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REVIEW

Collaborative Care: A Defining Characteristic for a Pulmonary Hypertension Center

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ABSTRACT

Introduction: Pulmonary hypertension (PH) is a complex, life-threatening disease. Development of pulmonary hypertension centers is recommended by pulmonary hypertension best practice guidelines and patient organizations to successfully care for patients. However, very little is published on the characteristics of these centers or the daily management required to care for this complex patient population.
Methods: This article, written by nurses who have extensive experience in managing patients with pulmonary hypertension, details the collaboration and workflows needed at a pulmonary hypertension center. Cohesive management among PH team members, patients, and caregivers is required for the long-term and successful care of patients.
Results: PH centers should have individual team members with clearly defined functional roles, policies and procedures that outline how to address the challenges of managing patients, effective communication between outpatient and inpatient teams, and well-educated staff to accurately diagnose and manage treatments, including accessing pulmonary arterial hypertension medications. Education of patients, concurrent with the support of caregivers and healthcare providers, plays a vital role in improving patients' understanding of their disease state and treatment options that best suit their lifestyle. PH team members assist patients in developing self-care skills to better recognize and respond to worsening symptoms and

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manage medication side effects to improve patient outcomes.
Conclusions: Defining collaborative care, work flows, and characteristics of a pulmonary hypertension center is essential to optimize care in this complex, chronic patient population.
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Keywords: Multidisciplinary care; Pulmonary arterial hypertension; Pulmonary hypertension care center; Pulmonary hypertension

INTRODUCTION

It is well documented in the heart failure literature that patient outcomes improve when multidisciplinary disease specific guidelines and self-care skills are implemented [1–3]. Disease management programs are designed to provide specialized care and follow-up while adhering to and implementing guideline driven practices. Research in the field of pulmonary hypertension (PH) has primarily focused on drug development with the goal of improving mortality and functional capacity. In addition to medication development, developing patient specific behaviors and management strategies may improve quality of life and prevent hospitalizations. Implementing individualized diagnostic and treatment plans for high risk patients with PH is a collaborative effort among healthcare professionals at PH centers.

Evaluation and treatment of patients with PH at a PH center is recommended by the European Society of Cardiology (ESC)/European Respiratory Society (ERS) guidelines [4] and endorsed by the Pulmonary Hypertension Association (http://www.phassociation.org). Establishment of expert referral centers is clinically and economically desirable and supported by patient organizations [4]. PH centers receive new patient referrals, evaluate causes of PH, manage treatments of patients with pulmonary arterial hypertension (PAH) and chronic thromboembolic pulmonary hypertension (CTEPH), and work with a multidisciplinary team to obtain the best outcomes for patients while focusing on research and patient education [4]. Collaborative PH multidisciplinary centers offer physicians who have expertise and experience with PH; nursing staff and support staff trained to assist in the diagnosis, education and management of complex treatment regimens of patients with PH; facilities with diagnostic capability and access to specialists; and clinical trials [5, 6]. PH centers have access to specialists including cardiologists, pulmonologists, rheumatologists, infectious disease specialists, hematologists, hepatologists, palliative care and transplant teams, geneticists and psychologists [5].

While characteristics may differ between PH disease management programs, common models and strategies exist in the multidisciplinary care of patients with PH. Criteria for regional and comprehensive PH centers have been well-defined [6, 7]. This review will describe the components needed for a collaborative model for the comprehensive care of primarily adult patients with PH. Team members and roles, policies and procedures, workflows, staff and patient education will be discussed. This article is based on previously conducted studies, and does not involve any new studies of human or animal subjects performed by any of the authors.

FUNCTIONAL ROLES AT A PH CENTER

Comprehensive PH care is provided by experienced healthcare professionals (Fig. 1). In the United States, a typical PH center model could consist of the following members:

PH Program Physician Specialist

The primary responsibility of the PH program medical director is to direct a multidisciplinary team to diagnose and provide comprehensive care to patients with PH. The director provides leadership, education, and direction to the team, community, and patients, and provides oversight for the PH team's growth and development. The medical director must be experienced in the care of patients with PH and board

