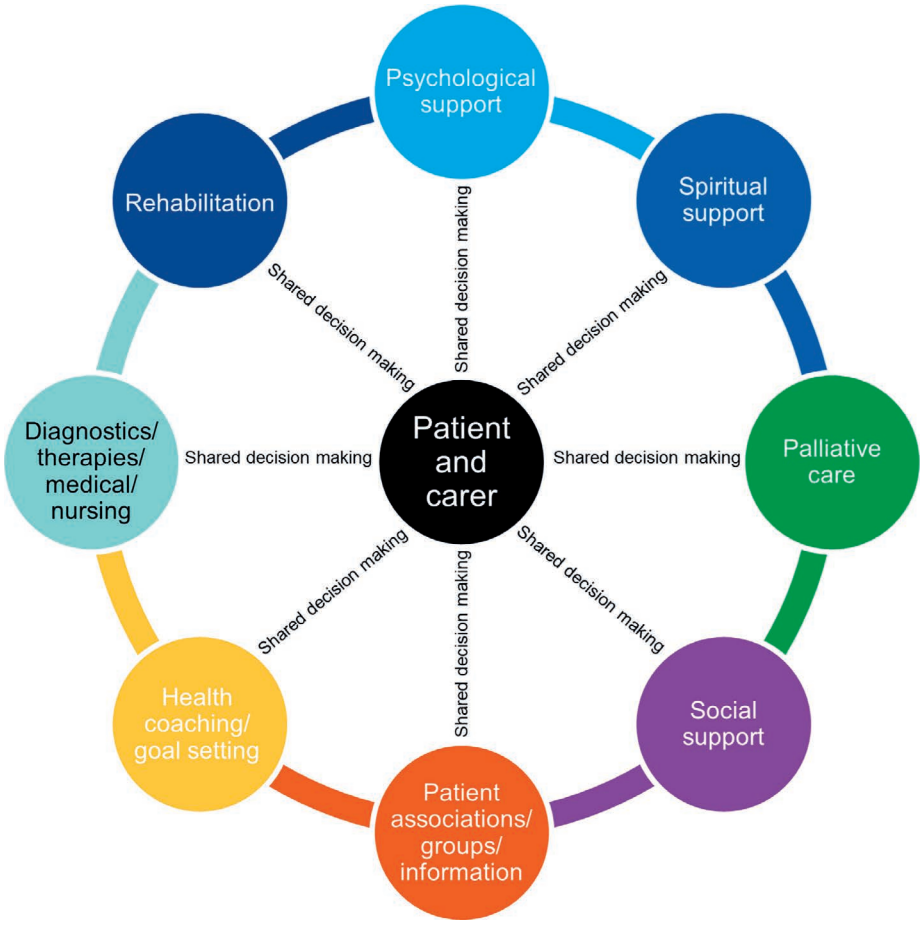

Figure 2: Components of a multidimensional approach to care of patients with pulmonary arterial hypertension, from McGoon et al.[3]

PC IN PAH

Despite the established benefit of PC in other cardiopulmonary diseases, there have been few studies addressing the role of PC in PAH. PC referrals are low for patients with PAH, both in the inpatient and outpatient setting, despite many of these patients having intensive support and symptom management needs.[11,32] Survey-based studies of PAH physicians and patients have identified several barriers to regular, early PC involvement, including patient and physician perceptions regarding the role of PC.[32,33] In a study on physician-perceived barriers to PC referral for patients with PAH, worry about the appearance of "giving up hope" was rated highly as a barrier, in addition to feeling the consult unnecessary in dealing with QOL or EOL needs for patients.[32] The misconception that patients could not receive PC along with active disease-modifying therapies for PAH was another clinician-perceived obstacle to referral. PC has also been incorrectly associated with only EOL and hospice.[32,33] Anxiety and declination from patients and caregivers surrounding PC referral also contribute to limited and late referrals.[32,33] Health care disparities and access to PC services may also represent barriers to PC referral, as suggested by an analysis of the National Inpatient Sample, which found that there were geographic, socioeconomic, and racial differences associated with PC utilization in PAH as well as lower rate of referrals for patients without private insurance.[11]

Despite limited evidence, there is an important rationale for earlier referral to PC for patients with PAH. PC referral early in the disease course can be beneficial for preparedness planning for both patients and caregivers, in addition to supporting shared decision making throughout the continuum of care. The prognostic uncertainty often associated with PAH, along with cost-related medication barriers and high rates of in-hospital death, make early and skilled discussions imperative to support patient autonomy and values.[4] Early PC referral in PAH can provide systematic advance care planning with discussions of surrogate decision makers, with a focus on what is a meaningful QOL to the patient. In addition, planning for acceptable transitional care plans and engagement in "what-ifs" discussions can be meaningful with early PC involvement.[4] The support of the PC interdisciplinary team in PAH is also very important in providing psychosocial support for both patients and caregivers, helping to acknowledge the challenges and emotions that come with a chronic life-limiting and debilitating illness.

The care of patients with PAH requires a multidimensional approach with a focus on alleviation of symptoms while supporting emotional, social, and

psychological wellbeing (Figure 2).[5] Many palliative treatment options exist, including pharmacologic, nonpharmacologic, and procedural. The use of low-dose opioids and benzodiazepines can be helpful for dyspnea and concomitant anxiety.[4] Nonpharmacologic treatments, such as supervised exercise regimens, meditation and guided imagery, and supplemental oxygen when appropriate, can be used to offset symptom burden as well.[34] Assessments for depression, anxiety, and insomnia, with appropriate screening and referrals for psychotherapy or psychiatric consultation, are also important, given the high prevalence of mood disorders among patients with PAH.[35] When patients with PAH have escalation of symptoms, have recurrent hospitalizations, and are closer to a more limited prognosis at EOL, having already been established with PC can help with patient familiarity of the team, making engagement in complex communication, in partnership with other primary teams, more beneficial and connected. Given the complexity of PAH management and the negative impact of PAH on HRQOL, PC has much to offer patients with PAH through the 8 domains of PC, which include supporting patients and families through structures and processes of care, management of physical and symptom needs, psychological, social, spiritual, and cultural aspects of care, care of the patient nearing EOL, and navigation of ethical and legal aspects of care.[36]

ROLES FOR PC IN PAH AND HOW TO INCORPORATE PC INTO CLINICAL PRACTICE

According to recent PAH treatment guidelines, there is no evidence to provide direct recommendations regarding utilization of PC services in PAH.[37] Thus, clinicians are left with little guidance to help determine the most appropriate timing for referral to PC specialists. In the absence of evidence or specific recommendations, referrals are delayed and typically occur at the EOL, when it is often too late to make meaningful improvement in an individual's HRQOL. To overcome barriers and facilitate earlier referrals, it can be helpful to have a standardized approach, either based on clinical or patient triggers. Clinical triggers could include escalation of therapy (initiation of parenteral prostacyclin therapy or triple therapy, for example) or events such as hospitalizations, lung transplantation referral, or clinical deterioration. Risk stratification can improve prognostic awareness and can also serve as a guide regarding timing of referrals. There are several tools for risk stratification in PAH, so one approach would be to refer patients who fall into a "high-risk" category.[38] These approaches based on clinical triggers would help to automate referrals, so they are part of routine care and longitudinal management. In conjunction with PC referrals, particularly those based on prognosis or clinical deterioration, it is imperative that PAH clinicians have ongoing discussions with patients to facilitate prognostic awareness and to provide education regarding the role of PC as an adjunctive therapy in the ongoing management of PAH.

