

# Integrated Transitions of Care for Patients With Rare Pulmonary Diseases

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## ABSTRACT

**Purpose/Objectives:** Many continuing education (CE) resources are available to support case management professionals in developing competencies in transitions of care (TOC) that apply generally across disease areas. However, CE programs and tools are lacking for advanced TOC competencies in specific disease areas. This article describes 2 projects in which leading TOC, case management, and CE organizations collaborated to develop CE-accredited interdisciplinary pathways for promoting safe and effective TOC for patients with rare pulmonary diseases, including pulmonary arterial hypertension (PAH) and idiopathic pulmonary fibrosis (IPF).

**Primary Practice Setting(s):** The interdisciplinary pathways apply to PAH and IPF case management practice and TOC across settings that include community-based primary care and specialty care, PAH or IPF centers of expertise, acute care and post-acute settings, long-term care, rehabilitation and skilled nursing facilities, and patients' homes.

**Findings/Conclusions:** Both PAH and IPF are chronic, progressive respiratory diseases that are associated with severe morbidity and mortality, along with high health care costs. Because they are relatively rare diseases with nonspecific symptoms and many comorbidities, PAH and IPF are difficult to diagnose. Early diagnosis, referral to centers of expertise, and aggressive treatment initiation are essential for slowing disease progression and maintaining quality of life and function. Both the rarity and complexity of PAH and IPF pose unique challenges to ensuring effective and safe TOC. Expert consensus and evidence-based approaches to meeting these challenges, and thereby improving PAH and IPF patient outcomes, are presented in the 2 interdisciplinary TOC pathways that are described in this article.

**Implications for Case Management Practice:** In coordinating care for patients with complex pulmonary diseases such as PAH and IPF, case managers across practice settings can play key roles in improving workflow processes and communication, transition planning, coordinating TOC with centers of expertise, coordinating care and TOC for patients with comorbidities, providing patient and caregiver education, promoting engagement between patients and the team, advancing the care plan, and improving ongoing adherence to treatment in order to maximize the patient's pulmonary function. Details regarding these interprofessional roles and responsibilities are provided in the full interdisciplinary TOC pathways for PAH and IPF.

**Key words:** continuing education, idiopathic pulmonary fibrosis, pulmonary arterial hypertension, transitions of care

Continuing education (CE) resources are available to support case management professionals in developing competencies in transitions of care (TOC) that apply generally across various disease areas. However, relatively few CE programs

and tools address advanced TOC competencies for specific diseases or conditions. This article describes two CE-accredited pathways developed to support case managers and other health care professionals in promoting safe and effective TOC for patients with

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*Pulmonary arterial hypertension symptoms include dyspnea, fatigue, weakness, chest pain, syncope, abdominal distention, and ankle swelling. Because it is a relatively rare disease and its symptoms are nonspecific, PAH is often misdiagnosed or diagnosed when patients have already progressed to stages of severe functional impairment.*

rare pulmonary diseases, including pulmonary arterial hypertension (PAH) and idiopathic pulmonary fibrosis (IPF).

The PAH and IPF interdisciplinary TOC pathways were developed through a collaboration among PRIME Education, Inc. (PRIME); the National Transitions of Care Coalition (NTOCC); and the Case Management Society of America (CMSA). CMSA is the leading membership association providing professional collaboration across the health care continuum to advocate for patients' well-being and improved health outcomes by fostering case management growth and development, impacting health care policy, and providing evidence-based tools and resources. NTOCC works with hundreds of organizations—including leading professional health care associations, medical specialty societies, standards bodies, and government agencies—to develop and

implement educational resources and tools for TOC. PRIME is a national health care CE company.

In this article, we describe our collaborative process for developing the interdisciplinary TOC pathways on PAH and IPF, summarize their contents and educational implications, and highlight their applications to professional case management. We encourage readers to download the complimentary full pathway documents, each of which is accredited by the Commission for Case Manager Certification and the American Nurses Credentialing Center for 2 hr of CE credits. The PDF documents are available at no cost from NTOCC's website at [www.ntocc.org](http://www.ntocc.org).

