Pulmonary Hypertension Patient Navigation: Avoiding the Perfect Storm

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Pulmonary arterial hypertension (PAH) is a progressive, incurable disease that presents a challenging journey for all involved. Specialized, complex care and treatment is needed for this population of patients, and should be provided in an organized, systematic manner to promote optimal patient outcomes. The concept of patient navigation can be used as a framework for the pulmonary hypertension (PH) center, so that care delivery is well structured and PAH patients have a guide to assist them through all aspects of the health care continuum. This article will focus on how a PH patient navigation program can be implemented and the role of a PH nurse navigator, using the Christiana Care Health System Pulmonary Hypertension Program in Newark, Delaware, as an example. There have been many advances in PAH diagnosis and treatment in the last 20 years, and the time has come to introduce a PH patient navigation model that can be used as a guide to structure PH programs.

PATIENT NAVIGATION BACKGROUND

The multidisciplinary team approach to caring for pulmonary arterial hypertension (PAH) patients has been documented as the accepted standard of care, and patients benefit from other disciplines that may be involved in the treatment plan in addition to doctors and nurses such as nurse practitioners, pharmacists, respiratory therapists, and social workers.[1] There can be many obstacles to proper PAH treatment that must be overcome by the pulmonary hypertension (PH) team to better control symptoms and achieve a longer life span for the patient. The path is not always clear, and there is often a great deal of time and energy expended to navigate the maze of all that is necessary to achieve effective management of the disease, especially for patients and their caregivers.

A growing number of resources are available to assist the PH team with different aspects of PAH care and treatment, but none that includes a model of all care components the PH center can use to help structure a planned or existing PH program. A PH patient navigation model includes principles for the PAH patient population that can be adopted by the center to enhance multidisciplinary efforts toward individualized treatment goals. The concept of patient navigation was founded in 1990, by Dr. Harold P. Freeman in Harlem, New York, to focus on health care disparities and effectively manage cancer patients across the health care continuum by using professionals to support individual patients.[2] Today, the Harold P. Freeman Patient Navigation Institute trains individuals to become patient navigators not only for cancer patients, but also for other chronic illnesses, and his program is the gold standard of patient navigation.[3] Trained patient navigators are better equipped to assist patients with barriers to care because there are well-defined roles and responsibilities and a set model to follow. Many cancer centers, such as the one at Christiana Care Health System (CCHS), use patient navigation to achieve better outcomes, increase patient satisfaction, and decrease institutional financial burdens by expanding timely access to care. Since 2015, all cancer programs accredited by the Commission on Cancer must have patient navigation as the standard of care.[2,4] Thus, PAH patients can likewise benefit from the consolidated efforts that a PAH patient navigation model can bring to overall care and treatment.

INCORPORATION OF PATIENT NAVIGATION INTO THE PH PROGRAM

The PH program at CCHS has the opportunity to incorporate patient navigation principles into the way patients are managed, using an adaptation of the Harold P. Freeman Institute's philosophies. In the model developed for the CCHS PH program, the basic continuum spans the period between diagnosing a patient with PAH through optimal symptom management and prolongation of lifespan (Figure 1). The overall goal of the model is access to PAH diagnosis and treatment, as statistics show that the average time between symptom manifestation and treatment remains at 2.8 years, and precious time is lost until the diagnosis is made.[5] Therefore, the first area of the model covers early recognition of patients at higher risk for developing PAH. Patients that are diagnosed at earlier stages and receive intervention have been proven to have decreased disease progression.[6] Although not uniformly accepted by the medical community, according to screening guidelines from the American College of Cardiology Foundation and the American Heart Association, patients with known familial PAH and/or BMPR2 mutations, systemic sclerosis,

