Can Prenatal Diagnosis of Parachute Mitral Valve be Achieved? A Case Report of Fetal Parachute Mitral Valve

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Case Report

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Abstract

A parachute mitral valve (PMV) is a common form of congenital mitral stenosis and is difficult to diagnose prenatally. We herein report a fetal case of PMV with coarctation of the aorta diagnosed at 25 weeks’ gestation by ultrasound and supported by autopsy. We also describe the use of a new sign to achieve a prenatal diagnosis of PMV.

Background

A parachute mitral valve (PMV) is a congenital anomaly of the papillary muscles (PMs) that frequently leads to mitral stenosis (MS). Fetal MS and fetal Shone’s syndrome but not absolute PMV have been reported, but PMV was confirmed after birth in some of these cases. We herein report a case of fetal PMV diagnosed by ultrasound and confirmed to be parachute-like asymmetric mitral valve (PLAMV) by autopsy, and we describe a previously unpublished prenatal sonographic sign of this rare manifestation.

Case Presentation

A 31-year-old pregnant woman (gravida 1, para 0) was referred to our hospital for fetal echocardiography (Voluson E10; GE Healthcare, Chicago, IL, USA) at 25 weeks’ gestation. The echocardiographic examination revealed a PMV associated with coarctation of the aorta (CoA) (Fig. 1). In the four-chamber view, the mobility of the mitral valve leaflets was obviously limited without thickening in two-dimensional (2D) echocardiography (Movie 1). The shape of the mitral valve and some chordae was similar to “Ω” in diastole, especially in four-dimensional (4D) echocardiography with volume contrast imaging (Fig. 1b and Movie 2). Further tracing of the chordae demonstrated that all the chordae inserted into the posteromedial PM. The short-axis view of the left ventricle only detected the posteromedial PM, the anterolateral PM was not identified. These findings led to the diagnosis of PMV. After a systematic scan of the fetal heart, CoA and a small left ventricle were demonstrated (Z score for aortic isthmus = -3.47, Z score for left ventricular end-diastolic dimension = -2.41, and Z score for left ventricular inlet length = -1.82). During follow-up, the parents decided to terminate the pregnancy at 30 weeks’ gestation after a prenatal consultation.

After termination of the pregnancy, autopsy of the fetal heart and copy number variation sequencing of the fetal skeletal muscle were performed under the informed consent of the parents. The copy number variation was normal, and autopsy revealed that all chordae were connected to the elongated posteromedial PM (Fig. 2a). The anterolateral PM was attached over its entire length to the ventricular wall with its tip located near the mitral valve annulus (Fig. 2b). CoA was confirmed at the same time (Fig. 2c). The diagnosis of PLAMV with CoA was therefore established.

Discussion
PMV is a rare congenital mitral valve malformation mainly involving the PMs and usually resulting in congenital MS. The specific incidence of PMV has not been mentioned in the literature to date. Congenital MS is a morphologically heterogenous lesion with a variable prognosis, and it is classically divided into four anatomic types\(^3\): typical congenital MS, hypoplastic mitral valve, supramitral valve ring, and PMV. Congenital MS is also divided into two functional types\(^4\): type A (with normal PMs) and type B (with abnormal PMs, including PMV and hammock mitral valve). Different types have different treatment strategies and prognoses. Although the long-term functional outcome of MV repair in children with congenital MS is satisfactory, surgical procedures for PMV are more complicated than other types, and repeat repair may be needed\(^5\).

Two subtypes of PMV have been described according to the underlying anatomy\(^6\): “true” PMV, which is characterized by a single PM that receives all chordae; and PLAMV, which has two PMs with unequally distributed chordae. PLAMV is further divided into three grades according to the degree of uneven distribution of the chordae. In grade II, the most common form, a few short chordae tendineae are attached to the elongated PM\(^6\). Grade III is the most severe\(^6\). The present case involved a grade III PLAMV with two PMs: one PM received almost all the chordae, and the tip of the other PM was located near the mitral valve annulus and was attached at its lateral side to the left ventricle wall. Aside from the anatomical differences in the PMs, both of these subtypes of PMV may have shortened or thickened chordae tendineae, and thickened mitral valve leaflets have been seen in some cases\(^7,8\). Additionally, there is no significant difference in the clinical manifestation, treatment, or prognosis between PLAMV and “true” PMV\(^9\); therefore, most scholars have classified them into one category in clinical research.