## COLLABORATIVE PROCESS FOR DEVELOPING THE INTERDISCIPLINARY TOC PATHWAYS

The interdisciplinary pathways on TOC for patients with PAH or IPF were developed through a collaborative workgroup model, with interprofessional clinical representation from physicians, pharmacists, nurses, and case managers representing diverse practice areas including acute care, preventative care, long-term care, home care, workers' compensation, and rehabilitation.

The workgroups were guided by the principle that *necessary care transitions for patients with PAH or IPF should be safe and unnecessary transitions should be prevented*. Moreover, in developing and supporting their recommendations, we used established evidence-based and expert consensus publications and other resources on TOC, care coordination, and quality improvement (see Table 1). Other guiding themes for developing the interdisciplinary

**TABLE 1**  
Publications and Resources Used to Develop the PAH and IPF TOC Pathways

Publication/Resource	Authors	Reference/Website
Transitions of Care Consensus Policy Statement	ACP/SHM/SGIM/AGS/ACEP/SAEM	Snow et al., 2009
Transitions of Care in the Long-Term Care Continuum Practice Guideline	American Medical Directors Association	<a href="http://www.paltc.org">www.paltc.org</a>
Care Transitions from Hospital to Home: IDEAL Discharge Planning	Agency for Healthcare Research and Quality	<a href="http://www.ahrq.gov">www.ahrq.gov</a>
NQF-Endorsed Care Coordination Measures	National Quality Forum	<a href="http://www.qualityforum.org">www.qualityforum.org</a>
NTOCC	NTOCC	<a href="http://www.ntocc.org">www.ntocc.org</a>
CMSA	CMSA	<a href="http://www.cmsa.org">www.cmsa.org</a>
VNAA Blueprint for Excellence	VNAA	<a href="http://www.vnaablueprint.org">www.vnaablueprint.org</a>
PHA	PHA	<a href="http://www.phassociation.org">www.phassociation.org</a>
PFF	PFF	<a href="http://www.pulmonaryfibrosis.org">www.pulmonaryfibrosis.org</a>
CHEST Practice Guidelines for PAH	<i>Chest</i>	Taichman et al., 2014
ESC/ERS Practice Guidelines for PAH	ESC/ERS	Galie et al., 2016
ATS/ERS/JRS/ALAT Practice Guidelines for IPF	ATS/ERS/JRS/ALAT	Raghu et al., 2015

*Note.* ACEP = American College of Emergency Physicians; ACP = American College of Physicians; AGS = American Geriatric Society; ALAT = Latin American Thoracic Association; ATS = American Thoracic Society; CMSA = Case Management Society of America; ERS = European Respiratory Society; ESC = European Society of Cardiology; IPF = idiopathic pulmonary fibrosis; JRS = Japanese Respiratory Society; NTOCC = National Transitions of Care Coalition; PAH = pulmonary arterial hypertension; PFF = Pulmonary Fibrosis Foundation; PHA = Pulmonary Hypertension Association; SAEM = Society for Academic Emergency Medicine; SGIM = Society of General Internal Medicine; SHM = Society of Hospital Medicine; VNAA = Visiting Nurse Associations of America; TOC = transitions of care.

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pathways were accountability and evaluation of TOC processes according to national standards and quality measures; effective communication, including timely and secure transfer of patient health information; patient and family/caregiver engagement; recognition of the patient and family/caregiver as part of the interprofessional team; recognition of the patient home as a care setting; and establishment of medical homes or coordinating community clinicians to support PAH or IPF patients and their families/caregivers in navigating the health care system.

