The Role of the Advanced Practice Nurse With Patients Undergoing Pulmonary Thromboendarterectomy and Balloon Pulmonary Angioplasty for CTEPH

Wendy Nelson, RN, MSN, CNS, ACNP Nurse Practitioner III UCLA Interventional Radiology Los Angeles, CA

Mary Miller, RN, MSN, ACNP-BC Nurse Practitioner III UCLA Cardiothoracic Surgery Los Angeles, CA

Shannon Salveson, RN, MSN, ACNP-BC Nurse Practitioner III UCLA Pulmonary Vascular Disease Program Los Angeles, CA Advanced practice nurses play an integral role in the management of chronic thromboembolic pulmonary hypertension patients undergoing pulmonary thromboendarterectomy and balloon pulmonary angioplasty. As integral members of the multidisciplinary team, advance practice nurses assist chronic thromboembolic pulmonary hypertension patients in the presurgical, postsurgical, and procedural settings by ensuring appropriate referral, workup, evaluation, and education.

INTRODUCTION

Chronic thromboembolic pulmonary hypertension (CTEPH) is a progressive pulmonary vascular disorder characterized by the deposition of chronic thrombotic material in the pulmonary arterial vasculature leading to obstruction of flow and ultimately remodeling of the vasculature and subsequent right heart failure if left untreated. Estimates vary, however the overall incidence of CTEPH following acute pulmonary embolism (PE) is approximately 3% (1% to 5%).¹⁻³ Notably, a substantial number of patients report no prior history of PE.¹ The management and treatment of CTEPH differs from other forms of pulmonary hypertension (PH) in that it is potentially reversible with pulmonary thromboendarterectomy (PTE) surgery.⁴ In addition, for patients who are not surgical candidates or those with persistent PH following PTE, medical

therapies and interventional therapies, specifically balloon pulmonary angioplasty (BPA), may play a crucial role in achieving improvement.

At our center, patients with PH and CTEPH are initially seen and managed by the Pulmonary Vascular Disease service, comprised of critical care pulmonologists and advanced practice nurses (APNs). Once diagnosed with CTEPH, patients are evaluated to determine candidacy for PTE or BPA by the CTEPH multidisciplinary team. This multidisciplinary team is comprised of additional disciplines including cardiothoracic surgery, interventional radiology (IR), and APNs in each of these specialties. The multidisciplinary team approach to assess candidacy for surgical PTE or BPA includes comprehensive review of the patient's medical history, presence of comorbid conditions, and right heart catheterization data, as well as imaging

modalities including ventilation-perfusion scans, echocardiograms, chest imaging, and pulmonary angiography.^{1,5-7} This process is critical in achieving positive surgical and procedural outcomes.

APNs play an integral role in the evaluation, management, and education of CTEPH patients, guiding them through an often complicated, and at times overwhelming, process from diagnosis to treatment. At our center, APNs provide patient education and management including evaluation for potential surgical and/or medical therapies, perform preprocedural evaluations, provide inpatient postoperative and postprocedure management, and provide continuity with outpatient follow-up after a given treatment strategy. Using a patient-centered approach in which the patient and family are involved, the APN is tasked to ensure patients and their families understand the diagnosis and treatment options and assist them in setting realistic expectations in terms of procedural outcomes and possible improvements in functional ability.^{1,7-9} The APNs are available to answer any questions that often arise and are a valu-

Correspondence: ssalveson@mednet.ucla.edu

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able component to a successful CTEPH program.^{6,7} In this article we will review the role of the APN, specifically the nurse practitioner, as part of the interdisciplinary team caring for CTEPH patients.

