Aortic forward flow in aortic atresia via ventriculo-coronary arterial connections

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A full-term male newborn weighing 2.7 kg was evaluated because of suspected cyanotic congenital heart disease on the day of birth. The echocardiographic study showed corrected transposition of the great arteries, severe tricuspid stenosis, right ventricular (RV) hypoplasia, aortic atresia, intact ventricular septum, and ventriculo-coronary arterial connections (VCACs) (Panel A, yellow arrow). RV contraction created reverse flow in the right coronary and left anterior descending coronary arteries via the VCACs, resulting in forward flow in the ascending aorta (Panels B and C, see Supplementary data online, Video S1). Computed tomographic angiography on the third day of life also demonstrated the VCACs between the hypoplastic RV and ascending aorta (Panels D and E). The patient was maintained on prostaglandin E1, and bilateral pulmonary arterial banding was performed on the 14th day of life. On the ninth postoperative day, echocardiography demonstrated a restrictive patent foramen ovale with accelerated flow velocity. Therefore, balloon atrial septostomy and right ventriculography were performed. The examination demonstrated the presence of VCACs consistent with the prior echocardiographic findings (Panels F and G). The patient underwent the Norwood procedure with an LV-PA shunt on the 55th day of life. This is the first report of this anatomic variant. Coronary insufficiency and critical ventricular dysfunction in the presence of VCACs is a well-described phenomenon in hypoplastic right and left hearts. The present case had a definitively different blood flow pattern in the ascending aorta and coronary arteries compared with these two representative anomalies with hypoplastic left or right ventricles.

Conflict of interest: None declared.

Supplementary data are available at European Heart Journal — Cardiovascular Imaging online.

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