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Case Report

North-south syndrome, a complication of circulatory assistance with Veno-Arterial ECMO. A case report

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ABSTRACT

We present the case of a 34-year-old male patient previously diagnosed with Marfan syndrome who was admitted for acute aortic insufficiency secondary to aneurysmal dilatation of the ascending thoracic aorta. In the postoperative period, post-cardiotomy cardiogenic shock was documented, so circulatory support was initiated with peripheral Veno-Arterial ECMO, which developed hypoxemia due to bacterial pneumonia and data compatible with North-South syndrome. We present a review, non-conventional cannulation strategies and a diagnostic alternative for this entity.

Keywords: Marfan Syndrome; ECMO, venoarterial; Shock, Cardiogenic (source: MeSH-NLM).

Introduction

Cardiogenic shock secondary to post-cardiotomy low cardiac output (CSPC-LCO) is a rare complication that develops after cardiac surgery. Some series report it in 6% [1], while others in up to 10% [2]. Among the supportive strategies for CSPC-LCO, is the use of venoarterial ECMO (Extracorporeal Membrane Oxygenation, V-A ECMO) [1-3]. Common complications associated with this device primarily involve vascular and hemorrhagic issues, however, in some cases, the development of refractory hypoxemia, known as the "north-south syndrome" [4], which has deleterious effects on the central nervous and cardiovascular systems, has been described. We present a case of north-south syndrome (NSS) in a patient with CSPC-LCO and review diagnostic and treatment strategies in this complication.

Case report

A 34-year-old male with a history of bilateral retinal detachment and a prior David's surgery (involving aortic root replacement and aortic valve resuspension) performed 12 years before the current admission, presented to the emergency department for dyspnea and angina. Physical examination revealed vital signs within normal parameters, a diastolic murmur consistent with aortic insufficiency, and a 12-lead electrocardiogram displaying signs of left ventricular volume overload. Transthoracic echocardiography (TTE) showed left ventricular dilatation with ejection fraction (EF) of 60% and severe aortic insufficiency. Thoracic aorta angiotomography revealed aortic dilatation of up to 76 mm and dilatation of the right coronary artery, with no evidence of aortic dissection. Subsequently, the patient was assessed by the local Heart Team, who decided to perform Bentall and Bono surgery and reconstruction of the right coronary artery.

Upon admission to the Cardiovascular Intensive Care Unit following the planned surgery, ST-segment elevation was observed in leads II, III, and AVF **(Figure 1)**. Transesophageal echocardiography (TEE) revealed akinesia in the inferior, inferoseptal, and inferolateral segments, spanning all three thirds of the heart, suggestive of myocardial infarction associated with cardiac surgery (Type V infarction). Therefore, an urgent diagnostic coronary angiography was performed, demonstrating adequate flow in the coronary arteries. Nevertheless, the patient's clinical course was complicated, marked by an increased use of inotropic and vasopressor agents, with no improvement in clinical or biochemical tissue perfusion parameters and the diagnosis of CSPC-LCO was established. Peripheral V-A ECMO with ventricular decompression using an intra-aortic balloon pump (IABP) was performed.

Three days after the start of mechanical circulatory support (MCS), hypoxia was observed in the upper part of the body while oxygenation values in the lower extremities remained normal **(Table 1)**. Suspected bacterial pneumonia was subsequently confirmed by a chest X-ray (presence of left basal consolidation), which led to a diagnosis of NNS. Ventilatory parameters were adjusted without resolution of hypoxemia, consequently, new configuration from V-A ECMO to Veno-Arteriovenous ECMO (V-AV ECMO) was performed. This change involved removing a vein and returning to the right heart and aortic arterial cavities. Antimicrobial treatment based on meropenem and vancomycin was also started.

Within 48 hours of implementing the new circulatory and venous support configuration, improvements in hemodynamic and cardiovascular function were documented. As a result, 72 hours after the initiation of this support, it was successfully removed without any complications. The patient was discharged home alive after a 25-day hospital stay.