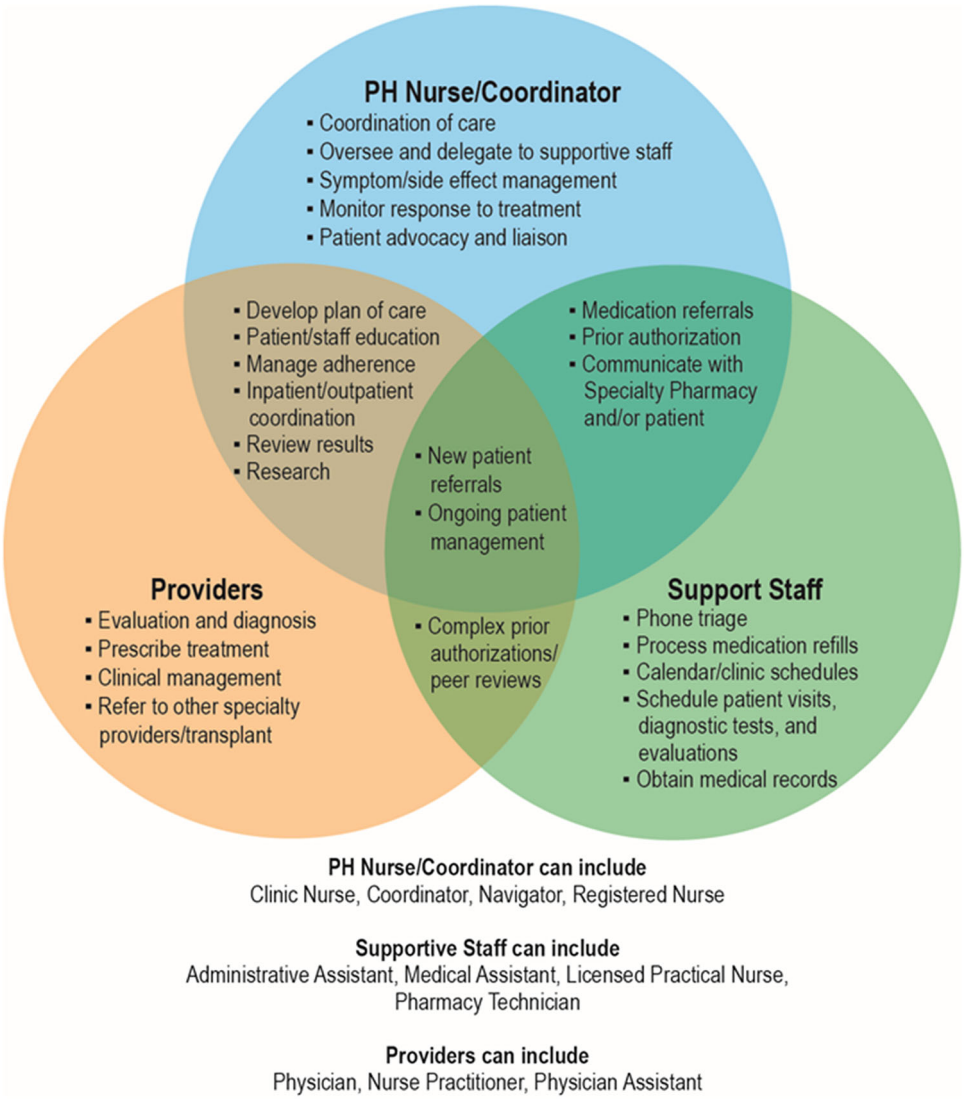


Fig. 1 The pulmonary hypertension team

certified in cardiology, pulmonology, and/or critical care medicine.

Medical Staff

The PH center may have additional licensed medical providers consisting of cardiologists, pulmonologists, hospitalists, fellows, critical care medicine physicians, nurse practitioners, and/or physician assistants that diagnose, treat, and manage PH. In the United States, these responsibilities and roles may be regulated by state or institutional licensure.

PH Program Coordinator

The PH program coordinator works closely with the medical staff in the management of the program's growth and development. The coordinator creates patient education materials, policy and procedures, and works with the team to create goals for optimal patient care. Nurses, nurse practitioners, physician assistants, respiratory therapists, or pharmacists that are experienced in the evaluation, treatment and management of patients with PH can serve as the program coordinators. The program

coordinator works closely with inpatient staff nurses and clinical educators to assure experienced and well-educated nursing staff care for patients with PH admitted to the hospital. The coordinator ensures that policy and procedures are in place and oversees communication processes to ensure patient safety.

PH Program Nurses

The PH program nurse(s) may or may not be the PH program coordinator. Nursing is responsible for the day to day management of patients with PH and coordination of their care (Table 1). The PH program nurse works closely with the medical staff, inpatient nursing, pharmacists, support staff and others to educate, coordinate care, and facilitate the treatment and management of patients with PH. The nurses are the liaisons for specialty pharmacy (SP) nurses and pharmacists.

Pharmacist

The PH pharmacist is well-educated on all PAH medication therapies and works closely with the PH team to provide current PAH medication therapy assistance. Pharmacists assist with monitoring for drug interactions, adverse effects and adherence. PH program pharmacists assist with monitoring long-term prostacyclin dose adjustments, monitoring the therapeutic effects, offering suggestions to adjust therapy, and providing clear instructions to the patient in collaboration with the healthcare team [8]. Specialty pharmacies that manage PAH therapies have been shown to improve adherence in patients with PAH and in so doing, improve disease state outcomes [8, 9].

Social Worker

The social worker assists the patient and family with psychosocial services and advocates for the patient. Social workers address issues including poverty, abuse, addiction, maladaptation, divorce, loss, unemployment, educational problems, disability, and mental illness. Social workers provide additional resources to patients and families to cope with the stresses of living with a chronic disease.

Research Staff

The PH program research staff is experienced in best practices and safety of patients participating in pharmaceutical and investigator sponsored clinical research. The principle investigator, research nurses, and/or research assistants collaborate to identify patients who can participate in clinical trials to further advance PH care, treatment, and disease management.

Support Staff

PH programs may have ancillary support staff as part of the PH team, such as medical assistants, pharmacy technicians, and clerical staff. The support staff assists the team by communicating with patients and team members, initiating medication or diagnostic test prior authorizations, processing refills, obtaining outside medical records and scheduling new patient evaluations and follow-up evaluations. Items to include in a new patient letter are found in Table 2. Dietitians and rehabilitation specialists provide additional patient education in their areas of expertise.

IMPORTANCE OF PH PROGRAM POLICIES

Once staffing of the PH program is established, providers must work together with the institution to create written documents that standardize the care of patients in the PH program. Written policies are essential to ensure adherence to published diagnostic evaluation and treatment guidelines, and to optimize patient safety. Polices must be easily accessible for staff and updated regularly. Consistent adherence to program policies also facilitates research activities both within the program and for multicenter industry or collaborative projects. The types of policies needed and questions a PH team should ask when developing a policy are found in Table 3.

PH STAFF EDUCATION

Staff education is essential to manage a chronic illness which requires complex medical

Table 1 Role of the nurse in daily patient management of patients with pulmonary hypertension

Coordination of care

- Direct and supervise care delivered by other pulmonary hypertension (PH) team support staff (medical and administrative assistants)
- Collaborate with various healthcare providers and specialists to ensure timely referral
- Interpret patient data, make decisions and take appropriate actions
- Facilitate effective inpatient and outpatient transitions
- Provide health promotion, disease prevention, health maintenance
- Medication reconciliation
- Ensure evaluation and follow-up testing is scheduled and reviewed
- Communicate/collaborate with PH care providers and others involved in PH care (specialty pharmacy, local providers, family)

Assessment and intervention

- Perform physical exams, nursing assessments, and health histories
- Telemanagement and advice
- Assess for heart failure and intervene appropriately
- Active listening, social perceptiveness and decision making
- Evaluate response to medications and manage side effects
- Assess for gaps in treatment and non-adherence
- Critically think, problem solve and anticipate patient needs

Education and research

- Provide patient and staff education on disease state, diagnostic evaluation, treatment options, medication action, benefit and anticipated side effects and general patient management
- Provide patient self-care skills, disease management (i.e., heart failure symptoms, fluid and sodium intake)
- Conduct research in support of improved practice and patient outcomes

Advocacy

- Help patients adapt to illness, develop coping skills, conserve energy
- Develop goals of care
- Serve as patient advocate and liaison
- Identify psychosocial, financial issues and refer to appropriate resources

