

Surgical Repair of a Unileaflet Mitral Valve: A Rare Congenital Abnormality and a Novel Surgical Approach



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INTRODUCTION

Congenital malformations of the mitral valve (MV) are rare, and the majority are detected in infancy and are associated with significant mortality.¹ In particular, descriptions of unileaflet MV (ULMV; either partial or complete leaflet agenesis) are limited to a few case reports.² We present the case of a 65-year-old woman who presented with exertional dyspnea. She was found on echocardiography to have an elongated anterior MV with complete absence of the posterior mitral leaflet, and she went on to undergo successful MV repair using a novel surgical approach. Imaging findings in this case provide unique insight into the embryologic basis of this abnormality and assisted in planning the surgical approach.

CASE PRESENTATION

A 65-year-old woman with long-standing systemic lupus erythematosus (diagnosed in 2003) that had been complicated by class III lupus nephritis (diagnosed in 2012) presented to her clinic appointment reporting progressive breathlessness on exertion and fatigue. She also had the comorbidities of hypertension and dyslipidemia and was usually taking the following medications: mycophenolate 1 g/day, prednisone 5 mg/day, furosemide 40 mg/day, aspirin 100 mg/day, and rosuvastatin 20 mg/day. On examination, the patient was comfortable, her vital signs were within limits, and she was noted to be in sinus rhythm. A cardiac examination revealed a harsh pansystolic murmur that radiated to her axilla and was louder with expiration. There was no evidence of fluid overload (namely, no peripheral edema), and her lung fields were clear. Electrocardiography documented sinus rhythm with no evidence of left ventricular (LV) hypertrophy.

Transthoracic echocardiography was performed and showed severe mitral regurgitation (MR). There did not appear to be a posterior leaflet but rather an elongated anterior leaflet of the MV coapting with a ridge of tissue where the posterior leaflet would normally reside (Figure 1, Video 1). There was severe MR, with an eccentric, predominantly anteriorly directed regurgitant jet with a proximal isovelocity hemispheric surface area of 9 mm and proximal isovelocity surface

area-derived effective regurgitant orifice area of 0.43 cm² associated with blunting of the pulmonary vein inflow S wave. The spectral intensity of the regurgitant jet was similar to forward flow, and there was a suggestion of a V cutoff sign to the spectral signal (Figure 2). Only a single papillary muscle (posteromedial) was noted on the parasternal short-axis view at the midcavity level. In association with the above, there was evidence of adverse remodeling, with moderate LV dilation (indexed LV end-diastolic volume 79 mL/m²; normal range, <62 mL/m²), severe left atrial dilation (indexed left atrial volume 63 mL/m²; normal range, <34 mL/m²), and moderate pulmonary hypertension (right ventricular systolic pressure 49 mm Hg). LV systolic function appeared preserved, with an ejection fraction of 63% (in the context of significant MR). No other morphologic abnormalities were apparent on echocardiography.

The patient was reviewed in the cardiology clinic, where it was clear that she was progressively breathless over a course of 12 to 18 months (specifically New York Heart Association functional class II dyspnea), worsening over the past 3 to 4 months. Her exercise capacity was limited because of her breathlessness, without features of paroxysmal nocturnal dyspnea, orthopnea, or ankle edema. Following clinic review, the following tests were scheduled: transesophageal echocardiography (TEE), exercise stress echocardiography, cardiac computed tomography, and cardiac magnetic resonance imaging.

TEE revealed absence of the posterior mitral leaflet (Figure 3). The anterior leaflet was slightly thickened and elongated (Video 2). There appeared to be multiple regurgitant jets along the line of coaptation between the anterior mitral leaflet and the rim of tissue along the line of the posterior leaflet. The largest jet had a proximal isovolumic surface area radius of 1.0 cm when the aliasing velocity (Nyquist limit) was reduced to 40 cm/sec. Continuous-wave Doppler through the mitral regurgitant jet showed a dense holosystolic envelope, with the spectral intensity of the regurgitant jet similar to the intensity of the antegrade flow, and the peak early mitral inflow velocity was elevated at 121 cm/sec. Systolic flow reversal was noted in the left and right upper pulmonary veins. The subvalvular apparatus linked to the posterior leaflet appeared deficient. LV systolic function was noted to be normal on TEE.