Additional research studies are needed to help determine the most appropriate patients and timing for referral to PC specialists. At Mayo Clinic Rochester, we are currently engaged in a patient-triggered randomized controlled trial of PC plus usual care versus usual care alone.[39] In this single-center study, we are enrolling patients with impaired HRQOL (regardless of disease status or prognosis) as assessed by the SYMPACT score, a PAH disease-specific and validated PRO tool, and randomizing them in a 1:1 fashion to PC plus usual care or usual care alone.[40] The primary outcome of this study is HRQOL. PRO-triggered PC referral represents an opportunity to address an individual's symptoms and concerns to facilitate living as well as possible through symptom management and coping support regardless of his or her disease prognosis or clinical status. The approach of PRO-triggered referrals also mitigates previously described patient and clinician barriers to utilization of PC services, such as concern that referral implies "giving up" or a poor prognosis.[32,33] The study is ongoing, and the results could help guide clinicians regarding an alternative approach to facilitate PC referrals but will need to be studied in a multicenter clinical trial.

FUTURE DIRECTIONS

Improved evidence is needed to help determine the impact of PC on HRQOL and other important patient-centered outcomes in PAH and to help guide clinicians regarding patient selection and appropriate timing of PC referrals. Without evidence and without education of providers and patients regarding the role of PC, early and regular utilization of PC services in the longitudinal management of PAH will continue to be limited. As PH clinicians, however, we owe it to our patients to use all the available tools we have, including PC, to alleviate their symptom burden, mitigate side effects of therapy, and ensure we are maintaining goal-consistent care that aligns with their values and wishes.

CONCLUSIONS

In summary, PAH is associated with impaired HRQOL and survival. Although current PAH-targeted therapies improve symptoms and pulmonary hemodynamics and delay disease progression, they are not curative, are commonly associated with adverse effects, and do not address all aspects of impaired HRQOL in PAH. PC, an interdisciplinary model of patient- and family-centered care with the goal of alleviating suffering and improving HRQOL, is underutilized in PAH but may be beneficial to address the unmet needs of PAH patients and the multidimensional aspects of living with a chronic and life-limiting illness.

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REGULAR ARTICLE

Perspectives on Mental Health Evaluation in Pediatric Pulmonary Hypertension: A Call to Action

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Background: The already high rates of anxiety and depression among healthy children have increased further since the COVID-19 pandemic began in early 2020. Prepandemic data suggest children with chronic disease were already experiencing increased rates of anxiety and depression as compared to their healthy peers. There is currently a paucity of literature on the prevalence of anxiety and depression in patients with pediatric pulmonary hypertension. In addition, there are no practice recommendations regarding mental health screening in this population.

Implications for Providers: We evaluated provider perception of mental health screening at 14 pediatric pulmonary hypertension care centers across the United States and Canada using a cross-sectional survey. Thirty-seven providers from 14 pediatric pulmonary hypertension centers in North America completed the survey.

Conclusion: Health care providers caring for pediatric pulmonary hypertension patients perceive their patient cohort to frequently experience anxiety and depression. In addition, these providers believe that their patients would benefit from formalized, and routine, assessment of anxiety and depression with referral to mental health services as appropriate. This highlights the need to better understand the prevalence of anxiety and depression in pediatric pulmonary hypertension patients and to establish formal mental health screening practices within pediatric pulmonary hypertension care centers.

PERSPECTIVES ON MENTAL HEALTH EVALUATION IN PEDIATRIC PULMONARY HYPERTENSION

Current studies on rates of anxiety and depression suggest an increased prevalence in pediatrics, with 25% of youth worldwide experiencing symptoms of depression and 20% of youth worldwide facing symptoms of anxiety.[1] This rate has increased from before COVID-19 pandemic times, highlighting the mental health toll of worldwide illness and isolation. There are few data on the pandemic's effect on the rates of anxiety and depression in children with chronic disease. Prepandemic data suggest children with chronic illness were already experiencing increased rates of anxiety and depression as compared to their healthy peers. In a meta-analysis, Pinquart and Shen evaluated 350 studies and 450 subsamples with the conclusion that children and adolescents with chronic illness have higher rates of depression than their healthy peers, which in turn can negatively influence the child's chronic medical condition.[2]

Pulmonary hypertension (PH) is a chronic disease characterized by a marked elevation of pressures within the pulmonary vasculature, associated with many diverse cardiac, pulmonary, and other systemic disorders.[3] It has

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significant morbidity and mortality with poor overall patient outcomes despite new therapies.[4] Studies have reported that pediatric PH patients have a significantly lower quality of life than other chronic disease cohorts as well as healthy norms, suggesting a need to assess and treat this patient cohort holistically rather than with a disease-specific approach.[5,6] One important aspect of quality of life is mental health. Currently, there are limited data on the prevalence of anxiety and depression among pediatric PH patients. In addition, there are no practice guidelines or recommendations regarding anxiety and depression screening practices in PH clinics. This paper examines current provider and care team perspectives on anxiety and depression in their pediatric PH patient cohorts.

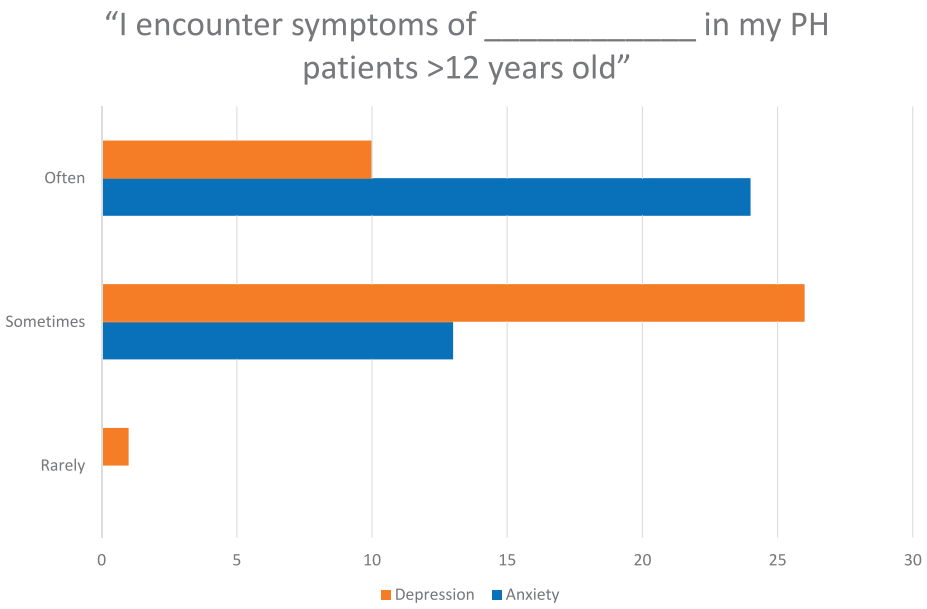

Figure 1: Providers encounter symptoms of depression and anxiety in pediatric pulmonary hypertension patients >12 years old.

