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Advances in PH Journal

Ask the Expert: Modalities Used to Help Prolong Survival in Patients With Advanced PAH

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What is the experience with ECMO and RVADs in the management of patients with advanced PAH?

Pulmonary arterial hypertension (PAH) is a relentless, progressive disease of the small and medium-sized pulmonary arteries that often culminates in development of right ventricular (RV) failure. Earlier detection and advances in pharmacotherapy have greatly enhanced quality of life and reduced clinical worsening, but a cure remains elusive, and many patients require lung transplantation. Even with close monitoring, the clinical course can be unpredictable and patients can present with pulmonary hypertensive crisis, characterized by profound RV failure leading to marked reductions in cardiac output with subsequent end organ hypoperfusion and systemic hypotension. Limited published data are available to guide decision making in this regard, and treatment focuses on optimization of volume status via diuretics and ultrafiltration, judicious use of inotropes, and the use of inhaled or intravenous pulmonary arterial vasodilators.

Mechanical circulatory support (MCS) is widely recognized as a therapy that can be used to treat a spectrum of pathologies affecting the lung parenchyma and the RV. Technological advances have greatly expanded the safety and capabilities of both percutaneous and surgically implanted devices. These devices are used as a bridge to either recovery of organ function or to transplantation. Published data regarding the use of these devices in PAH are largely limited to case reports. MCS typically takes the form of: right ventricular assist devices (RVAD, either percutaneous or surgically placed), which solely serve as a pump; or extracorporeal membrane oxygenation (ECMO), which can provide oxygenation (veno-venous [VV] ECMO); or pump function and oxygenation (veno-arterial [VA] ECMO).

PAH is pathophysiologically distinct from other more common conditions including acute respiratory distress syndrome (ARDS) and pneumonia by the location of disease in the precapillary vasculature, resulting in a markedly elevated pulmonary vascular resistance. Consequently, temporary surgically placed RVAD's routinely used for post cardiotomy RV failure,[1] which shunt blood from the right atrium to the pulmonary artery, are of limited use in pulmonary hypertensive crises. Published literature describes significant complications including anastamotic bleeding and hemotypsis.[2] Percutaneous RVADs, including the TandemHeart™ (Cardiac Assist Inc., Pittsburgh, PA) suffer from a similar complication of increasing blood flow into an already high resistance pulmonary circuit.[3]

VA-ECMO is a percutaneously inserted support which removes blood from the venous system, bypasses the heart and lungs, and returns oxygenated blood to the arterial system. The large lumen catheter requires a surgical cutdown approach and is typically placed in the femoral artery. Although highly efficacious, the primary limitation from this form of support stems from complications related to access (large bore arterial access leading to vascular complications) and the need for aggressive anticoagulation (increased risks of bleeding, stroke, and coagulopathy).[4] Despite advances in perfusion technology, this type of therapy cannot be used for expanded periods of time. This short duration may not be sufficient to allow for aggressive titration of pharmacotherapy, or to allow time for organ transplantation.

VV-ECMO provides oxygenation of blood, and is an accepted treatment of acute lung injury when parenchymal lung disease impairs oxygenation but RV function remains adequate.[5] Because pulmonary hypertensive crises are characterized by RV failure, this therapy would not provide the hemodynamic support necessary to improve organ perfusion. However, patent forame ovales (PFOs) are not uncommon in PAH, and can be life saving in pulmonary hypertensive crises by allowing a pop off mechanism to decompress the RV and to maintain cardiac output. In the presence of a PFO, it is conceivable that VV-ECMO could shunt blood across the interatrial septum, thereby maintaining cardiac output and improving tissue oxygenation, and stabilizing blood pressure, allowing for titration of PAH pharmacotherapy.[6] VV-ECMO has a lower incidence of hematologic and cardiovascular complications, and can be used for longer duration of time compared with VA-ECMO.

