

### Recurrent cardiopulmonary arrests in an infant with PA with VSD and MAPCAs: case report

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Introduction: Pulmonary atresia (PA) with ventricular septal defect (VSD) and major aorto-pulmonary collateral arteries (MAPCAs) is a rare congenital heart disease with a poor prognosis. Its management and treatment options heavily rely on the pulmonary blood flow pattern.

Clinical Case: A 2-month-old infant, postnatally diagnosed with PA/small-VSD/MAPCAs and confluent, but extremely hypoplastic PAs, was admitted to the emergency department due to dyspnoea, cyanosis (SpO<sub>2</sub>~50%) and hypotonia. She had a poor peripheral perfusion and the auscultation denoted only a continuous murmur, with the absence of the usual predominant systolic murmur. The echocardiogram, besides the known anatomy, evidenced a restrictive sub-aortic VSD, partially closed by aneurysmal tricuspid valve (TV) tissue protruding below the aortic valve, with an exclusive R-L shunt (70mmHg of gradient). A cardiac catheterization was performed, revealing multiple MAPCAs from supra-aortic branches and descending aorta to both lungs and hypoplastic PAs (nearly 1.7mm), no pressures in the LV were obtained. In the following days, she presented new episodes of hypoxemia and bradycardia while crying and subsequent cardiac arrests, requiring short courses of CPR. During the last, an echocardiogram was performed, showing the TV tissue protruding through the VSD into the left ventricle outflow tract (LVOT) limiting the flow through the aortic valve. This fact seemed to be the most reasonable explanation for the previous events, which appeared to be the cause of the recurrent cardiac arrests. She underwent a cardiac surgery with construction of a Melbourne shunt and fixation of the septal leaflet of TV to the right side of interventricular septum. The postoperative echocardiograms showed an adequate patency of the Melbourne shunt, small VSD (RV-LV gradient of 62 mmHg) and no LVOT obstruction. No more events occurred and the infant was discharged on oral aspirin with SpO<sub>2</sub> 75-80% and normal neurological evaluation. Future surgical intervention is still under discussion.

Conclusion: This case claims the attention to a rare cause of cardiac arrest in the setting of a PA with VSD, caused by dynamic LVOT obstruction.