Discussion

We present the case of a patient with CSPC-LCO who required circulatory and respiratory support with a non-conventional configuration due to the NSS complication.

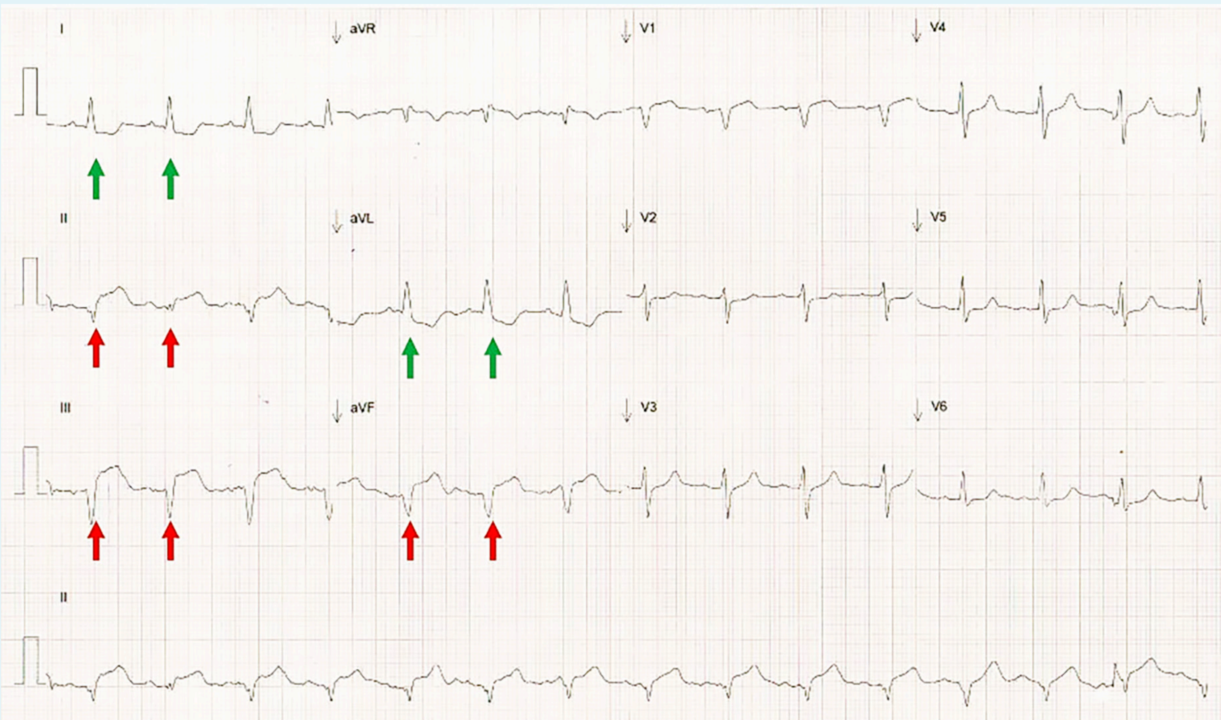

Figure 1. Immediate post-surgery electrocardiogram. A 12-lead electrocardiogram taken upon admission to the Cardiovascular Intensive Care Unit. ST-segment elevation in inferior leads (red arrows) with reciprocal changes in lateral leads (green arrows).

Table 1. Gasometric values of the patient at the time of the North-South syndrome diagnosis.

Sampling site		PO_2	PCO_2	SaO_2
ECMO	Pre-membrane	34 mmHg	50 mmHg	47%
	Post-membrane	227 mmHg	42 mmHg	100%
Lower extremities	Right pedal artery	201 mmHg	40 mmHg	100%
Upper extremities	Right radial artery	51 mmHg	49 mmHg	78%
IABP*	Descending thoracic aorta	52 mmHg	49 mmHg	79%

* The gasometry from the IABP is equivalent to that obtained from the left radial artery, due to the anatomical position of the distal lumen of the device..
IABP: intra-aortic balloon pump; ECMO: extracorporeal membrane oxygenation; mmHg: millimeters of mercury; pCO_2: partial pressure of carbon dioxide; pO_2: partial pressure of oxygen; SaO_2: oxygen saturation.

