

Original Article

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Repeated extracorporeal membrane oxygenation for support of an Adult with Congenital Heart Disease and reperfusion pulmonary oedema

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Abstract

We present the case of 28-year-old woman with a history of complex congenital cardiac surgery who developed cardiovascular collapse with reperfusion pulmonary oedema and right ventricular failure after surgical replacement of a severely stenosed right ventricle to pulmonary artery conduit. She required two separate episodes of support with extracorporeal membrane oxygenation and is alive and well 6 months after her initial cardiorespiratory crisis. We believe that consideration of a second period of support with extracorporeal membrane oxygenation is appropriate for select adults with CHD, provided they have a potentially reversible cause of postoperative cardiorespiratory collapse.

Our case provides several important lessons: (1) adults with CHD with severe postcardiotomy cardiorespiratory failure may potentially be salvaged even if they require multiple runs of extracorporeal membrane oxygenation; (2) adults with CHD with severe postcardiotomy respiratory failure with adequate cardiac function may potentially be salvaged with venovenous extracorporeal membrane oxygenation; and (3) patients supported with extracorporeal membrane oxygenation will benefit from care from a skilled multidisciplinary team who are able to focus on the support of the function of the organs of the patient whilst providing nutrition and mobilisation.

“Never give up, never surrender!”

Commander Peter Quincy Taggart, Galaxy Quest

Case presentation

A 28-year-old woman was born with dextro-transposition of great arteries with ventricular septal defect and subpulmonic stenosis, also known as discordant ventriculo-arterial connections with concordant atrioventricular connections with ventricular septal defect and subpulmonic stenosis. At the age of 3 months, she had a palliative Blalock – Taussig – Thomas shunt. Seven years later, she underwent a Nikaidoh procedure with aortic root translocation over left ventricle, closure of her ventricular septal defect, and Rastelli-type placement of a conduit from her right ventricle to her pulmonary artery.

At the age of 28, she presented to her local hospital with New York Heart Association Grade IV symptomatology with shortness of breath at rest, lower limb oedema, and cachexia. Computerized tomographic (CT) pulmonary angiography demonstrated severe calcification of her right ventricle to pulmonary conduit with severe conduit stenosis and multiple pulmonary emboli.

The patient was transferred to our institution for further evaluation. Upon arrival, the patient had echocardiographic evidence of right-sided heart failure, with severely depressed right ventricular systolic function and suprasystemic right ventricular systolic pressures. During her last follow-up, 6 months previously, right ventricular systolic function was normal. She was resuscitated with infusions of milrinone at 0.5 µg/kg/minute and adrenaline 0.03 µg/kg/minute. Cardiac catheterisation (Fig 1) confirmed the right ventricular hypertension and the conduit stenosis. Transoesophageal echocardiography showed a small patent foramen ovale, severely dilated right cardiac chambers, right ventricular hypertrophy, and severely depressed right ventricular systolic function. Additionally, a right lower lobe pulmonary infarct was identified on computerised axial tomography.

Catheter-based intervention was not felt to be possible due to the severe calcification of the conduit and very narrow residual lumen, which was almost completely obstructed by the

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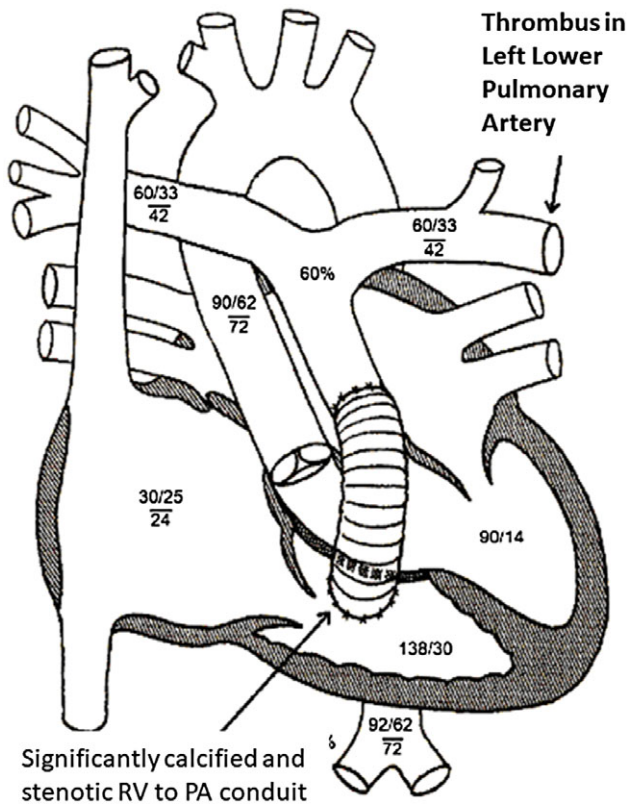