METHODS

With increasing recognition of anxiety and depression among pediatric PH patients and growing concern, a mental health subcommittee was formed within the Advanced Healthcare Provider Committee of the Pediatric Pulmonary Hypertension Network (PPHNet). We developed a formal survey (REDCap) to characterize health care provider practices and perspectives of anxiety and depression in pediatric PH patients (Supplemental Material). The REDCap survey was distributed via email to staff at all 14 PPHNet sites. Participants were invited to respond to the survey individually, and as such there was the potential for more than 1 survey from each center.

Aside from basic provider demographics including the name of the institution and participant's profession, the survey consisted of 18 questions. Four questions asked participants to place a mark on a sliding scale, from *disagree* to *agree*, regarding the perceived benefit of formalized depression and anxiety assessment as well as resources available should a referral be required. The remaining questions were polar questions about perceived frequency of depression and anxiety, mental health screening practices, and resources available to the center. Several open-ended questions allowed participants to specify tools used to screen for depression and anxiety, describe barriers associated with the assessment of mental health and referral for services, and comment on existing mental health resources available to the practice and any additional relevant comments about pediatric PH patients and depression and anxiety. The results were analyzed descriptively with categorical data reported as percentages.

FINDINGS

The survey was returned by 37 respondents from 14 pediatric PH centers in North America. Respondents included 13 MDs, 13 NPs, 6 RNs, 3 social workers, 1 psychologist, and 1 child life specialist. Many of the institutions had respondents from more than 1 profession. Most programs did not have representation from every profession.

When asked if they encountered symptoms of anxiety in PH patients >12 years old, 24 of the respondents replied *Often*, while 13 replied *Sometimes*. None of the respondents replied *Rarely*. When asked if they encountered symptoms of depression in their patients, 10 replied *Often*, 26 replied *Sometimes*, and 1 replied *Rarely*. (Figure 1) Respondents were then asked to rate several statements from 0 to 100, with 0 representing *disagree* and 100 representing *agree*. When asked to rate the statement "My PH patients >12 years old would benefit from a formalized, and routine, assessment for anxiety with referral to mental health services," respondents had a mean score of 94.24 (range 60-100). When asked to rate the same statement in relation to depression, the mean response was 91.82 (range 59-100). For the statement "If a PH patient screens positive for anxiety, there are appropriate mental health resources for referral," the mean score was 65.97 (range 10-100). The response for the same statement for depression had a mean score of 69.05 (range 10-100).

Only 3 of the 37 respondents implemented screening tools for anxiety in their clinical practice. One psychologist and 1 social worker used the GAD7 to screen for anxiety with clinical concern. A child life therapist screened for anxiety with each visit, the method of which is unclear. Only 2 respondents implemented screening tools for depression in their clinical practice. One psychologist uses the PHQ9 with clinical concern. One child life specialist implemented screening for depression with clinical concern but does not list the tool that is used. Of the 14 pediatric PH programs, 13 have access to a social worker, and 6 have access to a psychologist. Five of the programs had respondents from different professions, some answering *yes* and others answering *no* when

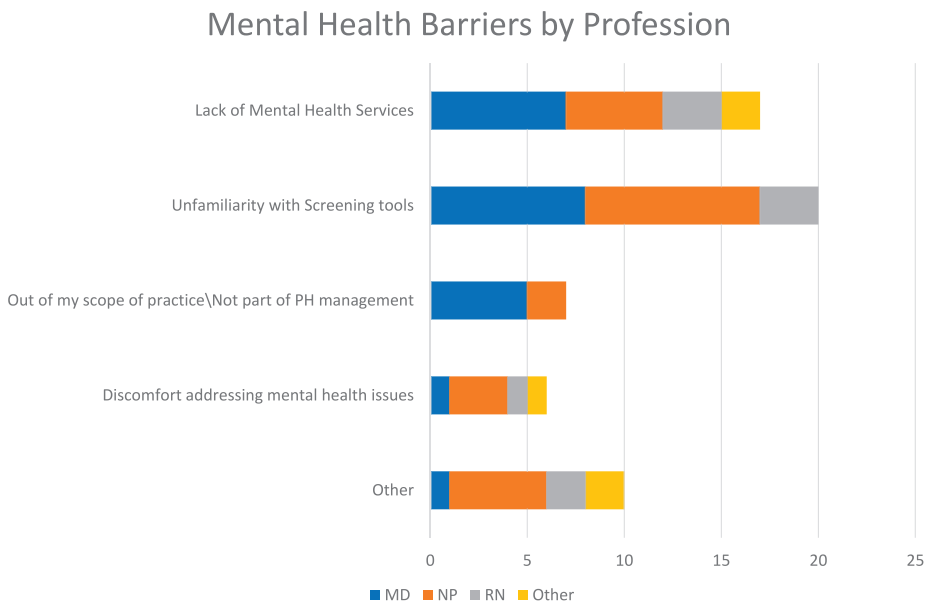

Figure 2: Barriers to accessing mental health services by profession.

asked about access to a psychologist. Two programs did not have access to a psychologist.

When asked about barriers to mental health services, respondents could check all that apply. Unfamiliarity with screening tools was the most frequently cited barrier (57%), followed by lack of mental health services (48%), out of my scope of practice/not part of PH management (20%), and discomfort addressing mental health issues (17%). Ten respondents identified other barriers with free text. Other barriers fit into 4 categories including clinic time, patient location, patient family buy-in, and immediate availability of services. (Figure 2)

Respondents were asked to comment via free text about the mental health services that are available to their practice. Seven respondents indicated that appropriate mental health services were available and accessible to their practice. Sixteen comments mentioned that services were available, however, there were access issues including long wait lists, limited referrals in the community, insurance issues, social work and psychology over-extension, and lack of direct mental health access within the PH clinic. Three comments directly pointed to a lack of available services, specifically citing paucity of services in the community, where it could be convenient for families and used for long-term follow up.

DISCUSSION

The pediatric PH care team members surveyed describe recognizing symptoms of anxiety and depression in their patients >12 years old. This is not surprising given the national rates of anxiety and depression among healthy adolescents and the knowledge that children with chronic illness have higher rates of anxiety and depression than their healthy peers.[1,2] More than half of the respondents recognize symptoms of anxiety in their patients *often* and symptoms of depression *sometimes*. While these providers agree that adolescent PH patients would benefit from regular assessment of anxiety and depression, there is lower confidence that there are appropriate mental health services to refer their patients to if they screen positive.

