In Utero Evolution of an Apical Left Ventricular Aneurysm

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Fetal echocardiography identified a large, thin-walled apical aneurysm communicating with the left ventricle (LV; LV aneurysm/LV area=1.4) in a male fetus at 22 weeks 4 days of gestation (Figure, A and Movie I in the online-only Data Supplement). Left ventricular size was normal (end-diastolic dimension [EDD]=0.93 cm; z=1.04) and the cardiothoracic (CT) ratio of circumferences was normal (0.50). Left ventricular function was borderline normal (shortening fraction [SF] = 28%), and right ventricular (RV) function was normal. The mitral valve (MV) and aortic valve (AV) both appeared structurally normal with no regurgitation. There was antegrade flow throughout the course of the aortic arch. Growth was appropriate for gestational age, no other anatomic concerns were identified, and there was no evidence of hydrops. There was no history of maternal viral illness.

At serial assessment, 7 weeks later, the CT ratio had increased to 0.75, the LV was severely dilated (EDD 3.08 cm; z=11.08), and the aneurysm was indistinguishable from the LV cavity (Figure, B and Movie II in the online-only Data Supplement). LV function was severely depressed (SF<10%); however, RV function remained normal. Color and pulsed Doppler interrogations of the AV and MV were abnormal, with trivial antegrade flow and moderate to severe regurgitation across both. Flow was reversed in the ascending and transverse aorta and across the atrial septum (left to right), creating a circular shunt. There was no evidence of evolving hydrops, and growth continued to be appropriate for gestational age.

Serial biweekly assessment was notable for cardiovascular stability, including CT ratio, LVEDD, and shortening fraction. Fetal growth continued at the 50th percentile for gestational age.

At 36 weeks of gestation, the fetus was listed as an in utero candidate for orthotopic heart transplantation (OHT). Spontaneous vaginal delivery of a 3.4-kg baby occurred at 39 weeks 4 days of gestation. Respiratory failure necessitated intubation and mechanical ventilation immediately after delivery. ECG confirmed normal sinus rhythm, with right atrial enlargement, nonspecific intraventricular conduction delay, and T wave inversions in the lateral precordium (Figure, C). Transthoracic imaging confirmed the prenatal diagnosis of a severe dilated cardiomyopathy (LVEDD 3.83 cm; z=9.27), with an estimated SF of 10%, an ejection fraction of 2% to 3%, and no identifiable antegrade flow across the aortic valve (Movie III in the online-only Data Supplement). Right ventricular function was preserved. The origins of both coronary arteries were normally located.

Both prostaglandin and heparin infusions were commenced, and on day of life (DOL) 1, the newborn was listed for OHT. Because systemic cardiac output was dependent on preserved RV function and persistent ductal patency, the prostaglandin infusion was maintained until DOL 11, when the infant underwent a hybrid procedure (ductal stenting and placement of bilateral pulmonary artery bands) to secure ductal patency and control pulmonary blood flow. The heparin infusion was continued and therapeutic levels maintained to prevent LV thrombus formation until DOL 14, when the infant underwent OHT. No intraventricular thrombus was identified before transplant nor at explant.

Histological evaluation of the explanted heart identified epicardial fibrosis and marked endocardial fibroelastosis within the LV and, to a lesser degree, the RV (Figure, D and E). At the junction of the aneurysm and myocardium, there was fragmentation of the myocardial fibers by fibrous tissue continuous with that of the endocardium.

The infant’s length of stay after transplant was 67 days (institutional median = 28 days after infant OHT), primarily because of a prolonged wean from ventilatory support. At 1 year, the infant is well, with no evidence of pulmonary hypoplasia despite in utero pulmonary parenchymal compression.

Congenital LV aneurysms are rare and poorly understood, with a total of 26 reported cases in the literature.1 In utero stability, after the development of severe LV enlargement and dysfunction, is unexpected. Serial images illustrate an apical LV aneurysm, which evolves into a dilated cardiomyopathy indistinguishable from other underlying etiologies. Congenital LV aneurysm is not recognized in the differential of fetal or neonatal dilated cardiomyopathy.2 The hybrid approach used was developed as an alternative to the Norwood procedure for high-risk neonates with hypoplastic left heart syndrome.3 Its application as a bridge to transplant for dilated cardiomyopathy is novel.

Disclosures

None.
References


Figure. **A**, Fetal 4-chamber view at diagnosis. There is a large left ventricular apical aneurysm (AN). The aneurismal portion is nearly akinetic, whereas the basilar portion of the left ventricle (LV) and the entire right ventricle (RV) show preserved function. The left and right atria (LA and RA) are of normal size. **B**, Four-chamber view at serial evaluation. The left ventricle (LV) is severely enlarged with increased endocardial echogenicity. Global function is poor. The apical aneurysm is indistinct from the basilar portion of the left ventricular cavity. The right ventricle (RV) appears of normal size with preserved ventricular function. Both atria are of normal size. **C**, ECG from day of life 1. **D**, Gross photograph: bisected heart with anterior aspect on left. External surface of aneurysm is inked red. The left anterior descending coronary artery is inked black. Ruler=0.5 inch. Entire chamber has extensive fibroelastosis. Mitral leaflets are thickened. **E**, Microscopic section: movat stain of the LV aneurysm, original magnification ×4. The epicardium is fibrotic, and the endocardium has the excess black elastic fibers of fibroelastosis. In the center, acute necrosis with hemorrhage was focal only in the aneurysmal area.
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Circulation. 2014;130:1982-1983
doi: 10.1161/CIRCULATIONAHA.114.011911

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