Figure 1. This figure documents the results of cardiac catheterisation. Cardiac catheterisation documented severe stenosis of the conduit connecting the right ventricle to the pulmonary artery, along with suprasystemic right ventricular pressures and elevated pressures in the pulmonary arteries. Thrombus was also visualized in the left lower pulmonary artery.

5 French pigtail catheter during cardiac catheterisation, with resultant haemodynamic compromise. The only remaining treatment option was a high-risk open-heart surgical replacement of the right ventricle to pulmonary artery conduit. Informed consent was obtained from the patient. In view of the haemodynamic compromise, the extent of the previous surgery, and the anteriorly translocated “Lecompte” position of the pulmonary arteries masking the aorta, elective left femoral cardiopulmonary bypass via cut-down was initiated prior to re-entrant sternotomy. The patient was cooled to 25°C. The calcified conduit was replaced with a 28 mm Vascutek tube graft containing a 25 mm Inspiris Resilia pericardial valve, without difficulty.

Shortly after separation from cardiopulmonary bypass, the patient developed some reperfusion pulmonary oedema, but this was stabilised with positive end expiratory pressure of 10cm of water, fraction of inspired oxygen of 60%, and inhaled nitric oxide of 40 parts per million. At the end of cardiopulmonary bypass, the right ventricular pressure was measured by direct puncture and was 5/9 systemic arterial pressure. Immediate postoperative transoesophageal echocardiography revealed right to left shunting across the patent foramen ovale. The patient was returned to the ICU on positive end-expiratory pressure of 10 cm of water for expectant management of the pulmonary oedema and reperfusion injury.

Within a few hours of arrival to the ICU, she became profoundly hypoxic with very poor lung compliance and systemic hypotension. Severe pulmonary oedema was noted on chest radiography (Fig 2) She was hand ventilated with 100% oxygen at high



Figure 2. Chest radiography before the first cannulation for extracorporeal membrane oxygenation demonstrates reperfusion pulmonary oedema.

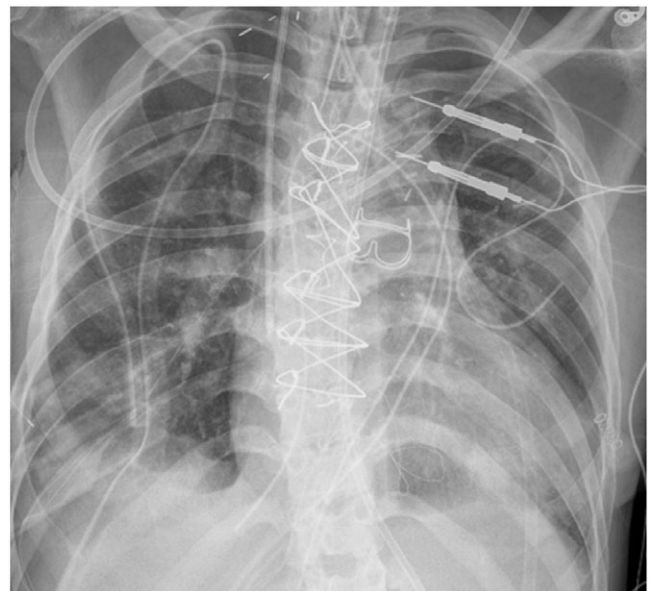


Figure 3. After 4 days of support with veno-arterial extracorporeal membrane oxygenation, our patient was successfully trialed off extracorporeal membrane oxygenation and decannulated. Chest radiography after decannulation demonstrates overall improvement, with substantial resolution of the reperfusion pulmonary oedema.