Despite provider perception of anxiety and depression in PH patients >12 years old, none of the MDs, NPs, or RNs surveyed screen for anxiety and depression with a validated tool with any regularity. It is possible that inclusion of more social workers or psychologists trained to care for PH patients as respondents in this survey would have resulted in increased reporting of screening practices. In a comparable chronic pediatric disease, the Cystic Fibrosis Foundation and the European Cystic Fibrosis Society have published guidelines to make screening for anxiety and depression part of general care for cystic fibrosis patients.[7] Liu et al published a QI project at Seattle Children's Hospital implementing the suggested guidelines into their cystic fibrosis practice. They found a prevalence of anxiety and depression in their population similar to other chronic disease states. Most importantly, they were able to leverage their findings to improve mental health services at their center.[8]

Of those surveyed, all but 1 of the programs have direct access to a social worker in their clinical practice. At least 1 of these social workers screens for anxiety in patients where there is clinical concern. The survey did not elicit more detailed information regarding the social worker's role in mental health screening within the PH practice. While a social worker seems to be a consistent member of a PH program, the presence of a psychologist to the multidisciplinary PH team varies among centers. More interestingly, different members from the same program at 5 of the 14 institutions gave conflicting answers as to access to a psychologist within their practice. At least 2 free-text comments alluded to having a psychologist within their division, however, the psychologist did not provide care for PH patients. This may speak to the fact that mechanisms of accessing mental health services at many institutions is unclear, leaving room for improvement.

All PH care team members surveyed recognized multiple barriers to providing mental health access to their PH patients >12 years old. Some of these barriers may be overcome by PH practice changes, while others are larger systems issues. Eight MDs, 9 NPs, and 3 RNs were not familiar with screening tools for anxiety and depression. Five MDs and 2 NPs noted mental health screening to be out of their scope of practice/not part of PH care. Six providers stated that they were uncomfortable addressing mental health issues. If PH providers followed the lead from their cystic fibrosis colleagues and integrated mental health screening into routine PH care, it would potentially increase provider familiarity with screening tools and their willingness to address anxiety and depression as part of routine clinical

practice. Additionally, routine screening will quantify prevalence of anxiety and depression and highlight the need for improved timely access to mental health services. While integrating routine mental health screening may not alleviate all of the barriers noted by providers, it may highlight faults within the system and serve as a call to action for hospital and governmental leadership to improve access to pediatric mental health care.

LIMITATIONS

One main limitation of this survey is the timing of distribution of the survey during the COVID-19 pandemic. It is not possible to determine how these answers may have been prior to the pandemic. In addition, the primary respondents were physicians, nurse practitioners, and nurses. There were only a few responses from social workers and psychologists, who may be more involved in the mental health screening process in PH clinics. Lastly, there was some variability between respondents at the same center (eg, 1 respondent indicated psychology resources were available while the other indicated they were not). It is unclear as to why some of these questions were answered with variability.

CONCLUSION

This cross-sectional survey highlights the need for improved mental health screening practices within the pediatric PH population. While the current prevalence of disease in this population remains unknown, providers perceive anxiety and depression to be very real issues for their patients. The COVID-19 pandemic has increased rates of anxiety and depression in healthy children, but the presence of an interval increase in pediatric PH patients is not known due to lack of formalized screening practices. As medical care of PH in children continues to improve, management strategies that focus on holistically treating the patient and improving quality-of-life efforts, including mental health evaluation, are important for the long-term health and wellbeing of this unique patient population and their families and caregivers.

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REGULAR ARTICLE

Social Media Participation Among Parents and Caregivers of Children With Pulmonary Hypertension

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Background: Engagement in social media has risen nearly 60% over the last decade with consumers engaging in social media not only browse social networking websites, but also to share health information, view health-related videos, and contribute to online support groups. For parents and caregivers (PCGs) of children diagnosed with pulmonary hypertension, participation in social media can be one way to associate with others in a similar position as individuals can connect with others around the world and share information.

Methods: In this study, parental and caregiver involvement was explored through a cross-sectional study utilizing a survey method to evaluate current practices in social media groups. PCGs of over 300 patients from a large pediatric pulmonary hypertension center were approached for participation in the survey via their child's web-based messaging portal connected to the electronic medical record. The survey was created in REDCap and given to parents electronically or on paper. The survey was comprised of 4 sections covering child demographics, PH medications and hospitalization encounters, social media utilization among PCGs, and the "Big 5 Personality Survey."

Results: Eighteen PCGs completed sections 1-3 of the survey, and 6 PCGs completed the entirety of the survey (sections 1-4). The children represented in the survey had been diagnosed for an average of 5.5 years. Seventy percent of PCGs in the survey reported using social media as a support resource, utilizing 2-5 different social media platforms. PCGs reported feeling overwhelmed, connected, discouraged, depressed, supported, seen, and good when participating. Among respondents, the three highest-scoring personality traits were agreeableness (31.9), conscientiousness (27.2), and openness to experiences (26.7).

Conclusions: This is the first study that seeks to understand social media's role as a support resource and its impact on PCGs of children diagnosed with pulmonary hypertension. It offers insight as to why PCGs may utilize social media platforms and emphasizes its significant role as a support resource for these families. Healthcare teams should consider evaluation of social media and support its role as a resource and source of education.

Over the past decade, social media platforms and applications have become increasingly popular with now 7 in 10 Americans using some form of social media for connecting with others.[1] Platforms and applications are constantly evolving to offer new functionalities and invoke new patterns of use among consumers.[2] The 2019 Health Information National Trends Survey (HINTS) reports that social media involvement has grown from 27% in 2009 to 86% in 2019.[2] The survey finds consumers engage in social media not only to browse social networking websites, but also to share health information, view health-related videos, and contribute to online support groups. With the rapid expansion of this technology over the last decade and the sharp rise in consumer engagement, the integration of social media technology must be further understood as it permeates various aspects of daily life and affects diverse demographics of people. For parents and/or caregivers (PCGs) of children diagnosed with a chronic illness, social media may be one way to associate with others in a similar position as individuals can engage with others around the world and share information.

Pulmonary hypertension (PH) is a chronic disease that can affect children of all ages. It is characterized by elevated blood pressure in the lungs and can be related to a variety of cardiac, pulmonary, and other systemic disorders.[3] As a chronic disease, it may entail many hospital admissions for medication conversions or illness exacerbations, outpatient office visits with multiple different specialty providers, and/or many different medications with frequent administration times. Families may have to travel far distances to receive care at a PH center,[4] and local communities may

Key Words—social media, pediatrics, parents, caregivers
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not understand the disease given its low prevalence worldwide.[5]

In addition to navigating the medical care of pediatric PH, PCGs must also navigate every other aspect of the child's life: school, development, family, and community life. PCGs are frequently faced with nonmedical dilemmas and may be unsure of resources for help. Such situations include schooling modifications like an individualized education plan and educating their child about PH and prognosis. Evaluating support strategies for PCGs of children diagnosed with PH is an important aspect of ensuring family-centered care and working to optimize the overall quality of life of the patient and their family. Health care teams may be aware that PCGs are turning to social media for support and information but may underestimate the extent of social media's impact and therefore miss an opportunity to support or guide social media as a positive resource. In this paper, PCG involvement was explored through a cross-sectional study using a survey method to evaluate current practices in social media groups.

METHODS

In this study, PCGs of over 300 patients from a large pediatric PH center were approached for participation in the survey via their child's web-based messaging portal connected to the electronic medical record. PCGs were also approached during their child's routine outpatient PH appointments and/or during their child's inpatient hospitalization. Surveys were available electronically and on paper. The surveys were completed in English. Inclusion criteria included a parent or caregiver of a child diagnosed with PH who was receiving care at the study site. Exclusion criteria included children not diagnosed with PH or cared for at another institution. The survey was limited to 1 per household, however PCGs were encouraged to complete the survey together, to reflect the collective feelings of all PCGs in the home. Data collection occurred from December 2020 to February 2021.

