

How to Manage an Arterio-Venous Extracorporeal Membrane Oxygenator Establishment in Case of Persistent Left Vena Cava Superior?

Persistan Sol Superior Vena Cava Varlığında Arteriyo- Venöz Ekstrakorporeal Membran Oksijenatör Kurulumu Nasıl Yönetilmelidir?

Hande İSTAR

Mugla Sıtkı Kocman University Faculty of Medicine, Department of Cardiovascular Surgery, Mugla, Turkey

Öz

Ekstrakorporeal Membran Oksijenatörü (ECMO), yenidoğan ve infantlarda vasküler yapıların ince oluşu nedeniyle sıklıkla v.jugularis interna ve ana karotid arter yoluyla hastaya bağlanmaktadır. Olgu sunumumuzda sağ v.cava superior'u ince kalibrasyonda olan konjestif kalp yetmezliği ve Ellis Van-Crevelde Sendromu tanımlı hastada, daha geniş çaplı persistan sol superior vena cava (PSSVC) yoluyla ECMO uygulaması sunulmaktadır.

Anahtar Kelimeler: ECMO, Ellis Van-Crevelde Sendromu, Persistan Sol Superior Vena Cava

Abstract

Extracorporeal membrane oxygenator (ECMO) can be established via v.jugularis interna and carotid artery in neonates and infants because of inadequate calibration of the other peripheral vessels. We aimed to introduce the ECMO establishment via persistent left v. cava superior in an infant patient whose right v.cava superior was inadequate in size.

Keywords: ECMO, Ellis Van-Crevelde Syndrome, Persistent Left Vena Cava Superior

Introduction

Extracorporeal membrane oxygenator (ECMO) can be established via v. jugularis interna, carotid artery in neonates and infants because of inadequate calibration of the other peripheral vessels. We aimed to introduce the ECMO establishment via persistent left v. cava superior (VCS) in an infant suffering congestive heart failure (CHF) and Ellis Van-Crevelde Syndrome, in case of inadequate size of right VCS.

Case

A four-month-old, 3.2 kg in weight female patient was admitted to our hospital because of CHF. She was hospitalized in the pediatric intensive care unit and intubated. Inotropic medical treatments were given. Echocardiogram evaluated CHF with an ejection fraction 40%, single atrium, complete atrioventricular septal defect and bilateral VCS. Atrioventricular valves had a severe incompetence. Pulmonary artery pressure was measured as 65 mmHg. Right and left VCS were 1.2 mm and 3.2 mm in diameter respectively. She had Ellis Van-Crevelde Syndrome; polydactyly in hands, abnormal toes of feet were detected (Figure 1C, 1D). Left VCS was

draining to the coronary sinus. Despite inotropic agents, cardiac decompensation occurred in 5 days. Before the decision of surgery, we decided to establish arterio-venous (AV) ECMO for life support. Informed consent was obtained from the patient's relatives in 2016.

Femoral veins were less than 1 mm in Doppler ultrasound investigation.

Under general anesthesia, with a cut-down method on the left side of the neck, left main carotid artery (LMCA), PLVCS were explored and cannulated with 8 F and 10 F cannulas (Medtronic) for the arterial and venous cannulations respectively. ECMO establishment was completed. To have a better venous return, the cannula was advanced forward to the coronary sinus (Figure 1A, 1B) following the measurement of the coronary sinus which showed an adequate diameter with echocardiogram. Coronary sinus damage was eliminated by screening with echocardiogram again, the venous return was adequate. Under ECMO support, we could diminish doses of inotropic agents in 10 days; however, thrombocytopenia occurred due to the destroying effect of ECMO circulation. Because of complications of massive transfusion, bleeding and bacterial sepsis, we lost our patient.

Discussion

ECMO is being widely used in case of congenital heart disease (CHD) and severe respiratory failure (1). PLVCS is frequently associated with CHD (2). PLVCS is found to be problematic in case of cardiac surgery while the cannulation process of cardiopulmonary bypass or insertion of pacemaker leads. Frequently, PLVCS is smaller in diameter

Hande İSTAR ORCID No
0000-0002-7150-0171

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Adres / Correspondence : Hande İSTAR
Mugla Sıtkı Kocman University Faculty of Medicine, Department
of Cardiovascular Surgery, Mugla, Turkey
e-posta / e-mail : handeistar@yahoo.com

rather than right VCS in CHD associations. In literature, right VCS agenesis in a sporadic manner (2).



Figure 1. A: ECMO establishment via left main carotid artery and PLVCS. B: X-Ray view of arterial and venous cannulas following the ECMO establishment. C: Polydactyly of Ellis Van-Creveld Syndrome in hands. D: Structural abnormalities in feet about Ellis Van-Creveld Syndrome.

Even though our patient was 4-month-old, she was 3.2 kg in weight. Moreover, she had a hypoplastic right VCS. Before ECMO cannulation, bilateral VCS existence, their diameters should be evaluated by Doppler ultrasound.

We believe that in case of low weight infant who requires ECMO support if PLVCS is larger than the right side, it is safe to perform the venous cannulation. However, the position of the venous cannula should be checked by echocardiogram and X-Ray particularly for possible rupture of coronary sinus and position of cannulas.

Written consent: Written consents of the patients were obtained on 10.10.2021.

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