Exercise stress echocardiography performed to objectively quantify the patient's symptoms, exercise capacity, and LV contractile reserve showed that the patient had mildly reduced exercise tolerance, reaching only 6.1 metabolic equivalents, with the test being limited by marked dyspnea. Poststress echocardiography showed that there was impaired contractile reserve with exercise, with mild global deterioration in LV ejection fraction at peak exercise.

Cardiac computed tomography showed normal coronary anatomy with right coronary dominance and normal coronary ostial position. There was evidence of mild coronary disease in the form of calcified atherosclerotic plaque in the left anterior descending artery and right

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VIDEO HIGHLIGHTS

Video 1: Three-dimensional transthoracic echocardiography. Three-dimensional view of the left ventricle in short axis at the level of MV showing single mobile leaflet anteriorly.

Video 2: Three-dimensional TEE. Surgical view of the MV seen en face through the left atrial aspect. Posterior mitral leaflet is noted to be absent.

Video 3: Cine magnetic resonance imaging of ULMV. Cine of the MV and left ventricle showing single mobile anterior leaflet with absent posterior leaflet.

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coronary artery, with only minor luminal irregularity. The mitral leaflet was abnormal, with a mildly thickened anterior leaflet and absent posterior leaflet. The papillary muscle architecture was abnormal, with trabeculations along the basal lateral part of the left ventricle with what appeared to be the remnants of a rudimentary papillary muscle

forming within a muscle meshwork. The posteromedial papillary muscle was more well formed than the anterolateral papillary muscle (Figure 4). There was also evidence of mildly thickened aortic valve leaflet with a mild central leaflet coaptation defect associated with trivial aortic regurgitation.

Cardiac magnetic resonance imaging demonstrated an elongated anterior leaflet with atresia of the posterior mitral leaflet (Video 3). LV trabeculations along the basal and mid lateral wall were noted to merge together with anterolateral papillary muscle to form a muscle band that inserted into the base of the basal anterolateral wall. There was atresia of the posterior MV leaflet. Therefore, the patient was characterized as having a single functioning elongated anterior MV leaflet that coapted with the muscle band along the basal anterolateral LV wall as described above. MR was further quantified on the basis of the integrated LV quantitative volume and aortic flow method (MR volume was 61 mL and regurgitation fraction approximately 46%, consistent with severe MR). Overall the left ventricle was moderately dilated, with normal LV systolic function. The basal septum measured 1.2 cm, with the rest of the myocardium normal in thickness. Quantitative values were as follows: LV end-diastolic volume index 119 mL/m², LV ejection fraction 64%, and LV myocardial mass index 66 g/m². There was no convincing evidence of myocardial edema on T2-weighted imaging. There was no convincing evidence of

