

CASE REPORT

Unusual consequence of a fetal atrial septal aneurysm

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Key Clinical Message

We describe the case of a significant fetal atrial septal aneurysm causing left ventricular inflow obstruction. Serial fetal echocardiograms demonstrated potential left heart hypoplasia. The fetal cardiologist guided the perinatal team to electively deliver the infant early and modify the course of developing hypoplastic left heart.

Keywords

Atrial septal aneurysm, fetal cardiology, fetal echocardiogram, hypoplastic left heart.

Introduction

A 29 year-old G2P1 woman with ventricular size discrepancy on perinatal ultrasound at 25 weeks gestation was referred for fetal echocardiogram at 27 weeks gestation. Her obstetrician had referred her to perinatology due to history of a bicornuate uterus and a prior preterm birth at 33 weeks. On the initial perinatology ultrasound, the right (RV) and left (LV) ventricles measured within normal range, but the LV was relatively smaller with an RV to LV end-diastolic dimension ratio of 1.3. On fetal echocardiogram using GE Vivid E9 machine with 4C-D transducer (GE Healthcare, Wauwatosa, WI), a large atrial septal aneurysm (ASA) prolapsed across the left atrium into the mitral valve orifice with minimal LV inflow obstruction (Fig. 1). Flow across the foramen ovale was bidirectional.

Follow-up fetal echocardiography focused on accessing the adequacy of growth of the left heart structures and was performed at 1 month, followed by 2 weeks, then weekly follow-up intervals. The standard measurements of fetal cardiac chamber and valve sizes, as well as color Doppler flow assessment, were obtained during each study [1]. Surrogates of left ventricular cardiac output adequacy, including direction of flow across the foramen ovale and aortic arch, were assessed. Serial fetal echocardiograms at 31, 33, and 34 weeks demonstrated increasing pulmonary venous and LV inflow obstruction (Fig. 1)

with the development of a mildly hypoplastic left ventricle and aortic arch. At 34 weeks, significant retrograde flow was seen in the aortic arch, raising concern for insufficient LV outflow. Premature atrial complexes were now noted as well. No placental circulation changes were observed; umbilical, ductus venosus, and cerebral Dopplers were within normal limits throughout gestation. Due to concern for progressive LV inflow obstruction causing evolving left heart hypoplasia, the perinatal team electively delivered the fetus early at 34 weeks.

After delivery, flow across the atrial septum shifted from left to right causing the ASA to bulge into the right atrium. There was no obstruction to systemic or pulmonary venous inflow, and a trivial low velocity left to right shunt persisted across the patent foramen ovale. The left heart structures measured at the lower limits of normal, with a mildly hypoplastic aortic valve. The infant is doing clinically well without intervention, with rare premature atrial complexes noted on routine electrocardiogram.

ASA is a localized redundancy of the interatrial septum that bulges into the right and/or left atrium. It can be seen in up to 40% of fetuses referred for fetal echocardiography and is most commonly associated with premature atrial complexes or atrial arrhythmias [2]. In up to 10% of children and adults, ASAs are usually isolated incidental findings or can be associated with interatrial shunts [3]. The location of the ASA bulging into the right or left atrium depends on the cardiac hemodynamics and

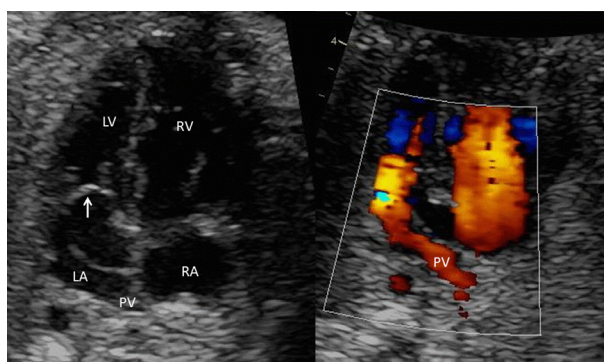


Figure 1. Fetal four-chamber view shows large atrial septal aneurysm (arrow) prolapsing into the mitral valve orifice, with color flow demonstrating mild obstruction to pulmonary venous (PV) flow into the left ventricle.

phase of the cardiac cycle. In normal fetal cardiac physiology, blood flows right to left across the foramen ovale directing oxygenated blood from the placenta into the left atrium. Therefore, fetal ASAs are commonly seen bulging into the left atrium, but typically do not cause flow obstruction.

Cases of ASA have been reported with associated atrial septal defects, atrioventricular valve prolapse, or hypoplastic right heart variants [4]. To our knowledge, this is the first report of an ASA causing LV inflow obstruction and potential hypoplastic left heart. Any obstruction to blood flow in the fetus reduces the *in utero* potential for growth in downstream cardiac structures. In this case, the large ASA caused significant LV inflow obstruction, as evidenced by the presence of left to right atrial flow, and created the potential for evolving hypoplastic left heart in later gestation. The RV became relatively dilated compared to the LV, leading to fetal echocardiography diagnosis of the obstructive ASA.

Careful follow-up demonstrated increasing ASA obstruction of pulmonary venous flow into the LV and decreasing rate of left heart structure growth. Increasing retrograde flow into the aortic arch signified inadequate

LV outflow, in this case due to inadequate LV inflow. The fetal cardiologist recommended early delivery based on this progression knowing that, after delivery, the normal increase in pulmonary blood flow and resultant elevation in left atrial pressures would shift the ASA into the right atrium and relieve LV inflow obstruction. Fetal ASAs are common, but this case demonstrates as an unusual, but modifiable, consequence of LV inflow obstruction, and potentially developing left heart hypoplasia. A comprehensive fetal echocardiogram evaluation is recommended in any fetus demonstrating abnormalities in the screening cardiac assessment (such as ventricular size discrepancy in this case) at any time during gestation.

Conflict of Interest

The submission is with the full knowledge and approval of the listed authors. None of the authors have any disclosures or conflicts of interest to report.

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