PH pulmonary hypertension

regimens. Inpatient nurses and physicians in training (residents and fellows) are educated on the management of PH and right ventricular failure. Educational in-services on emergency preparedness related to right heart failure and infused prostacyclins is discussed with specialty units and procedural areas caring for patients with PH. These areas include the emergency department, interventional radiology, general surgery, cardiac catheterization lab and

Table 2 Items to include in a new patient letter

Sending patients a letter prior to their initial appointment can very beneficial to the success of the initial pulmonary hypertension (PH) clinical meeting. Some helpful tips to include in the letter are:

- The appointment date/time/location. Arrive early to complete administrative paperwork
- Contact medical insurance company to investigate coverage for this visit. Request appropriate referral or pre-authorization, if required
- Bring all insurance cards, including prescription drug plan card
- Clinic may be some distance from parking. Bring walking aides, or wheelchair if needed
- Recommend having someone (close friend or family member) accompany patient to the appointment to take notes, ask questions and participate in the visit
- Request primary care provider or specialist to mail or fax copies of medical records (to include: progress notes and results of diagnostic test results such as; echocardiogram, right heart catheterization, lab tests, chest X-rays, chest computed tomography (CT) scans, and ventilation/perfusion (V/Q) scan, etc., to the PH center *prior* to the appointment for adequate record review) Note that the appointment may be rescheduled if records are not received prior to or at the time of the appointment. Include the PH center's phone and fax number in the letter
- Bring echocardiogram, chest x-ray, chest CT scan, and V/Q scan images on disc and hand carry to the appointment
- Bring all medications in their bottles and a medication list with medication strength and dose taken daily. Include initial start date or the year for medications if possible
- Bring a written list of the names, addresses, phone, and fax numbers of all physicians active in the patient's care
- Bring a written list of questions for the PH team
- May be asked to perform a 6 min walk test (6MWT) if physically able. Should wear, or bring comfortable shoes and clothes to complete this test
- Allow sufficient time for the appointment. The provider may decide to obtain testing if possible at the initial visit such as echocardiogram, labs or X-rays
- Allow sufficient travel time between scheduled tests

Additional information to consider:

- Map or visitors guide of the hospital/clinic
- Parking information, valet or self-parking costs and location
- Instructions specific to oxygen equipment for the evaluation
- Instructions on how to cancel or reschedule appointment and testing
- Contact number for questions prior to appointment

6MWT 6 min walk test, *CT* computed tomography, *PH* pulmonary hypertension, *V/Q* ventilation/perfusion

digestive disease procedure personnel. Staff nurses on inpatient step-down and intensive care units providing care for patients with PH are in-serviced regularly to ensure competency. Avoidance of bolus and gaps in prostacyclin treatments, transfer of infusions between venous access sites and outpatient infusion pump training are all important patient safety considerations that require initial and ongoing training [10, 11].

Hospital pharmacists need to know how to order and prepare PAH medications. In order to

Table 3 Types of policies needed and questions to ask when developing a policy

Types of policies needed

Essential:

- Standard pulmonary hypertension (PH) evaluation
- Right heart catheterization with vasodilator testing (VDT) including specifics about type of vasodilator, dosing, and types of patients that will require VDT
- Standard program treatment algorithm that includes type and timing of repeat testing and office visits
- Hospital policies for management of prostacyclins
- Training plan and schedule for hospital staff about the disease process, treatment and complex medical regimen
- Monitoring system for patients receiving medications that require routine lab monitoring
- Pharmacy order sets for pulmonary arterial hypertension (PAH) medications that include verification of dosing and type of diluent, if applicable

New patient procedures:

- Records that are required for a new patient to be scheduled
- Testing that is scheduled with or after the first patient visit (Fig. 2)
- Use of a customized new patient letter (Table 2) and information packet

Policies to guide management of specific clinical situations:

- New onset of atrial arrhythmias
- Bacteremia in central venous catheters
- Perioperative management of patients with PAH
- Contraception options for women of childbearing potential
- Anticoagulation

Questions to ask when developing a policy

What to think about when creating a hospital policy for administering PAH therapies.

- Who is responsible for insurance approval for the medication and the hospitalization?
- What type of central line will be used? What department will place it?
- What units will be trained to care for the parenteral therapy patient and how will you ensure admission is restricted to those units?
- Who will enter dosing and titration orders?
- Who will monitor appropriate device programming, patient symptoms and side effects?
- Who will teach the patient and family and who will assess safety and competence for discharge and during subsequent admissions?
- Who will be responsible for routine medication cassette changes?

Table 3 continued

- Will the patient remain on their home pumps or be converted to hospital pumps?
- How will patients on oral or inhaled prostacyclins be transitioned to other administration methods in the event they are unable to take their therapy (NPO, intubation)?
- What patient issues should trigger immediate notification of the PH team and who is notified?
- Are there pumps other than the patient's pumps available for emergency situations and where are they kept?
- Who will stock associated supplies (tubing, batteries, ice packs) the unit, central service or the pharmacy?
- Who should hospital staff call for assistance with troubleshooting pump issues?
- Who will arrange discharge medication and supply shipments?
- Where will back-up cassettes be kept on the unit?
- Will you standardize cassette change times while inpatient or allow patients to remain on their home schedules?
- How will you ensure that the patient has a back-up pump at the bedside at all times?
- How will the Emergency Department staff be trained to manage patients on prostacyclin therapies?
- What is the procedure for evaluating and managing dysfunctional central lines or suspected central line infections?

NPO Nil per os, Latin for "nothing by mouth", *PAH* pulmonary arterial hypertension, *PH* pulmonary hypertension, *VDT* vasodilator testing

prevent medication errors upon admission, concentration and infusion rates must accurately reflect the patient's outpatient dose. A consistent patient dosing weight is used for prostacyclins and is documented in the electronic medical record (EMR) with the dose, concentration, pump rate and device used for the infusion.