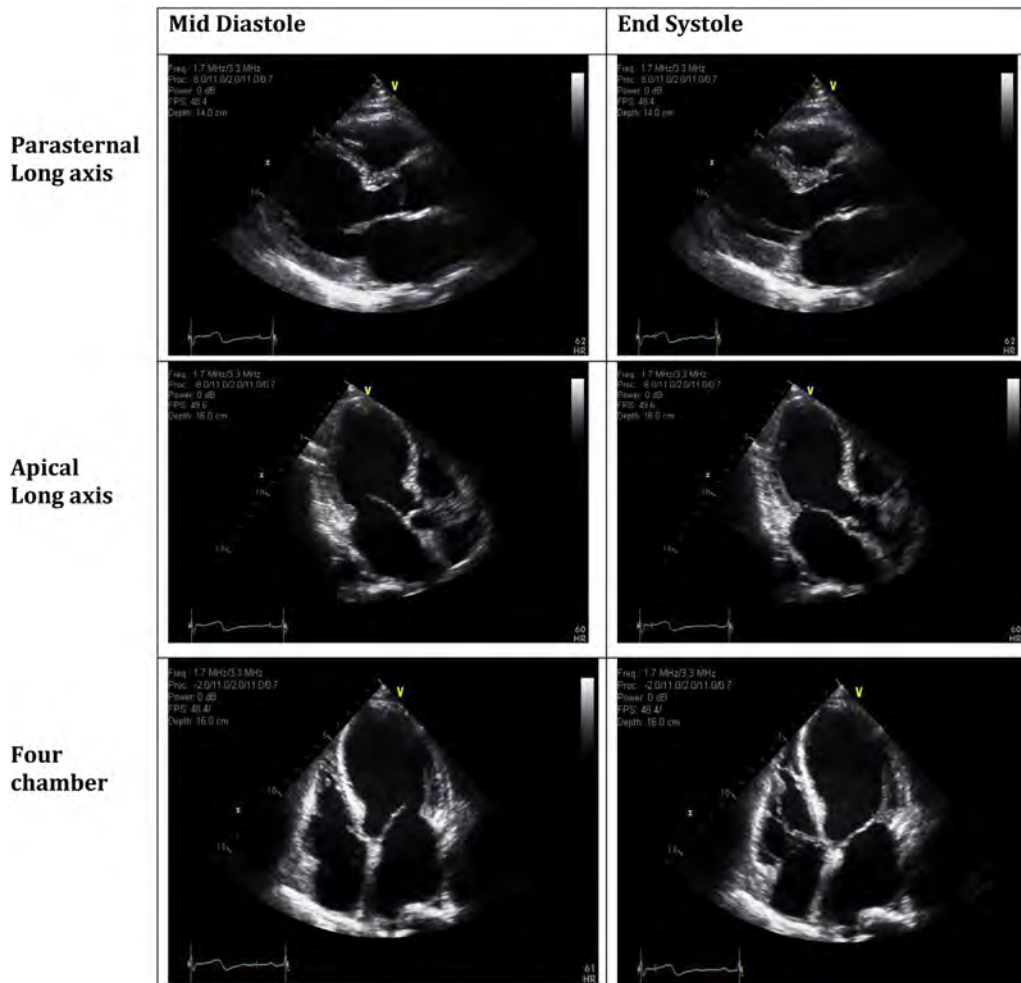


Figure 1 Echocardiogram with parasternal and apical long-axis and four-chamber views illustrating the ULMV. Transthoracic views showing mid-diastolic and end-systolic frames to illustrate ULMV. The posterior mitral leaflet is not seen in any of the projections.