pressure and emergently cannulated for veno-arterial extracorporeal membrane oxygenation via the left femoral vein and left femoral artery. Prior to emergent cannulation, her peripheral oxygen saturation was 58%, and her mean arterial pressure was 48 mm of mercury. Initial flow was established at 4 L/minute with a sweep gas of 100% oxygen. An anterograde distal perfusion catheter was placed in the left common femoral artery. Her right ventricular function and appearance on chest radiography improved (Fig 3). After 4 days of support with veno-arterial extracorporeal membrane oxygenation, she was successfully trialed

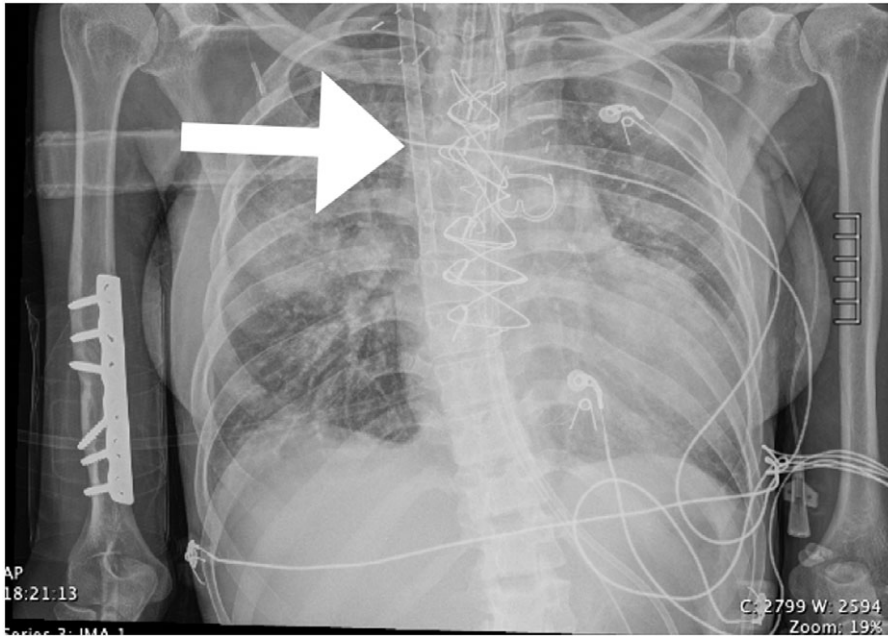


Figure 4. Chest radiography after the second cannulation for support with extracorporeal membrane oxygenation. The white arrow points to the venous drainage cannula in the right internal jugular vein.

off extracorporeal membrane oxygenation and decannulated, and her femoral vessels were reconstructed.

After separation from her first extracorporeal membrane oxygenation run and decannulation, she made good progress initially; however, 1 week after decannulation, she again decompensated and collapsed with hypoxia, acute right ventricular failure, and near cardiac arrest. She was recannulated for veno-arterial extracorporeal membrane oxygenation via the right jugular vein and the left femoral artery proximal to the previous arterial reconstruction (Fig 4). Once flow was established and her physiology was stabilised, a retrograde limb perfusion catheter was inserted via the left posterior tibial artery. She was treated for presumed pneumonia. In a view of the anticipated need for prolonged respiratory support, a tracheostomy was performed on the sixth day of the second period of extracorporeal membrane oxygenation. Transthoracic echocardiography during reduced veno-arterial extracorporeal flow demonstrated excellent right ventricular function. However, attempts to wean the patient completely off of support with veno-arterial extracorporeal membrane oxygenation resulted in inadequate gas exchange despite good lung mechanics; therefore, on the twelfth day of the second period of support with extracorporeal membrane oxygenation, she was converted from veno-arterial extracorporeal membrane oxygenation to veno-venous extracorporeal membrane oxygenation, in order to facilitate early mobilisation and physical therapy.