Recent advances in MCS have improved our ability to care for a spectrum of patient with advanced cardiovascular and pulmonary pathology. Pulmonary hypertensive crisis remains a unique therapeutic challenge, and the potential to utilize MCS in this setting offers a promising approach for these acutely compromised patients. Additional studies are needed to determine the optimal type of MCS, to establish safety, and to improve our understanding of overall efficacy.

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With the choices of therapies available for PAH, what are the current indications for atrial septostomy? What are the absolute contraindications and risks? Who would be the "ideal" candidate for this procedure?

Julio Sandoval, MD

Untreated idiopathic pulmonary arterial hypertension (PAH) is characterized by a progressive elevation of pulmonary artery pressure, followed by worsening right ventricular failure (RVF) and ultimately death. The development of specific disease-targeted therapy for PAH has offered major advances in the management of patients, and these medical treatments have been shown to improve both quality and quantity of life. Nevertheless, many patients still deteriorate with time and require consideration of other approaches such as atrial septostomy (AS) and lung transplantation.[1],[2]

In severe PAH, AS creates a right-to-left shunt reducing right ventricle preload. It results in an increased cardiac output (CO) and an augmentation of systemic oxygen transport (SOT) despite the fall in systemic arterial oxygen saturation (SaO_2).[3],[4] Although the precise role of AS in the treatment of PAH patients remains uncertain, the performance of the procedure has steadily increased over the years. The last review of the worldwide experience records 223 reported cases.[3] Mean age was 28 ± 17 years and 70% were female patients with idiopathic PAH (82%). The mean NYHA functional class of the group was 3.56 ± 0.4. Indications for the procedure were worsening RVF (43%), effort syncope (38%), or both (19.4%), with bridge to transplantation in 14% of cases. Ninety-six (43%) of these patients had failed to respond or had deteriorated following targeted medical treatment.

The performance of AS in severely ill patients may explain a still high immediate procedure-related mortality rate (?7%). On the other hand, from a total of 186 surviving patients, syncope and RVF improved in 88%. Hemodynamic response after septostomy was characterized by a significant fall in right atrial pressure (RAP), and an increase in CO and in SOT. Hemodynamic improvement is contingent on baseline RAP; patients with a RAP between 11-20 mm Hg had the best risk: benefit ratio. In the 128 patients with follow-up available, the median survival after septostomy was 60 months.[3]

At present, AS should be considered for advanced NYHA Class III and Class IV patients with recurrent syncope and/or RVF despite all available medical treatments (including combination therapy). It should also be considered either as a bridge to lung transplantation or as the sole treatment modality when other therapeutic options are not available.[1–3]

Absolute contraindications for the procedure include patients with impending death. Patients with resting SaO_2 <90% on room air and severe RVF appear more likely to worsen or die after atrial septostomy. A RAP >20 mm Hg has been repeatedly identified as the most significant risk factor for procedure-related mortality.

To minimize the risk of death or complications, stepwise balloon-dilatation AS (BDAS) is the procedure of choice.[3,4] There are no current guidelines for the optimal size of the defect. Massive right-to-left shunting, as a result of a “too large orifice,” may result in inadequate pulmonary blood flow and severe, refractory hypoxemia. The aim is to achieve a fall in SaO_2 ?10%. An increase in left ventricular diastolic pressure to 18 mm Hg should also preclude further dilation. BDAS should be performed only in centers experienced in both interventional cardiology and pulmonary hypertension.

There is no question that BDAS has a place as a therapeutic modality for advanced PAH. I believe, however, that it is performed too late in the course of the disease, when the risk of death is high. The preferred timing for BDAS should be elective rather than rescue. The “ideal” candidate for the procedure would be a Class III patient with syncope or persistent RVF, RAP between 10-18 mm Hg and SaO_2% >90%, who has failed medical therapy in a goal-oriented strategy.[5] Many patients listed for lung transplantation fulfill these criteria and BDAS should be used more frequently in this group in an attempt to prolong survival.

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