

Prenatal Diagnosis of Left Ventricular Aneurysm and Diverticulum

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We report two cases of localized left ventricular (LV) protrusion, an entity that has been described in the literature as aneurysm or diverticulum. Both cases had different outcomes from those previously reported. Little is known on the incidence and natural history of these rare anomalies, whose progression may be asymptomatic or lead to severe complications and death in the prenatal period. The therapeutic approach should be customized.

Case Report 1

A 34-year-old patient, gravida 2, para 1, on the 16th week of gestation, was referred to the maternal-fetal medicine department due to the finding of pericardial effusion on routine prenatal echocardiography. The fetal anatomy was otherwise normal. Fetal echocardiography showed significant pericardial effusion and a finger-like protrusion with a narrow neck located in the LV apex, measuring approximately 5 x 5.7 mm (Figure 1). No thrombi or other cardiac abnormalities were found. Pericardiocentesis was indicated and successfully performed at 20 weeks of gestation. Drainage of 6 mL of serosanguineous fluid was followed by immediate pulmonary expansion. A mild increase of the pericardial effusion was observed in post-intervention control tests in relation to post-puncture tests, however always with preserved left ventricular function and good pulmonary expansion. On the thirty-seventh week, fetal death was diagnosed on a control echocardiogram. Necropsy was not performed because of family's refusal.

Case Report 2

A 25-year-old patient, gravida 1, para 0, on the 30th week of gestation was referred for fetal echocardiography due to suspected cardiac malformation. The study showed dilated LV with decreased systolic function (fractional shortening = 25%) and a sac-like protrusion with a wide neck in the LV lateral wall below the mitral valve (Figure 2). Paradoxical systolic bulging of this structure, which measured 12 x 13

Key Words

Prenatal care; ultrasonography prenatal; pericardial effusion.

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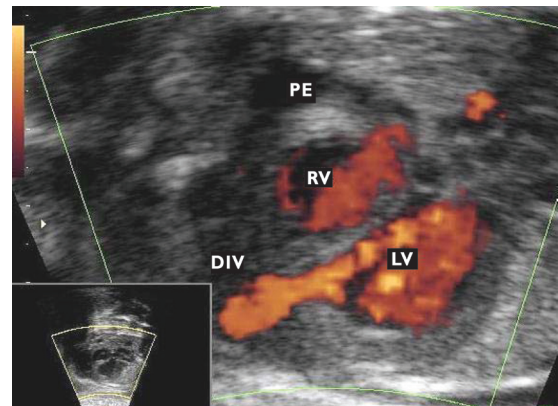


Figure 1 – Fetal echocardiogram showing a diverticulum in left ventricular apex and pericardial effusion. DIV – diverticulum; RV – right ventricle; LV – left ventricle; PE – pericardial effusion

mm, was observed. Color and pulsed Doppler demonstrated flow inside the structure, thus confirming continuity with the LV. Watchful waiting during pregnancy did not show either hemodynamic decompensation or worsened ventricular function. Cesarean section was performed on the 36th week due to maternal infection (pyelonephritis), delivering a preterm infant weighing 2.5 kg with a one and five-minute Apgar score of 8 and 9, respectively. Neonatal echocardiography confirmed the malformation and showed preserved LV systolic function. Watchful waiting was chosen, and, to date, the seven-month-old infant progresses asymptotically, not requiring any therapeutic intervention.

Discussion

Congenital protrusions of the heart ventricular walls have been described in the literature as diverticula or aneurysms. Their incidence is unknown, because they can progress asymptotically or lead to hemodynamic complications and death in the prenatal period¹. The underlying pathophysiological processes are little understood; however, it is hypothesized that they result from a localized weakening of the ventricular wall due to an interruption in the development during embryogenesis, infection, or ischemia^{2,3}. Although there is no definitive consensus for the differentiation between these entities, some criteria have been proposed.