APN ROLE: PTE

PTE is a potentially curative treatment option for CTEPH. The surgery is relatively young and has improved throughout the years. The first pulmonary endarterectomy was performed in 1957, approximately 30 years after the disease was first noted.¹⁰ That patient lived for just a few hours.¹⁰ The introduction of cardiopulmonary bypass during PTE allowed for greater success, and by 1989, 250 surgeries had been performed. At that time, the mortality rate was around 20%.¹⁰ Today the mortality rate is less than 5%.¹⁰ Techniques devised at the University of California San Diego, including the use of circulatory arrest during PTE and the creation of specific surgical instruments that allow for more distal retrieval of fibrous clots, have resulted in lower mortality rates.¹⁰ Perhaps most important in the success of PTE is using a multidisciplinary approach at dedicated CTEPH centers that include the cardiothoracic surgeon, pulmonologist, interventional radiologist, and APNs. This section will highlight some of the roles of the APN in the care of PTE patients.

First and foremost, any patient with a history of PE and chronic dyspnea should be referred to a CTEPH center. Early referral can prevent the development of secondary small vessel vasculopathy that will result in suboptimal outcomes post-PTE (ie, residual PH) and reduce the increased risk of morbidity and mortality.¹¹ At our institution, patients are referred directly from our pulmonary colleagues. The APN in the cardiothoracic setting ensures each patient has undergone a thorough workup and orders appropriate testing that has not yet been completed. These tests include a transthoracic echocardiogram to evaluate right ventricular function and systolic pressure, a computed tomography of the chest to evaluate for presence and extent of thrombotic disease, a ventilation-perfusion scan to confirm

and determine the amount of perfusion and ventilation mismatch, and a pulmonary angiogram to delineate disease location and provide baseline left and right pulmonary artery pressures. Other important preprocedural considerations the APN includes are evaluating anticoagulation, obtaining a complete history and physical, and determining if the patient has a history of recurrent deep vein thrombosis or PE. To minimize complications, the APN refers any patient with recurrent deep vein thrombosis, PE, or suspected coagulopathy to hematology prior to surgery. A patient with concurrent deep vein thrombosis and evidence of anticoagulation failure is referred for inferior vena cava filter placement prior to surgery.¹² IR places the inferior vena cava filter, and given documented low rates of retrieval, follow-up by the APN is necessary to prevent potential complications such as filter migration, thrombosis, or inferior vena cava fracture and embolization.13

Surgery for PTE is approximately 8 hours in duration. The patient recovers in the intensive care unit and is followed closely by the pulmonary and cardiothoracic teams. Careful monitoring for reperfusion injury or pulmonary edema is imperative in the immediate postoperative period. The APN will order nitric oxide, allowing for decreased pulmonary vascular resistance and improved oxygenation.¹⁴ PTE is performed with deep hypothermic circulatory arrest, a cooling of the body to less than 20 degrees to reduce harm to the central nervous system. A neuro exam is performed by the APN to evaluate for possible neurological sequela. Transient neurological dysfunction, such as confusion, motor weakness, seizure, agitation, and/or delirium can occur after PTE and is typically seen in patients with longer deep hypothermic circulatory arrest time and in individuals greater than 64 years of age.¹⁵ Additional management considerations include right ventricular support with inotropes and early mobilization and ambulation to improve patient functionality and reduce the risk of postoperative deep vein thrombosis, lung infection, and pleural effusion.¹⁶ The length of stay for a PTE patient at our

institution is approximately 5 days. Patients are discharged on warfarin anticoagulation with a goal international normalized ratio (INR) between 2 to 3. The APN must emphasize adherence to lifelong anticoagulation as it remains integral to the treatment of the CTEPH patient.¹⁷ Before discharge, it is common practice at our institution to obtain an echocardiogram. Approximately 25% of PTE patients develop a pericardial effusion postsurgery, and 5% require surgical drainage.¹⁸ Zhang and colleagues reviewed 502 patients that underwent PTE between 2018 and 2020 and determined that younger, taller males were more likely to experience postoperative pericardial effusion as well as patient patients with re-entry sternotomies.¹⁸ Once a pericardial effusion is found, the patient is serially monitored with echocardiogram to ensure stability or improvement. The APN will need to monitor and maintain tight control of the patient's INR to prevent supratherapeutic levels that may result in potential worsening effusion or tamponade physiology.