Prenatal ultrasound diagnosis of PMV is challenging. Instead, this condition is usually diagnosed postnatally. In fetuses, MS has been commonly reported, but PMV has not\(^1,2,10\). A restrictive opening of the mitral valve is an important clue for fetal MS\(^10\). Further differential diagnosis of MS has great value for prenatal consultation, but it is difficult. In children and adults, the echocardiographic characteristics of PMV include a single PM at the mid-papillary level and parachute leaflets at the basal level in the left ventricular short-axis view, doming of the elongated chords in diastole, and a pear-like shape of the left atrium in the four-chamber view\(^8\). In the fetus, however, the anatomy of the PMs has been considered to be inaccurately identified at the mid-papillary level in the left ventricular short-axis view\(^1,10\). In our case, fetal echocardiography failed to demonstrate the anterolateral PM. Thus, diagnosis of PMV by only observing the number and position of the PMs is unreliable. However, we found that observing the morphology and movement of the valve leaflets and chordae, especially in the four-chamber view, was a useful method for prenatal diagnosis of PMV. 4D echocardiography is more visual than 2D. The mitral valve “Ω” sign described in this case was a key diagnostic clue and is formed by the anterior leaflet, posterior leaflet, and several chordae. In diastole, the tips of mitral valve leaflets exhibits limited opening because of the narrowed interchordal spaces. The converging chordae and the tips of the leaflets form the arch-shaped portion of the “Ω” sign, and the left parts of the leaflets resemble the horizontal portion. 2D The mitral valve “Ω” sign can also be used to differentiate PMV from typical congenital MS and hypoplastic mitral valve. Typical congenital MS is characterized by thickened and rolled leaflets,
thickened and shortened chords, and restrictive opening of the mitral valve\textsuperscript{10}. In a hypoplastic mitral valve, all components of the mitral valve are miniature versions of those of a normal valve\textsuperscript{11}.

**Conclusion**

We have herein reported a case of PMV with CoA diagnosed by echocardiography and confirmed by autopsy. This is the first report of the mitral valve “Ω” sign in a fetus with PMV. This sign provides a strong basis for the diagnosis of fetal PMV in the second trimester of pregnancy, and it may help to advance the diagnostic rate of PMV.

**Declarations**

Ethical Approval and Consent to participate

We further confirm that the data collection covered in this manuscript that has involved our patient has been approved by the ethics committee of West China Second University Hospital of Sichuan University. And written informed consent was obtained from the parent of the patient.

Consent for publication

Written informed consent for publication of the clinical details and images was obtained from the parent of the patient.

Availability of data and materials

The data and materials are available from the corresponding author on reasonable request.

Competing interests

The authors declare that they have no competing interests.

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Authors’ contributions

Xiaohui Dai and Jiao Chen wrote the main manuscript text. Xiaohui Dai, Jiao Chen and Fumin Zhao prepared figures and movies. Hanmin Liu and Lin Wu participated in the follow-up of the patient. All authors read and approved the final manuscript.
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Figures
Figure 1

Fetal echocardiography images. (a) The two-dimensional four-chamber view showed that limited opening mitral valve in diastole (arrow). (b) Four-dimensional volume contrast imaging showed the shape of the limited opening mitral valve in diastole was similar to "Ω" (arrow) in four-chamber view. (c) The thickened chordae tendinea converged into a single PM (arrow) in four-dimensional volume contrast imaging. (d) The aortic arch long-axis view showed that the aortic arch and the isthmus (arrow) were remarkably narrowed.

LV, left ventricle; RV, right ventricle; LA, left atrium; RA, right atrium; IA, innominate artery; LCA, left common carotid artery; LSA, left subclavian artery; DAO, descending aorta.

Figure 2

Autopsy of the fetal heart. All chordae tendinea inserted into the posteromedial papillary muscle (a). The anterolateral PM was elongated with its lateral side attached to the LV wall without chords connection (b). The aortic arch was narrowed (c), especially the isthmus (arrow).

P, posteromedial papillary muscle; A, anterolateral papillary muscle; AML, anterior mitral valve leaflet; PML, posterior mitral valve leaflet; AAO, ascending aorta; IA, innominate artery; LCA, left common carotid artery; LSA, left subclavian artery; DAO, descending aorta.
Supplementary Files

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