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Venopulmonary ECMO Improved Hypoxemia and Supported the Right Ventricle in a Patient with Decompensated Eisenmenger Syndrome

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Abstract

Mechanical circulatory and/or respiratory assistance with extracorporeal membrane oxygenation (ECMO) has become a standard of care for patients with circulatory (venoarterial) and/or respiratory (venovenous) failure refractory to standard therapies. Adult patients with congenital heart disease are an increasingly recognized and growing population and include various groups, such as undiagnosed cases in childhood and palliated and/or corrected cases, which require subsequent care because of residual lesions, cardiac arrest/insufficiency, and arrhythmias, among other conditions. In addition, these patients are prone to developing pathologies that are typical of adulthood with a generally increased risk of morbidity and mortality because of their low reserves and organic damage associated with the underlying heart disease, which makes them candidates for ECMO. These patients represent an additional challenge in this therapy because malformations and the presence of a shunt can generally affect the usual cannulation methods and hemodynamic and oximetry monitoring. Thus, the configuration decision must be made on a case-by-case basis. Here, we present a cannulation method, venopulmonary artery ECMO, which provides hemodynamic and respiratory support, and may be ideal for patients with shunts and/or right ventricular dysfunction. To our knowledge, this is the first report of this configuration in patients with congenital heart diseases.

Keywords

Eisenmenger Syndrome, Venopulmonary Artery ECMO, Refractory

Hypoxemia, Right Ventricular Dysfunction

1. Introduction

Extracorporeal membrane oxygenation (ECMO) has gained increasing popularity as an effective therapy for managing cardiac and/or respiratory failure refractory to conventional medical management, or when the parameters of these conventional therapies are extremely high, preventing organic recovery. Patients with adult congenital heart disease (ACHD), whether palliated, fully corrected, or uncorrected, represent a growing population with a high morbidity rate that requires high demands for care. As patients with ACHD age, there is an increased prevalence of heart failure and other noncardiac comorbidities. Healthcare utilization among ACHD patients has grown steadily in inpatient and outpatient settings, and those patients with complex forms of ACHD have the highest rates of emergency department utilization, hospitalization, and critical care needs [1]. In parallel with survivorship, the complexity of ACHD has increased substantially over the past two decades, and there is an increased need for options of respiratory (venovenous (VV) ECMO) and/or mechanical circulatory support (venoarterial (VA) ECMO) [2]. ACHD patients often have a complex anatomy and may have undergone multiple prior interventions, resulting in limitations to vascular access [3]. Because of its special configuration, venopulmonary artery (VPA) ECMO (**Figure 1**) appears to be an attractive option in patients with ACHD. Changing the configuration of the extracorporeal therapy by placing the return cannula toward the pulmonary artery (PA) can improve the patient's clinical condition for two main reasons: 1) by injecting oxygenated blood directly into the PA, the degree of recirculation is significantly reduced, thereby improving the systemic oxygenation of the patient; and 2) mechanical right ventricular unloading allows the application of lung and right ventricular protective ventilation [4]. This therapy could be particularly attractive in the management of patients with congenital heart disease and/or Eissenmenger syndrome, since its decompensation usually includes severe right heart failure, pulmonary hypertensive crisis, and worsening hypoxemia, as a "bridge to recovery". Here, we present the case of a young patient diagnosed with Eisenmenger syndrome (ES) who developed severe refractory hypoxemia and pulmonary hypertensive crisis associated with a respiratory infection and postpartum state, who received respiratory and hemodynamic support from VPA ECMO. To our knowledge, this is the first report of a case of ACHD supported with this modality, which may be beneficial in this population, specifically in those with intracardiac shunts.

2. Case Description

A 28-year-old woman with a medical history of a heart murmur since childhood was diagnosed with perimembranous ventricular septal defect (VSD) with a