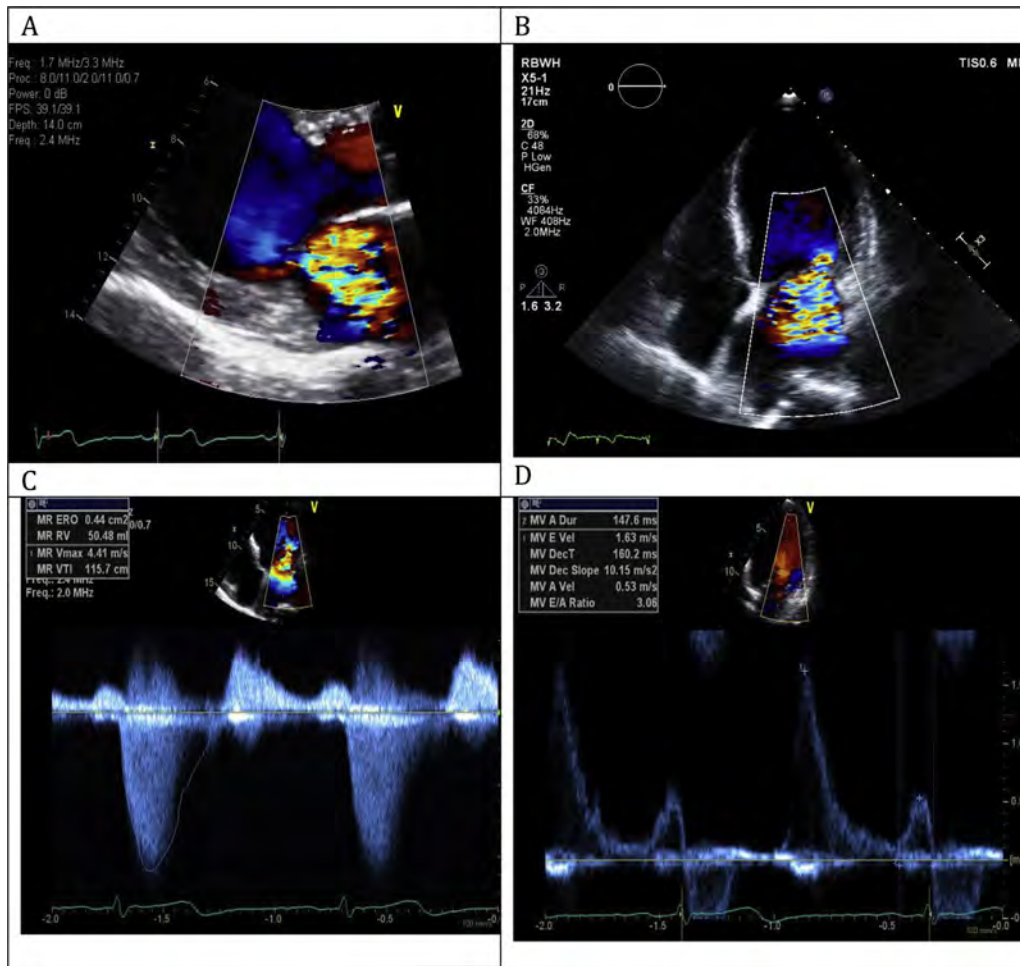


Figure 2 Severe MR on Doppler echocardiography. **(A, B)** Color Doppler showing a broad, eccentric regurgitant jet in parasternal long-axis and apical four-chamber views. **(C)** Continuous-wave Doppler showing dense MR envelope with V cutoff sign. **(D)** Pulsed-wave Doppler showing elevated E-wave velocity consistent with severe MR.

LV infiltration, fibrosis, or prior ischemic damage on gadolinium delayed enhanced imaging. Normal vascular connections were noted.

On the basis of the investigations described above, an assessment of severe symptomatic MR with evidence of LV dilatation and impaired contractile reserve associated with moderate pulmonary hypertension was made, and the patient was referred to the cardiothoracic department for consideration of MV surgery. Given the MV anatomy and the patient's age, a replacement with a prosthetic MV was considered the most likely surgical alternative. However, the patient was extremely reluctant to take warfarin because of complications of warfarin use in family and friends, and she was keen for repair if feasible. The risk for bleeding calculated with the HAS-BLED score was noted to be low in the patient, however. The likelihood of surgical repair was considered low, but the surgeon agreed to attempt repair in the first instance after a detailed review of the available imaging. After detailed discussion of the surgical possibilities, the patient provided informed consent for attempted MV repair, the success of which would be assessed intraoperatively, and failing which a prosthetic replacement would be deployed.