In the peripheral V-A ECMO mode, the configuration has a mixing point for the flow coming from the heart and the return from the device, which depends on the inotropism of the cardiac muscle and the flow returning from the ECMO. If the cardiac output from the left ventricle is normal, the mixing will occur distally compared to reduced cardiac output, where mixing can occur at the level of the brachiocephalic trunk. The mixing point can be

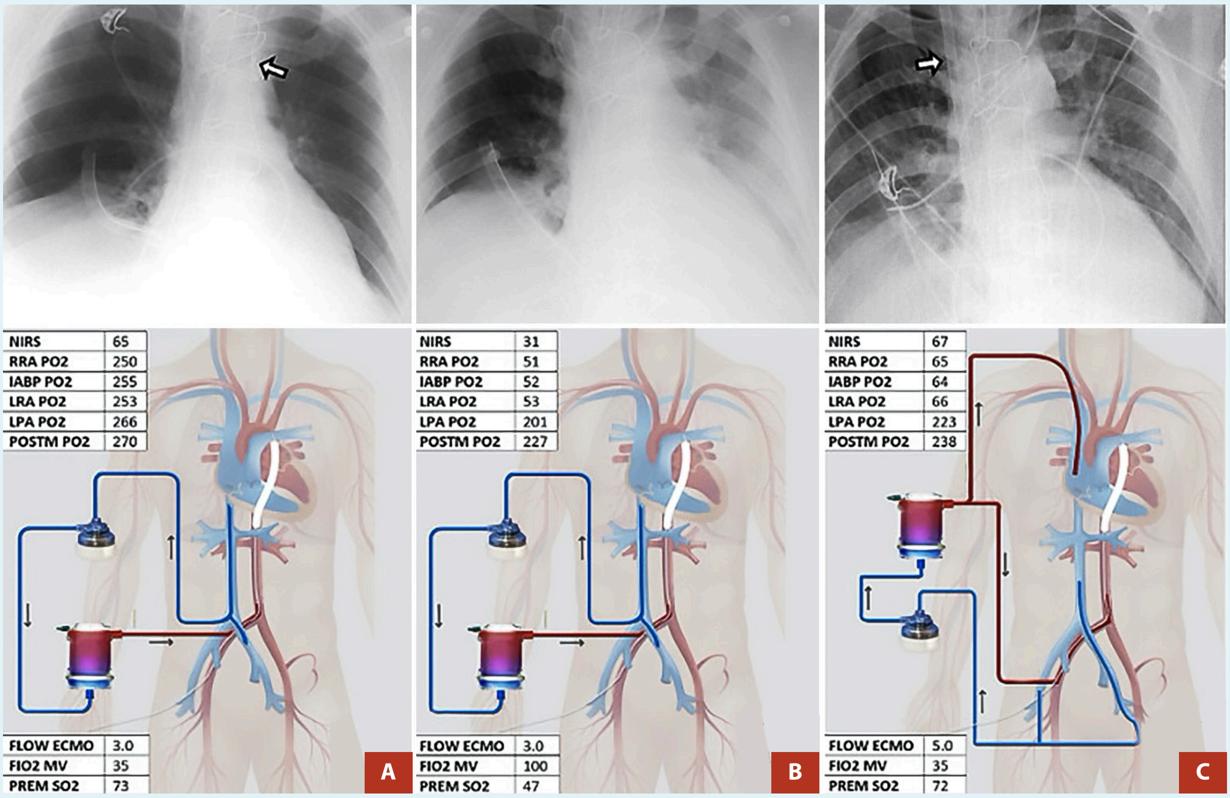

Figure 2. North-South Syndrome. Transition from V-A ECMO to V-AV ECMO. Correlation between chest X-ray and gasometric findings. **Panel A.** Patient in cardiogenic shock with circulatory support using V-A ECMO. **Panel B.** Same configuration as Panel A; however, the chest X-ray shows left basal opacity and evidence of gasometric abnormalities. The arrow indicates the tip of the intra-aortic balloon pump. **Panel C.** Configuration of V-AV ECMO for the treatment of the gasometric abnormalities in Panel B. The arrow indicates the position of the return cannula in the right jugular vein.

V-A ECMO: venoarterial extracorporeal membrane oxygenation, V-AV ECMO: veno-arteriovenous extracorporeal membrane oxygenation, NIRS: near-infrared spectroscopy, RRA: right radial artery, PO_2: partial pressure of oxygen, IABP: intra-aortic balloon pump, LRA: left radial artery, LPA: left pedal artery, POSTM: post-membrane, FLOW ECMO: ECMO flow, FiO_2 MV: fraction of inspired oxygen in mechanical ventilation, PREM SO^2: Pre-membrane oxygen saturation.

identified by measuring blood gases in the right and left radial arteries [5].

Under normal conditions, the anterograde flow from the heart has oxygen concentration within normal values (60-90 mmHg) and depends on the integrity of the pulmonary parenchyma for proper oxygen exchange. If this mechanism is compromised, it can lead to hypoxemia. If the oxygen mixing point is distal to the aortic trunks, there is a risk of hypoxemia in the heart, brain, and upper extremities [5,6] **(Figure 2)**. This condition is known as NSS or also referred to as Harlequin Syndrome, because the lower limbs continue to receive oxygenation from the ECMO, resulting in a cyanotic appearance in the upper extremities and a hyperemic appearance in the lower extremities.

