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Case Report

Extracorporeal Membrane Oxygenation in a Heterotaxy Patient Presenting with Cardiogenic Shock

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Abstract

26-year-old male not known to have any previous medical history presented with picture of cardiogenic shock to emergency department. The patient underwent emergency VA ECMO for 4 days. Investigations showed Large Atrial Septal Defect (ASD) and Interrupted Inferior Vena Cava (IVC) and left atrial isomerism. Two months later the patient underwent successful surgical repair of his ASD. He was discharged home in stable conditions.

Keywords: Cardiogenic shock; Extracorporeal Membrane Oxygenation (ECMO); Heterotaxy; Interrupted inferior vena cava

Case Report

A 26-year-old male recently immigrated to Canada, presented to the emergency department complaining of a 2-day history of increasing abdominal pain, hematemesis, he also complained of decrease exercise capacity over last 3 months. He was found to be in right sided heart failure with ascites. Chest radiograph showed significant cardiomegaly (Figure 1A). Later on, the patient developed uncontrolled atrial fibrillation and became hemodynamically unstable requiring electrical cardioversion. After cardioversion, the patient continued to have hemodynamic instability requiring high doses of inotropes. Echocardiogram showed severe biventricular failure appearance suggestive of noncompaction, an enlarged right atrium and severely dilated right ventricle. Moderate Tricuspid Regurgitation, Moderate Mitral Regurgitation.

The patient continued to show signs of cardiogenic shock with anuria and elevation in lactate despite the high dose of inotropic support. The decision was to manage the patient with mechanical circulatory support. Transesophageal Echocardiogram (TEE) echo showed severe biventricular failure, Left Ventricular Ejection Fraction < 10%, large atrial septal defect with a left to right shunt and evidence of pulmonary hypertension (Figure 1C). Heparin was given, activated clotting time was 250s, ECMO cannulation was performed percutaneously using a Seldinger technique in the right femoral vessels, Cannula sizes: R arterial: 17 French(F), right venous 19(F). (Bio Medicus-Medtronic).

The positions of the cannulas were confirmed with TEE. The cannula was connected to the ECMO machine, achieving adequate flows of 4L/min. on TEE there was evidence of right ventricular decompression and mild improvement in the RV systolic function after initiation of ECMO. The patient stayed 8 days in the cardiovascular surgery intensive care unit. The patient showed signs of improvement and was successfully weaned of inotropes and ECMO on the 4th day. The patient was fully investigated during his hospitalization. TEE showed improvement of heart function ejection fraction improved to 52%. Cardiac MRI showed biventricular dilation with preserved ventricular function, pulmonary and systemic venous return to the heart appeared conventional. Atrial situs is normal, interrupted IVC with azygous continuation on the right, a small spleen (Figure 1B).