Her hospital course was complicated by her pre-existing cachexia: her preoperative body mass index was only 17.37 kg/m². Initially, she received total parenteral nutrition, then she was transitioned to nasogastric feeding, and then to normal food. She was mobilised aggressively, walking every day with the supervision of physiotherapy whilst on veno-venous extracorporeal membrane oxygenation (Figs 5 and 6, Video 1). After 36 days of her second period of support with extracorporeal membrane oxygenation, she was successfully decannulated. Her sweep gas was disconnected for the last 3 days of support with extracorporeal membrane oxygenation to ensure that she was ready for decannulation. She continued to improve, her tracheostomy was removed on postoperative day

91, and she was discharged on postoperative day 95 (Fig 7). Six months after discharge from hospital, the patient reports that she has been feeling well and has resumed her normal activities.

Discussion

Our adult with CHD recovered after two runs of extracorporeal membrane oxygenation totalling 40 days:

- ECMO run 1 was 4 days of veno-arterial extracorporeal membrane oxygenation
- ECMO run 2 was 36 days of extracorporeal membrane oxygenation (12 days veno-arterial extracorporeal membrane oxygenation followed by 24 days veno-venous extracorporeal membrane oxygenation)

Successful extracorporeal membrane oxygenation use was first reported in 1972.¹ In 2009, the Conventional Ventilatory Support versus Extracorporeal Membrane Oxygenation for Severe Adult Respiratory Failure trial (CESAR trial)² reported a multicentre randomised controlled trial of adults with severe acute respiratory failure who were randomised to treatment with extracorporeal membrane oxygenation versus maximal conventional ventilatory support and management (e.g., steroids, prone positioning, bronchoscopy, and inhaled nitric oxide). In the CESAR trial, 63% of patients (57/90) allocated to consideration for treatment using extracorporeal membrane oxygenation survived to 6 months without disability compared with 47% (41/87) of those allocated to conventional management. The CESAR trial² demonstrated a statistically significant improvement in survival without severe disability at 6 months for patients in the treatment arm where they were transferred to a single extracorporeal membrane oxygenation centre, compared to patients in the control arm who received continued conventional ventilation in a tertiary intensive care unit.

An analysis of the Extracorporeal Life Support Organization registry of repeated runs of extracorporeal membrane oxygenation documented that “Across all age groups, patients receiving pulmonary support on the last run tended to have higher survival rates

