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# Glenn shunt as a rescue strategy for acute right ventricular failure after right ventricular myocardial infarction.

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## Abstract

We present the case of a 52-year-old woman with cardiogenic shock and refractory right ventricular failure due to spontaneous dissection of the right coronary artery. She remained dependent on mechanical support for several weeks. Both a right ventricular assist device implant and a bidirectional cavopulmonary anastomosis were explored as long-term support options. A history of malignancy and possible right ventricular functional recovery resulted in a decision in favour of the bidirectional cavopulmonary anastomosis and concomitant tricuspid valve annuloplasty. Postoperatively her clinical condition improved significantly, and she could be discharged home. Echocardiography showed normalization of right ventricular dimensions and slight improvement of right ventricular function.

**Keywords:** heart failure • right ventricle • cardiogenic shock • Glenn shunt • bidirectional cavopulmonary anastomosis • mechanical circulatory support • RVAD • LifeVest

## ABBREVIATIONS

BCPA	bidirectional cavopulmonary anastomosis
ECMO	extracorporeal membrane oxygenation
HIT	heparin-induced thrombocytopenia
IVC	inferior vena cava
LA	left atrium
LV	left ventricle
PDE-5	phosphodiesterase-5
RA	right atrium
RPA	right pulmonary artery
RV	right ventricle
RVAD	right ventricular assist device
SVC	superior vena cava
VA	veno-arterial
VT	ventricular tachycardia

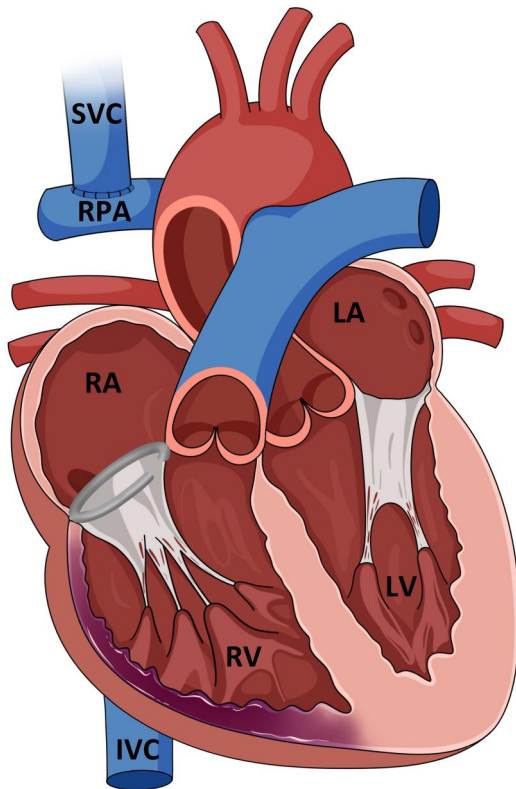
## CASE REPORT

A 52-year-old woman was admitted with acute myocardial infarction due to a spontaneous right coronary artery dissection. Her medical history was notable for a conservatively managed left posterolateral branch myocardial infarction at the age of 39 with normal left ventricular (LV) function. At 51, she was treated for breast cancer with a curative intent.

At presentation, an emergency percutaneous coronary intervention of the right coronary artery was performed. The post-procedural course was complicated by ventricular tachycardia (VT) and fibrillation for which she was successfully resuscitated. During the following hours she developed cardiogenic shock due to failure of the right ventricle (RV) that was refractory to fluid resuscitation and inotropes. She was transferred to the intensive care unit of a tertiary heart failure referral centre and was stabilized using peripheral veno-arterial (VA) extracorporeal membrane oxygenation (ECMO). Echocardiography showed a dilated RV with severely impaired function, paradoxical septal motion with signs of RV volume overload and severe tricuspid regurgitation with elevated filling pressures. The LV function was slightly impaired.

She developed heparin-induced thrombocytopenia (HIT) and was rejected for an urgent heart transplant listing due to the recent oncological history. The VA-ECMO was exchanged for percutaneous veno-pulmonary ECMO in order to allow for recovery of the RV. Five weeks after the start, weaning was initially successful.

