Prenatal Diagnosis of Coronary Artery Fistulae – A new case series

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Objectives

Congenital coronary artery fistula (CAF), especially when isolated, is a rare cardiac abnormality with a reported incidence between 0.2 to 0.6%. We present a new case series of prenatally diagnosed CAF focusing on prenatal imaging characteristics, prenatal and perinatal management as well as outcome.

Methods

A database search of fetal echocardiograms form 2012-2022 in our institution was performed for identification of cases carrying the diagnosis of coronary artery fistulae. Emphasis was given to the gestational age at diagnosis, prenatal and postnatal clinical and imaging characteristics, genetic associations as well as prenatal and perinatal management and short-term outcome.

Results

Three cases with the diagnosis of CAF were identified. All cases were referred for suspected congenital heart disease: case A at 20+1/7 weeks gestational age (GA), case B at 35+2/7 weeks GA and case C at 25+4/7 weeks GA. Abnormal flow jet from the coronary sinus to the right atrium (RA) in case A, in the atrial septum and to the RA in Case B and from the coronary sinus (CoS) to the left ventricle (LV) in case C, raised the suspicion of a CAF. The feeding coronary was identified due to its dilation. The CAF originated from the left coronary (LC) cusp in Case A and connected to an enlarged CoS; ventricular size discrepancy (borderline left sided structures), low normal systolic function and significant diastolic runoff in the descending aorta, were present. In Case B the origin was from the LC cusp with an interatrial septal defect (dilated Kugel’s artery) and exit to the RA. RA and right ventricular enlargement were present with preserved systolic function. In case C the origin was from the LC cusp connecting to the coronary sinus which had coronary ostial atresia and an abnormal connection to the LV – a right aortic arch with a left ductus arteriosus and a membranous ventricular septal defect (VSD) were also present (complete vascular ring). All diagnoses were confirmed postnatally by echocardiography; two had additional coronary angiography and one had a tomographic angiography (CTA). Case A and B required cardiac intervention in the catheterization laboratory for fistula closure soon after birth. Case A at one year follow-up revealed no residual fistula flow, normal biventricular size and function and mild-moderate supra-mitral stenosis secondary to enlarged ‘stiff’ coronary sinus containing the vascular plugs, that led to surgical intervention at almost two years of age. Case B at age 12 months had no residual flow and case C at 7 months follow-up has a decrease in fistula size and is awaiting surgical correction of the vascular ring and VSD.

Conclusion

Congenital CAF are rare defects, but their prenatal diagnosis is feasible, with accurate identification and mapping of the involved vascular structures. In utero surveillance of ventricular function and growth of cardiac structures as well as growth of the fistula is imperative, particularly in presence of large shunts and aortic insufficiency given the risk for myocardial ischemia. Parental counseling, delivery planning and perinatal management will be based on the prenatal echocardiographic findings. Prostaglandins, immediately after birth in order to support the systemic circulation might be considered. Congestive heart failure postnatally is not uncommon and usually cather or surgical closure needs to be performed in hemodynamically significant lesions. Long-term outcome is favorable even in large CAF causing significant shunts in utero.

References

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