CASE REPORTS

HYPOVOLEMIC SHOCK AND THE NEED FOR INVASIVE MECHANICAL VENTILATION ON A PATIENT WITH CONGENITAL HEART DISEASE

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Abstract

Techniques of venoarterial-extracorporeal membrane oxygenation (VA-ECMO) have improved over the decades, with numerous applications.1 Those with reversible low cardiac output benefit most from this support.[1]

Case of 21-year-old male, history of congenital heart disease (severe right ventricle hypoplasia and pulmonary artery stenosis with extracardiac cavo-pulmonary shunt (Fontan surgery), and atrial septal defect). Brought to the Emergency Department due to a Mallory-Weiss syndrome, upper-endoscopy "laceration at esophagogastric junction (EGJ) with active bleeding", clips were applied. However, worsening shock, repeated melenas and hematemesis, hemoglobin drop, lactate 2.8mmol/L, and needing noradrenaline (1.21mcg/kg/min). Due to active blood loss and worsening shock, the patient was intubated to maintain airway protection. Repeated upper-endoscopy "voluminous live red clot at EGJ, 4-clips and active bleeding of mucosa between, injection of polidocanol". Despite the implemented strategy, high risk of rebleeding remained. Following invasive mechanical ventilation (IMV), sustained hypotension having to increase noradrenaline (1.52mcg/kg/min) and lactate (5.8mmol/l), despite fluid resuscitation. Echocardiogram evidenced severe ventricular dysfunction, and fixed inferior vena cava (IVC) of 20mm. The heart defect combined with positive intrathoracic pressure, contributed to the worsened shock, as Fontan circulation is dependent on low vascular resistance to maintain output2. Needing VA-ECMO and admitted to ICU, volemia optimization, adjusting ventilation to lower intrathoracic pressure and started on milrinone and sildenafil. Another upper-endoscopy showed laceration at EGJ, with placement of clips. Echocardiogram revealed "Normal left ventricle. Hypoplastic right ventricle. Mild mitral regurgitation; aortic VTi 19cm. IVC 22mm. RV/RA gradient 70mmHg. Interatrial bidirectional shunt". Favorable evolution permitted extubation, suspension of milrinone and sildenafil, followed by decannulation.

With rescue ECMO, congenital heart disease are salvageable despite sudden decompensation[3]. This case, positive intrathoracic pressure impairs the Fontan circulation, dependent on preload and higher central venous pressure to maintain cardiac output, as the ventricle is unable to compensate increased demands[2], and worsening shock.

Keywords: *Congenital Heart Disease, venoarterial-extracorporeal membrane oxygenation, VA-ECMO*

INTRODUCTION

Techniques of venoarterial-extracorporeal membrane oxygenation (VA-ECMO) have evolved and improved over the last two decades, allowing numerous applications of this life saving support.[1] Most of the patients who benefit from such support have reversible low cardiac output, or sudden cardiac arrest from a correctable cause.[1]

DISCUSSION:

Here we describe a clinical case of a 21-year-old male with a history of congenital heart disease; characterized with severe right ventricle hypoplasia and pulmonary artery stenosis with a surgical total extracardiac cavo-pulmonary shunt (Fontan surgery), and an atrial septal defect post-septostomy. The daily medication regimen consisting of 100mg of acetylsalicylic acid. Brought to the Emergency Department due to vomiting that progressed to abundant hematemesis, which started on that same day. Diagnosis of a Mallory-Weiss syndrome, with an upper-endoscopy showing a "laceration at the esophagogastric junction with active bleeding", clips and hemospray were applied to control bleeding. The patient remained hemodynamically stable despite abundant blood loss and hemoglobin drop from 19g/dl to 12.7g/dl. However, the patient's condition worsened on the second day, with hypovolemic shock, due to repeated melenas and hematemesis, associated with a continued significant drop in hemoglobin from 12.7g/dl to 9g/dl, presented a lactate level of 2.8mmol/L, capillary refill time 3 to 4s and needing noradrenaline (1.21mcg/kg/min) to maintain mean arterial pressure (MAP) of 60 to 65mmHg. In the meantime, resuscitation was continued, completing a total of 5.5l of crystalloids and 4 units of packed red blood cells (PRBC), coagulation tests and fibrinogen were normal, therefore no other blood products were transfused. In addition, the patient was submitted to a computed tomography angiography that showed "endoluminal extravasation by the contrast product, suggesting an active hemorrhagic focus". Furthermore, due to active blood loss and worsening shock, the patient was intubated to maintain airway protection. The upper endoscopy was then repeated, evidencing "voluminous live red clot at the esophagogastric junction, adrenaline injected, observed area without continuity with the presence of 4 clips and active bleeding on the mucosa between the clips, injection of polidocanol with resolution of active bleeding". Despite the implemented hemorrhage control strategy during the endoscopy and apparent control at the time, the patient remained at high risk of rebleeding. Following invasive mechanical ventilation (IMV) a significant deterioration was observed, particularly from the hemodynamic perspective, with sustained hypotension having to increase noradrenaline (1.52mcg/kg/min) and maximum lactate was at this point of 5.8mmol/l, despite ongoing fluid resuscitation. An echocardiogram was done, evidencing severe ventricular dysfunction, and fixed inferior vena cava (IVC) of 20mm (with IMV). The patient possesses a Fontan circulation, consisting of loss of the subpulmonary pump with elevated pressure in the caval system, non-pulsatile blood flow in the pulmonary circuit and reduction of the systemic output, dependent on a passive cavo-pulmonary flow and low pulmonary vascular resistance to maintain adequate cardiac output[2]. Given the patient's congenital heart defect and altered circulation, combined with the hypovolemic state and the positive intrathoracic pressure from the IMV, both factors contributing to the worsening of the shock. The rescue ECMO team was contacted to consider ECMO as a potential strategy for managing the worsening shock. Initially, the plan was to continue fluid resuscitation, assess the feasibility of reducing positive-end expiratory pressure (PEEP) and even extubate, as these factors were contributing to the reduced cardiac output. However, on evaluation, this plan could not be conducted in a timely manner that would allow for a cautious and prompt stabilization. The PEEP was reduced from 6cmH_2O to 3cmH_2O, further reduction of PEEP was not possible without compromising adequate oxygenation. Although extubating was pondered, due to the suboptimal hemorrhage control and high risk of rebleeding, airway protection through invasive ventilation was essential. This case presents a clear hypovolemic component, making ECMO an unsuitable supportive option. However, due to the significant obstructive shock component, ECMO was deemed the best option to optimize the patient and wean it from invasive ventilation, despite its own possible complications. Therefore, needing rescue VA-ECMO in possibly an obstructive and hypovolemic shock, while maintaining fluid resuscitation. The patient was admitted to the Intensive Care Unit (ICU) ECMO unit, where initial treatment included volemia optimization, adjusting ventilation parameters to lower intrathoracic pressure and started on milrinone and sildenafil to decrease pulmonary vascular resistance. On admission, the patient had noradrenaline at 1.52mcg/kg/min for a mean arterial pressure (MAP) of 60 to 65mmhg, heart rate (HR) of 150bpm, end-expiratory CO2 of 18mmhg, pulse pressure variation of 26% and lactate of 4.2mmol/l. In consequence of hemorrhage losses and fluid ressucitation, hemoglobin fell from 10 to 6.1g/dl, platelets of 104 to 70x10^3/ul and coagulation with activated partial thromboplastin time (aPTT) of 57s, prothrombin time (PT) 32s and fibrinogenio 106mg/dl, with some blood loss around catheters. An echocardiogram showed "Preserved left ventricular systolic function (FS), hyperdynamic left ventricle with kissing walls, right ventricular (RV) atresia and IVC 13mm". On admission, the patient was started on albumin 20% and completed 150ml, 1.5l of crystalloids, 3 units of PRBC, 6 units of plasma, 1 unit of platelets, 1 unit of prothrombin complex, 10mg of vitamin K and 2g fibrinogen. Within the first 24hours noradrenaline went from maximum to 0.24mcg/kg/min for MAP 65mmhg, HR decreased to 81bpm, EtCO2 increased to 32 and lactate levels normalized. The transthoracic echocardiogram revealing "Left heart cavities with normal dimensions, hypoplasic right heart. Mild mitral valve insufficiency. Left ventricle function conserved with an aortic velocity-time integral of 19cm. Fixed IVC of 22mm. A right ventricle-right atrial gradient of 70mmHg." The upper endoscopy was repeated once again, confirming laceration at the gastroesophageal junction, continued hemorrhage between clips

with one thought to be superficially applied, which was removed, followed by the correct placement of clips with hemorrhage control. The clinical evolution was favorable permitting extubation by day 6, followed by percutaneous ECMO decannulation and suspension of milrinone and sildenafil by day 7 and 8 in the ICU, respectively. An echo-doppler was performed by vascular surgery which documented right common femoral artery pseudoaneurysm and right femoral deep vein thrombosis, needing surgical correction with patch and started on anticoagulant for 3 months. Due to coagulopathy and an episode of melenas still occurring on day 7 and day 10 of ICU stay, anticoagulation was delayed until day 12, started on unfractionated heparin, and then switching to enoxaparin. Even though the ongoing blood loss did not have hemodynamic repercussions, due to the severity and difficulty in hemorrhage control presented initially, the decision was to delay anticoagulation until it could be safely introduced.

CONCLUSION

Extracorporeal support has now clearly become a significant tool in the care of complex congenital heart disease[1]. The results of rescue ECMO have shown that many patients with congenital heart disease are salvageable despite sudden decompensation[3], such as in the clinical case described; however, it remains only a form of mechanical support. In this case, considering the severity of ongoing bleeding, a predominantly hypovolemic mechanism for shock worsening cannot be excluded. Still, given the order of events and noticeable deterioration with intrathoracic positive pressure, it is thought that it also contributed to the decline, and because these contributing factors could not be removed, the decision was made to cannulate the patient for a prompt stabilization, while also optimizing volemia. Management with ECMO seemed to be the most suitable support option for this multifactorial shock, not due to the hemorrhagic, but due to the obstructive component. The VA-ECMO support was essential, while the patient was optimized with effective hemorrhage control, volemia ressucitation, and weaned from invasive ventilation, despite the possible complications, such as hemorrhage, in an already hypovolemic hemorrhagic case. Furthermore, a reminder that patients with congenital heart disease, with altered circulation such as in Fontan circulation, rely on passive cavo-pulmonary flow and tolerate less increases in pulmonary pressure. Since the ventricle only pumps the volume supplied by the cavo-pulmonary system, it cannot compensate for increased demands, making procedures like intubation potentially detrimental to the clinical condition[2].

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