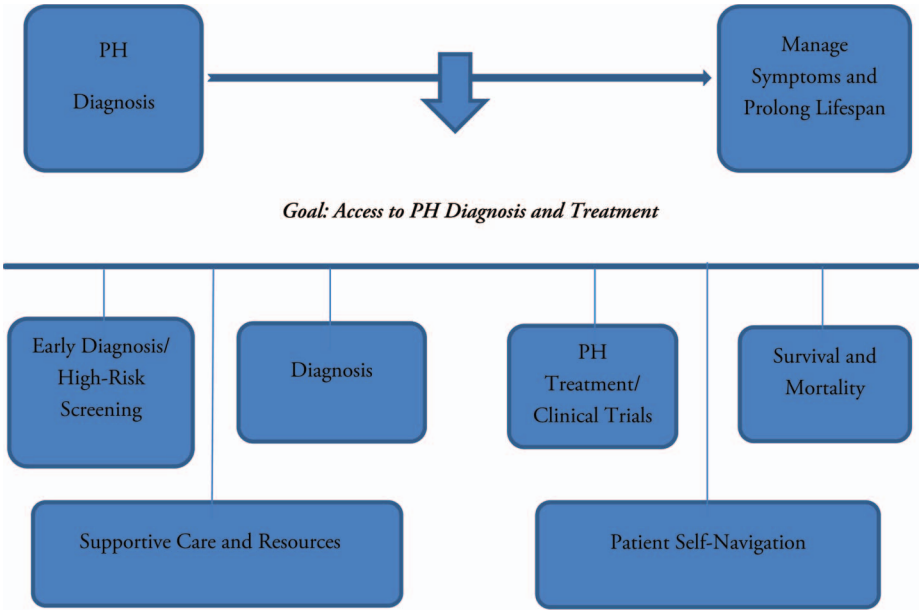

Figure 1: Christiana Care Health System PAH Patient Navigation Model.

HIV, portal hypertension, recent acute pulmonary embolus, congenital heart disease with shunt, sickle cell disease, and prior use of appetite suppressants warrant periodic screening for PAH.[7] Medical professionals are likely to see patients with these conditions that have symptoms of unexplained dyspnea, syncope, and lower-extremity edema that could be PAH, so these patients would benefit from definitive testing by a PAH specialist to rule out the disease.[5,8] Outreach efforts by the PH center in the form of written materials and education to health care providers regarding the symptoms of PAH can assist with early diagnosis of PAH.

DIAGNOSIS AND TREATMENT OF PAH

Evidence-based guidelines have been established to diagnose PAH and include obtaining a thorough medical history and symptomatology. Diagnostic testing such as an echocardiogram, electrocardiogram (ECG), chest x-ray (CXR), computed tomography (CT) scan of the chest, ventilation perfusion lung scan, pulmonary function tests including a 6-minute walk test, a sleep study and lab work, and also a right-heart catheterization (the definitive assessment for making the diagnosis of PAH) will be required.[9] Most, if not all, of these tests will be ordered to rule out other causes of the patient's symptoms and to distinguish them from other types of PH that is not PAH.[10] The PH team must explain to the patient what PAH is, as well as the specifics of what each test means and identify their importance in making the diagnosis of PAH.

Due to the complexity of diagnosing and treating PAH, patients are better managed in a PH center with PAH specialists or in a practice that collaborates closely with a PH center so that patients have the advantage of accessing all modes of PAH treatment, research, and other resources for support.[1,5] Patients that are not involved with a PH center have been shown to be misdiagnosed or not treated properly with PAH medications, and follow-up is often not consistent with established PAH standards of care.[11] Patients with a PAH diagnosis are better served in specialized centers that can offer the full range of treatment as the disease progresses, and have the opportunity to be involved in industry-sponsored clinical trials. The PH team must communicate test findings and treatment plans to all practitioners involved with the patient so that care is a seamless, timely, collaborative effort.