Several factors relevant to a repair were taken into account from a review of the available imaging as well as a literature review of previous attempts at surgical repair in ULMV. In terms of the few previously reported repairs in the literature ($n = 5$), several common elements

were noted. A restrictive annuloplasty was used in the majority of cases.¹ Augmentation of the posterior leaflet with a bovine pericardial patch was reported in some studies. Mobilization of the posterior leaflet had been performed only occasionally. Failure of successful repair necessitating MV replacement with a prosthetic valve was also reported in several cases.^{1,2} In terms of cardiac imaging, careful review of echocardiography, cardiac computed tomography, and cardiac magnetic resonance imaging suggested a non-delaminated leaflet in this case, offering the possibility of freeing up and mobilizing the posterior leaflet by dissecting in the plane of non-delamination, as shown in [Figure 1](#). This was anticipated to create a native leaflet without the need for patch augmentation. The anterior leaflet was elongated, and the potential for resection of the anterior leaflet with bileaflet repair was also planned for in case of postrepair prolapse.

At surgery, MV repair with augmentation of the posterior mitral leaflet with a CardioCel patch (LeMaitre Vascular, Burlington, MA) and annuloplasty with a 38-mm Carpentier-Edwards Physio II annuloplasty ring (Edwards Lifesciences, Irvine, CA) was performed. Surgery was performed with a routine median sternotomy on extracorporeal circulation. The posterior leaflet was noted to be rudimentary and laminated to the LV wall following cardiotomy, confirming the findings on cardiac imaging. Rudimentary papillary muscle and chords were also noted to be attached along the posterior wall. The