CARE OF THE PH PATIENT

Diagnosis of PH

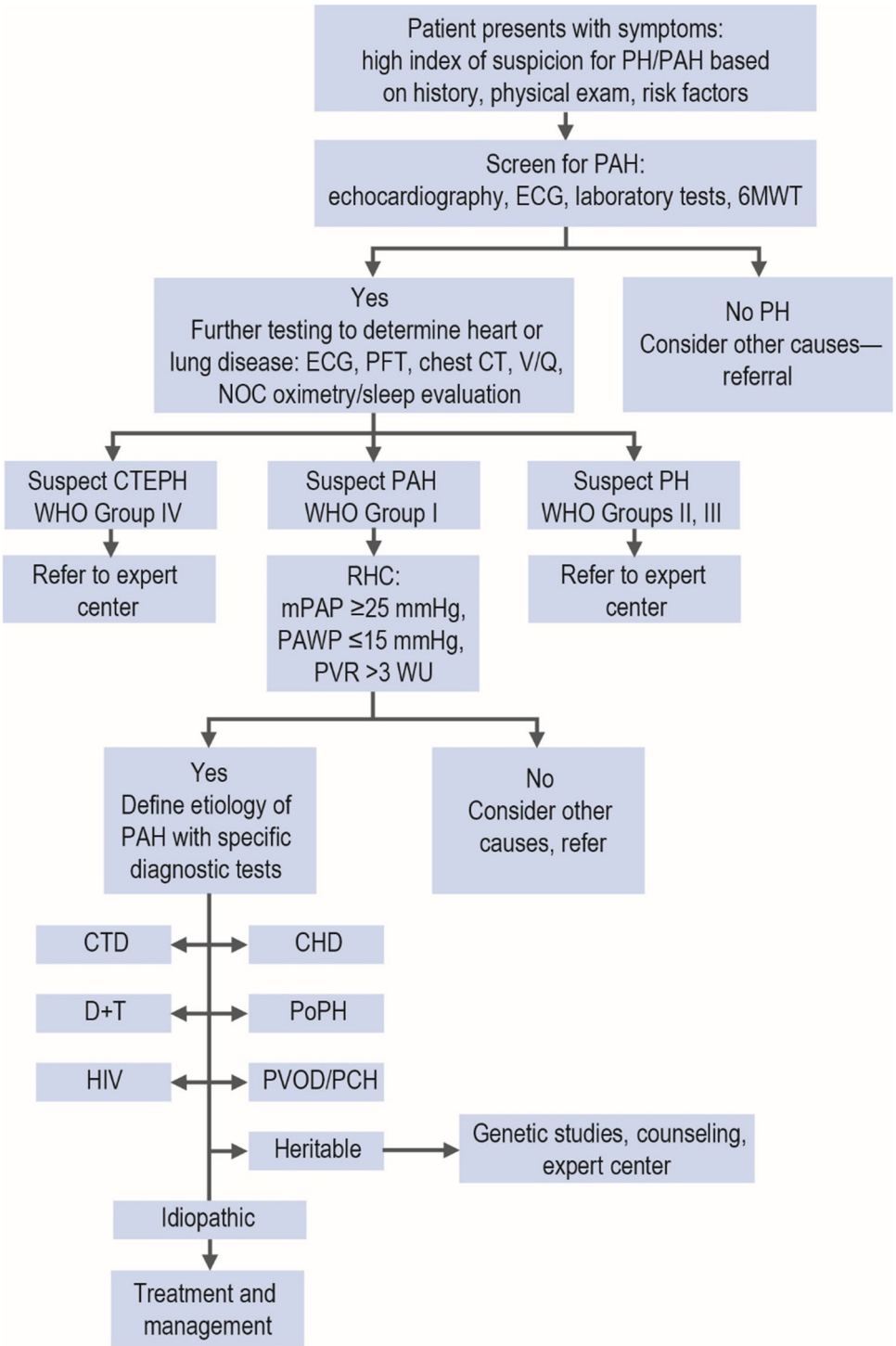
PH is defined by a mean pulmonary artery pressure (mPAP) ≥25 mm Hg at rest determined by right heart catheterization [4, 12] and is a debilitating, progressive disease. According to clinical presentation, hemodynamic characteristics, and pathological findings, the World Health Organization (WHO) has characterized PH into five groups (Table 4) [13]. The patient with WHO Group 1 PAH is characterized by an increase in mPAP ≥25 mm Hg, pulmonary vascular resistance (PVR) >3 Wood Units, and a pulmonary artery wedge pressure (PAWP) ≤15 mmHg [12, 13]. PAH is caused by cellular changes in the pulmonary vasculature associated with inflammation, oxidative stress, fibrosis, apoptosis, and metabolic remodeling [14]. These changes can lead to increased pulmonary vascular resistance, right ventricular failure and death [5, 15].

Patients referred to PH centers for diagnosis and treatment often are referred late (with functional class III or IV symptoms), receive misdiagnoses, and are inappropriately prescribed PAH specific medications [16]. Patients often report not being able to obtain PAH specific medications or had gaps in treatment prior to being referred to a PH center. A complete description of the diagnosis and treatment options for PH is beyond the scope of this paper, and the reader is referred to published guidelines [4, 12, 17] for a more complete description. Briefly, a comprehensive PH center must have the resources, personnel and protocols to perform a complete diagnosis and prescribe and manage complex medication regimens. In diagnosing patients with PAH, more common causes of PH must be excluded. Patients who present with symptoms and risk factors for the

Table 4 World Health Organization (WHO) pulmonary hypertension classification and functional assessment Adapted with permission from McLaughlin [19] and Simonneau [13, 19]

WHO pulmonary hypertension (PH) diagnostic groups	
Group 1	Pulmonary arterial hypertension (PAH)
Group 2	PH due to left heart disease including valvular disease
Group 3	PH due to lung disease and/or hypoxia
Group 4	Chronic thromboembolic pulmonary hypertension (CTEPH)
Group 5	PH with unclear or multifactorial mechanisms
WHO pulmonary hypertension functional classification	
Class I	Patients with PH in whom there is no limitation of usual physical activity; ordinary physical activity does not cause increased dyspnea, fatigue, chest pain, or presyncope
Class II	Patients with PH who have mild limitation of physical activity. There is no discomfort at rest, but normal physical activity causes increased dyspnea, fatigue, chest pain, or presyncope
Class III	Patients with PH who have a marked limitation of physical activity. There is no discomfort at rest, but less than ordinary activity causes increased dyspnea, fatigue, chest pain, or presyncope
Class IV	Patients with PH who are unable to perform any physical activity and who may have signs of right ventricular failure. Dyspnea and/or fatigue may be present at rest, and symptoms are increased by almost any physical activity

CTEPH Chronic thromboembolic pulmonary hypertension, *PAH* pulmonary arterial hypertension, *PH* pulmonary hypertension, *WHO* World Health Organization



development of PAH undergo diagnostic testing (Fig. 2) in order to classify the patients into the appropriate diagnostic category, which will drive patient referrals to specialty care and definitive treatments. The multidisciplinary PH team assures that the appropriate diagnostic tests are ordered, completed and reviewed to arrive at the correct diagnosis.