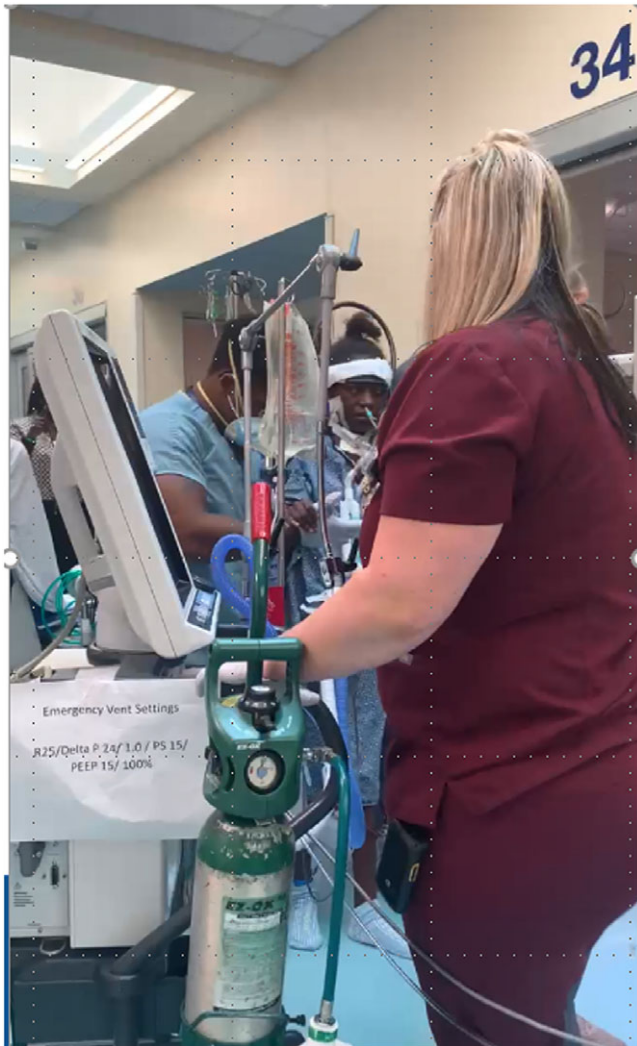


Figure 5. The photograph and the accompanying video (Video 1) show our patient ambulating while supported with veno-venous extracorporeal membrane oxygenation and demonstrate the incredible TEAM effort necessary to facilitate her recovery. (This photograph and video are reproduced with permission of our patient.)

regardless of support type on the first run. The only first run complication independently predictive of mortality on the final run was renal complications (OR: 1.60, 95%CI: 1.28-1.99).³ This analysis of the Extracorporeal Life Support Organization registry concluded that “Though the use of multiple runs of ECMO (extracorporeal membrane oxygenation) is growing, outcomes remain poor for most cohorts. Survival decreases with each additional run. Patients requiring additional runs for a pulmonary indication should be considered prime candidates. Renal complications on the first run significantly increases the risk of mortality on subsequent runs, and as such, careful consideration should be applied in these cases.”

Regardless of these important studies, each patient should be evaluated individually with respect to expected outcomes and allocation of resources. In the presented case, we hypothesised that the acute lung injury was caused by a combination of early reperfusion injury and right ventricular failure secondary to reversible residual pulmonary hypertension. Our case provides several important lessons:



Figure 6. This picture shows our patient on the roof of University of Florida Shands Hospital while supported with veno-venous extracorporeal membrane oxygenation. (This photograph is reproduced with permission of our patient.)

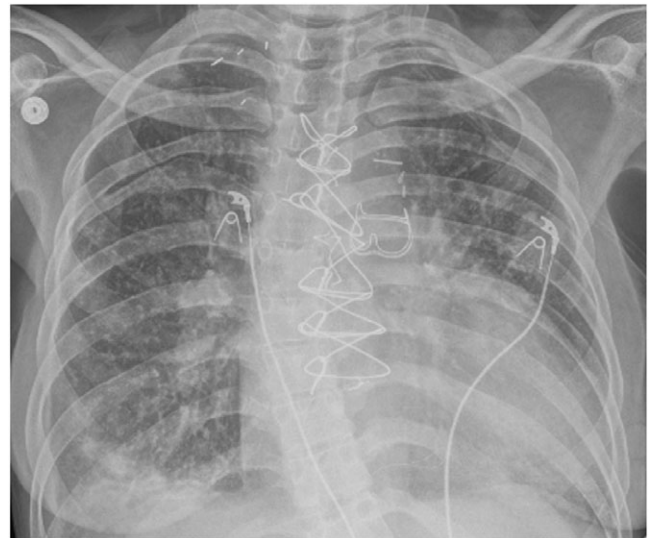


Figure 7. This final chest radiograph before discharge demonstrates improvement of the patchy bilateral alveolar opacities.

- Adults with CHD with severe postcardiotomy cardiorespiratory failure may potentially be salvaged even if they require multiple runs of extracorporeal membrane oxygenation.

- Adults with CHD with severe postcardiotomy respiratory failure with adequate cardiac function may potentially be salvaged with veno-venous extracorporeal membrane oxygenation.
- Patients supported with extracorporeal membrane oxygenation will benefit from care from a skilled multidisciplinary team who are able to focus on the support of the function of the organs of the patient whilst providing nutrition and mobilisation.

Conclusion

A second run of support with extracorporeal membrane oxygenation may be appropriate for patients who have a potentially reversible cause of cardiorespiratory collapse. However, in order for this strategy to be successful, a second run of extracorporeal membrane oxygenation requires a skilled multidisciplinary team who are able to focus on the support of the function of the organs of the patient whilst providing nutrition and mobilisation.

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Conflicts of interest. None.

Ethical standards. The authors assert that all procedures contributing to this work comply with the ethical standards of the relevant national guidelines on human experimentation and with the Helsinki Declaration of 1975, as revised in 2008.

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