However, within days after discontinuation of the ECMO, she developed fast recurrent VTs that were haemodynamically poorly tolerated and resulted in hypotension and elevated lactate levels. Intravenous administration of amiodarone resulted in further progression of RV failure due to the negative inotropic effects and the development of a slow junctional rhythm that was poorly tolerated haemodynamically, limiting further pharmacological options.



**Figure 1.** Schematic overview of the bidirectional cavopulmonary anastomosis in this patient. IVC: inferior vena cava; LA: left atrium; LV: left ventricle; RA: right atrium; RPA: right pulmonary artery; RV: right ventricle; SVC: superior vena cava.

Escalation in the inotropic regimen in the days after ECMO weaning further contributed to the incessant VTs and resulted in a persistent decrease of organ perfusion (INTERMACS profile 2).

After careful consideration and discussion with the electrophysiology team, an ablation procedure was deferred due to the proposed haemodynamic triggers of the VTs and the estimated low chance of a curative procedure. Invasive haemodynamic evaluation ([Supplemental Table 1](#)) revealed a cardiac output of 3.05 L/min (CI 1.61 L/minute/m<sup>2</sup>) and low pulmonary pressures (mean pulmonary arterial pressure 9 mmHg, pulmonary vascular resistance 1.9 WU).

Because of persistent inotropic dependency and recurrent VTs in the setting of progressive RV failure, VA-ECMO was re-initiated, and both a right ventricular assist device (RVAD) and a bidirectional cavopulmonary anastomosis (BCPA) with TV repair were explored. In a BCPA, the superior vena cava (SVC) is disconnected from the right atrium and anastomosed end-to-side to the right pulmonary artery (Fig. 1), redirecting a substantial proportion of the venous return directly to the lungs, bypassing the RV. Due to the recent HIT and the chance of recovery of RV function in the near future, a decision was made to perform a BCPA with a concomitant TV annuloplasty.

Two months after her initial presentation, the patient underwent BCPA and restrictive TV annuloplasty. She had an arterial oxygen saturation of 95% in ambient air and could be weaned off VA-ECMO and inotropes within 3 weeks after the operation. Echocardiography showed normalization of RV dimensions, moderate to severely impaired RV function and trivial TV regurgitation.

Because there was an indication for an implantable cardioverter-defibrillator (ICD) (secondary prevention), a subcutaneous ICD (S-ICD) seemed most appropriate because the BCPA now limited transvenous lead access. However, due to a sternal wound infection, implanting the device was postponed. One month after surgery, she was discharged home with guideline-dictated medical therapy and a LifeVest Wearable Defibrillator (Zoll Medical Corporation, Chelmsford, MA, USA) as a bridge to an S-ICD. The peripheral oxygen saturation at discharge was 99%, and she subsequently completed a cardiac rehabilitation program. Her New York Heart Association functional class IV improved to III (INTERMACS profile 2 to 7) at the 6-month follow-up examination, and there have not been any heart failure-related admissions.

## DISCUSSION

To our best knowledge, we describe for the first time a case of successful rescue using BCPA for refractory RV failure in a 52-year-old patient with acquired heart disease. The concept of unloading the RV in the setting of severely reduced systolic function and the inability to cope with the full venous return and maintaining pulmonary flow and left ventricular preload are important and can provide valuable lessons for managing patients with cardiogenic shock.

BCPA is a technique typically used in patients with staged Fontan palliation of functionally single-ventricle congenital heart disease in whom the SVC is anastomosed end-to-side to the right pulmonary artery, thereby allowing for SVC blood flow to both lungs [1]. This procedure increases systemic oxygen saturation and partially redirects the venous return directly to the pulmonary circulation. Although good outcomes have been reported for the paediatric population, the literature on the utilization of BCPA in adults with acquired heart disease is limited. Several essential pathophysiological issues should be considered. Low LV end-diastolic pressures, low mean pulmonary artery pressure and low pulmonary vascular resistance are imperative for the functioning of a BCPA. The contribution of SVC flow to total venous return in adults is smaller than in young children (around 55% at the age of 2.5 years and 35–40% at 7 years and older) [2]. It is therefore essential to consider the expected haemodynamic effect and to what degree a BCPA may unload an RV in an adult patient. Data on concomitant BCPA in adult patients with Ebstein malformation and right ventricular heart failure demonstrate mid-term survival and beneficiary effects on quality of life, when performed in a selected group of patients who fulfil the above-mentioned criteria [3].

