# Innovations in the Management of Chronic Thromboembolic Pulmonary Hypertension

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### Introduction

Chronic thromboembolic pulmonary hypertension (CTEPH) is a rare but serious condition that occurs when blood clots become lodged in the pulmonary arteries, leading to high blood pressure in the lungs. CTEPH can lead to symptoms such as shortness of breath, chest pain, and fatigue, and can ultimately result in right heart failure if left untreated. While CTEPH was once considered a largely untreatable condition, recent innovations in the management of the disease have led to improved outcomes for patients [1].

## Description

Diagnosis of CTEPH typically involves a combination of imaging studies, such as computed tomography (CT) scans and pulmonary angiograms, and pulmonary function tests. Once a diagnosis is confirmed, treatment options may include medical management, balloon pulmonary angioplasty (BPA), and pulmonary endarterectomy (PEA). Medical management of CTEPH typically involves the use of pulmonary arterial hypertension (PAH) medications, such as riociguat and prostacyclins [2,3], which work to dilate the pulmonary arteries and reduce pulmonary artery pressure. While these medications can be effective in improving symptoms and hemodynamics in some patients, they are not curative and may have significant side effects.

BPA is a relatively new treatment option for CTEPH that involves using a catheter to deliver a balloon to the site of the blood clot in the pulmonary artery, where it is inflated to reopen the blocked vessel. BPA has been shown to be effective in improving pulmonary artery pressure and functional capacity in patients with CTEPH who are not candidates for PEA. However, BPA is a technically challenging procedure that requires specialized training and may not be available at all medical centers.

PEA is considered the gold standard treatment for CTEPH and involves surgically removing the blood clots from the pulmonary arteries [4]. PEA is typically reserved for patients with proximal, surgically accessible clots and good right ventricular function. PEA has been shown to be highly effective in improving symptoms and survival in patients with CTEPH, with up to 90% of patients experiencing significant improvement in pulmonary hemodynamics. In addition to these traditional treatment options, recent innovations in the management of CTEPH have expanded the treatment armamentarium for patients. One such innovation is the use of percutaneous mechanical thrombectomy (PMT) in the management of CTEPH. PMT involves using a catheter-based device to mechanically break up and remove blood clots from the pulmonary arteries. This technique has been shown to be effective in

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improving pulmonary hemodynamics and functional capacity in patients with CTEPH who are not candidates for PEA.

Another innovative treatment option for CTEPH is the use of extracorporeal membrane oxygenation (ECMO) as a bridge to PEA. ECMO is a life support technique that involves pumping blood outside of the body, oxygenating it, and then returning it to the body. In patients with CTEPH who are too sick to undergo PEA immediately, ECMO can be used to support the patient's cardiovascular system while they are stabilized and prepared for surgery.

Finally, advances in imaging techniques, such as magnetic resonance imaging (MRI) and positron emission tomography (PET), have improved the ability to diagnose and monitor CTEPH. These techniques can provide more detailed information on the location and extent of blood clots in the pulmonary arteries and can help guide treatment decisions. Recent innovations in the management of CTEPH have expanded treatment options for patients and have led to improved outcomes. While medical management and PEA remains the mainstay of treatment for CTEPH, techniques such as BPA, PMT, ECMO, and advanced imaging have provided additional options for patients who may not be candidates for these traditional therapies [5].

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Not applicable.

### **Conflict of Interest**

There is no conflict of interest by authors.

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