ROLE OF THE PH NURSE NAVIGATOR

While there may be patient navigators in other chronic care programs that are not nurses, a nurse in the role of the PH program navigator is most suitable. The PH nurse navigator is a pivotal team member that can serve as the triage person for the PH team and coordinate all patient care activities once the PAH diagnosis is made. This nurse should obtain a comprehensive patient assessment at the initiation of treatment, and these activities remain at the forefront of patient navigation.[2] Key information regarding patient support systems can be gathered at this time so that any barriers to PAH treatment can be identified and addressed from the start. The PH nurse navigator gathers information regarding drug enrollments for PAH medications (which are all very expensive) and obtains necessary prior authorizations. An essential skill for this nurse is experience in drafting appeal letters to insurance companies with the physician that include clinical studies in support of the drug's medical necessity, as some insurance companies will deny increased doses of certain medications or some treatments altogether. The PH nurse navigator also provides education about PAH medication along with providers, pharmacists, and specialty pharmacies. Patients should be given information on copay assistance programs to help them obtain the prescribed drugs and any other resources that they may need. The nurse navigator position is crucial to the team as a whole because this is where the responsibility for patient navigation lies.[9] The nurse navigator assists with patient care throughout the process, including all inpatient and outpatient services, and is the liaison between care settings so that vital information regarding the PAH treatment plan is not lost as the patient transitions from one setting to another. The relationship of the PH team with inpatient providers and nursing staff requires regular communications to ensure that all are addressing common treatment goals for the patient. The PH nurse navigator role confirms this care association via face-to-face meetings, telephone calls, or exchange of office notes and discharge summaries from inpatient stays. The PH team must create a notification mechanism when a patient is admitted to the hospital, especially those patients on parenteral

PAH therapies to ensure up-to-date dosing is ordered.

Besides the array of medication currently available to treat PAH, patients should still be considered for lung transplant evaluation based on prognosis and response to prostanoid therapy.[12] An established relationship with area facilities should exist if the PH center does not have lung transplantation available at its facility. Also, patients with chronic thromboembolic pulmonary hypertension (CTEPH) should be referred to facilities that can evaluate for a pulmonary thromboendarterectomy, as this surgery has been shown to improve hemodynamics and survival in these patients.[13]

SUPPORT SERVICES AND END-OF-LIFE CARE

The care of PAH patients involves much more than prescribing medications and treatments, and most patients require access to support such as financial, social, and logistical services. Resource information should be readily available for patients so treatment obstacles can be overcome if possible. Barriers to PAH treatment including lack of insurance, patient education level, logistics of transportation, inability to work, gaps in language skills, and other cultural issues can make it difficult for the patient to navigate health care systems and receive adequate care.[14] Many PAH patients need guidance on how to apply for disability; others require assistance in applying for medical transportation to come to appointments; and some have social concerns that must be addressed. Issues related to a patient's inability to comprehend a complex drug regimen, lack of housing, or poor family support can greatly affect the PAH treatment plan, and the PH team must be prepared to assist patients in dealing with these matters. Having a social worker or financial counselor as part of the PH team offers valuable assistance for patients in these areas. In addition, anxiety and depression are common comorbidities in PAH patients associated with the debilitating symptoms and drastic lifestyle changes that arise as the disease progresses.[15] Consequently, PH centers should have the ability to provide mental health services or a mechanism to refer patients to outside resources. Every PH center should provide access to a PH support group or deliver information regarding area support groups associated with the Pulmonary Hypertension Association (PHA), including the PHA telephone support group if the patient cannot attend in person. Patients and caregivers alike should be encouraged to join a support group for education, encouragement, and communication with other patients—all of which may help patients gain skills to cope with PAH. PHA offers a considerable amount of resources in all of these areas for patients, caregivers, and providers.

