# An arcade in the heart: Multimodality imaging

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## ABSTRACT

**Congenital mitral stenosis (MS) is a spectrum of anomalies that result in functional and anatomic obstruction of inflow into the left ventricle. Mitral arcade is one of the varieties of congenital MS where there is an abnormal development of chordae tendineae, resulting in stenosis, regurgitation, or both. Here, we describe the case of a mitral arcade in a child, which was diagnosed on echocardiography and confirmed with other imaging modalities.**

**Keywords:** Anomalies, congenital, hammock, mitral arcade

## INTRODUCTION

Mitral arcade is a less frequent congenital malformation of the mitral valve apparatus, first described by Layman and Edwards. This anomaly is characterized by enlarged and elongated papillary muscles connected to each other and to the free edge of the anterior mitral leaflet by a bridge of fibrous tissue which appears as an arcade. This malformation can lead to stenosis, regurgitation, or both. The defect can be diagnosed by various modalities, including echocardiography, catheterization, and radiological imaging.

## CASE REPORT

A 9-year-old girl, incidentally found to have congenital heart disease at the age of 3 years but lost for follow-up, presented with easy fatigability, exertional palpitations, and dyspnea from the past 4 years. She was born by normal vaginal delivery to nonconsanguineous parents. She had recurrent respiratory tract infections from birth. Growth and developmental milestones were normal for her age. She had no history suggestive of cyanotic spells. There was no history of interrupted feeding in infancy. The physical examination demonstrated normal hemodynamic parameters, normal breathing pattern with a respiratory rate of 18 breaths/min, and the lungs were clear. Saturation was normal in all the

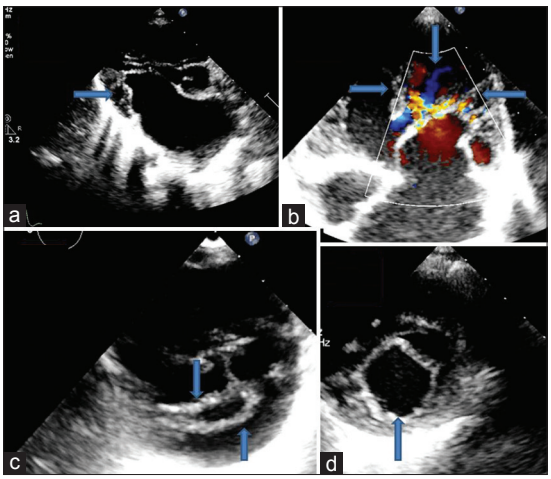


**Figure 1: Echocardiogram images – (a) Parasternal long axis showing short, thick chordae forming fibrous band. (b) Color Doppler in apical four-chamber view showing splaying appearance. (c and d) Parasternal short axis showing normal mitral valve area at annular level and hammock appearance at subvalvular level**

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four limbs and there was no clubbing. On cardiovascular examination, there was palpable P2, louder first heart sound, and normal split with louder pulmonary component of the second heart sound. There was a 2/6 grade mid-diastolic murmur at the left ventricular apex. There was a 3/6 grade pansystolic murmur in the left parasternal area. There was no jugular venous distension or hepatic engorgement.

On workup, blood parameters were within normal limits. Electrocardiography showed normal sinus rhythm, normal axis, left atrial enlargement, and right ventricular hypertrophy. Chest roentgenogram showed normal cardiothoracic ratio, straightening of the left cardiac border (suggesting left atrial appendage enlargement), pulmonary venous congestion, and plethoric lung fields. Transthoracic echocardiography showed short, thick chordae forming fibrous band with almost direct attachment of elongated papillary muscles to mitral leaflets on parasternal long-axis view [Figure 1a]. On parasternal short-axis view, there was mild thickening of mitral leaflets with normal orifice area at annular level (at valve level) and hyperechoic mass between two papillary muscles representing interconnecting fibrous band (hammock mitral valve, below the valve level) [Figure 1c and d]. On apical four-chamber view, arc-like configuration of the papillary muscles constrained by interconnecting fibrous band was noted along with a large muscular ventricular septal defect with inlet extension. On Doppler color mode in four-chamber view, there was "splaying appearance" of multiple jets through the reduced interchordal spaces involving both leaflets [Figure 1b].

On catheterization study, left ventriculogram in the right anterior oblique 30° view showed contrast negative, radiolucent area extending from the papillary muscles to mitral leaflet which represents the fibrous band [Figure 2]. On cardiac magnetic resonance imaging, diagnosis of mitral arcade was confirmed with demonstration of anatomic details [Figure 3].

