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Case report

ROLE OF EXTRACORPOREAL MEMBRANE OXYGENATION IN PEDIATRIC MYOCARDITIS

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ABSTRACT

Fulminant myocarditis in children is rare, but it has the potential for high morbidity and mortality. In this report, we review articles demonstrating the role of venoarterial extracorporeal membrane oxygenation (VA-ECMO) in the fulminant myocarditis, and describe the clinical course of a healthy 11-year-old girl who rapidly deteriorated into cardiogenic shock. The patient was weaned from mechanical ventilation on day 7 and she was discharged from the ICU on day 12. VA-ECMO is considered as a life-saving therapy in patients with hemodynamic instability prior to recovery or other escalation therapy.

I. INTRODUCTION

Myocarditis is an inflammatory disease of the myocardium with various etiologies, including viruses, bacteria, toxins, and systemic disorders [1]. Affected patients can manifest a broad clinical spectrum of signs and symptoms, ranging from subclinical disease to cardiogenic shock, arrhythmias, and death. The severe form of myocarditis that necessitates aggressive and timely treatment to prevent rapid hemodynamic collapse, is termed acute fulminant myocarditis (AFM). Although an estimated annual incidence of myocarditis is only 1 to 2 per 100000 children [2-6], the diagnosis can be difficult due to the nonspecific symptoms and the lack of a sufficiently sensitive and specific diagnostic test for myocarditis. The prognosis of pediatric patients depends on the severity of clinical manifestations. In fact, myocarditis patients developing cardiogenic shock refractory to optimal medical treatment may benefit from the use of veno-arterial extracorporeal membrane oxygenation (VA-ECMO) as a life-saving method. The aim of this report is to review articles demonstrating the role of VA-ECMO in the specific context of cardiogenic shock due to myocarditis in children and to present a case study of fulminant myocarditis in Hue Central Hospital.

II. CASE PRESENTATION

A previously healthy 11-year-old girl was admitted to Hue Central Hospital, with a one-day history of chest pain. Upon arrival at our emergency department, the patient complained of continuous pain located behind her sternum, accompanied by a burning sensation. She denied any other symptoms including dyspnea, fever, or palpitations. Her vital signs were as follows: blood pressure 108/61 mmHg, heart rate 108 bpm, respiratory rate 23 rpm. Initial laboratory tests were significant for Troponin I of 12,34 ng/ml (normal < 0,015), pro BNP of 4033 pg/ml (normal < 125). Also, an emergency electrocardiogram revealed ST - segment depression in inferior (II, III, aVF) and anterolateral leads (V4, V5, V6) (Figure 1). Our initial treatment for this patient included furosemide and Enalapril. One day after admission, the patient suddenly lost palpable pulses and became unresponsive, with the monitor showing a sinus rhythm of ventricular tachycardia at a rate of 239 beats per minute (Figure 2). After intubating the patient, we performed cardiopulmonary resuscitation, immediate defibrillation (80J and 160J), and administrated Lidocaine, Amiodarone.

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Bedside ultrasound showed a decreasing Left Ventricular Ejection Fraction (31%). Subsequently, the ventricular tachycardia attack was non-responsive to medical treatment and a decision was made to institute veno-arterial extracorporeal membrane oxygenation (VA-ECMO).