The web-based survey was created and housed through Research Electronic Data Capture (REDCap, v10.6.12, Vanderbilt University, Nashville, Tennessee). REDCap is a secure web application used for survey data collection. An e-consent was embedded into the survey for electronic signature and was stored within the database. Electronic signatures were obtained by participants typing in their full legal name or signing directly on the REDCap page under *signature*. For PCGs who preferred to complete a paper copy of the survey, the survey was printed out from REDCap, written consent was reviewed, and signatures were obtained in real-time. Data was then directly entered into REDCap from the surveys by the research team.

The survey was divided into 4 sections. Section 1 of the survey included information on demographics including the child's current age, number of individuals living in the household, number of caregivers assisting with the child's care, and the role of the individual(s) who was completing the survey. Section 2 asked about the child's PH including the number of medications for PH taken, length of time of diagnosis, and the number of hospitalizations related to PH or respiratory illness in the past year. Section 3 evaluated the support resources used, the social media platforms used, time spent each day on social media for personal use, changes in use patterns related to the COVID-19 pandemic, how the PCG finds social media groups, benefits of involvement, emotions when participating in social media groups, and other ways the parent or caregiver finds support. Section 4 involved the completion of the 50-question "Big Five Personality Survey."[6] The "Big Five Personality Survey" is based on the 5 broad traits of personality: extroversion, agreeableness, conscientiousness, neuroticism, and openness to experience.[7,8]

LIMITATIONS

There were several limitations identified in this study. One limitation is that majority of the respondents were female and listed themselves as the mother of the child. There were also only 24 responses available for data analysis. Some incomplete surveys were still included in the results of the study as some participants completed the entire survey except for the 50-question "Big Five Personality Survey." This may be because of the length of the "Big Five Personality Survey" when combined with sections 1-3. The survey did not evaluate for the classification of PH of the child such as congenital heart disease or bronchopulmonary dysplasia, which may provide a deeper understanding for other comorbidities of the child and complexity of care needs. Finally, the survey was completed during the COVID-19 pandemic. This limited the number of PCGs present with the child because of visitation restrictions. While the survey attempted to assess how the caregiver or parent's involvement has changed because of the pandemic, it can be difficult to fully interpret how the pandemic has affected social media use in this population.

Table 1. Social Media Platform Use and Time Spent

Type of platform (N = 24)	n (%)
Facebook	21 (87)
Twitter	1 (4)
YouTube	9 (38)
LinkedIn	2 (8)
Instagram	7 (29)
Snapchat	3 (12.5)
Reddit	1 (4)
Other	3 (12.5)
Daily time spent (N = 24)	n (%)
Less than 15 minutes	3 (12.5)
15 minutes to 1 hour	4 (16.7)
1 to 2 hours	10 (41.7)
2 to 4 hours	5 (20.8)
More than 4 hours	2 (8.3)
COVID-19 use changes	n (%)
Did not change	8 (33)
Decreased	2 (8.3)
Increased	15 (58)

RESULTS

Twenty-four PCGs completed the survey. Eighteen of the surveys had all 4 sections completed, and 6 surveys only had the first 3 sections completed.

Section 1: Demographic Data

Among respondents, the mean age of the child receiving PH care was 7 years with a range of 6 months to 16 years. Twenty-three of the respondents were

mothers, and 1 respondent was a grandmother. The average number of household members among all 24 respondents was 4 persons, and most children had a mean of 2 caregivers.

Section 2: PH

Of the 24 responses, children had been diagnosed with PH for a mean of 5.5 years (range 3 months to 15 years.) Children had an average of at least 1 hospitalization in the preceding year (range 0-5, standard deviation 1.25.) Children were taking 0 to 5 medications to treat PH, mean 2 medications, standard deviation 1.27.

Section 3: Social Media Use

Social media platform use is summarized in Table 1. Respondents were asked about the type of support platforms they used. Seventy percent (17 of 24 responses) of the respondents reported using social media as a support resource. Of this 17, 12 respondents reported using social media alone as a resource, and 5 respondents reported using both social media and support groups as a resource. Twenty percent (5 of 24 responses) of the respondents denied using either support groups or social media as a support resource. Three respondents reported support groups alone as their support resource. The use of particular social media platforms including Facebook, Twitter, YouTube, LinkedIn, Instagram, Snapchat, and Reddit was assessed. Fifty percent of the participants (12 of 24 responses) marked use of 2 to 5 platforms. Three respondents marked "other" and reported using TikTok.

PCGs reported finding social media groups through the search function of the social media platforms and recommendations from their health care team and other PCGs. A free-text area was provided for respondents to write how they feel when participating in social media groups. PCGs reported feeling overwhelmed, connected, discouraged, depressed, supported, seen, and good. They also said involvement was helpful and useful. One respondent stated that involvement provided "guidance in decision-making and information gathering from those with actual experience [with the disease]." Another stated they are "always able to find a resource or answer to a question from those with real-life experience." Another free-text area was provided to assess how involvement in social media groups may help others. Responses included gathering learning tips from others, getting support and encouragement, and helping others to know they are not alone. In addition to social media, PCGs were also asked about other support resources they rely on. Family and friends were reported as a support resource by 15 of 24 respondents (62.5%). Other support resources included the Pulmonary Hypertension Association, counseling, and the PCG's faith community.

Table 2. Big Five Personality Survey Data (N = 18)[a]

	Mean	Standard deviation
Extroversion	17.0	8.4
Agreeableness	31.9	4.9
Conscientiousness	27.2	6.2
Neuroticism	19.3	7.4
Openness to experience	26.7	5.1

[a]Scores can range from 0 to 40 on each dimension.

Section 4: "Big Five Personality Survey"

Of the 24 respondents, only 18 respondents completed the "Big 5 Personality Survey." From these responses, scores for extroversion, agreeableness, conscientiousness, neuroticism, and openness to experience were calculated. Table 2 provides an overview of the mean and standard deviation for the "Big Five Personality" scores for this group of respondents.

DISCUSSION

The primary objectives of this study were to (1) determine what resources PCGs use for support of the child's illness, (2) determine common social media platforms used by this population, and (3) understand how social media platforms are perceived by PCGs. All objectives were met for this study, though this study does raise several questions for future investigation.

Support Resources

Pediatric PH is a heterogeneous disease with significant morbidity and mortality. The etiology of disease can vary greatly and carry with it quality of life challenges due to the burden of illness. In their study evaluating the impact of PH on quality of life on children diagnosed with the disease and their caregivers, Mullen et al[9] reported that parents of children diagnosed with PH encountered more stressful events than published norms of parents of children diagnosed with cancer. Parents in Mullen et al's study[9] reported using coping strategies more often than a normative sample, and parental stress correlated inversely with the quality of life of the child.

The data in this survey illustrates the reliance of PCGs on social media as a support resource. While some PCGs reported that involvement can be overwhelming and discouraging, the majority of respondents indicated that involvement was a source of support and encouragement. PCGs appreciated hearing advice from peers who were also dealing with the real-life manifestations of the disease and learning advice to make things easier. Twenty percent of participants noted they do not use social media as a support resource, but later noted they felt supported and less alone when participating in social media groups. This suggests that some participants may experience a lack of recognition or an unwillingness to name social media as an actual support resource.