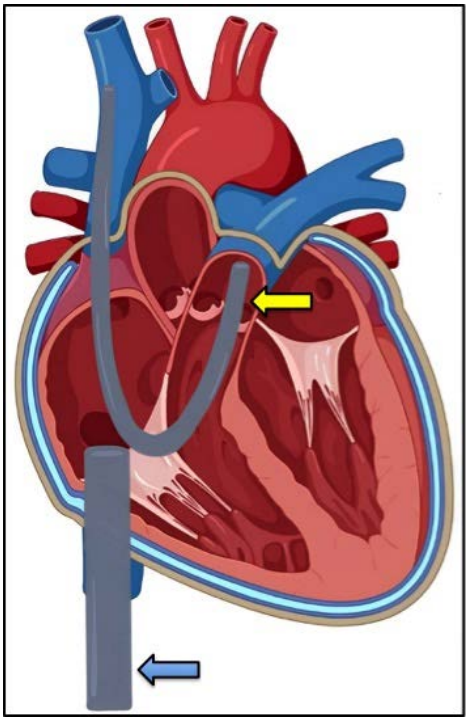

Figure 1. Venopulmonary artery ECMO, double cannulation technique. The drainage venous cannula (blue arrow) is positioned in the right atrium at the cavoatrial junction. The return venous cannula (yellow arrow) passes through the superior vena cava, the right atrium, and the right ventricle to the main pulmonary artery (final position).

QP/QS ratio of 2.2. At 8 years of age, she underwent cardiac catheterization, being excluded from surgical treatment because of suprasystemic pulmonary hypertension, and she was diagnosed with ES. She received medical treatment with digoxin, captopril, and aspirin until the age of 18. She could carry out her daily activities without difficulty and was classified as NYHA functional class II. The patient did not know her baseline SpO_2.

The illness that brought her to the hospital began with a dry cough, myalgias, and rhinorrhea. Two days later, she developed sudden-onset dyspnea. A pregnancy of 25 weeks of gestation was documented, and it was decided to end it abdominally (cesarean section). The patient then developed acute respiratory distress syndrome, requiring FiO_2 of up to 100% despite positive end-expiratory pressure titration, septic shock, dual vasopressor management (norepinephrine and vasopressin), and broad-spectrum antibiotic therapy. Because of refractory respiratory and circulatory failure, transfer to our center was requested for evaluation of ECMO therapy.

The patient's vital signs were as follows: blood pressure of 80/50 mmHg, heart rate of 110 bpm, respiratory rate of 18 bpm, temperature of 38.1˚C, and SpO_2 of 40%. These readings were accompanied by clinical evidence of poor perfusion and elevated lactate levels. Portable chest X-ray showed grade 2 cardiomegaly, dilatation of the main PA, and bilateral diffuse infiltrates (**Figure 2**). A transthoracic echocardiogram was performed, showing a 15 mm perimembranous VSD with

a right-to-left shunt, dilated right cavities with a decreased circumferential function (fractional area change of 31%) (Figure 3). A pulmonary artery systolic pressure (PASP) of 163 mmHg and a mean pulmonary artery pressure (mPAP) of 112 mmHg were quantified. Computed tomography was performed, where pulmonary thromboembolism was ruled out, but multiple bilateral infiltrates suggestive of pneumonia were documented.

The patient was transferred to the hemodynamics department for the procedure. A Teflon-coated exchange guidewire (260 cm) was advanced to the superior vena cava through the right femoral route, and a 21 French venous cannula (Medtronic, Minneapolis, MN, USA) was advanced to the cavoatrial junction without complications. Subsequently, a 0.021 Teflon-coated guidewire was

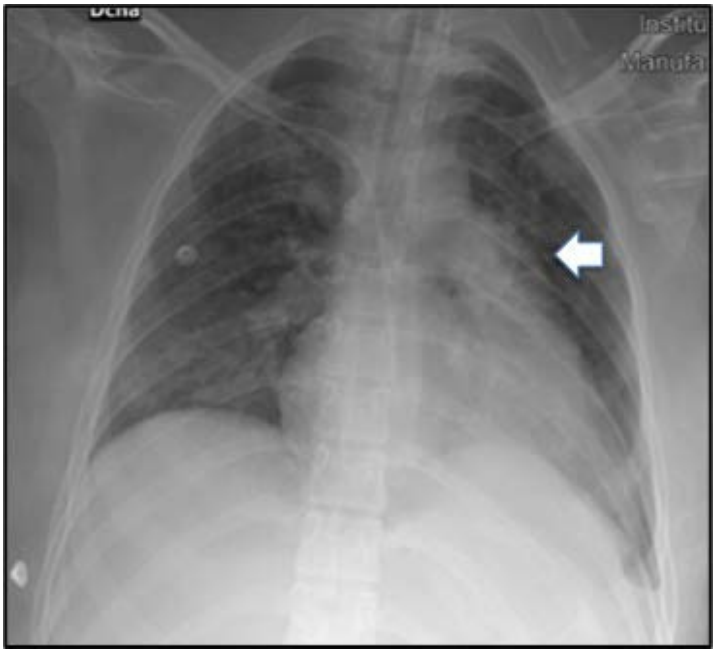

Figure 2. Portable chest X-ray showing diffuse pulmonary infiltrates, dextrorotation, and pulmonary artery dilation (white arrow).