Following discharge, the PTE patient will have a return follow-up visit in the cardiothoracic outpatient clinic. The surgical APN provides recovery expectations and continues to closely monitor for complications through an in-person follow-up in addition to telehealth monitoring. At our institution, it is common for the APN to manage the patient's INR in the outpatient setting until the INR reaches a therapeutic level. The pulmonary team will schedule a visit and perform baseline testing postsurgery that allows for an objective assessment of pulmonary artery pressure, right ventricular function, exercise capacity, and quality of life.11

Early referral of patients with unexplained dyspnea following PE with appropriate anticoagulation to a CTEPH center remains a critical first step in successful treatment. PTE is an option for some patients with CTEPH and can be curative. The APN assists the CTEPH patient in the presurgical and postsurgical settings by ensuring appropriate referral, surgical workup, postoperative monitoring, and medication management.

APN ROLE: BPA

BPA is a safe and effective catheter-based therapy for select CTEPH patients. BPA is an option available to CTEPH patients who are not eligible for surgical intervention with PTE for a variety of reasons including certain comorbidities, persistent symptoms despite optimized surgical or medical therapy, patient request for a less invasive procedure, or for residual PH post-PTE.^{1,2,4,16,19-21} The survival rate for nonsurgical PTE candidates has significantly improved since the availability of BPA as well as newer medical therapies.²² Taniguchi and collegues looked at the survival rates of nonoperative CTEPH patients that underwent BPA and found 1-year and 3-year survival rates of 98.8% and 92.9%, respectively.²³

BPA was first attempted in 1988 to treat acute PE in patients that did not respond to treatment with anticoagulation or thrombolytic therapy. The first reported case of BPA for CTEPH occurred in the same year and resulted in improved cardiac function, pulmonary pressures, and exercise tolerance, however, it was not widely adopted because of high complication rates. Work continued refining the procedure and once BPA was performed in a staged manner, the risk of complications was significantly reduced.^{21,24}

At our institution, BPA is performed by vascular interventional radiologists. As the CTEPH patient volume and case complexity grew, IR reevaluated their resources which lead to the addition of an APN to their multidisciplinary team to assist with preprocedural evaluations and postprocedure follow up.^{25,26} Preprocedure, the APN reviews the patient's medical history, pertinent laboratory work, and evaluates for any possible contrast allergies which would indicate a need for premedication orders. Coagulopathies may be identified which require the APN to involve hematology in the case.^{1,2,17} A transthoracic echocardiogram is reviewed to evaluate pulmonary artery pressure and RV function.^{1,7} In addition to echo data, the severity of PH is evaluated by review of the patient's right heart catheterization data and the patient's functional status including current symptoms and limitations. A

thorough medication reconciliation as well as documentation of baseline activity tolerance and oxygen requirements are also evaluated prior to BPA. Imaging review with the IR and multidisciplinary team is an integral step to identify the location and nature of thromboembolic lesions. Distal lesions in the segmental and subsegmental vasculature, down to small pulmonary arteries of 2 to 5 mm in diameter, that are considered inaccessible to PTE may be amenable to BPA.^{5,7}

As an educator, the APN has an important role within the multidisciplinary team providing patient and family education to enhance understanding of the disease state, explaining the BPA procedure, and assisting with care coordination to help the patient navigate successfully through the process.^{7,28} Quality preprocedural education empowers the patient to make decisions that will meet their needs and goals of care. The APN provides this education through the creation of patient-specific education materials and helps to educate hospital staff in the care of these complex patients to optimize outcomes.²⁹

BPA is performed in a staged, stepwise manner to reduce radiation exposure and to decrease the risk of complications such as reperfusion pulmonary edema and renal impairment due to contrast medium over 1 or multiple sessions.^{1,19,21} Multiple vessel locations may be addressed during a single BPA session and are generally limited to 1 lung. At our institution, 2 separate sessions are usually performed during 1 hospitalization, allowing for at least 24 hours in between sessions. Postprocedural monitoring takes place in the medical intensive care unit and involves cardiac monitoring and ongoing evaluation of oxygenation with close attention to increased oxygen requirements that may indicate lung injury. The IR APN also monitors the patient for potential procedural complications, all of which require prompt intervention, including reperfusion pulmonary edema that may manifest immediately or several days postprocedure, hemoptysis, contrast associated renal dysfunction, and access site issues.²¹ Postprocedural labs including CBC and BMP are monitored closely.