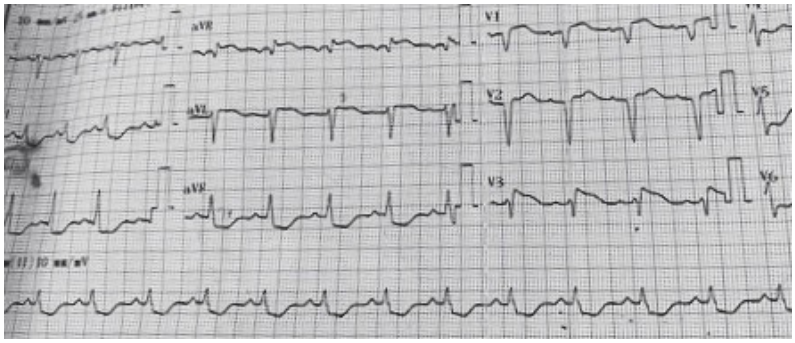

Figure 1: ECG at admission

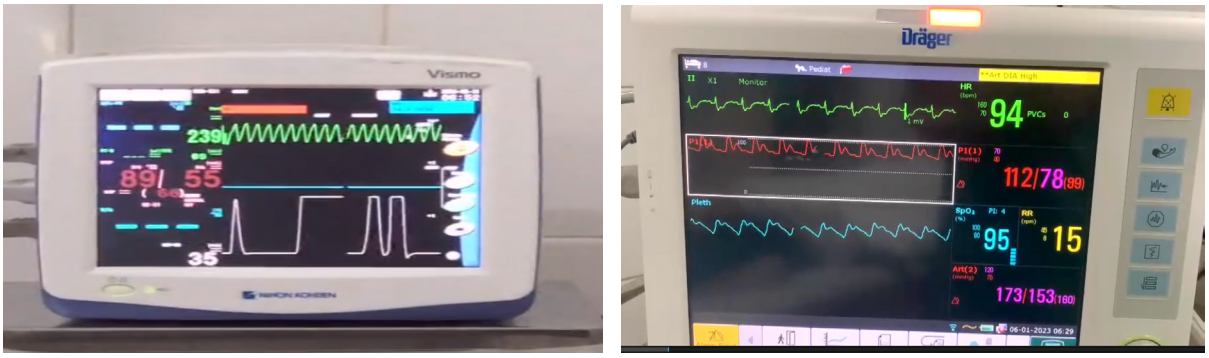

Figure 2: Ventricular Tachycardia on day 2 of admission, disappeared on day 3 of hospitalization

Figure 3: Monitoring after decannulation of VA-ECMO

The patient underwent peripheral ECMO cannulation (Left femoral vein, left common femoral artery, and distal perfusion catheter). The ECMO circuit included TERUMO Cardiovascular Systems, Tokyo, Japan. Due to the support of VA- ECMO (2000 rpm, Blood Flow 2400 ml/min, sweep gas: 2 l/p, FiO2: 60%) with standard heparin for ECMO, the patient's circulation stabilized (Dobutamin 5 mcg/kg/min). However, refractory ventricular tachycardia continued after placing on ECMO, so we steadily increased the dose of both Lidocaine and Amiodarone infusion to 30 mcg/kg/min and 15 mcg/kg/min respectively. In addition, we also used intravenous immunoglobulin (IVIG) in the

treatment of this patient. On day 3 of hospitalisation, VT disappeared (Figure 3) and the patient's circulation improved. After 5 days, we decided to discontinue vasoactive drugs, antiarrhythmic drugs, and lowered the ECMO parameters as much as possible (Blood flow: 1,5 l/min, Sweep gas: 1 l/min, FiO2: 50%). The echocardiogram showed the increasing Left Ventricular Ejection Fraction (50%), so weaning from ECMO was attempted. Therefore, the patient was decannulated, with the support of VA-ECMO for 5 days (Figure 4). The patient ứa weaned from mechanical ventilation on day 7 and she was discharged from the ICU on day 12. However, one month later, follow-up revealed arterial thrombosis at the site of cannulation. Therefore, surgical revascularization was performed and anticoagulant therapy with Acenocoumarol was initiated in preventing another thrombosis.

III. DISCUSSION

Myocarditis is characterized by myocardial inflammation, injury or necrosis, and ultimately fibrosis [1]. Myocyte injury, lysis and myocardial necrosis sequently lead to cardiac enlargement and diminished systolic function, which cause the typical signs of congestive heart failure, atrial or ventricular arrhythmias and sudden death.

The etiology of myocarditis includes viruses, bacteria, toxins and immune disorders, in which viral myocarditis is the most common causative agents [1].

The true incidence of viral myocarditis is unknown because some affected children may have subclinical disease and be undetected.

The pathophysiological sequence of monocyte injury in acute myocarditis occurs in 2 separate phases [7]. During the first phase, viral infection phase is often reported as a prodromal presentation of fever, myalgia, respiratory and gastrointestinal symptoms several days prior to the onset of symptoms of heart dysfunction [2, 8, 9]. Subsequently the agents trigger the host immune system, resulting in T cell and cytokine activation. Ultimately, this leads to myocyte injury and impaired ventricular function.

Primary therapy for acute myocarditis is supportive, including Beta-blockers and ACE inhibitors. Acutely, the use of inotropic agents and diuretics should be considered. Mechanical ventilatory support may be necessary in extreme cases to augment cardiac output by reducing the work of breathing and left ventricular afterload. However, when patients are not adequately responding to maximal medical therapy, ECMO can be employed as a bridge to recovery, or a bridge to transplant [10, 11].

