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Use of Intravenous Prostaglandins as a Bridge Therapy for Chronic Thromboembolic Pulmonary Hypertension Patients Awaiting Pulmonary Endarterectomy

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Introduction

The gold standard for treatment of chronic thromboembolic pulmonary hypertension (CTEPH) has been surgical pulmonary endarterectomy (PTE) [1]. However, pharmacologic therapy has been used in CTEPH patients in the following scenarios: (i) those deemed inoperable due to significant co-morbidities or extensive distal precapillary vasculopathy and (ii) in those with residual pulmonary hypertension following PTE [2]. There are a limited number of randomized control trials that have evaluated the efficacy of medical treatment in these select CTEPH patients. Small open label trials with bosentan, treprostinil, epoprostenol and sildenafil have been conducted in CTEPH patients but their conclusions are limited by small cohort of patients, lack of blinding and randomizationin their study design as well as absence of a control arm [3]. Presently, there is no strong recommendation regarding appropriate use of medical therapy for these CTEPH patients [3]. Furthermore, there is no evidenceto support medical therapy as a bridge to PTE. In fact there is hesitation as it is thought to cause delay in surgical referral.

Discussion

When pharmacologic therapy is used as a bridge to PTE, no appreciable significant difference in postoperative hemodynamics has been noted [4]. However, intravenous prostacyclin therapy in patients undergoing PTE has shown a significant reduction in preoperative pulmonary venous resistance that has been associated with improved operative mortality [5]. Moreover, the landmark CHEST-1 and CHEST-2 clinical trials proved that soluble guanylate cyclase stimulator, (riociguat) can improve exercise capacity and pulmonary vascular resistance in inoperable CTEPH patients and those with recurrent pulmonary hypertension after PTE [6].

Case Presentation

A 51 year old male with coronary artery disease status post multiple percutaneous coronary interventions (PCI), hypertension, type 2 diabetes and history of chronic right ventricular (RV) failure along with pulmonary hypertension (PH) was referred for evaluation of CTEPH in

June 2018 following a hospitalization for acute on chronic respiratory and RV failure. His echocardiogram revealed severely dilated and functionally-reduced RV with large pericardial effusion but without tamponade features, left ventricular ejection fraction (EF) 58%, bi-atrial enlargement, enlarged inferior vena cava and ascites. He underwent a therapeutic paracentesis and a V/Q scan was performed which showed high probability for pulmonary embolism. He was started on Apixaban and referred to us for possible PTE and PH treatment. Prior to this admission, the patient noticed a progressive decline since January 2018. He was seen in April 2018 for dyspnea on exertion and EF at that time was 35%. He underwent a left and right heart catheterization (LHC/RHC) with coronary angiogram along with multiple PCIs at that time. The RHC showed elevated pulmonary artery pressures. His EF improved status post PCI but his RV systolic pressure and RV function did not. He was placed on home oxygen therapy. The patient had no family history of autoimmune disease, cardiac disease or venous thromboembolism.

His workup for CTEPH was completed with pulmonary angiogram and RHC which showed severely elevated pulmonary artery pressures (110/7 mmHg) with mean pressure of (40 mmHg), high right atrial pressure (16 mmHg) and low cardiac output and index by Fick method (3.47 Liters/minute and 1.68 Liters/minute/meters squared respectively). There was a total occlusion of the right lower lobar artery with scattered web lesions throughout right middle and upper lobe vessels (3rd and 4th generation) as well as scattered web lesions in the 4th generation vessels of the leftpulmonary artery.

The patient was started on intravenous treprostinil as well as riociguat to try to optimize his hemodynamics and he subsequently underwent PTE two months later. Echocardiogram immediately prior to PTE demonstrated an improvement in RV function from severely to moderately depressed and improvement in pulmonary artery pressures to 50 mmHg. Post procedure pressure on post-operatively day 1 off of intravenous treprostinil was 25/3 mmHg with normal cardiac output. His post-operative echocardiogram showed further improvement in RV parameters (RV size 5.0 cm initially, now 4.4 cm with RV function now reported as mild to moderately reduced). Warfarin was initiated for lifelong anticoagulation therapy.

Conclusion

Management of CTEPH patients prior to PTE is mainly supportive care with systemic anticoagulation, optimization of fluid status and supplemental oxygen for resting hypoxemia. However, in symptomatic patients who are PTE candidates, the use of aggressive pharmacotherapy using parenteral prostanoids can be beneficial [7]. Our case suggests the off license use of PH therapy with utilization of intravenous treprostinil in CTEPH as a bridge to PTE. This is supported by improvement in results of PTE surgery and potential association with long-term hemodynamic and clinical outcomes. However, there is very limited data to support these conclusions1. This is a growing area of interest andneed for expansion of clinical research so that medical therapy can be validated for CTEPH patients pre-PTE.

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