Serial chest x-rays are reviewed to survey for reperfusion pulmonary edema and/or pulmonary hemorrhage that may occur as a result of vascular injury during BPA.²¹ At our institution, resumption of oral anticoagulation is multifactorial, taking into consideration the patient's overall stability, and is usually resumed the evening after the procedure if there are no bleeding complications or subsequent procedures scheduled. Close monitoring for signs and symptoms of bleeding after restarting anticoagulation is imperative. The APN ensures that patients understand the importance of taking oral anticoagulation as directed with a discussion of warning signs to contact their provider post-BPA. The CTEPH multidisciplinary team collaborates to determine if a second session is appropriate during a given hospitalization based on the patient's response to the initial BPA including change in PA pressures post-BPA, and overall stability without complications or increases in oxygen requirements. Similar criteria are used to determine timing of discharge with most patients being discharged as early as 24 to 48 hours post-BPA. In the event future procedures are warranted, the risks and benefits of additional interventions will be reviewed by the multidisciplinary CTEPH team as an outpatient.

APN ROLE: FOLLOW UP POST-BPA

Post-BPA follow up with Pulmonary Vascular Disease clinic is essential and should be arranged during the hospital admission to ensure a seamless discharge and continuity of care in the outpatient setting. At our institution, like other expert BPA centers, 4-week follow-up includes repeat perfusion imaging, echocardiography, functional class assessment, 6-minute walk testing, and evaluation of brain natriuretic peptide levels.^{30,31} At this first posthospital visit, discussion about timing of the next staged BPA procedure, if indicated, occurs. The quantity of procedures performed will vary from patient to patient and is determined by severity of disease, target lesion morphology and accessibility, as well as the expertise of the BPA center.^{30,32} If additional targeted areas

are identified, a repeat BPA session is scheduled within 4 to 6 weeks.³⁰ On average, anywhere from 2 to 10 sessions are required.^{30,32}

Medical therapy to treat inoperable CTEPH, both pre-BPA and post-BPA, is also discussed in the outpatient setting. Riociguat is a soluble guanylate-cyclase stimulator that works to promote vasodilation and decrease pulmonary vascular remodeling and inflammation, and is the only drug approved for treatment of CTEPH.^{31,33} Current research is investigating the use of riociguat and BPA in combination therapy to improve patient outcomes. These data suggest that patients treated with medical therapy prior to BPA decrease the risk of complications in the postprocedure setting, indicating the benefit of sequential treatment methods to improve functional capacity and hemodynamics, and reduce potential complications associated with BPA.30-34

Anticoagulation in this patient population is lifelong, usually with use of either vitamin K antagonists or direct oral anticoagulants, however there is limited data to make formal recommendations in regards to direct oral anticoagulants therapy specifically.^{31,32,35} Current guidelines recommend vitamin K antagonists for CTEPH patients, with a target INR range of 2 to 3, though there is clearly a need to evaluate the safety of direct oral anticoagulants therapy compared to standard of care in this population with further prospective data review.³¹

Ultimately, further research is needed to define the standards of practice across institutions and provide data on longterm outcomes in BPA.

SUMMARY

Research has consistently demonstrated that APNs provide high-quality care with excellent outcomes that are safe, patient-centered, and cost effective. Involvement of an APN on the multidisciplinary team for patients with complex illnesses such as CTEPH has been shown to lead to improved outcomes and greater patient satisfaction.^{8,22,36-43} At our center, the addition of APNs from the pulmonology, cardiothoracic surgery, and IR specialties enhances multidisciplinary collaboration among the CTEPH team and is aimed at improving care and outcomes in this patient population.²⁹

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