Although fulminant myocarditis is relatively uncommon [1, 4, 6, 8, 10] and only known to be present in 10-15% patients with myocarditis [12], the high mortality rate of the disease has been reported. Therefore, ECMO has become a life-saving therapy until palliative or corrective intervention. Currently, some selected publications showed the survival rate of 50% for AFM patients to hospital discharge, which has remained consistent over decades in spite of advances in mechanical support [13].

In the world, the number of neonatal and pediatric VA-ECMO runs annually increases; howerver, the survival rate has not changed clearly (Chart 1).

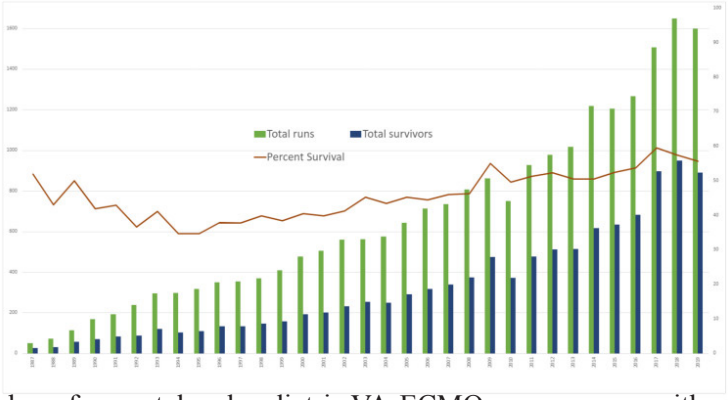

Figure 4: Number of neonatal and pediatric VA-ECMO runs per year with percent survival (Updated from ELSO) [14].

In Vietnam, according to a study at Vietnam National Chidren's Hospital, in the 5-year period from 2016 to 2021, there are 37/54 patients with acute myocarditis received ECMO, in which the survivors acounted for 68% [15].

Some common indications for VA-ECMO consist of cardiogenic shock with high inotropic requirements, dysarrhythias, cardiac arrest (Table 1).

Table 1: Indications for ECMO [14]

Indications of VA-ECMO
Cardiogenic shock with high inotropic requirement
Dysrhythmia Multiple ventricular premature complexes Ventricular tachyarrhythmia Ventricular bradyarrhythmia Atrioventricular block
Cardiac arrest (Post-CPR, E-CPR)

Some researches on using VA-ECMO in myocarditis over the period of 10 years showed that the number of children received ECMO was different in many centers. This depends on the clinical circumstances and experience in these centers. Although, in the severe patients requiring ECMO support, the survival rate was reported with over 60%, leading to the increase in the overall survival rate of myocarditis [16].

Indicating timely in supporting VA-ECMO in myocarditis patients is still a challenge in clinical practice. In order to stabilise hemodynamics, organ perfusion; mean arterial pressure (MAP), vasoactive-inotropic score (VIS), LVEF, or creatinine level are often considered as factors for supporting VA-ECMO and preditors of in-hospital mortality during treatment [17-20].

The VIS was calculated as follows: dopamine dose (mcg/kg/min) + dobutamine dose (mcg/kg/min) + 100 × epinephrine dose (mcg/kg/min) +10 × milrinone dose (mcg/kg/min) + 10.000 × vasopressin dose (unit/kg/min) + 100 × norepinephrine dose (mcg/kg/min) [21].

A decrease in VIS and an increase in LVEF at the 24th and 48th hour post to ECMO are associated with survival rate in acute fulminant myocarditis, while a rise in creatinine level is considered as a predictor of mortality [18], [20, 22].

Though there is no convincing data demonstrating a clear survival benefit of IVIG in myocarditis [23, 24], IVIG used to limit the immune reponse is still considered a potential therapeutic strategy. In this case, we also prescribed IVIG for the patient.

In this case report, ECMO played a crucial role in stabilising the patient's hemodynamics and ensuring adequate organ perfusions.

IV. CONCLUSION

Myocarditis is rare in children and the diagnosis can be difficult to establish given the nonspecific symptoms and lack of a sufficiently sensitive and specific diagnostic test for myocarditis. In addition, the mortality rate of acute fulminate myocarditis is still high. Therefore, VA-ECMO is considered as a life-saving therapy in patients with hemodynamic instability prior to recovery or other escalation therapy.

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