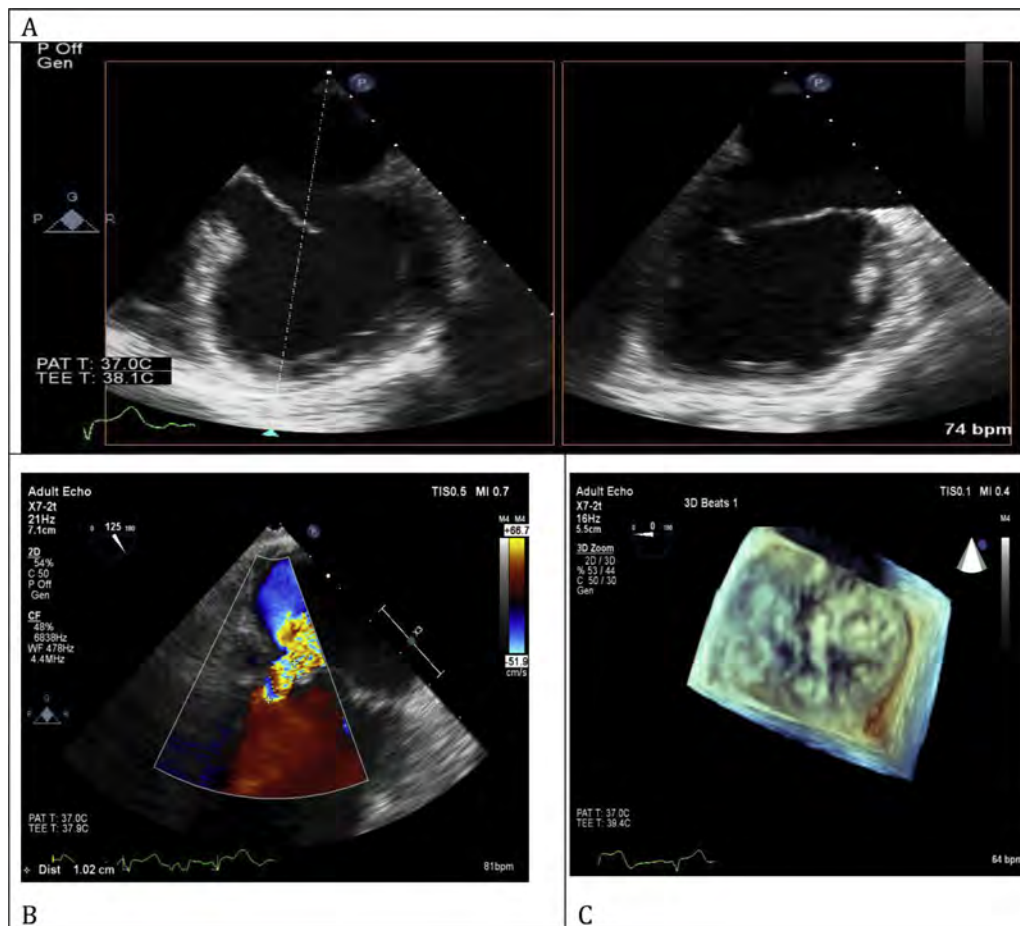


Figure 3 Transesophageal echocardiography. **(A)** Transesophageal midesophageal images showing the elongated anterior mitral leaflet and absence of significant posterior leaflet tissue. **(B)** Color Doppler illustrating significant regurgitant flow. **(C)** Real-time three-dimensional short-axis view echocardiography with en face visualization of the MV.

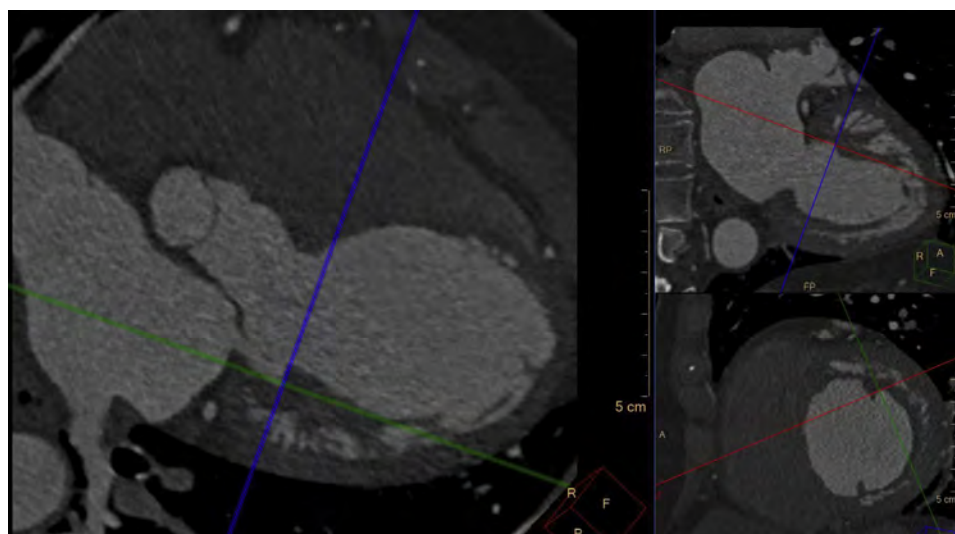


Figure 4 Cardiac computed tomography demonstrating non-delaminated posterior leaflet. Three projections through the left ventricle showing non-delaminated posterior leaflet with rudimentary remnants of subvalvular apparatus.

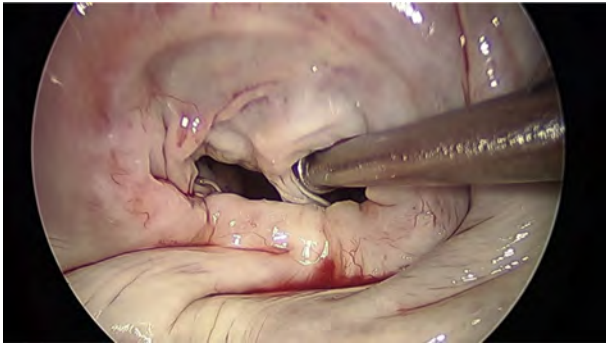


Figure 5 Intraoperative view before intervention highlighting the ULMV. En face surgeon's view through the left atrium shows anterior MV leaflet with attached chordae anteriorly with complete absent posterior leaflet and subvalvular apparatus.