In the direct postoperative period, notable hypoxia can be indicative of elevated mean pulmonary artery pressures and pulmonary vascular resistance. Inhaled nitric oxide and phosphodiesterase-5 (PDE-5) inhibitors (e.g. sildenafil) may be considered in this situation, but there are no robust data on the safety and efficacy of PDE-5 inhibitors in patients with low pulmonary pressures. Given the favourable postoperative course of this patient with improved haemodynamics and normal oxygen saturations at ambient air, adjunctive treatment with vasodilators or PDE-5 inhibitors was not further considered.

No data are currently available on the changes in pulmonary pressures in cases of improvement of RV function and increase in pulsatility of the flow entering through the RV outflow tract.

Obtaining such data requires meticulous follow-up and low threshold invasive haemodynamic evaluation of the patient presented.

A solitary RVAD implant for isolated RV failure is rare, and robust data on outcomes are limited [4]. There have been a few studies with limited patient numbers in which the HeartMate 3 left ventricular assist device (Abbott, Abbott Park, IL, USA) was implanted off-label as an RVAD. Compared to implanting a left ventricular assist device, implanting an RVAD is technically more challenging due to specific anatomical characteristics of the RV, its close relation with the sternum and the limited thickness and enhanced pliability of the RV and right atrial myocardium compared to the LV, which may predispose the patient to malalignment and suction events. Because the RV afterload is generally lower than the LV afterload, the pump typically operates at a lower speed or needs tapering of the outflow graft in order to obtain adequate afterload. In addition, there may be a higher incidence of pump thrombosis [5].

In this case, the patient was initially weaned from temporary mechanical circulatory support because she appeared to be stable based on her clinical course, haemodynamic parameters and echocardiographic findings. However, the recurrent RV failure after weaning necessitated new ECMO support. Because longer stays in the intensive care unit were only expected to increase the patient's frailty and because she had already developed ECMO-related complications, we deemed that her optimal window for clinical recovery.

The first and most important reason for our team to choose the option of BCPA with TV repair as the long-term management strategy for this patient was the reasonable chance that the RV might recover in the not-too-distant future, making the RVAD redundant and stressing the importance of weighted decision making in guarding the patient from VAD-related complications. The second reason for choosing this option was a patient-related factor: She developed a (polymerase chain reaction-confirmed) HIT syndrome, which would have obliged the surgical team to use argatroban for anticoagulation during surgery and during the time it would be necessary to obtain an adequate international normalized ratio, potentially complicating her recovery with haemorrhagic complications. Furthermore, implanting an RVAD would have been a technically challenging procedure, because the patient was very small (160 cm). The final reason to choose the BCPA was the sobering outcome of isolated RVAD implants based on patients in the EUROMACS database, especially in the destination therapy and bridge-to-candidacy population. In our patient, the transplant option was not an option for the next 5 years because of a recent malignancy.

This case illustrates that BCPA might be a good alternative for RVAD in refractory RV failure in selected patients.

## SUPPLEMENTARY MATERIAL

[Supplementary material](#) is available at *EJCTS* online.

## CONFLICTS OF INTEREST

The Department of Cardiology reports receiving unrestricted research and educational grants from Boston Scientific Corporation, Medtronic and Biotronik. The funders were not involved in any part of this manuscript or the procedures described.

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## ETHICS STATEMENT

All procedures performed involving the human participant were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

## CONSENT

The authors confirm that written consent for submission and publication of this case report has been obtained from the patient in line with COPE guidelines.

## DATA AVAILABILITY

No new data were generated or analysed in support of this research.

## Reviewer information

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