Right Coronary Artery to Right Ventricle Fistula and Congenital Coronary Ostial Atresia in a Neonate

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PURPOSE

- Both coronary artery fistula (CAF) and congenital coronary ostial atresia are rare diagnoses.
- We present a case of an infant with both a right coronary artery to right ventricle (RCA-RV) fistula and right coronary ostial atresia who presented with a murmur, ST-segment changes on EKG, and elevated troponin.
- We discuss the exam findings associated with CAFs, the clinical significance, and management options.

CASE DESCRIPTION

- A 2-week old 35 week neonate was noted to have a continuous murmur on exam.
- EKG showed diffuse ST-segment changes (figure 1).
- Echocardiogram demonstrated an RCA-RV fistula with normal biventricular function (figures 2a-2d).
- Troponin was elevated to 191 (LLN < 22).
- Coronary angiography demonstrated right coronary ostial atresia and retrograde filling of the right coronary distribution via a dilated left circumflex coronary artery, with a RCA-RV fistula (Figure 3).
- Troponins improved and ST-segment changes resolved, though with new deep Q waves in inferior leads.
- He remained asymptomatic and was discharged after 1 week.
- At two month follow-up, he was doing well, with normal EKG and normal function on echo.

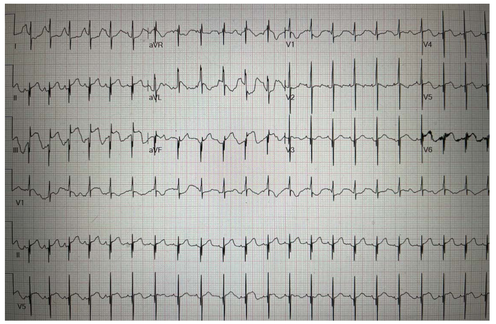

Figure 1: Initial EKG, demonstrating diffuse ST-segment elevation

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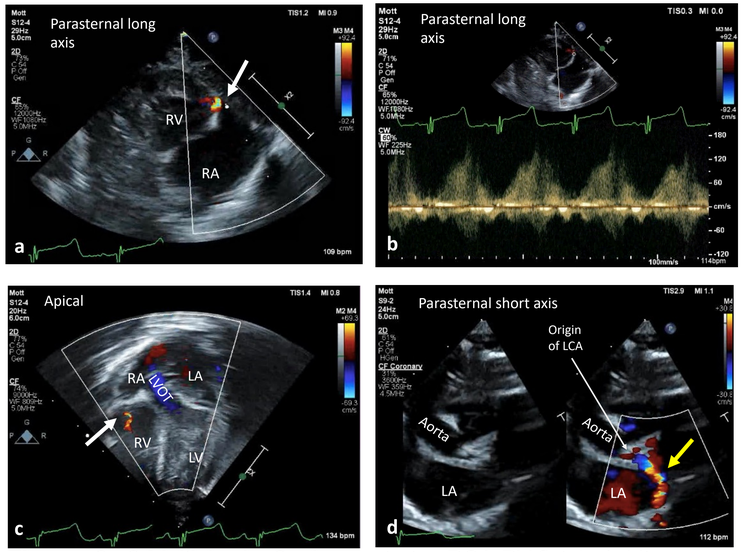

Figures 2a-2d: Initial echo **2a** demonstrates RCA-RV fistula (white arrow), with continuous flow shown by doppler (**2b**). The RCA-RV fistula is seen again in figure **2c** (white arrow). Figure **2d** demonstrates increased flow from the left circumflex branch of the LCA (yellow arrow)

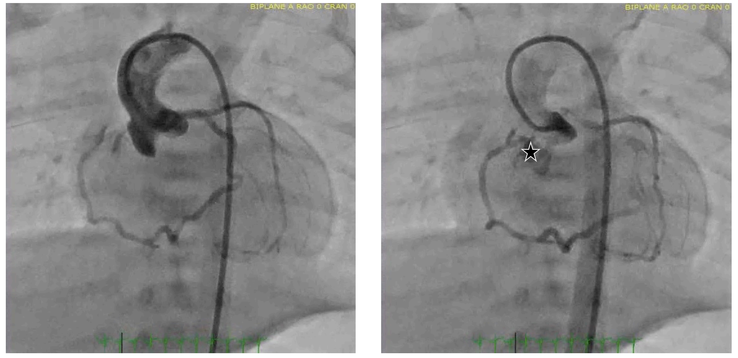

Figure 3: Selective angiography through a 3.3 French JL1 catheter positioned within the left coronary os demonstrates a dilated left circumflex coronary artery, with retrograde filling of the right coronary and a fistulous connection between the RCA and the RV (★)

DISCUSSION

- CAFs are an abnormal communication between the coronary artery and a cardiac chamber or great vessel.
- They are relatively rare, with an estimated prevalence of 0.002%, comprising 0.4% of all cardiac malformations.
- CAFs are more often congenital than acquired and are typically asymptomatic and found incidentally on echo or coronary angiography.
- If hemodynamically significant, they can present with heart failure.
- CAFs can grow, and the majority of patients over 20 are symptomatic.
- The typical exam finding would be a continuous low-pitched murmur heard best at the left sternal border due to continuous flow from the coronary artery into the low-pressure RV or PA.
- Treatment options include transcatheter occlusion versus surgical-based approaches such as endocardial closure and epicardial ligation.
- Both surgical and catheter-based interventions are overall successful with minimum complication rates in both neonates and older individuals.
- Right coronary ostial atresia is exceedingly rare and typically congenital.
- Due to the extensive network of collateral vessels, patients typically do not experience ischemia and intervention is rarely required

CONCLUSIONS

- This neonate had initial troponin elevation and EKG ST-segment changes.
- Given the finding of RCA-RV fistula in addition to RCA ostial atresia with right coronary fed from the left coronary, presentation was concerning for coronary steal phenomenon.
- Due to his size, this patient was not a good candidate for catheter based intervention. Given the lack of symptoms and resolution of initial abnormal findings, the decision was made to discharge with close follow-up.
- Based on the current literature, CAFs are typically incidental findings that rarely require intervention, but it is uncertain how right coronary ostial atresia impacts that prognosis.
- With concomitant finding of right coronary ostial atresia, this patient's CAF may become clinically significant over time and require intervention.