Social Media Platform Use

Facebook was reported as the primary social media platform used among respondents. This platform as well as most of the others listed in the survey allows participants to actively participate in discussion, read current and historical postings, and show support for discussion through *liking* a particular posting. Participants in this survey were not asked whether they actively participate

through conversations with others or approach social media more passively by reading conversations rather than directly contributing. This would be helpful to understand in future studies, to better characterize the PCG's role in support forums and understand optimal methods of providing supports: passive vs active.

The COVID-19 pandemic also affected the use of social media platforms with over half of participants stating their use increased. This finding was expected given that in addition to social posts, social media is often used as a source of current events and news information. Furthermore, the combination of social isolation, and lack of entertainment during quarantine or lockdown periods through the COVID-19 pandemic, was a likely impetus for more people to scroll through social media. Even during isolating moments, through an internet connection, social media could offer a quick connection to the outside world.

Perception of Social Media Platforms

This study highlights how PCGs feel when participating in social media platforms. PCGs noted the importance of hearing information from others going through a similar experience. Nicholl et al[10] discuss how parents in their study felt empowered when engaging in social media, and their engagement allowed them to better understand their child's condition. Their study also highlighted the importance for health care teams to understand what information PCGs seek in these platforms, which may be areas of missed opportunities for discussion and education.

Personality Assessments

The "Big Five Personality Survey" was included in this study to better understand the types of personalities who may be more inclined to participate in social media groups. Kircaburun et al[8] conducted a similar study seeking to understand the correlation between social media use and the Big Five personality traits among college-aged students. They discovered that differences in social media use were tied to differences in personality traits. For example, persons scoring highest in agreeableness may be more likely to maintain relationships found on social media and seek information and education. Persons scoring highest in extroversion may use social media as a task management tool and may exhibit more self-disclosure. Conscientious individuals may feel more gratified maintaining relationships on social media.

Among respondents, the 3 highest-scoring personality traits were agreeableness (31.9), conscientiousness (27.2), and openness to experiences (26.7). These were scored out of 40, thus a higher score may suggest a stronger personality trait.[8] However, interpretation of the results must be done with caution. The results could indicate an unintentional but inherent selection bias as persons with these traits may be more amenable to completing the research survey in its entirety. Unfortunately, the small sample size limits application to the general PH PCG community.

For the PCGs who completed the survey, the mean extroversion score was below the overall mean among the 5 personality traits. It is unclear if this suggests that those who are not extroverts can use social media for support or that everyone uses social media. The mean agreeableness score was highest among the 5 personality traits. As noted above, this may be a reflection of an inherent selection bias given that more agreeable people may agree to participate in the survey when asked.

An additional limitation to this study is the lack of socioeconomic context for the survey. It would be helpful to also understand demographics and socioeconomic differences among those involved and if these pose barriers to using social media (for example, age of user, access to internet, access to computing devices, literacy, language).

CONCLUSION

This is the first study that seeks to understand social media's role as a support resource and its impact on PCGs of children diagnosed with PH. It offers insight as to why PCGs may use social media platforms and emphasizes its significant role as a support resource for these families. With the rising rates of social media use over the last decade, it remains evident that this resource will continue to be used by PCGs. Health care teams should work to understand the impact of social media use by PCGs and explore opportunities for education on topics PCGs may be wanting more information about. The role of social media as a resource should be promoted with efforts to promote health education and virtual support options.

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Transitions in Pulmonary Hypertension

In January 2022, Claire Parker, RN, MSN, CPNP-AC, Pediatric Nurse Practitioner at UCSF Benioff Children's Hospitals, San Francisco, California; Nancy Bair, RN, MSN, APRN, CNS-BC, Pulmonary Hypertension Advanced Practice Provider at the Respiratory Institute, Cleveland Clinic, Cleveland, Ohio; Neal Chaisson, MD, Director of the Critical Care Medicine Fellowship at the Respiratory Institute, Cleveland Clinic, Cleveland, Ohio; Liza Bonpin, San Francisco, California; and Maddie Bonpin, San Francisco, California, gathered to discuss the lifetime transitions in pulmonary hypertension care.

Claire Parker: Hello. My name is Claire Parker. I'm a nurse practitioner with UCSF Pediatric Pulmonary Hypertension. Today, we're here to discuss some transitions throughout the lifespan of a pulmonary hypertension patient. We've got some wonderful representatives from various PH teams, including a parent and patient. Liza and Maddie, do you want to go ahead and introduce yourself? Maybe, Maddie, you can tell us how old you are and what you've been up to?

Maddie Bonpin: Hi, I'm Maddie I'm 16 years old. I have PH, and I've had it for my entire life.

Liza Bonpin: Hi, I'm Maddie's mom, Liza. I'm Maddie's super fan since she was born. I can't believe she's almost going to be going to college! Nice to be with you all and talk about possible transitions in the near future.

Neal Chaisson: My name is Neal Chaisson. I'm an adult pulmonologist and PH provider. I often deal with adolescent transitions because at the Cleveland Clinic, each provider has a specific subset of PH that we specialize in. The primary patient population that I see are patients with congenital heart disease and associated pulmonary hypertension. Obviously, many of these folks transition up through the pediatric cardiology pathway and into our clinic.

Nancy Bair: I'm Nancy Bair. I'm the pulmonary hypertension Clinical Nurse Specialist at the Cleveland Clinic with Dr Chaisson and 8 other physicians. I've been with the group in PH for about 18 years now.

Claire: Wonderful. We'll start with a question for Liza and Maddie. We would like to know, from your perspective, what is exciting about your transition to adult pulmonary hypertension care and maybe what makes you a little bit nervous or what is unknown about that transition for you?

Maddie: I don't know. I haven't really thought about it because it's not really something I've thought about with the transition to adult care. I don't really have anything that I'm excited or nervous about.

Liza: Are you excited about almost college?

Maddie: Oh, yes, that. I know I'll be in college in a year and a half.

Liza: That's part of the transition, I think.

Maddie: Yes, it is. I'm excited about college because I'm a junior in high school now. I'll be applying soon.

Claire: Maddie, what do you think will be different for you when you come to visits with us versus visits with an adult provider?

Maddie: Probably the same thing with different doctors.

Claire: Are your parents going to continue to come with you to clinic visits?

Liza: It'll probably just be you, I'm thinking. I'm more than happy to accompany you maybe.

Claire: Liza, have you thought about what it will look like with the transition to adult care and more independence for Maddie?

Liza: A little bit. We've been training with getting to know your meds and knowing what you take and not yet how to order the meds. Just those nitty-gritty details. Then just recently talking about what happens when you're in college, if you're living away from home needing to remember to eat right and take your meds. She doesn't really cook or have any interest in cooking. I know she knows how to buy food, as she said. Having a stable job, which she does have a stable job right now, while at school with income to buy your food if you're not going to cook it. Not everyone cooks. That's okay, I told her. Being financially independent as well, which is I think really important. Just knowing how to take care of herself to face herself. That's still a work in progress, but those are all hopes and concerns that she can be independent and know also when to ask for help and who to ask for help with. It's exciting. She's come a long way since 16 years ago. We're looking forward to it. Obviously, start planning now, start practicing now.