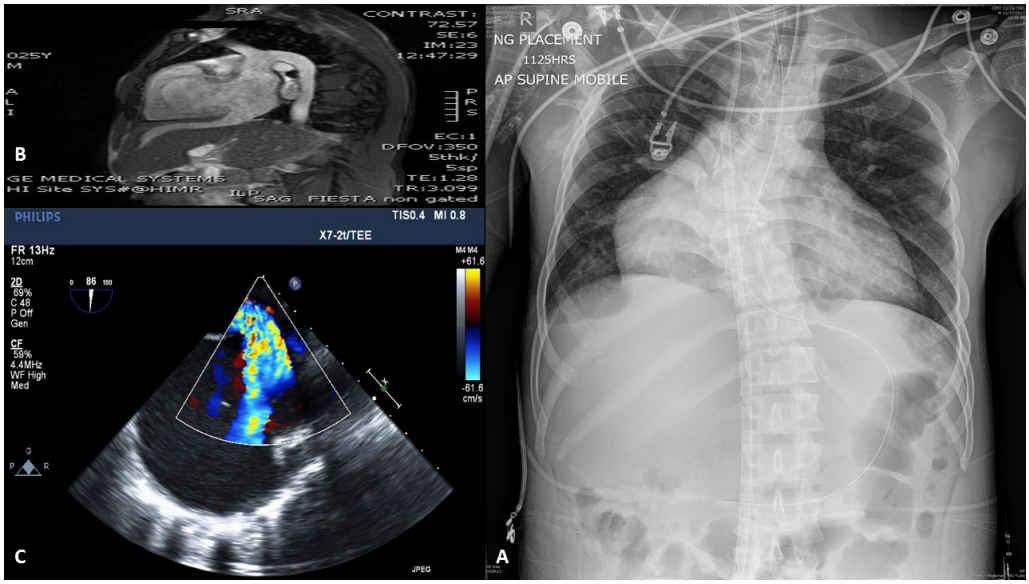

Figure 1: 1A-Chest Xray post ECMO insertion, 1B-MRI showing interrupted IVC, 1C-TEE showing ASD.

The patient had a good clinical course and was discharged home in stable condition. Two months later, he underwent ASD closure using a bovine pericardial patch and a De Vega tricuspid valve annuloplasty, intra operative findings showed 2 fingers like right atrial appendages confirming the diagnosis of right atrial isomerism. An AAI single chamber pacemaker was implanted because of persistent requirement of temporary pacing on a background of rhythm alternating between junctional bradycardia, sinus pauses, rapid atrial fibrillation and sick sinus syndrome. Patient was discharged home in stable conditions.

Discussion

We presented a case of refractory cardiogenic shock secondary to biventricular failure in an adult patient who was subsequently found to have a large ASD. It is part of the natural history of ASDs to cause right sided heart failure in older patients usually in the context of significant tricuspid regurgitation secondary to severe right heart and tricuspid annular dilation and often with coexistent pulmonary arterial hypertension [1]. Left ventricular failure on the other hand has been described as a possible complication of ASD closure particularly in patients with diastolic dysfunction because of the acute volume loading after closure.

Although Veno-arterial ECMO is frequently used in adults with refractory cardiogenic shock for recovery or as a bridge to VAD or heart transplantation, it is less commonly used in patients with Adult congenital heart disease presenting with cardiogenic shock. Reports in the literature of the utilization of ECMO in children and adults with congenital heart disease as a bridge to surgery are limited [2]. Uilkema et al. [3] presented two patients with adult congenital heart disease requiring veno-arterial ECMO as a bridge to surgery. It was indicated for right sided heart failure in one patient and a failing Fontan circulation in the other. In a more recent publication Maybauer et al. [4] presented a case series of four patients with adult congenital heart disease requiring veno-arterial ECMO, only one of the four patients were placed on ECMO for cardiogenic shock as a bridge to surgery where he underwent replacement of his failing tricuspid valve prosthesis. This was originally implanted for a diagnosis of Ebstein's anomaly.

Heterotaxia is characterized by poor or no differentiation between the left- and right-sided organs, with resulting congenital malformations of multiple organ systems. Polysplenia syndrome (left atrial isomerism) is characterized by multiple splenic tissues, absence of the sinus node [5], in our case we inserted An AAI single chamber pacemaker because of persistent requirement of temporary pacing on a background of rhythm alternating between junctional bradycardia, sinus pauses, rapid atrial fibrillation and sick sinus syndrome. The most significant differential point is the Inferior Vena Cava (IVC), which is mostly normal in patients with asplenia syndrome but is interrupted (with azygos continuation) in 85% of polysplenia patients [5].

Given the acuity of our patient's presentation, he was placed on ECMO before making a diagnosis of heterotaxy or having abdominal imaging to rule out an interrupted IVC. During the procedure TEE was used to guide cannula position. The azygous was mistaken for an inferior vena cava. The patient was fortunate that the venous cannula did not injure his liver or perforate the inferior vena cava but rather made coursed to the azygous continuation. We advise that in patients with adult congenital heart

disease requiring veno arterial ECMO, abdominal imaging could be helpful prior to peripheral cannulation to rule out such vascular anomalies. Similar to previous reports [3,4], we found that veno-arterial ECMO is a life-saving intervention in patients with adult congenital heart disease presenting with cardiogenic shock.

Conflict of Interest

The authors declare that there is no conflict of interest regarding the publication of this paper.

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