

# The Effect of Extracorporeal Membrane Oxygenation in the Management of Refractory Ventricular Tachycardia Developed after Fontan Procedure (Case Report)

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**For citation:** Olga S. Anikina, Ilya A. Soynov, Ilya A. Velyukhanov, Olga A. Suzdalova, Yuri Yu. Kulyabin, Stanislav A. Sergeev, Alexey N. Arkhipov, Igor A. Kornilov. The Effect of Extracorporeal Membrane Oxygenation in the Management of Refractory Ventricular Tachycardia Developed after Fontan Procedure. *Obshchaya Reanimatologiya = General Reanimatology*. 2024; 20 (5): 77–80. <https://doi.org/10.15360/1813-9779-2024-5-77-80> [In Russ. and Engl.]

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## Summary

**Aim:** to evaluate the effect of extracorporeal membrane oxygenation (ECMO) as a life support in the treatment of a patient with refractory ventricular tachycardia developed after Fontan procedure.

**Patient and treatment.** A 4-year-old child developed refractory ventricular tachycardia (up to 250 bpm) and hemodynamic depression 18 hours after the Fontan procedure. After the failure of cardiopulmonary resuscitation and antiarrhythmic therapy, resectionotomy with central venoarterial (VA) ECMO support was performed, followed by diagnostic angiocardiology. Contrast-enhanced cavopulmonary angiography revealed stenosis of the left pulmonary artery, which was treated with balloon angioplasty and stenting.

**Results.** Ventricular tachycardia resolved and sinus rhythm was restored within 24 hours after left pulmonary artery stenting, supported by continuous ECMO and antiarrhythmic therapy. On day 3, transthoracic echocardiography showed good single ventricle contractility after a trial weaning from ECMO. As a result, the ECMO support was removed and the sternum sutured. The patient was discharged from the hospital on day 47 in stable condition.

**Conclusion.** The prompt initiation of VA ECMO support in a 4-year old patient with refractory ventricular tachycardia post-Fontan procedure along with the complex management of post-procedural residual tachycardia using a combination of antiarrhythmic agents helped restoring sinus rhythm and could contribute to preventing neurological complications.

**Keywords:** ECMO; refractory ventricular tachycardia in children; Fontan procedure

**Conflict of interest.** The authors declare no conflict of interest.

**Funding.** This study was performed under the State Contract No. 124022500251-0 of the Russian Ministry of Health.

## Introduction

Refractory ventricular tachycardia (VT) is a rare and life-threatening complication after cardiac surgery [1]. The main causes of postoperative refractory VT in children with congenital heart disease include ineffective myocardial protection and residual lesions after cardiac surgery [2, 3]. The only emergency treatment option is the use of venoarterial extracorporeal membrane oxygenation (ECMO), which can serve as a bridge to either recovery or heart transplantation [1, 2].

Here we present a clinical case of sinus rhythm restoration as a result of effective use of venoarterial ECMO for refractory ventricular tachycardia in a patient 4 years after cardiac surgery.

The aim was to evaluate the outcome of a patient with refractory ventricular tachycardia receiving ECMO.

## Clinical Report

A 4-year-old patient with a body weight of 16 kg was admitted to E. N. Meshalkin Research

and Medical Center for Fontan's surgery with the initial diagnosis of congenital heart disease: hypoplastic left heart syndrome. Echocardiography showed normal contractility of the systemic ventricle, bidirectional cavopulmonary anastomosis without deformation, interatrial communication of 3.4 cm, accelerated flow in the neo-aorta. A stent was identified in the descending aorta; peak gradient at this level was 54 mmHg. Significant tricuspid regurgitation and a vena contracta width of 0.79 cm were noted.

The child underwent a complete extracardiac cavopulmonary anastomosis with a Gore-Tex 18 mm vascular graft (GORE-TEX® Vascular Grafts, W. L. Gore & Associates, Inc, Flagstaff, AZ, USA) with a 4 mm fenestration, we performed pulmonary artery branch repair, aortic arch repair with a Vascutek vascular prosthesis flap (Terumo, Renfrewshire, United Kingdom), De Vega suture angioplasty of the tricuspid valve. Aortic arch reconstruction was performed under antegrade cerebral perfusion (total time 43 minutes). Cardioplegic solution Custodiol (650 ml Custodiol, Dr. Franz Köhler Chemie GmbH, Ger-

many) was used for myocardial protection. There was spontaneous recovery of cardiac activity. Cavopulmonary anastomosis was performed under parallel cardiopulmonary bypass (CPB) for 189 minutes. Weaning from CPB was performed with the infusion of minimal doses of norepinephrine and epinephrine without cardiac arrhythmias. Intraoperative transesophageal echocardiography (TEE) demonstrated a functional cavopulmonary anastomosis without accelerated blood flow. Single ventricle contractility was normal (ejection fraction 58%).

18 hours after surgery, the child developed ventricular tachycardia with ventricular rate up to 250 beats/min and hemodynamic compromise. Cardiopulmonary resuscitation was performed with multiple electrical defibrillations at 5-minute intervals.

Antiarrhythmic therapy with a triple bolus injection of amiodarone 5 mg/kg followed by infusion at 5 mg/kg/h and a double bolus of lidocaine 1 mg/kg was administered without beneficial effect. Blood electrolytes were within normal ranges. At the time of resuscitation, the inotropic index was 34 points with high doses of dopamine, epinephrine, norepinephrine, and phenylephrine. During refractory ventricular tachycardia, echocardiography was performed and showed reduced contractility of the single ventricle. During cardiopulmonary resuscitation, resection with central veno-arterial extracorporeal membrane oxygenation (ECMO) was performed, followed by diagnostic angiography. A cavopulmonary contrast study revealed a left pulmonary artery stenosis (Fig., *a*), and balloon angioplasty was

performed with a CP Stent L 34–45 mm (NuMED Inc, Hopkinton, New York, USA) (Fig., *b*). Coronary angiography showed no deformation or abnormal compression of the coronary arteries.