The two interdisciplinary pathways are designed for health care professionals across the care continuum; however, they emphasize the central roles of case managers in care coordination and TOC, engaging the entire clinical team and respecting the needs and wishes of patients and their families. In addition to presenting foundational knowledge about PAH and IPF disease process and treatments, the pathways include recommendations and supporting evidence for promoting safe and effective TOC. These recommendations address transitions between key settings, including primary care, specialty care, PAH or IPF centers of expertise, acute care, long-term care, post-acute care, and patients' homes. The pathway documents also include practical TOC tools, including checklists for essential care transition interventions and handouts that patients can use to keep track of their medications and prepare for appointments with their health care providers.

## **KEY FOUNDATIONS OF KNOWLEDGE ABOUT PAH AND IPF**

The interdisciplinary PAH and IPF pathways emphasize the importance of professional development and CE for building current evidence-based knowledge about the diseases to inform appropriate TOC practices. The pathways thus include information about PAH and IPF epidemiology, common health care gaps and barriers, and unmet treatment needs. In addition, evidence-based guidelines are reviewed for the diagnosis, treatment, and management of PAH and IPF. In the following sections, we summarize key aspects of PAH and IPF knowledge that are addressed more comprehensively in the interdisciplinary TOC pathways.

## **PAH Epidemiology and Care Gaps**

Pulmonary arterial hypertension is a type of pulmonary hypertension that, as diagnosed through right-sided heart catheterization, is characterized by elevated pulmonary arterial pressure and vascular resistance (McGoon & Kane, 2009). If left untreated, PAH leads to right ventricular heart failure and death. The U.S. prevalence of PAH is estimated to be 12.4 cases per million adults (Frost et al., 2011). Mean age at diagnosis is approximately 50 years, and the 5-year survival rate is estimated to be 60% (Frost et al., 2011; Theppan, Ryan, & Archer, 2012). Pulmonary arterial hypertension is associated with poor quality of life and high rates of morbidity, hospitalization, and mortality; in addition, health care costs and resource utilization for PAH patients are significant (Sikirica, Iorga, Bancroft, & Potash, 2014).

The symptoms of PAH include dyspnea, fatigue, weakness, chest pain, syncope, abdominal distention, and ankle swelling. Because it is a relatively rare disease and its symptoms are nonspecific, PAH is often misdiagnosed or diagnosed when patients have already progressed to stages of severe functional impairment (Galie et al., 2016). A definitive diagnosis is often delayed as numerous tests are conducted to rule out other, more common diseases (Galie et al., 2013). In an analysis of 2,525 patients with PAH in the REVEAL Registry, the mean length of time from onset of symptoms to diagnosis was 2.8 years (Badesch et al., 2010). With this delay, patients miss key opportunities for early treatments that can significantly improve outcomes (Theppan, Shah, Rich, & Gomberg-Maitland, 2007). Moreover, as many as 75% of patients are readmitted within 1 year of a PAH-related hospitalization, with 20% of these patients being readmitted within 30 days of discharge (Lacey, Buzinec, & Hunsche, 2013). Common comorbidities add to PAH case management and TOC challenges. In a REVEAL Registry analysis, 40% of patients had hypertension, 29% had collagen vascular disease/connective tissue disease, 25%

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had clinical depression, 22% had thyroid disease, and 12% had diabetes (Badesch et al., 2010).

IPF Epidemiology and Care Gaps

Idiopathic pulmonary fibrosis is a chronic and progressive disease of unknown cause that affects interstitial tissue in the lungs. The U.S. prevalence of IPF is estimated to be 18.2 cases per 100,000 adults (Raghu, Chen, Hou, Yeh, & Collard, 2016). Mean age at diagnosis is approximately 66 years, and the median survival is 3–5 years from diagnosis (Puglisi et al., 2016). IPF is characterized by:

- 1. nonspecific symptoms that also affect patients with other pulmonary diseases, as well as cardiovascular and rheumatological diseases;
- 2. various comorbidities, including chronic obstructive pulmonary disease, lung cancer, pulmonary hypertension, venous thromboembolism, sleep apnea, gastroesophageal reflux disease, depression and anxiety, and cardiovascular disease; and, when inadequately treated,
- 3. rapid deterioration and severely debilitating effects on function and quality of life. (Fulton & Ryerson, 2015)