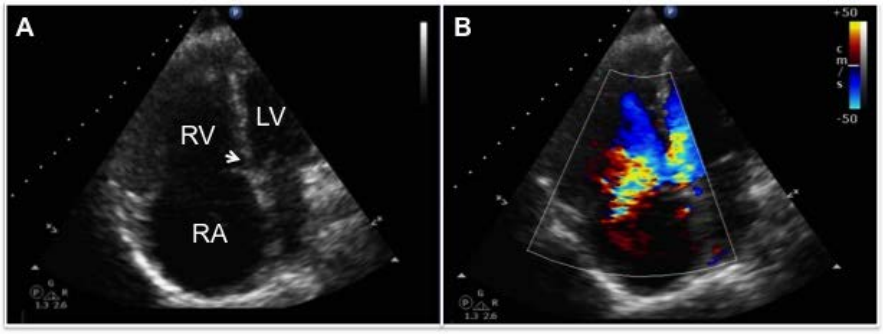

Figure 3. Transthoracic echocardiogram. (A) Apical 4-chamber view, showing dilation of the right atrium and ventricle, interventricular septal defect, and collapse of the left heart chambers due to exacerbated biventricular interdependence. (B) Apical 4-chamber view with color Doppler mode, showing a right-to-left ventricular shunt. LV: left ventricle, RA: right atrium, RV: right ventricle.

advanced to the right branch of the PA through the right internal jugular vein; over the guide, a 5 French multipurpose catheter was advanced, through which a Lunderquist extra-stiff wire guide (Cook Medical, Bloomington, IN, USA) was introduced. Then, a 17 French venous cannula (Medtronic) was advanced to the right branch of the PA (Figure 4). Support was started with 100% extracorporeal oxygen fraction, blood flow of 4 l/min, and sweep gas flow of 4 l/min, improving SpO_2 (from 40% to 100%). Subsequently, the oxygen supply was titrated until reaching a SpO_2 of 75% - 80% according to the patient's baseline characteristics and pathophysiological substrate (Table 1). PASP was reduced to 98 mmHg, mPAP to 60 mmHg (Figure 5) and the right ventricular fractional normalized

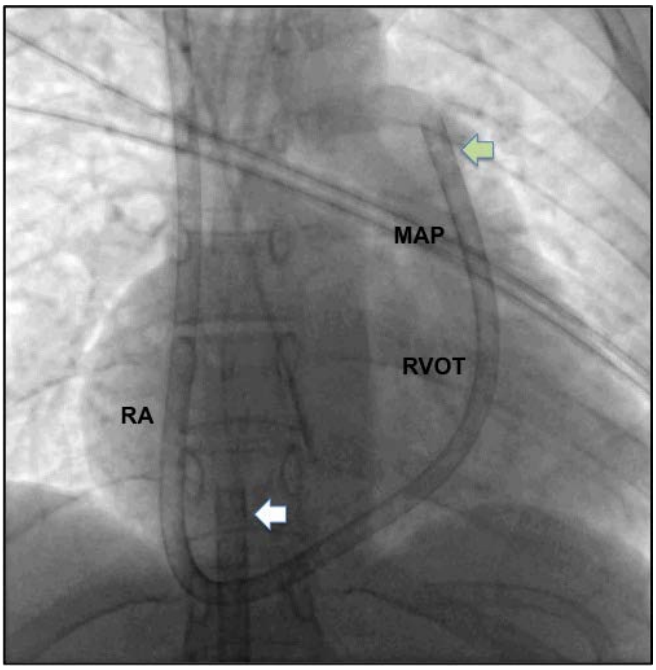

Figure 4. The return venous cannula (white arrow) passing through the superior vena cava, the right atrium, and the right ventricle to the main pulmonary artery (final position). The drainage venous cannula is positioned in the right atrium at the cavoatrial junction (green arrow). MPA: main pulmonary artery, RA: right atrium, RVOT: right ventricular outflow tract.

