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A case study of Harlequin syndrome in VA-ECMO

Said Ali Masoud Al Hanshi, Farhana Al Othmani

Address for Correspondence:
Said Ali Masoud Al Hanshi
Pediatric Cardiac Intensive Care Unit, National Heart Center, the Royal Hospital, BOP 1331, PC 111, Muscat, Oman
Email: abushatha50@hotmail.com

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Background: Harlequin syndrome is a rare autonomic disorder, characterized by unilateral diminished sweating and flushing of the face in response to heat or exercise.[1]
Harlequin syndrome is described in patients receiving peripheral veno-arterial extracorporeal membrane oxygenation (VA-ECMO), where differential oxygen saturation is observed between the upper and lower parts of the body.
It is a phenomenon related to cannulation, where upper body hypoxia occurs due to compromised arterial return despite an initial correct bifemoral cannula insertion.
Case report: A 9-month-old child presented to the Royal Hospital, Oman, with a 1-day history of tachypnea and potential case of chocking, which rapidly progressed into ARDS and refractory respiratory failure despite providing maximum supportive measures: different modes of conventional mechanical ventilation and high-frequency oscillatory ventilator (HFOV), steroids, inhaled surfactant, inhaled iloprost, and prone positioning. Inhaled nitric oxide was not available. Rigid bronchoscopy excluded foreign body aspiration and echocardiography showed normal heart structure and function. As all measures failed and oxygen index remained low, ECMO was initiated. She was initially cannulated for VA-ECMO; two femoral venous cannulas of sizes 16F and 10F were inserted into both sides for adequate drainage and a size 8F femoral artery cannula was inserted into the right side; VV-ECMO cannulas were not available. The ECMO machine delivered a flow rate of about 80 ml/kg/min, BP 90/46, HR 130, CVP 14, Hb 12.6 g/dl, and milrinone 0.5 μg/kg/min. There was differential oxygen saturation between the upper and lower parts of the body; saturation was 50 – 60% in the upper part and 100% in the lower part, and FiO_2 was 100%.
To solve the problem, VA-ECMO was changed from peripheral to central through insertion of a venous

cannula into the right atrium and an arterial cannula in the aorta with a flow rate of about 110 ml/kg/min. The child was successfully decannulated after 10 days of ECMO support and discharged home 2 months after admission. Respiratory viral panel was positive for adenovirus.

Discussion: Harlequin syndrome is a rare complication of peripheral VA-ECMO. However, it can be as high as 8.8%.[2]

It occurs when the heart function is preserved or recovering, but the lungs are still poorly functioning, so the native cardiac output flows against the pumped blood, usually in the aortic arch region.[2] The reinfusion jet flows retrograde up the aorta and may meet resistance from antegrade flow generated by the left ventricle.

Depending on the amount of native cardiac function, the location of the interface between antegrade and retrograde flow will vary, and in circumstances where there is impaired native gas exchange with a significant amount of poorly oxygenated blood ejected from the left ventricle, the oxygenated reinfused blood may not reach the aortic arch.[3] Subsequently, the coronary arteries, and to a variable degree the supra-aortic vessel as well, are provided with hypoxic blood, and the heart and brain can be affected.[2]

Therapeutic options consist of relocation of the arterial cannula into the right subclavian artery or aorta,[2] or converting to central VA-ECMO. It can also be solved by converting the system into a VA-V setting, where an additional return cannula may be added to the configuration with a "Y" connection off the femoral arterial reinfusion cannula, with insertion into an internal jugular vein.[3]

Conclusion: Harlequin syndrome is a known complication of peripheral VA-ECMO, where the upper part of the body is poorly oxygenated. It occurs when the native heart function is preserved but the lungs are poorly functioning. Therapeutic options include converting to central VA-ECMO or VA-V-ECMO.

Keywords: Harlequin syndrome, VA-ECMO, differential oxygen saturation, case study

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