These characteristics underscore the unique systems- and team-based challenges involved in effectively diagnosing, treating, and managing patients with IPF. In addition, they account for well-documented gaps in clinical processes, coordinated care, and care transitions in this field (Spagnolo, Tonelli, Cocconcetti, Stefani, & Richeldi, 2012). Studies have indicated that the duration between IPF symptom onset and diagnosis ranges between 1 and 4 years (Lamas et al., 2011; Spagnolo et al., 2012). Accurate diagnosis requires a multidisciplinary, interprofessional approach in TOC to appropriately transition patients through multiple levels of care and specialists including, among others, pulmonologists, radiologists, pathologists, surgeons, rehabilitation spe-

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Current Medication Management for PAH

Although PAH remains incurable, recent advances in vascular-targeted pharmacotherapies have significantly improved patient outcomes (Hill, Cawley, & Heggen-Peay, 2016). As shown in Table 2, the main Food and Drug Administration (FDA)-approved vascular-targeted pharmacotherapies include calcium channel blockers for vasoreactive cases, endothelin receptor antagonist, phosphodiesterase Type 5 (PDE-5) inhibitors, prostacyclin analogues, a guanylate cyclase stimulator, and a prostacyclin receptor agonist. As detailed in the TOC pathway for PAH, appropriate therapy decisions depend on assessing many complex factors associated with patients’ risk for mortality and issues related to the different routes of therapy administration (Galie et al., 2016). The pathway also summarizes findings from clinical trials on vascular-targeted PAH pharmacotherapies that have demonstrated endpoints including improvements in exercise capacity and 6-min walk test distance, functional status, quality of life, hemodynamics, time to clinical worsening, and survival.

TABLE 2  
Vascular-Targeted Pharmacotherapies for Pulmonary Arterial Hypertension

Pharmacotherapy Class	Agents
Calcium channel blockers for vasoreactive cases	Amlodipine, nifedipine, and diltiazem
Endothelin receptor antagonists	Bosentan, ambrisentan, and macitentan
Phosphodiesterase Type 5 inhibitors	Sildenafil and tadalafil
Guanylate cyclase stimulator	Riociguat
Prostacyclin analogues	Intravenous (epoprostenol and treprostinil) Subcutaneous (treprostinil) Inhaled (iloprost and treprostinil) Oral (treprostinil)
Prostacyclin receptor agonist	Selexipag



A major recent advancement in PAH therapeutic strategies emphasizes early and aggressive use of an initial combination of medications. This represents a paradigm shift in PAH treatment and management, which have traditionally involved stepwise additions of single agents (Galie et al., 2016). This change in treatment practice has required coordinated care and medication management protocols established by pharmacists, nurses, primary care and specialty physicians, and case managers, among others. On the basis of positive results from the AMBITION clinical trial, in 2015, the FDA approved combination ambrisentan and tadalafil therapy for the first-line treatment of PAH (Galie et al., 2015). In the most recent international guidelines for PAH, the AMBITION trial results are cited as evidence for the recommended use of combination therapy as a first-line option for PAH patients in selected groups (Galie et al., 2016).

### **Current Medication Management for IPF**

Until recently, efforts to develop safe and effective pharmacotherapies for IPF have been marked by unsuccessful clinical trials (Spagnolo et al., 2012). Indeed, therapies that are effective in treating other forms of fibrotic lung disease, such as the combination of prednisolone, azathioprine, and N-acetylcysteine, are potentially harmful in IPF patients, underscoring again the importance of accurate diagnosis (Raghu et al., 2015). In 2014, however, successful results were reported from Phase 3 clinical trials on two new oral medications: pirfenidone (King et al., 2014) and nintedanib (Richeldi et al., 2014). Pirfenidone, which has antifibrotic and anti-inflammatory properties, significantly reduces rates of IPF-related declines in lung function, as measured by forced vital capacity (FVC), and 6-min walk test distance. In addition, pirfenidone has been demonstrated to improve progression-free survival in IPF patients (King et al., 2014; Puglisi et al., 2016). Nintedanib, which acts by inhibiting fibrotic processes in IPF pathogenesis, also significantly reduces the rate of FVC decline and, among some patients, prolongs the time to acute exacerbations (Richeldi et al., 2014, 2016). On the basis of their efficacy data and acceptable safety profiles, pirfenidone and nintedanib were approved by the FDA in 2014. Recent evidence supports treatment initiation with pirfenidone or nintedanib soon after diagnosis, when lung function is relatively preserved (Albera et al., 2016; Raghu et al., 2015). These advances in treatment require case managers to work more closely with pharmacists and the entire clinical team than ever before to manage and support medication adherence, medication reconciliation, patient education, and overall medication management as part of effective care transitions.