◀ **Fig. 2** Diagnostic algorithm of patients with pulmonary hypertension (adapted with permission from Hoeper 2013) [12]. *6MWT* 6 min walk test, *CHD* congenital heart disease, *CT* computed tomography, *CTD* connective tissue disease, *CTEPH* chronic thromboembolic pulmonary hypertension, *D + T* drugs and toxins, *ECG* electrocardiogram, *HIV* human immunodeficiency virus, *mPAP* mean pulmonary artery pressure, *NOC* nighttime, *PAH* pulmonary arterial hypertension, *PAWP* pulmonary artery wedge pressure, *PCH* pulmonary capillary hemangiomatosis, *PFT* pulmonary function tests, *PH* pulmonary hypertension, *PoPH* portopulmonary hypertension, *PVOD* pulmonary venoocclusive disease, *PVR* pulmonary vascular resistance, *RHC* right heart catheterization, *V/Q* ventilation/perfusion

Management of PAH

Medical therapies recommended for the treatment of PAH [4] target three distinct pathways (endothelin, nitric oxide and prostacyclin), [18] identified as pathologic. PAH specific therapies including endothelin receptor antagonists (ERA), phosphodiesterase type 5 (PDE-5) inhibitors, soluble guanylate cyclase (sGC) agonists, prostacyclin analogues, and prostacyclin receptor agonists are approved by the FDA for monotherapy or combination therapy for the treatment of PAH [4].

An algorithm for the treatment and management of patients with PAH is presented in Fig. 3. Once the diagnosis of PAH has been confirmed, a risk assessment is made to guide treatment choices depending on a low–high risk stratification tool [19] or the REVEAL risk calculator [20]. Evidence based guidelines [4, 17] can be consulted for guidance about initial and ongoing therapy. The PH team's expertise with PAH treatments, patient's lifestyle, beliefs and support systems, as well as insurance and financial resources must be considered. The treatment plan is created by a shared decision-making process involving the patient, family and caregivers, and members of the PH team. Select patients may qualify for clinical trials and be approached to consider research options. It can be difficult to reach a decision on the appropriate therapy; therefore, patients may benefit from contact with an experienced patient mentor or a home visit and assessment by the SP nurse. This is particularly important for patients who will receive complex, parenteral therapies.

Close clinic follow-up and reassessment is necessary for ongoing patient management [4]. At regular intervals, response to therapy is assessed and additional therapy is discussed if goals are not met. The PH team considers the appropriate timing for transplant referral and reviews the goals of care with patient and family.

PAH Medication Access

Once a treatment strategy has been chosen, access to PAH specific medication is initiated (Fig. 4). The following outlines typical PAH medical access processes in the United States. Inhaled and parenteral options are typically reimbursed under the medical benefit, since they require a delivery device while oral formulations are generally covered under the pharmacy benefit and authorized by contacting the Pharmacy Benefit Manager (PBM) [8]. Patients without prescription drug coverage may receive assistance from the pharmaceutical company's patient assistance program (PAP). These patients should also receive counseling on how to obtain prescription drug coverage in the future. There is a limited distribution network for ERAs, sGC agonists and prostacyclin type medications. Although PDE-5 inhibitors [oral sildenafil (Revatio®), generic sildenafil, oral tadalafil (Adcirca®)] may be obtained through local pharmacies, these medications may be requested through a SP due to their rarity, expense in stocking and dosing complexity. The SP provides valuable experience and counseling on adherence and side effect management.

Most PAH medications will require prior authorization. If denied, the PH team must decide whether or not they will appeal the denial. The reasons for prior authorization denial may include that the request does not meet the insurance plan's medical necessity criteria, the plan may have a step edit in place (patient must try and fail formulary alternatives first), the medication is not on the plan's formulary or the medication is a plan exclusion.