Claire: It's been a long journey with your pediatric providers. It's going to be a shift, and it's going to be different. I can tell you from a pediatric provider perspective, we're so excited for when we're able to transition some of our patients to the adult group, and it almost feels like we're bringing you towards graduation. We are both excited and nervous for you to be more independent.

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Sometimes it's a little hard even for your providers to deal with that transition knowing that we won't be involved in your medical care moving forward. Neal, what challenges do you see when young adults transition into your practice?

Neal: I think back to 2 recent experiences, and I think Nancy has worked with me on both of these patients. One patient came of age to transition and was encouraged by her mom to become more independent, but was very scared to leave the *nest*, if you will. This wasn't a bad thing, but she emphasized to me the need to be patient and help her make the transition from dependence on her parents to independence on her terms. Many pediatric pulmonary hypertension patients have travelled this journey shoulder to shoulder with their parents, especially when they're on IV therapies. So navigating things can be a real challenge. We had to really sit down with my patient and establish goals together. Sometimes they worked, and sometimes we had to regroup and provide reassurance and another shot at boosting her confidence.

On the flip side, I had another patient with idiopathic pulmonary arterial hypertension since age 4 who transitioned easily. This individual went to college and insisted on being so independent that they just decided the medications weren't necessary. This obviously created some issues for him. Regrouping and helping his parents to rebuild trust in their son was a challenge. I think that the smoothest transitions occur when we help families, parents and patients alike, to anticipate and discuss what a transition looks like. What are the pitfalls of being too dependent or too independent? Questions like, "How do I get a new provider if I move? How do I choose the right insurance once I leave home? How do I interact with the specialty pharmacy?" These are all important topics and frankly, most adolescents have very limited knowledge of this side of patient life.

Claire: Neal, do you have a social worker or someone that helps with that transition in insurance?

Neal: We have a full-time pulmonary hypertension coordinator, Kasi Timmerman. Her job is to help us with navigating many of these issues, including insurance prior authorizations, obtaining patient copay assistance or grant funding, working with the specialty pharmacies to make sure that patients have as many options as possible etc. She is fabulous, especially given the current challenges we are facing with a lack of funding for patient copay assistance.

Claire: Yes. It's challenging on our end as well. We have a wonderful social worker and nurse coordinator that helps prepare for all of these transitions. Sometimes as prepared as we can be, we still face obstacles. Nancy, do you have any thoughts on challenges and excitement around transition in general?

Nancy: I think the social work issue is definitely an issue for the adult patients, and we do have access to a social worker group that we can find through Epic. They're very good about contacting the patients and following up more for financial kinds of support, not for the medication piece. We have that pretty well nailed with our program coordinator. Compliance is an issue when you have the young adults come into the adult practice and maybe not having mom or dad with them to help them get to their labs, remember to keep their appointments, remember to call us with issues or call the pharmacy back when they call them 50 times to refill their medicine. I think those are the biggest issues.

Claire: Liza asked about the age that we actually transition pediatric patients to adult care. I told her that in our practice it's a bit of a range. Sometimes it depends upon the developmental level of the patient or family dynamics. It sometimes depends upon insurance. In California, that may not be until the age of 21 because we have California's Children's Services, which covers pulmonary hypertension patients until they reach 21. Sometimes our young adults decide that when they transition to college, it would be better to have an adult PH provider which is close to their new residence rather than traveling back to wherever their home pediatric provider is. There is no absolute age cutoff or one age for transition. Do you find that as well?

Nancy: Yes. We have never had a big pediatric presence in the pulmonary hypertension world. That's an interesting point about insurance coverage through the state programs. I am not familiar with Ohio's programs to allow for transition to adulthood. For many years, we have worked together with our Pediatric Cardiology group to comanage the PH patient. The pediatric social workers have helped me with many issues surrounding medication support through our Medicaid programs.

Neal: Our pediatric cardiology group has followed several patients with congenital heart disease throughout their pediatric period and then transitioned them to our group as adults. We recently welcomed a new pediatric cardiologist to focus further on pediatric PH at the Cleveland Clinic. Prior to her arrival and even now, we work together with the pediatric cardiology program on a case-by-case basis to discuss cases that are particularly difficult. As those kids start to transition toward adulthood, they often transition to our group for more specific management of their PH. In some cases, our pediatric cardiologists continue to follow them for their congenital heart disease care. They assume the cardiology portion, we assume the PH portion.

Claire: I think that's great, comanagement over several years. Maddie, whether you've known it or not, we've been thinking about transition for even a few years now, and when we go to appointments and talk to you about what your meds are and having you tell us more, what doses you're taking, and asking if you're starting to call your specialty pharmacy. This is all part of this lead up to more independence and having part of your visit without having your mom in the room, too.

Liza: I do agree with that. That is, I think, maybe the most challenging is

the insurance component. When you become an adult, you don't really realize how important insurance is or how much you need it. A lot of millennials or younger people nowadays think, "Oh, I'm just going to go see the urgent care in the clinic. I don't need to deal with insurance." For Maddie, it's a different story. She needs insurance no matter what. Transitioning from CCS, which has been really amazing regarding prior authorizations and getting the specialty pharmacies to help with the meds and making sure they come in time. Getting all this stuff in line and delivered.

Neal: Maddie, what are the things that you worry about the most or that you hope that your adult physician would be able to provide or continue for you?

Maddie: I haven't really thought about that.

Claire: Maddie's very in the now.

Neal: Yes.

[laughter]

Claire: Are there any tools that you like to see young adults have when they transition to your practice, Neal?

Neal: Yes. I have learned to have the frank discussion with adolescents about the fact that even if mom or dad is still at the appointment, that's not who the appointment is about anymore. I think it's really important to focus my attention on the adolescent patient as the patient and to start moving the focus of communication away from mom and dad. A progression must occur where responsibility for the PH care moves from parent to child. As I said earlier, depending on where you are, we have to meet you there and work with you to make that transition work.

That can be really hard for some people, and it can take some time. For others, they're on the opposite end of the spectrum and they don't want mom and dad to know anything, which is totally fine. We get to work with the consequences of both approaches.

Liza: That's why I encourage practicing now at least with Maddie to encourage her independence. She does get a lot of the appointments and reminders on her phone, right, Maddie?

Maddie: Yes.

Liza: I don't receive the reminders. She's the one that reminds me. That's a start, I think.

Claire: That's a great start. I think Liza, too, we've seen over the last at least year plus that I think you do a really nice job of when we ask a question in clinic, you look to Maddie. She's the one answering her health questions. That's a great move toward independence, and you can help fill in when it's needed. Maddie is getting to be the age where she's really in charge of her body and her medications and knows what's going on better than the rest of us.

Liza: Exactly.

Claire: In thinking about other types of transitions, sometimes we get to the point where we need to transition to aggressive IV or sub-q prostacyclin therapy. Sometimes the medication management support isn't there, whether it's from the patient's own capacity or lack of supportive caregivers. If you run into this, what options do you consider?