As is also true for PAH, the medication management of patients with IPF may require physical exercise, pul-

monary rehabilitation, and supplemental oxygen therapy. In addition, patients with moderate to severe disease may be eligible for lung transplantation. These treatments, along with coordinated approaches to palliative care, are addressed in more detail in the TOC pathways.

### **Barriers to Safe and Effective TOC for Patients With PAH or IPF**

In general, vulnerable TOC points for patients with PAH or IPF can include hospital admissions and readmissions, transfers to and between the post-acute care entities, or discharge to home with coordination of care for equipment and supplies. Specific TOC barriers are related to the unique characteristics of patients with PAH or IPF, including their age, comorbidities, special therapy needs, and potential therapy-related complications. As an example, for patients with PAH, long-term care facilities and skilled nursing centers for rehabilitation may not have the expertise or equipment to administer prostanoid therapy. Delays in medication are also a common issue in care transitions. Seamless transitions to self-care at home can pose major challenges for PAH or IPF patients with low health literacy, inadequate self-management skills, lack of caregiver support, special cultural needs, physical comorbidities, or mental health disorders. The interdisciplinary TOC pathways for PAH or IPF include expert recommendations and evidence-based strategies for overcoming these common barriers.

### **ROLES OF CASE MANAGERS IN TOC FOR PATIENTS WITH PAH OR IPF**

As emphasized in the two TOC pathways, case management professionals play key roles in PAH and IPF education, advocacy, patient and family caregiver engagement, development of care plans, and care coordination. The following sections summarize major points about these roles and the related supportive evidence and resources that are presented in the pathway documents for utility by case managers.

### **Coordinating Workflow Processes and Communication of Interprofessional Care Teams**

To ensure effective care transitions for PAH and IPF patients, and to reduce risks of negative outcomes such as preventable rehospitalizations, extensive interprofessional coordination is required in processes that include transition planning, reconciling medications, communicating patient health information in a timely fashion, educating and empowering patients and caregivers, monitoring treatment outcomes, and managing comorbidities. Given the complex nature of PAH and IPF, the roles and responsibilities for these processes are

distributed across different health care settings. Within each setting, specific care transition responsibilities should be identified for each team member. However, without oversight and coordination of processes across settings, care transitions are likely to be compromised. This point addresses the key roles that case managers can play in coordinating key processes of interprofessional PAH or IPF care. Moreover, case managers are ideally positioned to provide training and education to interprofessional teams on the coordinated management of care transitions of PAH and IPF patients.

Safe and effective care coordination and TOC for patients with PAH or IPF depend on comprehensive, clear, and timely communication among interprofessional team members. The interdisciplinary pathways on PAH and IPF provide recommendations for establishing and maintaining effective bidirectional communication and transfer of patients' medical records, care plans, and TOC plans (see Table 3).

Transition Planning

Because of the incurable and progressive nature of PAH and IPF, exacerbations of comorbid diseases, and interposition of complications such as pneumonia, many patients undergo multiple care transitions throughout the course of their disease. As emphasized in the interdisciplinary pathways, effective transition planning requires clear identification of the roles of care team members and processes for addressing family/caregiver needs. The pathways include detailed recommendations for TOC planning, accounting for key roles of case managers in:

- 1. conducting comprehensive assessments of patients' clinical and nonclinical needs;
- 2. participating in the development and documentation of TOC plans;

- 3. ensuring that all care team members, including patients and caregivers, understand the plans and have the necessary supportive resources to carry them out; and
- 4. coordinating systems- and team-based processes to ensure the timely, safe, and effective completion of TOC stages.

