A tale of two co-existing congenital mitral pathologies: arcade & parachute

Short title: Mitral valve parachute and arcade

Rishabh Khurana¹ MD, Aro Daniela Arockiam¹ MD, Elio Haroun¹ MD, Ankit Agrawal¹ MD, Suma Thomas¹ MD, Rukmini Komarlu¹ MD, Zoran Popovic¹ MD PhD, Leonardo Rodriguez¹ MD, Gosta Pettersson¹ MD PhD, Brian Griffin¹ MD, Shinya Unai¹ MD, Tom Kai Ming Wang¹ MBChB MD

Affiliations:

1. Section of Cardiovascular Imaging, Heart and Vascular Institute, Cleveland Clinic, Ohio, United States
2. Department of Thoracic and Cardiovascular Surgery, Heart and Vascular Institute, Cleveland Clinic, Ohio, United States

Corresponding author: Tom Kai Ming Wang MBChB, MD

9500 Euclid Avenue, Main Campus, J1-5, Cleveland Clinic, Cleveland, Ohio 44195, United States.

Phone +1 2164448130; Fax: +1 2164456158; E-mail: wangt2@ccf.org

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Glossary of Abbreviations:

MA: Mitral Arcade

PMV: Parachute mitral valve

TTE: Transthoracic Echocardiography

TEE: transesophageal echocardiogram
Central Picture legend:
Echocardiography (TTE, TEE) evaluation of parachute and arcade mitral valve case

Central Message:
Resolving a diagnostic dilemma of a rare mitral valve anomaly (arcade & parachute) by multimodality imaging, to guide further cardiothoracic surgical management as well as follow up assessment.

Clinical Summary
A 26-year-old female presents with heart murmur since childhood and occasional palpitations. Physical examination was notable for soft pan-systolic murmur at the apex. Electrocardiogram was normal. Transthoracic echocardiography (TTE) revealed suspected parachute mitral valve (Figure 1, Central Image) with moderate mitral stenosis with mild-moderate mitral regurgitation, normal bi-ventricle systolic function, moderately dilated left atrium and mild pulmonary hypertension. Detailed transesophageal echocardiogram (TEE) was performed, showing mitral valve parachute (dominant anterolateral papillary muscle with majority of attachments) and arcade (shortened, thickened and rudimentary chordae), both contributing to severe sub-valvular mitral stenosis (gradients 26/13 mmHg) with moderate mitral regurgitation. Cardiac CT performed for coronary and thoracic anatomy evaluation, using Siemens Definition Force dual source 2x192-slice scanner showed similar mitral valve anatomy (Figure 2). IRB approval was not required; the patient provided informed written consent for publication of her anonymized data for this case report.
Operative findings include most of the chords were from the antero-lateral papillary muscle, the anterior head and the posterior heads were fused together, and the posterior head was flat and arcade-like providing short chords to P1 and lateral P3 (Supplemental Figure 1). The chords were individually identified and fenestrated to elongate and create a more natural papillary orientation. The anterior head was separated from the posterior head to increase mobility and opening. A secondary chord to the posterior head, which was limiting the motion, was cut. Subsequently, the P1/P2 chords were separated to further improve mobility, and Duran mitral valve annuloplasty ring (size #35) was placed to stabilize. Post-operative course was uncomplicated. Pre-discharge transthoracic echocardiogram showed stable mitral valve repair with unremarkable gradients (mitral valve gradients of 11/6 mmHg) and trivial mitral regurgitation, along with normal biventricular size, systolic function.