Neal: Fortunately, this has not happened in my clinical practice very often. However, in some cases, we see patients who travel from overseas to see us. Sometimes, those folks don't have access to the same medications in their country as we have here. That can be challenging and our options in these cases can be really limited, which is frustrating. We live in a world where access to health care is very unequally distributed.

In other cases, because of religious or social customs, some patients do have access to care but opt against taking certain medications. This issue most commonly arises with patients who I recommend to start parenteral prostacyclin therapy. Fortunately, in the last few years, we've been blessed to have oral prostacyclin or prostacyclin-like pathway medications as an alternative to IV or subcutaneous therapy for these folks. I don't know if this is the ideal alternative, but at least the alternative exists. When I first started, the oral prostacyclin pathway was not available.

Claire: I agree. We actually run into issues like this in pediatrics as well. Sometimes it's not an ideal social situation. There's not a secondary caregiver that can help monitor an IV or sub-q prostacyclin. We hope to give them the best option that aligns with their lifestyle and social situation knowing that it may not be your first choice.

Nancy: I think of one of Neal's patients, actually, that we met for the first time in the hospital and required a parenteral prostacyclin. We had no idea how terrible her living situation was. A specialty pharmacy nurse went to see her and told us that she had no running water. Even though you asked these social questions in the hospital, those critical details were not asked. Often times the patients are too embarrassed to talk about their living situations, whether it is the physical surrounding issues or family support. I think that's the problem when we get the emergency admissions to the hospital. We really don't know the patient.

Claire: I think that's such a good point. Our therapies are not without risk in day-to-day management. I think we need to be very vigilant of all situations. On the flip side, have you had issues where a patient tells you that they would like to transition away from a more aggressive therapy like IV or sub-q prostacyclin now that they're older or maybe their living situation has changed or any other circumstances? When you make this decision, do you tend to get another cardiac cath? Do you follow them more closely? What's your general approach?

Neal: The answer is definitely yes. There's not many people who love IV or subcutaneous therapy. At least none that I know of. I think about all the things that I did as an adolescent that are restricted for these patients simply because of the mechanics of the pump and the

inability to get it wet et cetera. There's a big push in a lot of adolescents, especially as they go off to college, to get off of IV prostacyclins or sub-q prostacyclins. There are obvious limitations to that.

Sometimes we have tried to get people on oral therapy and it's worked well. Sometimes it hasn't. I don't have extensive experience transitioning these patients, but with the last few approaches we've done, what I've said to the patient is, "Let's get a baseline right-heart cath. Let's transition you. Then about 3 months after that transition is complete, we'll repeat your right-heart cath." That's where we've been and it seems to work well for everyone.

Claire: Yes. We've had similar strategies. We've had a handful of adolescents especially that have not done well with sub-q or IV therapy. I think it makes us very nervous to transition away from it, knowing that the other options are not as effective. We also really value the patient's quality of life and want to improve med adherence and prevent bad site pain. I agree, I think we've had some good experiences transitioning from parenteral to oral or inhaled therapy. We've had other kids that did not do well. We've had to have those hard discussions that maybe it's best, even though it's not their favorite, to go back to IV or sub-q therapy.

Nancy: When I was thinking about that question, I was thinking about our elderly population that all of a sudden, whether there may be a change in their mental status and they can't remember to change their cassettes or they no longer have a family member that lives with them, or perhaps they just voluntarily want to move to an assisted living situation. We try to accommodate that because it's not a safe situation that they're going into.

Claire: I think that speaks to the fact that quality of life is a spectrum which balances with medical management.

Nancy: I think it's important to have serious discussions with the patient and family. If you have an elderly patient that you meet first time that has a bad heart cath in the hospital, the first thing everyone wants to do is be really aggressive. We have all likely had to deal with a situation when a patient needs care beyond the hospital. I think it's important to have that discussion if the family thinks that at some point their loved one is going to need an assisted living or skilled nursing facility. Parenteral prostacyclins are generally not accepted in those facilities, which puts everyone in a stressful situation. Some of our oral medications can be quite complex because the patient has to put together different doses to make the final dose. In someone with even early dementia, an error can be life threatening.

Claire: That's a good point. The last question we have is in regard to transitioning to hospice. How have you dealt with urgent hospice situations for patients who are on parenteral medications? Or when the patient is leaning toward hospice, but the family is reluctant to stop the pulmonary hypertension medications?

Neal: Oh, boy. This is a hard one. I think that this is a very personal decision. I think first and foremost, we have to remember to put away whatever is on our agenda and recognize that we're there to support a family who's in a state of crisis. Sometimes that takes a little bit of time and listening and working on a timetable that isn't our own. I heard a quote one time that before patients can accept your care, they need to know you care. I think that that's part of the key in hospice discussions.

If a patient doesn't trust that you have their best intentions in mind, then the ability to navigate conversations that are delicate, such as those related to hospice, tend to be fraught. Stopping to just listen to what the concerns of a patient and family are can be really helpful toward assuring them that you're working on their timeline. Sometimes that requires months, or years, or sometimes it requires very little time. At the end of the day, I have never pushed somebody against their will to come off of medication. What I have done when the situation presents itself is help to try to explain what the purpose of hospice is and how that's different from what the purpose of the medications are.

Giving patients and families the space to recognize that hospice is not trying to treat the disease, but helping to treat the patient provides an alternative to the narrative we're just withdrawing support. In fact, we're actually providing a different level of support that is wholly dedicated to empowering patients to pass away on their terms, with dignity and without suffering. That is powerful. That can't happen until families trust that in fact, the support you're proposing to provide is actually in their love one's best interest.

Claire: I absolutely agree. Aligning with your patient and really listening to what they're asking for in life, and how they want to live out whatever time they have left, can really change your strategy on how to best treat your patient. That may mean aggressive medical management or more comfort care and time outside of the hospital. It is a broad spectrum.

Nancy: I think this is always such a difficult situation. I know there've been times where we've had patients who wanted to transition to hospice but did not want to abruptly stop their PH medications. Before finalizing the hospice contract, the patients ordered their medications which allowed them to transition more comfortably. It is important to discuss with hospice and patient to develop the plan of care.

Claire: Does anybody have any last thoughts? I think we're just coming to the end of our time.

Neal: There isn't a magic formula for the transition between pediatric and adult care. It's not like, once you hit 18, there's some other human being inside that body that magically changes or flips a switch which necessitates care by one provider or another. It's a continuum. In my opinion, the best programs provide a continual off-ramp and an on-ramp. At the end of the day, recognizing that the patient is the customer,

and it's our job to work with them and not the opposite, is important. This was engrained in me when I first showed up at the Cleveland Clinic. But in the hustle of our jobs, it is easy to lose sight of why we're here. As Nancy said, our pediatric PH program is pretty small. The patients who we have had transition and the congenital heart disease patients we have had transition have been an opportunity for us to build fantastic relationships with patients and their pediatric providers. Over the years as those relationships have grown, we've built mutual trust. This, in turn, helps build rapport among patients who have been with that practitioner for years and years and are now transitioning.

Claire: Yes. I agree. Absolutely. I think that is a wonderful way to wrap up our roundtable discussion. Thank you all so much for taking the time out of your day to join us.

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