Coordinating TOC With Centers of Expertise

As emphasized in leading clinical practice guidelines and by medical societies for PAH and IPF, patients should receive care for these diseases from clinicians in established centers of expertise (Galie et al., 2013; Raghu et al., 2015; Taichman et al., 2014). In 2011, the Pulmonary Hypertension Association (PHA), which is the leading advocacy organization for patients with PH and PAH, launched an initiative to accredit centers of experience across the United States. The goal of this initiative is to improve the quality of care and outcomes in patients with all types of pulmonary hypertension by promoting comprehensive patient care, patient education, standardization of care, patient information flow, and coordinated care involving all health care providers of PAH patients. A list of accredited centers is available on the PHA website. For patients with IPF and their caregivers, the Pulmonary Fibrosis Foundation has collated a network of care centers with experience and expertise in treating patients with fibrotic lung disease. Although PH and IPF centers of expertise provide comprehensive disease-specific care, circumstances such as hospital admissions or clinic visits for comorbid conditions require expertise in TOC. For case managers, opportunities may exist to work with PAH or IPF centers of expertise to ensure safe and effective transitions across these settings. The TOC pathways clarify potential roles and responsibilities

TABLE 3  
TOC Pathways Recommendations for Shared Accountability and Communication

<i>Clear and timely communication of the patient's plan to all health care team members and across all sites of care during transitions</i>
<ul style="list-style-type: none"><li>• Health care provider must communicate plan of care to the patient and the receiving provider before handoff is completed</li><li>• The sending provider must be available to the receiving provider for any questions and clarifications regarding the patient's care after the handoff<ul style="list-style-type: none"><li>○ Notify the receiving entity of the patient diagnosis of PAH or IPF, that specialty care is involved, and the need to involve the expert care center as appropriate</li><li>○ Inform the receiving entity of the contact information for the patient's expert care center</li></ul></li></ul>
<i>Ensuring that a health care provider is responsible for the care of the patient at all times</i>
<ul style="list-style-type: none"><li>• The sending health care provider must remain responsible for patient's care until the receiving provider has acknowledged that he/she can effectively assume the care of the PAH or IPF patient</li><li>• The receiving provider has to acknowledge the receipt of transferred information in a timely manner, understand the plan of care for the patient, and be prepared to assume responsibility for patient's care</li></ul>
<i>Assuming responsibility for the outcomes of the care transition process by all relevant health care providers</i>
<ul style="list-style-type: none"><li>• If the provider who has assumed care of the patient determines that the patient should go to another level of care than provided, the provider is responsible for communicating with the receiving provider before handoff and notifying the PAH or IPF treatment chain of care<ul style="list-style-type: none"><li>○ Be involved in the patient's transition safety and outcomes report, utilizing quality indicators including disease-associated indicators</li></ul></li></ul>

Note. IPF = idiopathic pulmonary fibrosis; PAH = pulmonary arterial hypertension TOC = transitions of care.

of case managers in working with PAH or IPF centers of expertise.

For patients who may not have access to centers of expertise, it is essential that the established centers maintain, whenever possible, the potential for coordination of care and oversight to local clinical caregivers. This effort has the potential to provide PAH and IPF patients the best care practicable to geographical areas and patients without direct access to the centers.