Table 1. Blood gas values before and after ECMO cannulation.

Time (hours)	Pre-cannulation	Post-cannulation				
		0	6	12	24	48
pH	7.47	7.22	7.38	7.61	7.49	7.48
PaO_2 (mmHg)	31	149	54	47	42	58
CO_2 (mmHg)	87	58	35	37	51	49
HCO_3 (mmol/L)	35.5	29	23	34	34	33
Lactate (mmol/L)	5.6	13.2	3.2	2	1.6	2
Oxygensaturation (%)	38	98	78	82	71	80

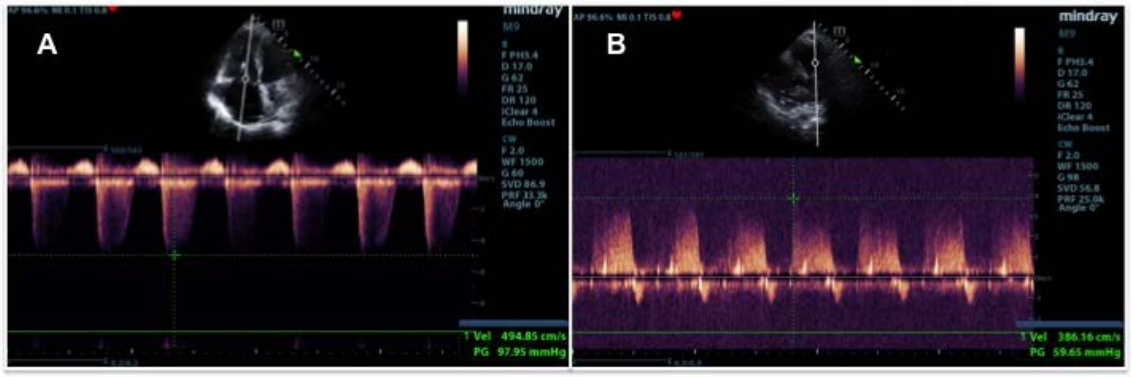

Figure 5. Transthoracic echocardiogram. (A) Using continuous wave—CW Doppler, a maximum tricuspid regurgitation gradient of 98 mm Hg is documented. (B) A maximum gradient of the initial maximum velocity of the pulmonary regurgitation flow of 60 mm Hg is quantified.

after starting the therapy.

Concerning the complications during ECMO, the patient developed cardiac tamponade (resolved with a pericardial window, with 300 ml of hematic fluid drained) and bleeding from the left main bronchus, which caused total lung atelectasis (resolved with bronchoscopy). These complications prolonged the extracorporeal assistance, making weaning impossible. Notably, the patient did not develop mechanical, bleeding, or infectious complications related to the cannulation site.

On day 20 of extracorporeal assistance, the patient developed a fever, neurological status changes, and acute kidney injury. Intravascular hemolysis was documented, and hemolytic uremic syndrome was suspected (negative direct Coombs test), for which steroid therapy, plasmapheresis, and renal replacement therapy were started.

The patient's clinical condition progressively deteriorated despite circulatory and respiratory support with ECMO for 34 days, continuous renal replacement therapy, and plasmapheresis. She required higher levels of oxygen provided by the ECMO; she continued with renal and hepatic dysfunction, persistent hemolysis, elevation in serum lactate levels, and refractory metabolic acidosis. Finally, the patient developed pulseless electrical activity and died, despite cardiopulmonary resuscitation maneuvers.

3. Discussion

There are multiple indications for utilizing VA ECMO in ACHD, and it may be a useful tool in stabilizing ACHD patients who present with cardiogenic shock, low cardiac output state, or cardiomyopathies due to acute coronary injury. VA ECMO may also be used as rescue therapy in ACHD patients who are unable to be weaned from cardiopulmonary bypass or in those who need periprocedural support. Finally, it may serve as a bridge to decision or bridge to transplant [5], and may be appropriate for reversible causes of ventricular dysfunction. For patients with ACHD undergoing cardiac arrest, VA ECMO should be considered

early in resuscitation efforts [6]. Furthermore, some ACHD patients may have isolated respiratory failure and benefit from VV ECMO [7]. ACHD patients often have different baseline arterial and mixed venous oxygen saturations secondary to congenital or acquired intracardiac shunts and systemic and pulmonary circulations that are partially or completely parallel. Our patient presented two indications of extracorporeal support: severe refractory hypoxemia and right ventricular dysfunction.