Diverticula are characterized by a narrow connection with the ventricle and presence of myocardial fibers in the composition of their walls, thus enabling efficient systolic

Case Report

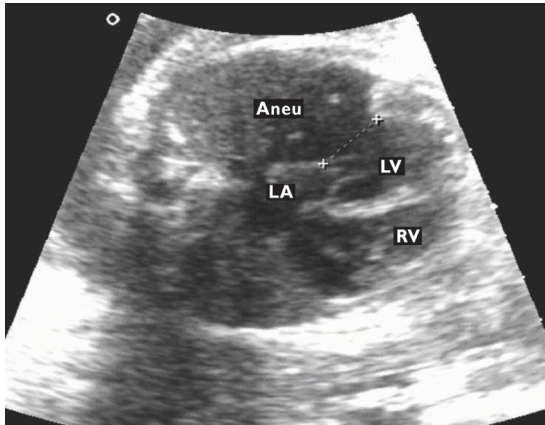


Figure 2 – 4-chamber-view fetal echocardiogram. Asterisks point to the neck of the left ventricular aneurysm. Aneu – aneurysm; LA – left atrium; RV – right ventricle; LV – left ventricle.

contraction in some cases¹. They may be isolated³ or associated with interventricular⁴ and defects of the mid-thoracoabdominal line². They affect either the right^{3,4} or the left ventricular chamber¹, more commonly in the apex^{1,3,4}. Most cases reported in the prenatal period are from findings of tests performed during the second trimester of pregnancy, because diverticula are usually smaller structures than aneurysms and do not cause cardiovascular compression. Thus, the prognosis seems to be favorable, and most of the neonates are asymptomatic at birth⁵. However, depending on their location and association with other anomalies, diverticula may trigger heart failure, arrhythmia, rupture, large pericardial effusion, and hydrops fetalis^{1,2,5,6}. In two cases with pericardial effusion diagnosed in early phases of pregnancy, the risk of pulmonary hypoplasia led to termination of pregnancy, a procedure that is legally permitted in the country where these cases were reported^{5,6}. It is worth pointing out that, in the absence of hemodynamic impairment, pericardial effusion usually has a favorable outcome and spontaneous resolution². Some authors have reported the benefits of pericardiocentesis in cases with pericardial effusion associated with heart failure, permitting normal lung development and a favorable outcome^{2,3,7}. This was not observed in case 1 reported here, which was diagnosed as a diverticulum. Pericardiocentesis was performed because the pregnancy was in its early stages. Based on previous experience with good outcomes after pericardiocentesis, watchful waiting was recommended after puncture; however, fetal death occurred close to term. Since there were no signs of hemodynamic decompensation in the serial tests, the major hypothesis is of ruptured diverticulum, despite its unchanged size. Our experience, therefore, contrasts with the usually favorable outcome attributed to diverticula. Another aspect that should be pointed out is that the finding of isolated pericardial effusion with no obvious

cause requires the investigation of uncommon abnormalities, such as ventricular or atrial diverticulum⁸.

On the other hand, aneurysms are characterized by a unilobulated or multilobulated bulging of a ventricular wall segment, with a typical wide neck, fibrotic tissue and akinesia⁹. They can also be asymptomatic⁴, but lead to a higher risk of complications such as rupture, embolism, arrhythmia, pericardial effusion, heart failure and hydrops fetalis¹⁰. Indication of surgical treatment after birth is controversial, with some authors suggesting surgical excision only in symptomatic patients, and others supporting surgery regardless of symptoms⁹. Case 2, diagnosed as an aneurysm, showed an association with dilatation and impaired LV systolic function in the initial moment, thus suggesting a worse outcome. Again, unlike previous reports of the literature, we observed a good outcome, with no hemodynamic decompensation during the follow-up until delivery. Additionally, the LV systolic function returned to normal and a good clinical outcome was observed after delivery. Again, the experience with this specific case is different from the usually poor outcome attributed to aneurysms. In our opinion, these peculiarities result from the fact that these two entities are distinct presentations of the same anomaly. Although differences exist in relation to the macroanatomical and histological characteristics, both represent dysplastic myocardial areas with different degrees of severity, probably acquired during early phases of embryogenesis. Additionally, the lack of consensus on the definition of terms (diverticulum versus aneurysm) and their interchangeable utilization may have led to imprecise diagnoses in the past, thus affecting the perception of the clinical outcome of these conditions. The treatment proposed in the cases reported (serial assessment, pericardiocentesis, watchful waiting) followed the few recommendations available in the literature, which are always based on the experience with small case series.

In conclusion, the diagnosis and differentiation between ventricular diverticula and aneurysms may be made in the prenatal period, from the early phases of pregnancy. Although a better prognosis has been attributed to fetuses with diverticula, adverse outcomes may result from any of the two entities. The true natural history of these rare conditions is little known and further interventions in fetuses with similar conditions should be customized.

Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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Study Association

This study is not associated with any post-graduation program.

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