Since PAH is an incurable disease that is progressive and often fatal, the PH team must be prepared to assist patients and caregivers with living with a chronic illness, in addition to facilitating end-of-life discussions and decisions. The PAH specialist should introduce the recommendation for palliative care as an adjunct to symptom control and enhancement of the patient's quality of life when appropriate.[16] If all treatment efforts have been exhausted, hospice services should be offered, and the PH team must support the patient and family through this experience. The nurse navigator role is particularly important to assist patients in identifying palliative care or hospice programs that will fit their needs. PAH patients that elect hospice usually have to terminate prostacyclin or other therapies due to the cost of medication, unless there is another terminal diagnosis other than PAH. PAH medications and treatments are considered aggressive therapies and are in contrast with hospice concepts and criteria, so they are not covered by insurance.[17]

PATIENT SELF-NAVIGATION

Most importantly, a climate supporting patient self-navigation is essential within the PH model to ensure patients are empowered to be involved in their own care and treatment decisions as much as possible.[18] Through the development of the patient-care team relationship, the patient's attitude and understanding of PAH should be determined, along with adherence to treatment directions and testing, and awareness of when to reach out to the PH practitioner with questions or concerns. The PH nurse navigator can triage patients' needs and work with them on an individual basis to offer suggestions for tools to help them stay organized with treatment and progress, such as weight charts, fluid management guides, and symptom diaries. Patients should be encouraged to participate in treatment decisions, and self-efficacy must be fostered as treatment progresses.

CCHS PH PROGRAM

During the process of establishing a PH patient navigation program, the PH team at CCHS has experienced many changes in the way PAH patients are cared for. The CCHS program sees approximately 90 adult PAH patients, and the hospital has a separate heart failure clinic. All diagnostic testing is provided at CCHS, as well as all forms of PAH treatment: oral, inhaled, subcutaneous (SQ), and intravenous (IV) therapies. There is a dedicated outpatient clinic within the hospital, and patients are cared for and treated by the same team members whether they are inpatient or outpatient. PAH patients are referred to outside facilities for evaluation for lung transplant or pulmonary thromboendarterectomy as applicable.

The CCHS PH program was established in the late 1990s with Dr Gerald O'Brien, a physician who was board certified in pulmonary disease and critical care and had been a PAH specialist for 20 years. There was also a nurse clinical specialist, a clinical pharmacy specialist, and a few staff nurses on the pulmonary step-down unit. Today there is a full team with the addition of a nurse practitioner, nurse navigator, and research nurse. The present medical director is Dr Jeffrey Stewart, who is board certified in pulmonary disease, critical care, and internal medicine. Social work services are available on an as-needed basis from the pulmonary step-down unit. The PH team provides after-hours coverage for patients, and there is a pager notification system in place when one of the IV/SQ/inhaled therapy patients is seen in the emergency department (ED). All patients on IV/SQ or inhaled therapy

are admitted to one unit where the nurses have ongoing, specialized training. The intensive care unit nurses are also trained, and there is a PAH patient algorithm for the ED nurses to follow. The PH team meets every month to discuss all aspects of the program and is continuously developing new systems to enhance patient care. The team has also worked diligently in educating staff at the hospital and within the community. A quarterly PH support group meeting is held at the hospital, and patients are offered the opportunity for inclusion in clinical trials through the PH program. A patient navigation assessment tool is used to help identify areas of patient concern and barriers to PAH treatment and care. There is access to a dietitian and mental health professionals within the CCHS hospital system, and reporting tools are being developed to assess program quality. The presence of a central PH nurse navigator has been an important feature in decreasing fragmentation of care, and patient satisfaction has been enhanced in the CCHS program by anecdotal information from patients and caregivers.

Instituting a patient navigation system can support those responsible for taking care of PAH patients by providing a springboard from which policies, procedures, and other program attributes can be formulated. PH nurse navigators that have clearly outlined responsibilities and duties can enrich a PH program and streamline care. Patient safety is paramount to every PH program, and there must be systems in place to protect patients from unintended harm that can be caused by health care personnel not trained to care for and treat PAH patients. These patients require the regular support of the PH team to guide them through all aspects of care and treatment, and the PH team needs the type of structure a patient navigation model provides. There is evidence that when patient navigation services are received, patients are more satisfied with care and more likely to adhere to their treatment plan.[2,19] Therefore, both the PAH patient and the PAH team benefit from the presence of a PH patient navigation model, and the partnership between the two can create a solid foundation for ideal treatment outcomes.