PA hypertension affects 5% - 10% of ACHD patients and is associated with significant morbidity and mortality [8]. Chronic exposure of the pulmonary vasculature to increased flow occurs, resulting in the remodeling of the vascular bed, which causes an increase in pulmonary vascular resistance until ES is finally established [9]. The risk of developing ES varies depending on the type of heart disease; in patients with uncorrected VSD, it reaches 50% [10]. Vasoconstriction, thrombosis in situ, and the abnormal vascular remodeling condition of the obstruction of the pulmonary vessels at the microvascular level explain the increase in vascular resistance, as a consequence of which the sustained increase in right ventricular afterload leads to right ventricular failure and eventually death. Hypoxemia in ES is related to the shunt reversal in the presence of systemic pulmonary arterial pressures. It ranges from mild to severe and typically worsens with exertion, affecting exercise capacity and the ventilatory response [11] [12]. Hypoxemia is typically associated with an increase in hemoglobin concentration, red cell count, and hematocrit (secondary erythrocytosis); it is a compensatory mechanism that increases the oxygen-carrying capacity of the blood by acute extracardiac conditions (infectious processes) or simply by progression secondary to the natural history of the disease that causes an imbalance in the relationship between oxygen supply and consumption and organic damage. When the usual therapy with invasive or noninvasive ventilatory support and the use of vasoactive drugs are not sufficient to restore oxygen supply demand and achieve reversal of tissue hypoxia, ECMO should be considered as a bridge to recovery, a bridge to decision, or a bridge to transplant [13], as in the case of our patient.

Regarding the type of configuration, there is more experience in pediatric patients. VA ECMO can be performed via peripheral cannulation (most commonly drainage via right internal jugular vein, return via right common carotid artery) or central cannulation (most commonly drainage via right atrium, return to ascending aorta). Central cannulation is generally easily accessible and preferable in the early post-cardiotomy period or if peripheral vessel size and cannula selection would be prohibitive to achieving adequate ECMO flow. This is especially important to consider in the setting of shunt-dependent pathology (*i.e.* hypoplastic left heart syndrome following Norwood procedure with modified Blalock-Taussig shunt). In these patients, the position of the arterial cannula relative to the mBTS and parallel circulations can result in pulmonary overcirculation and inadequate systemic flow. There is no experience to our knowledge with VPA ECMO or any specifically suggested setting in patients with Eissenmenger

syndrome [14].

Concerning the use of ECMO in ACHD patients, baseline arterial and mixed venous oxygen saturations must be considered when setting goals and expectations. The anatomy and pathophysiology of congenital cardiac malformations must be thoroughly understood. Canulation can be extremely challenging in ACHD secondary to multiple factors, including congenitally abnormal systemic and pulmonary venous connections, residual systemic-to-pulmonary communications, and the loss of patient blood vessels secondary to multiple catheterizations and cardiac surgical procedures. In patients with residual systemic-to-pulmonary communications, the return cannula position relative to the shunt is critical to avoid residual shunting of deoxygenated blood in pulmonary hypertension and/or right ventricular failure [15]. However, this last approach has not been reported in this population.

In our patient, when using VPA ECMO, the presence of refractory hypoxemia was managed by passing the shunt site at the level of the right ventricle, and injecting blood directly into the PA and toward the systemic circulation. This configuration also prevents recirculation. In addition, performing a "bypass" of the right ventricle prevents its failure, which is common during ECMO assistance, despite correcting the gas exchange and implementing less harmful ventilatory parameters (with the consequent improvement in pulmonary hemodynamics). These mechanisms include pulmonary vascular dysregulation, microvascular thrombosis, atelectasis, and continuous nonpulsatile flow [16]. This strategy has yielded encouraging results in nonpediatric populations diagnosed with severe acute respiratory distress syndrome, with and without associated right ventricular dysfunction [17] [18]. In our case, PA cannulation was performed with a conventional extracorporeal circulation venous cannula. Therefore, this strategy can be applied in centers with limited resources or where the usual cannulation devices for right ventricular assistance are unavailable or scarce.