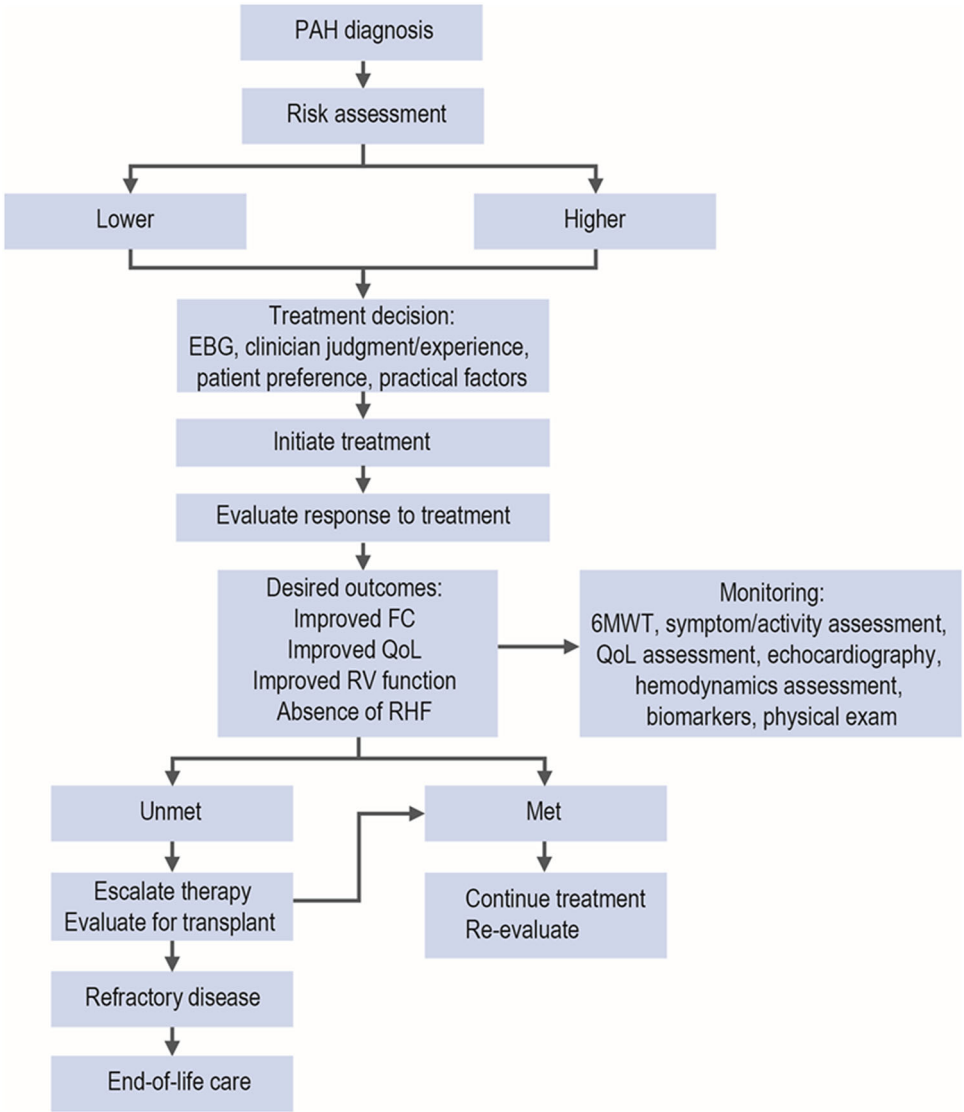


Fig. 3 Treatment and management of patients with pulmonary arterial hypertension. *6MWT* 6 min walk test, *EBG* evidence-based guidelines, *FC* functional class, *PAH* pulmonary arterial hypertension, *QoL* quality of life, *RHF* right heart failure, *RV* right ventricular

The rationale for medical necessity and counter response to the denial must be addressed in the appeal. If the denial is upheld, either a second level appeal or a peer-to-peer discussion with the provider and reviewer can be initiated. Some pharmaceutical companies may accept an application to PAP after a second denial, or the PH team may elect to discuss an alternate treatment with the patient.

Once the medication has been approved, the SP will contact the patient to provide medication counseling, process the claim and discuss the co-insurance or co-pay. The PH team must be familiar with the various co-pay assistance options available to commercial-insured as well as federally-insured patients and guide them appropriately.

PAH specific medications are assigned to higher tiers by the PBM resulting in larger co-pays. The prescriber may request a tier reduction to reduce the amount of the patient's co-pay and out-of-pocket expenses. As more PAH medications become available as generics, costs will decrease, but step edits may become

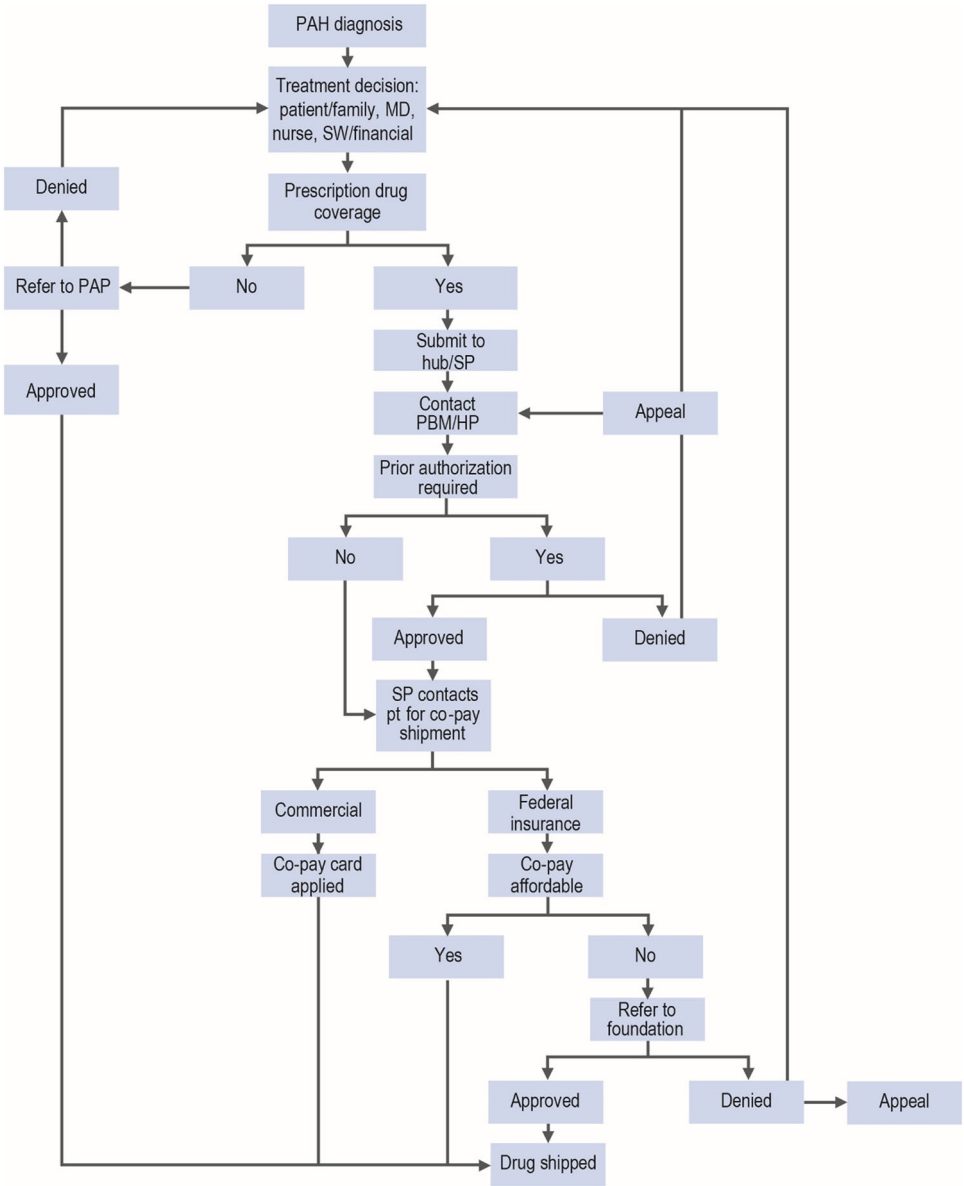


Fig. 4 Pulmonary arterial hypertension medication access. *HP* health plan, *MD* medical doctor, *PAP* patient assistance program, *PAH* pulmonary arterial hypertension, *PBM* pharmacy benefit manager, *pt* patient, *SP* specialty pharmacist, *SW* social worker

more common, requiring patients to try and fail generic alternatives before accessing more expensive non-generic medications.

