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Follow the MAPCA: an unusual source of right pulmonary artery flow

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Abstract

Major aortopulmonary collateral arteries in the setting of pulmonary atresia with intact ventricular septum are very rarely encountered, having been documented in only a handful of case reports. We present the right ventriculogram of a patient found to have this rare combination of findings along with right ventricular-dependent coronary artery circulation and unusual supply of the right pulmonary artery.

We present a case of a cyanotic newborn found by echocardiogram to have pulmonary atresia with intact ventricular septum and major aortopulmonary collateral arteries. Cardiac catheterisation was performed to assess the coronary artery circulation and major aortopulmonary collateral artery supply. The right ventriculogram demonstrated large coronary sinusoids arising from the right ventricle, supplying all major branches of the coronary artery circulation (Fig 1, Supplementary Videos S1-S2). There was no contrast seen entering the aorta from the coronary circulation as the ostia were atretic, consistent with right ventricular dependence of the coronary arteries. Additionally, there is a vascular connection arising from the left main coronary artery that supplies the right pulmonary artery. Thus, flow from the right ventricle travels through a coronary sinusoid to the left coronary artery before coursing through the collateral vessel to supply the right pulmonary artery. While major aortopulmonary collateral arteries in the setting of pulmonary atresia intact ventricular septum are quite rare, having only been reported in a handful of published reports,[1] the novelty of this angiogram lies in the well-displayed, unusual supply of flow to the right pulmonary artery. To our knowledge, this particular anatomy has never been displayed or reported in the literature.

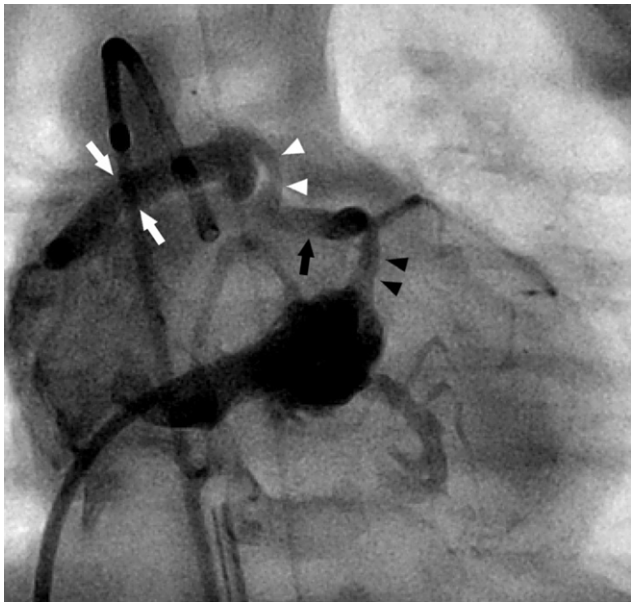


Fig. 1 Still frame of right ventricular angiogram demonstrating unusual course of blood supply to the right pulmonary artery (black arrow heads: coronary sinusoid; black arrows left main coronary artery; white arrow heads: collateral vessel; white arrows: right pulmonary artery)

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Conflicts of interest. None.

Ethical standards. The authors assert that all procedures contributing to this work comply with the ethical standards of the relevant national guidelines on human experimentation (please name) and with the Helsinki Declaration of 1975, as revised in 2008, and have been approved by the institutional committee.

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