There is a lack of direct comparison randomized studies of VPA ECMO versus other ECMO modalities. Current VPA ECMO literature is limited to observational data. A recent systematic review of five observational studies including 194 patients with coronavirus disease 2019 (COVID-19)-related ARDS receiving VPA ECMO showed that application of VPA ECMO, using single-site dual-lumen cannula was associated with high survival rates. VPA ECMO was also associated with reduced incidence of acute kidney injury and need for renal replacement therapy [19]. However, there is no experience of the use of this care modality in other populations, including patients with ACHD.

Despite achieving respiratory and circulatory support goals, our patient died because of a complication associated with prolonged exposure to extracorporeal circulation. In this case, it was thrombotic microangiopathy, which, although rare, usually has a fatal outcome [20]. However, it is important to note that, despite the long period of extracorporeal support (34 days), the patient did not develop complications related to cannulation (infection, bleeding, thrombosis, dis-

placement, or malposition).

Compared with the general population, patients with ACHD are exposed to a greater risk of complications because they present a higher rate of renal [21], neurological (stroke, neurodevelopmental disorders, cognitive impairment, psychiatric disease, and epilepsy) [22], pulmonary (patients who have undergone prior thoracotomy may have restrictive lung disease, and obstructive sleep apnea is underdiagnosed and undertreated in ACHD), and hepatic [23] disorders. The highest prevalence of liver disease associated with ACHD includes Fontan circulation, Ebstein anomaly, tetralogy of Fallot with severe pulmonary regurgitation, transposition of the great arteries with atrial switch repair, and ES. Furthermore, hematologic issues in ACHD include coagulopathies, iron deficiency, and bleeding disorders.

The advantages of VPA ECMO to limit complications include the possibility of being able to perform peripheral venous cannulation, with the lowest risk of bleeding and vascular complication, in addition to favoring the early mobilization and extubation of patients, limiting the risks of ventilator-associated pneumonia and myopathy of the critically ill patient secondary to the need for sedatives, analgesics and vasopressors for long periods of time. In our particular case, however, the patient's critical condition limited these possibilities of extubation, early mobilization and rehabilitation, secondary to the complications she developed (cardiac tamponade, bleeding from the left main bronchus with total lung atelectasis and hemolytic uremic syndrome), which finally ended up complicating the evolution and limiting the possibility of successful weaning.

The predictors of adverse outcomes of ECMO in patients with ACHD have been primarily studied in postcardiotomy patients. Precannulation risk factors for mortality include Fontan physiology, weight > 100 kg, female sex, delayed cannulation, and neuromuscular blockade. In contrast, postcannulation risk factors for mortality include renal complications, neurological complications, and pulmonary hemorrhage [24].

4. Conclusion

ACHD patients may be candidates for ECMO assistance if they present respiratory and/or circulatory failure refractory to conventional medical management. The VPA approach seems to be an attractive option in this population, as it provides a "protective" strategy for both ventilation and hemodynamics. In addition, in patients with shunts, it is possible to provide oxygenated blood directly into the PA, thereby reducing the venous mixture. Complications should be closely monitored during therapy because they are more prone to development given the higher morbidity of this population.

5. Highlights

- ECMO is an effective therapy for refractory cardiac and/or respiratory failure.

- Adult patients with congenital heart disease have a high rate of morbidity.
- Venopulmonary artery ECMO appears to be a life-saving option in this population.

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Ethics Approval and Consent to Participate

The local institutional research and ethics committees waived approval for this study.

Consent for Publication

The patient or a legally authorized representative provided written informed consent for patient information and images to be published.

Data and Material Availability

The data that support the findings of this study are available on request from the corresponding author (DMS).

Authors' Contributions

DMS: Original idea, image acquisition, analysis and writing the original draft, review and editing. **GMJR:** Original idea, interventional procedure and editing. **EGC:** Image acquisition and analysis. **RES**: Writing the original draft and editing. **EYRM:** Writing the original draft. **YMTL:** Writing the original draft. **JLES:** Editing. **GRV:** Review.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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Abbreviations

ACHD: adult congenital heart disease
ECMO: extracorporeal membrane oxygenation
ES: Eisenmenger syndrome
NYHA: New York Heart Association
PA: pulmonary artery
PASP: pulmonary systolic pressure
QP/QS: ratio of the pulmonary cardiac output to the systemic cardiac output
VA: veno-arterial
VPA: veno-pulmonary artery
VSD: ventricular septal defect
VV: veno-venous