## Results

After 24 hours on ECMO and continuous infusion of amiodarone 5 mg/kg/h, sinus rhythm was restored. No arrhythmias were observed for the next 3 days. On day 3, VA-ECMO was discontinued and transthoracic echocardiography showed good contractility of the single ventricle. The patient was then weaned from ECMO and the chest was sutured. The total ICU stay was 10 days and no neurological abnormalities were noted in the child. The patient was discharged from the hospital on day 47 in stable condition.

## Discussion

The conventional approach to the treatment of pediatric VT is medical therapy [4]. However, VT refractory to medical therapy and triple ineffective defibrillation often leads to hemodynamic compromise and the need for cardiopulmonary resuscitation [1–3]. In such cases, the only emergency treatment option is ECMO [2]. The causes of refractory VT or ventricular fibrillation include ineffective myocardial protection, electrolyte disorders, and congenital heart rhythm disorders such as long QT syndrome, short QT syndrome, and Brugada syndrome, as well as residual lesions after cardiac surgery [2, 5]. In our case, electrolyte disturbances and myocardial ischemia (absence of ST-segment elevation in the early postoperative period

and normal recovery of sinus rhythm after surgery) were excluded. Veno-arterial ECMO with central cannulation was initiated during cardiopulmonary resuscitation to stabilize hemodynamics and to identify further causes of arrhythmias. According to the ECMO protocol used in our center, the diagnosis of residual lesions is performed after cardiac surgery [5].

Echocardiography is the gold standard for ruling out residual lesions [6], but in single ventricle patients on ECMO, diagnosis can be challenging and invasive diagnostic modalities are more effective [7]. Additional tests, such as computed tomography or

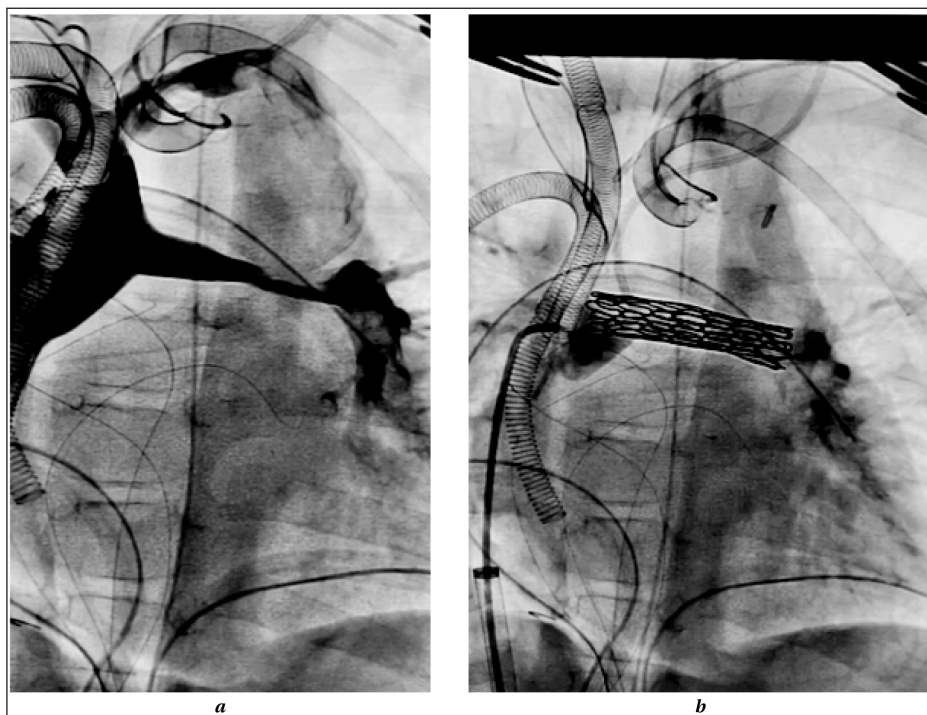


Fig. Cardiac catheterization in a patient on VA-ECMO: stenosis (*a*) and stenting (*b*) of the left pulmonary artery.

cardiac catheterization, can identify the causes of arrhythmias and the need for veno-arterial ECMO [5, 8]. Cardiac catheterization revealed an extensive stenosis of the left pulmonary artery and compression by the aorta.

After stenting of the left pulmonary artery within 24 hours, the ventricular tachycardia resolved and sinus rhythm was restored.

ECMO helped to maintain stable hemodynamics throughout its administration and to identify the cause of the residual lesion. Once the abnormal conditions have been corrected in patients with a single ventricle heart, the time to weaning from ECMO depends on several factors, such as the duration of cardiopulmonary resuscitation, global systolic and diastolic ventricular function, and the presence of ECMO-related complications [9,10]. The use of a pre-conditioned ECMO circuit shortened the duration of

cardiopulmonary resuscitation and reduced the risk of neurological complications. Single ventricle ejection fraction was restored immediately after control of the rhythm disturbance, and careful surgical hemostasis helped to avoid ECMO-related complications. The implemented action algorithm allowed to discontinue venoarterial ECMO after 56 hours and to discharge the patient in stable condition in sinus rhythm and without neurological deficit.

## Conclusion

The timely initiation of venoarterial ECMO in the treatment of refractory ventricular tachycardia, together with detection and elimination of postoperative residual lesions and combined drug antiarrhythmic therapy, allows restoration of normal heart rhythm and helps to avoid neurological complications.

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Received 29.05.2023  
Accepted 03.09.2024