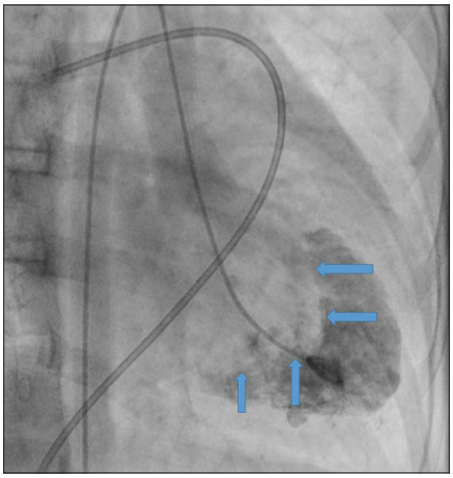

**Figure 2: Left ventriculogram showing arcade as radiolucent area**

## DISCUSSION

Congenital mitral stenosis (MS) involves abnormalities at different levels of mitral valve apparatus. Ruckman and Van Praagh[1] proposed a simple classification based on pathologic findings of autopsy specimens. Carpentier *et al*.[2] described a functional classification based on the location of the major lesion.

Mitral arcade is a less frequent malformation and was first described by Layman and Edwards in 1967. This anomaly is characterized by enlarged and elongated papillary muscles connected to each other and to the free edge of the anterior mitral leaflet by a bridge of fibrous tissue which gives appearance of an arcade. This anomaly which looks like an arcade from the left ventricular view gives an appearance of hammock when viewed from the left atrium and was well appreciated in our case. The exact developmental origin of the mitral arcade is not known, but suspected to represent an arrest of mitral valve development at a stage after loss of muscle in chordae and leaflets but before the final attenuation and elongation of mitral chordae.[3] According to Layman and Edwards,[4] the salient features of an anomalous mitral valve arcade include (a) an adequately sized mitral valve orifice; (b) short, thick, and poorly differentiated chordae with direct union of the papillary muscles to the anterior

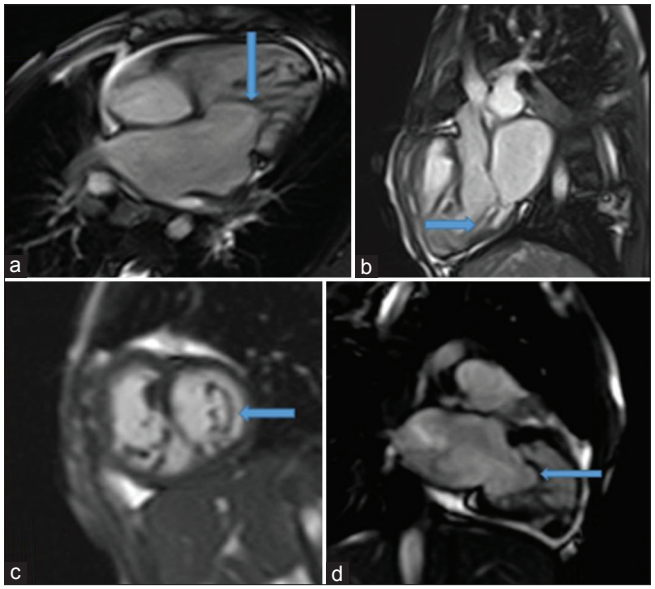


**Figure 3: Cardiac magnetic resonance imaging – four-chamber (a) and three-chamber (b) steady-state free precession images demonstrate the elongated, mildly thickened anterior and posterior mitral valve leaflet with absent chordae. Short-axis oblique (c) and two-chamber (d) images demonstrate the fibrous band between the anterolateral and posteromedial papillary muscles. (c) Hammock-like appearance with two leaflets between the papillary muscles**

leaflet; (c) narrow or nearly nonexistent spaces between the abnormal chordae; and (d) greater differentiation of the chordae attached to the posterior papillary muscle. Our case had all the features.

Diagnosis is made on echocardiocardiography, but can be supplemented by catheterization and radiological imaging. The classical echocardiographic findings of the mitral arcade were first described by Parr *et al.*in the pediatric population.[5]

Cardiac catheterization will give additional information for diagnosis. The abnormality was described as a rectangular radiolucent defect (fibrous band) extending between the mitral valve and papillary muscles on ventriculogram.[6] Radiological imaging has a higher spatial resolution than echocardiography for assessing the anatomical details of the tensor apparatus and may be used as an adjunctive diagnostic tool in selected patients.[3] The patient was referred for surgical correction after catheterization study.

## CONCLUSION

Mitral arcade is a rare but important cause of progressive mitral regurgitation (MR) or MS. This anomaly should be considered in patients in whom the mechanism of MR or MS is not clear. Echocardiography is an excellent imaging technique to establish the diagnosis. Angiography is supportive for the diagnosis. Radiological imaging offers definitive supplementary information regarding anatomic details.

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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### Conflicts of interest

There are no conflicts of interest.

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