Discussion

Parachute mitral valve (PMV) is formed by specific malformations of the mitral leaflets per se, as well as various sub-valvular structures, giving rise to a funnel configuration of the mitral valve. The chords are short and thick with a convergent papillary insertion, whether a single papillary muscle receives all chords (true PMV), or two papillary muscles with all chordae inserting into one muscle and the other being hypoplastic, and commissures barely distinguishable and frequently underdeveloped. Mitral arcade (MA), first described in 1967, is an extremely rare congenital anomaly of the mitral chordal apparatus so diagnosis is often not considered. It is characterized by an “arc” like configuration of two papillary muscles, due to an interconnecting band of fibrous tissue that runs along the line of closure of the mitral leaflets. There is severe shortening or complete absence of chordae tendineae such that the leaflets insert almost directly into the papillary muscle. When viewed from atrial surface, the mitral valve (MV) appears like a
hammock\textsuperscript{3,4}. The fibrous band interferes with normal valvular motion, which prevents the typical apposition of mitral leaflets, thereby resulting in mitral regurgitation. Additionally, mitral stenosis may co-exist due to presence of mechanical obstruction related to prominent papillary muscles blocking the sub-valvular apparatus and inadequate opening of the mitral valve.\textsuperscript{4} Symptomatic patients present with worsening dyspnea and congestive heart failure from mitral valve dysfunction. An incidental cardiac murmur may be auscultated in asymptomatic people, years before clinical presentation, as seen in the index case.

In patients when etiology, severity and/or need for intervention of significant MR is uncertain by TTE alone, other multi-modality imaging techniques provide supplementary roles. TEE is critical in better characterizing these features. Contemporary 3D-imaging techniques with multiplanar reconstruction on TEE can accurately delineate the precise cause, location and severity of MR in the majority of cases.\textsuperscript{3} Exercise stress echocardiography can provide complementary information. Cardiac CT angiography can also precisely assess the complete anatomical details of the mitral valve apparatus including papillary muscles and chordae as demonstrated in our case. Finally, cardiac MRI can be useful to quantify MR severity by regurgitant volume and fraction, chamber quantification, and has tissue characterization abilities to assess myocardial scar and concomitant cardiomyopathy.

Adults with mitral arcade seem to have milder mitral valve dysfunction and can escape detection in early life and therefore present later from degenerative valvular apparatus over time. Medical treatment is limited to symptomatic relief of heart failure and arrhythmias if present. Intervening
on the mitral valve is similar to other etiologies when there is severe mitral stenosis and/or regurgitation, with concomitant symptoms or other secondary indications, and surgery remains the definitive strategy. Although mitral valve repair is typically preferred over replacement for MR, our case is one of the few that has reported a successful repair for mitral arcade and parachute, while replacement remains the mainstay, and transcatheter interventions have not currently been reported.³

References


Figures and Legends

Figure 1: Transthoracic (TTE) and transesophageal (TEE) echocardiography evaluation of parachute and arcade mitral valve case

A) TTE apical 2 chamber view without and with color showing mitral valve restricted opening and stenosis (white arrow) from parachute mitral valve (red arrow)

B) TEE mid-esophageal left ventricular long axis views with biplane: left image 3-chamber view showing parachute mitral valve (red arrow) with restricted opening (white arrow) and right image bicommissural view showing arcade subvalvular apparatus (yellow arrow).

C) TEE transgastric left ventricular view with biplane: left image short axis view and right image long axis view both showing parachute mitral valve (red arrow).

D) TEE three-dimensional rendered image of the mitral valve: left image en-face short axis view from left atrial side above the valve and right image short axis view from left ventricular side below valve, showing the restricted opening/obstruction (white arrow) mainly below the valve from arcade and parachute mitral valve (yellow arrow).

Figure 2: Cardiac computed tomography evaluation of parachute and arcade mitral valve case.

A) Apical 4 chamber view showing parachute mitral valve with prominent anterolateral papillary muscle attached to both leaflets (red arrow)

B) Apical 2 chamber view showing arcade of subvalvular apparatus of the mitral valve (arcade morphology – yellow arrow). Short and rudimentary chordae are seen (blue arrow)

C) Left ventricular short axis view mitral valve level showing prominent leaflet and subvalvular muscle thickening with restricted opening (orange arrow)

D) Left ventricular short axis papillary muscle level showing one prominent anterolateral papillary muscle (red arrow).
Supplemental Figure 1

Intra-operative image of the parachute and arcade mitral valve case from the left atrial aspect. It shows narrowed mitral valve orifice (orange arrow) with arcade subvalvular apparatus (*)