### **Coordinating Care and TOC for Patients With Comorbidities**

As described earlier, comorbidities can pose significant challenges in care transitions for patients with PAH or IPF. Across health care settings, team members should be made aware of patients' comorbidities, current medications, and drug contraindications or interactions. For example, some front-line therapies for PAH are contraindicated for certain patients, such as those with Eisenmenger syndrome or portopulmonary hypertension who should not take calcium channel blockers (Galie et al., 2016). PDE-5 inhibitors and prostacyclins may interact with nitrates, nitric oxide donors, antihypertensives, and other vasodilators; thus, these therapies may be contraindicated for patients with PAH and heart disease (Galie et al., 2016). Addressing these considerations, the TOC pathways provide essential knowledge about PAH and IPF comorbidities as well as potential drug contraindications and interactions. In addition, the pathways provide guidance for medication reconciliation in the context of TOC.

### **Providing Education and Promoting Engagement for PAH and IPF Patients**

Leading health care organizations, including The Joint Commission, have recognized the importance of patient/caregiver education and engagement in supporting safe and effective TOC (The Joint Commission, n.d.) In addition, recent studies have identified the key roles that families and caregivers play in TOC,

*Early in PAH or IPF disease progression, patients and families/caregivers should be provided with education, regular discussions on their disease progression, and support in advance care planning decisions. Ideally, these discussions should begin early, evolve over time, and be revisited at the beginning of each level of treatment.*

such as supporting medication management and arranging posttransition care plan tasks (Coleman & Roman, 2015; Coleman, Roman, Hall, & Min, 2015). These developments underscore the essential roles that case managers may play, with appropriate training, in providing education and promoting engagement for PAH and IPF patients. Topics may include risks and benefits of available treatment options, medication use and adherence, maintaining accurate medication lists, self-care practices, and establishing and maintaining relationships with centers of expertise, advocacy organizations, and support groups.

### **Advance Care Planning**

Early in PAH or IPF disease progression, patients and families/caregivers should be provided with education, regular discussions on their disease progression, and support in advance care planning decisions. Ideally, these discussions should begin early, evolve over time, and be revisited at the beginning of each level of treatment. In the setting of shared patient decision making and fully informed consent, patients considering late-stage therapies should be informed about the challenges they will likely need to navigate and the impact of opting out of treatments or starting and later stopping them. In a survey study, many caregivers of PAH patients who died from the disease reported that they were not aware of palliative

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care and hospice service options (Grinnan et al., 2012). Physician-reported barriers to referrals to palliative care for patients with PAH include lack of approval by patients or their families/caregivers and concern that it represents “giving up hope” (Fenstad et al., 2014). Similarly, a recent retrospective analysis found that few IPF patients received a referral to palliative care, and when referrals were made, it was often late in the disease course (Lindell et al., 2015). Considering this evidence for some hesitation among physicians and patients/families/caregivers to discuss advanced planning, case managers may play supportive and coordinating roles in planning and referrals to specialists in palliative and end-of-life care. Case managers play an important role in ensuring that advance directives and patient preference for end-of-life care are shared and transferred at each level of care.

## IMPROVING PAH AND IPF CARE COORDINATION AND TRANSITIONS: FUTURE DIRECTIONS FOR CASE MANAGERS

Both PAH and IPF are profound pulmonary diseases requiring proactive and effective TOC to ensure that affected patients are receiving timely and efficacious treatment in the least invasive level of care and in the most cost-effective manner. Because of the number of diverse providers and settings often involved in PAH and IPF care, coordination of services is paramount. Case managers and all involved health care professionals can benefit from the stepwise protocols and TOC resources available in the PAH and IPF NTOCC pathways, including the “7 Essential Interventions” developed by the NTOCC and adapted specifically to PAH and IPF in each guide. Health systems and their health care teams may also realize improved patient outcomes through implementation of the pathways in interprofessional practice.

With current national emphasis on care coordination via health policy enactments, including the Centers for Medicare & Medicaid Services’ Medicare Access and CHIP Reauthorization Act of 2015 (MACRA), case managers have a renewed opportunity to demonstrate value. The case manager plays a central role in TOC that can fundamentally improve patient and caregiver outcomes, as well as systems-based outcomes in complex diseases such as PAH and IPF. As national health policies give impetus to case management services, case managers can give impetus to effective TOC.

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