References

1. Gin-Sing W. Pulmonary arterial hypertension: a multidisciplinary approach to care. *Nurs Stand.* 2010;24(38):40-47.
2. Swanson J, Strusowski P, Mack N, et al. Growing a navigation program: using the NCCCP navigation assessment tool. *Oncology Issues.* 2012; 27(4):36-45.
3. Harold P. Freeman Patient Navigation Institute. http://www.hpfreemanpni.org/. Accessed April 22, 2013.
4. Rosenthal ET. Patient Navigation in Action if Not in Name. *Oncology Times.* 2012;34(9):31-32.
5. Pulmonary Hypertension Association. Recommendations for Improving Patient Outcomes. http://www.phaonlineuniv.org/files/pha_white_paper_final.pdf. Accessed May 22, 2013.
6. Leary PJ, Edelman JD. Screening for pulmonary arterial hypertension. *Adv Pulm Hypertens.* 2012;11(2):78-83.
7. McLaughlin VV, Archer SL, Badesch DB, et al; ACCF/AHA. ACCF/AHA 2009 expert consensus document on pulmonary hypertension: a report of the American College of Cardiology Foundation Task Force on Expert Consensus Documents and the American Heart Association: developed in collaboration with the American College of Chest Physicians, American Thoracic Society, Inc., and the Pulmonary Hypertension Association. *Circulation.* 2009;119(16):2250-2294.
8. Tartavoulle, T. Evaluation and management of the adult patient with pulmonary hypertension. *J Nurse Pract.* 2011;7(5):409-416.
9. Mughal MM, Mandell B, James K, Stelmach K, Minai OA. Implementing a shared-care approach to improve the management of patients with pulmonary arterial hypertension. *Cleve Clin J Med.* 2003;70(Suppl 1):S28-S33.
10. Preston IR. Properly diagnosing pulmonary arterial hypertension. *Am J Cardiol.* 2013;111(8 Suppl):2C-9C.
11. Deaño RC, Glassner-Kolmin C, Rubenfire M, et al. Referral of patients with pulmonary hypertension diagnoses to tertiary pulmonary hypertension centers: the multicenter RePHerral study. *JAMA Intern Med.* 2013;173(10):887-893.
12. Long J, Russo MJ, Muller C, Vigneswaran WT. Surgical treatment of pulmonary hypertension: Lung transplantation. *Pulm Circ.* 2011;1(3): 327-333.
13. Rahnavardi M, Yan TD, Cao C, Vallely MP, Bannon PG, Wilson MK. Pulmonary thromboendarterectomy for chronic thromboembolic pulmonary hypertension : a systematic review. *Ann Thorac Cardiovasc Surg.* 2011;17(5):435-445.
14. D'Anna LH. Considering the impacts of health disparities, inequalities, and inequities on early diagnosis of pulmonary arterial hypertension. *Adv Pulm Hypertens.* 2012;11(2):73-77.
15. McCollister DH, Weintraub P, Badesch DB. Depression: beyond description as an important comorbidity in PAH—the next steps. *Adv Pulm Hypertens.* 2012;10(4):227-232.
16. Sg2. Clinical Intelligence: Cancer Care Coordination With Nurse Navigators. https://www.sg2.com/wp-content/uploads/2014/05/Cancer-Care-Coordination-with-Nurse-Navigators.pdf. Accessed January 22, 2016.
17. Grinnan DC, Swetz KM, Pinson J, Fairman P, Lyckholm LJ, Smith T. The end-of-life experience for a cohort of patients with pulmonary arterial hypertension. *J Palliat Med.* 2012;15(10): 1065-1070.
18. Kennedy A, Rogers A, Bower P. Support for self care for patients with chronic disease. *BMJ.* 2007;335(7627):968-970.
19. Esperat MC, Flores D, McMurry L, et al. Transformacion Para Salud: a patient navigation model for chronic disease self-management. *Online J Issues Nurs.* 2012;17(2):2.