NSS is a complication associated with peripheral V-A ECMO due to retrograde flow (not observed in central ECMO due to anterograde flow) [7,8]. Among the most frequently described etiologies of NSS are acute pulmonary edema due to left ventricular distension from afterload associated with ECMO support and pulmonary infections associated with mechanical ventilation. The prevalence of NSS varies from 8.8% [9] to 13.3% in recent studies [10].

The classic description of the NSS was made in neonates, where the immature skin allowed for the identification of color changes in tissues. In this particular case, due to the inability to obtain arterial access in the left upper extremity (due to anatomical complexity), a distal lumen sample of the IABP was obtained as an analog to the left radial artery. Through this sample, the diagnosis of SNS was successfully made. The significance of early diagnosis of NSS lies in optimizing myocardial recovery and preventing cerebral ischemia, which can result in neurological deficits [8].

Currently, there are no clear guidelines for the treatment of NSS. In this case, cannulation to V-AV ECMO was chosen because conventional maneuvers (optimizing ventilator parameters, increasing V-A ECMO flow along with left ventricular unloading) were unsuccessful. Other modalities include converting from peripheral to central V-A ECMO, as well as dual-pump support (utilizing an additional pump to direct the flow back into the jugular vein), which appears to reduce the risk of clot formation, consumption of coagulation factors, and pulmonary embolism, etc. [10].

In conclusion, the timely identification of respiratory complications associated with mechanical circulatory support in patients with cardiogenic shock should be a primary goal for healthcare teams dedicated to the care of these patients, in order to improve clinical outcomes.

Ethical aspects: Written consent was obtained for scientific disclosure purposes. The images and information presented do not allow for the patient's identification.

Author contributions: EALD: Writing - Review & Editing. KSHB: Conceptualization, Methodology, Writing - Original Draft, Writing - Review & Editing. DRN: Writing - Original Draft. APM: Writing - Original Draft. DMS: Visualization, Writing - Review & Editing. LABH: Supervision. GRV: Supervision, Validation.

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