The SP contacts the patient monthly to arrange their medication shipments. The PH team must order and monitor labs to assure that risk evaluation and mitigation strategy (REMS) requirements are met as well as educate patients about the risks, monitoring and precautions to take, and particularly, the appropriate contraception and pregnancy avoidance for women of child-bearing potential on applicable PAH medications. During the authorization and approval process, PH team members will collaborate with SP to ensure timely shipment and adequate medication supply for patients, requesting new authorizations when they expire or the patient's insurance changes.

Communication Between Inpatient and Outpatient Teams

Patients with PH require admission to the hospital for acute decompensation, or for the management of other co-morbidities. Elective admissions occur for initiation of certain high-risk medications where close monitoring and specialized care are necessary. Many centers have dedicated units trained to provide care for patients with PH. Collaboration between the inpatient and outpatient teams requires effective interpersonal communication during these admissions. Inpatient reports may be relayed daily to the outpatient team or weekly via group meetings. A liaison or navigator may serve as a constant team member and facilitate communication between the care teams.

When patients are admitted to the hospital, they may be cared for by medical professionals who may not be familiar with their disease state or outpatient plan, it is important for the PH team to oversee the medical management during admissions. Medicare and Medicaid require that the facility (hospital or skilled nursing facility) provides all medications to the patient while admitted. Using the patient's home supply of medication during admission can create problems when trying to obtain the next home shipment. Due to the complexity of PH and medications required, it is imperative to maintain ongoing communication between the inpatient and outpatient teams of physicians, nurses, pharmacists, and respiratory therapists during transitions of care. The PH team members involved will vary among centers, however, ensuring good communication will lead to the best outcomes for patients and continuity of care for patients in the outpatient setting.

Patient Education

Pulmonary hypertension team members collaborate to educate patients and develop patient self-care skills [21, 22]. Patient education begins at the initial contact and is reinforced at each encounter via the teach-back method. New patients are educated about diagnostic procedures and PH WHO group classification. Upon diagnosis of PAH, patients are educated on disease state, prognosis and disease progression. Therapeutics options and their risks, side effects and benefits are described to allow patients to make therapy decisions which meet their lifestyle needs and goals of care. Medication education starts with the PH providers and continues with the SP members. Training patients on PH infused and inhaled medications can be completed by PH coordinators, PH pharmacists, and/or SP nurses and requires demonstration and teach-back methods. Discussing the purpose and dosing of the medications, the goals of treatment and potential side effect management will encourage patient adherence.

Patients are taught strategies to recognize and manage or prevent heart failure symptoms. Home management strategies to monitor for fluid retention including daily weights, sodium and fluid intake monitoring, and evaluating food labels, can be taught in individual and group classes. Patients need to know when and how to contact the PH center for questions, worsening symptoms or side effects. Patient education should include medications to avoid or prevent worsening symptoms or which may interact with PH specific medications. Reinforcement of patient education should be ongoing and the goal of all PH team members.

Discussion of the importance of adherence to PH treatments to prevent worsening symptoms is essential. Ensuring patients are trained on aseptic technique, emergency preparedness, side effect management, and safe practices enables patients to adapt these therapies into their everyday routines with the goal of improved quality of life.

Patient Empowerment

Patient-centered care requires that patients have an integral role in their healthcare team (Fig. 5). Patients who take an active role in their healthcare may experience long-term benefits. Patients need to learn self-care management and to set goals for their healthcare. Discussing goals of care with their providers is an ongoing process for patients and families.

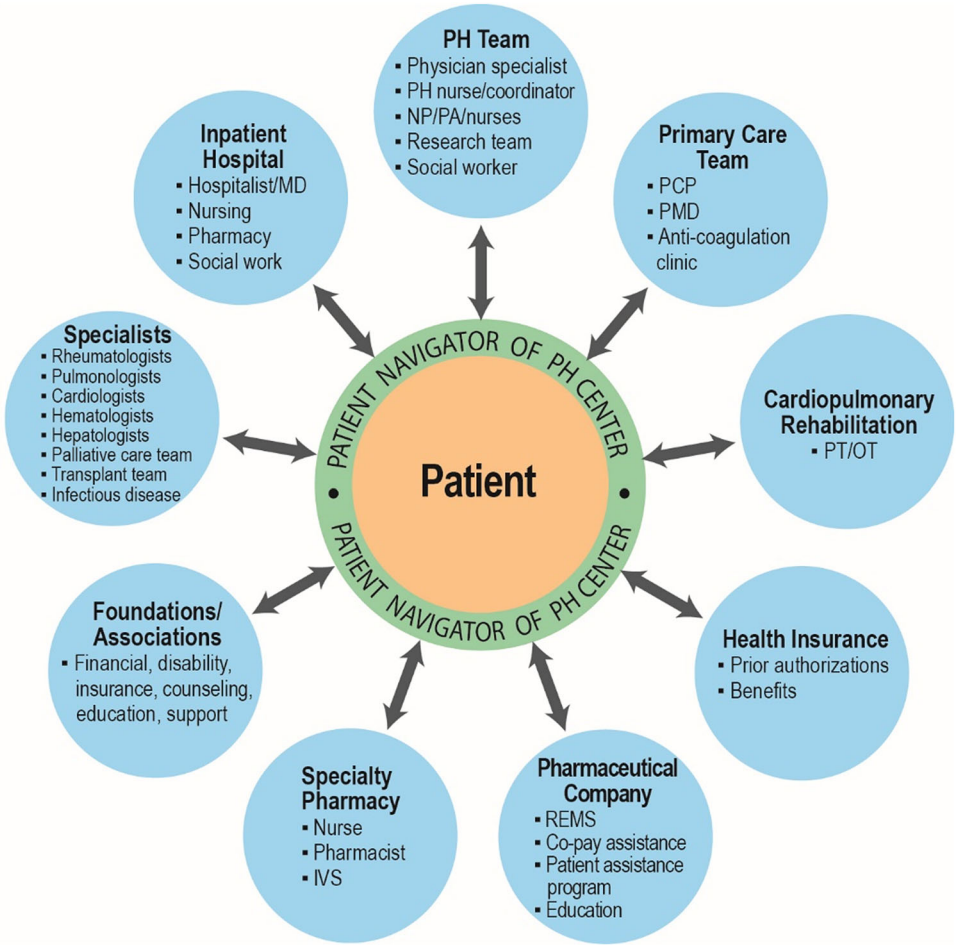


Fig. 5 Coordinating care of patients with pulmonary hypertension. The function of the navigator may be performed by one or more people at the pulmonary hypertension center. The navigator facilitates interaction with various groups and services to access comprehensive care for patients with pulmonary hypertension. *IVS* insurance verification specialist, *NP* nurse practitioner, *OT* occupational therapy, *PA* physician assistant, *PCP* primary care physician, *PH* pulmonary hypertension, *PMD* primary medical doctor, *PT* physical therapy, *REMS* risk evaluation and mitigation strategy

Self-care management includes monitoring patient health, identifying patient goals for quality of life and following recommended health maintenance guidelines. Monitoring health is an ongoing process. Encouraging patients to keep a health journal is a great way for them to be able to follow their response to therapy. A health journal might include daily weights, sodium intake, activity or exercise and general feelings of well-being for the day. During clinic visits, this information can be invaluable and can make the most of the face-to-face visit with the PH provider.

Each person has a different perspective on what is important and gives them good quality of life. Patients who identify what is most important in their lives can learn how to prioritize their activities, such as pacing themselves with activities of daily living, planning major events with frequent rest periods, and exercising at the time of day they feel the best. Quality of life questionnaires are frequently used by palliative care and the PH team to assess many different aspects of a patient's life. Responses can identify areas where resources are needed to help patients cope with life

Table 5 Resources for patients and healthcare professionals

For patients:

Pulmonary Hypertension Association (PHA):

- PHA classroom
- Biennial PHA International Conference
- Regional 'PHA on the road' conferences
- PHA Facebook groups, mentor programs, group chats
- Telephone support line
- Pulmonary Hypertension (PH)—a patient's survival guide
- Pulmonary arterial hypertension (PAH) medication sheets
- New diagnosis packet, living with PH; tips and brochures

Advocacy and awareness programs

Industry patient mentor programs

Patient education materials:

- Industry
  - Medication guides and brochures
  - Disease state pamphlets
- Specialty pharmacy
- Institutional PH center materials

For healthcare professionals:

- Pulmonary Hypertension Association (PHA)
- Biennial PHA International Conference and Scientific Sessions
- Medical education programs (preceptorship program, on demand)
- PHA Online University
- Advances in Pulmonary Hypertension (quarterly medical publication)
- PH Clinicians and Researchers membership network
- Pulmonary Hypertension Professional Network (PHPN):
  - List-serve
  - Symposium
  - Mentor program
- Continuing medical education courses and webinars
- Professional networking
- Organizations dedicated to PH/CTEPH research, advocacy and awareness

Table 5 continued

International, National and Regional Conferences (examples: American College of Chest Physicians (CHEST), American Thoracic Society (ATS), The International Society for Heart and Lung Transplantation (ISHLT), Neonatal and Childhood Pulmonary Vascular Disease conference
• Published PH consensus guidelines
• PH center preceptorships
• Grand rounds
• Sponsored dinner programs
• Medical and nursing journals/textbooks

CTEPH chronic thromboembolic pulmonary hypertension, *PAH* pulmonary arterial hypertension, *PH* pulmonary hypertension, *PHA* Pulmonary Hypertension Association, *PHPN* Pulmonary Hypertension Professional Network

challenges and where support from a social worker may be needed.

Health maintenance is important for patients with a chronic illness. The Centers for Disease Control and Prevention [23] recommends regular check-ups, influenza and pneumococcal immunizations, mammograms, and colonoscopies. These healthcare checks are recommended at certain intervals for disease screening and prevention and are essential as patients live longer on PAH therapy.

Resources for patients and healthcare professionals are found in Table 5. The Pulmonary Hypertension Association (http://www.phassociation.org) provides many resources for patients, family members and healthcare providers. Services include information on diagnosis, treatments, support groups and mentoring programs.

DISCUSSION

Despite advances in care and treatment, PH remains a complex, challenging and life-threatening disease. Guidelines and patient organizations have recommended the development of PH centers to successfully care for patients with PH. A well-educated, collaborative multidisciplinary team with clearly defined functional roles offers the expertise and experience to care for patients with PH.

Policies and procedures outline how to address the challenges of managing patients with PH in the PH center. Education of staff is essential to accurately diagnose, manage treatment and improve patient outcomes.

Early diagnosis and appropriate management of patients is essential for long-term survival. The majority of patients followed at PH centers are often diagnosed with PAH and CTEPH. Obtaining difficult-to-access treatments for PAH can have its own challenges. Coordinated care with effective communication within the PH team and between the team and other providers will allow for smooth transitions of care.

With the support of caregivers and healthcare providers, the patient plays a vital role in their own care. Education of patients will aid in improving patient understanding of their disease, selecting therapeutic options that best suit their lifestyle and goals of care, practicing self-care skills for early recognition and management of worsening symptoms and effectively managing PAH medication side effects.

Limitations of this article include a lack of published data on the effects of multidisciplinary care and self-care management on PH patient outcomes. There is also a lack of research on non-medical management of this complex, progressive, chronic illness. Ratios of the optimal number patients to PH team members have not been established but would provide support for requests to hospital administration to build appropriately manned PH centers. Each institution will have a finite amount of resources to devote for team members at a PH center; therefore, more data is

needed to define the complexity of treatment for these patients and their care.

CONCLUSIONS

Long-term care of patients with PH requires collaborative management between specialty care centers, patients, and caregivers. Applying best practices leads to successful PH patient care. This article provides a model of collaborative, multidisciplinary care to develop a comprehensive primarily adult PH care center.

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