anterior leaflet was noted to be slightly thickened but otherwise satisfactory (Figure 5). A CardioCel patch was applied, sutured anteriorly to the liberated papillary muscle tissue and posteriorly to the MV annulus. The leaflet was then delaminated and detached from posterior annulus, forming a neoleaflet with an annular and papillary attachment and reinforced with initially a 34-mm annuloplasty ring. Intraoperative TEE following this initial repair revealed a bulging pericardial patch associated with significant systolic anterior motion of the MV and moderate to severe MR. A decision was made to revise the repair, and the patient was reopened. The revision was performed with a 38-mm annuloplasty ring and quadrangular resection of part of the posterior neoleaflet. Repeat intraoperative TEE following these modifications revealed trivial MR and good coaptation at the A1-P1, P2-A2, and A3-P3 coaptation points (Figure 6). This was accepted as a successful repair, and the patient was successfully closed and weaned off bypass. The patient had an unremarkable postoperative course. Repeat echocardiography before discharge showed intact repair with only trivial MR.

Outpatient echocardiography at 3 months revealed only trivial MR, and the patient reported alleviation of her symptoms. Echocardiography at 12 months demonstrated a durable repair, with only mild residual regurgitation. The previously observed adverse remodeling and hemodynamic features had normalized (LV end-diastolic volume 51.2 mL/m², right ventricular systolic pressure 28 mm Hg), and



Figure 6 Intraoperative photograph of a successful repair with pericardial patch. En face surgeon's view after repair shows the annuloplasty ring in situ and refashioned posterior neoleaflet coapting with the anterior leaflet.

most important, the patient continued to report marked reduction of her exertional symptoms.

Clinic review at 24 months revealed sustained reduction of symptoms, with no reported complications from the procedure and durable repair on follow-up echocardiography. Repeat echocardiography at this time showed an ejection fraction of 61%, 2/4 aortic regurgitation, and a satisfactory repair with 1 to 2/4 MR, a mean gradient of 6 mm Hg, and 1/4 TR. Annual clinical and follow-up echocardiography is planned for the patient.

DISCUSSION

Clinically significant congenital MV lesions are rare, and in particular, ULMV is the rarest of these congenital anomalies, with only 21 previously reported cases.³ Congenital mitral stenosis, mitral atresia, accessory valvular tissue, and cleft MV are much more commonly reported.^{2,4} Complete absence of the MV is usually considered to be incompatible with life past the neonatal period and associated with severe MR. The prevalence of ULMV was estimated to be 1:8,800 in a German population,⁴ but the true prevalence is likely far less. The cause of the abnormality is unknown, but imaging findings in this case provide insights and clues to an embryologic basis. The normal development of the posterior MV structures begins in the sixth week of gestation with loosening of sheets of mesenchymal cells from the compact mesenchyme of the left lateral wall of the atrioventricular canal, in a process termed delamination. The delaminated tissue then undergoes differentiation, cranially becoming connective tissue forming the posterior valve leaflet and chordae tendineae and caudally becoming muscular tissue forming the anterolateral papillary muscle.⁵ Imaging in our patient demonstrated rudimentary valvular structures, including the remnants of a rudimentary anterolateral papillary muscle, laminated to the posterior LV wall, suggesting an arrest in posterior leaflet delamination as the pathologic abnormality. Although ULMV is most commonly described in isolation, it also has been associated with bicuspid aortic valve, atrial septal defect, and Williams syndrome. Two familial clusters have been reported, with a mother and two siblings in one family and two siblings in another. All other cases are sporadic, with male and female individuals equally affected.³

To the best of our knowledge, this case remains the single documented report of successful repair of ULMV in a symptomatic adult with subsequent documented follow-up at 24 months. MV repair is the preferred surgical procedure for MR and is currently the most commonly performed procedure for MR in North America.⁶ Surgical alternatives include MV replacement with either a prosthetic or a bioprosthetic valve. Generally speaking, MV repair is desirable and should be attempted, as the subvalvular apparatus and ventricular geometry are conserved, thereby preserving LV function.⁶ In addition, there is associated lower operative mortality and avoidance of prosthetic valve-related complications such as thromboembolism, anticoagulation-related hemorrhage, and endocarditis.⁷ There is emerging evidence attesting to the improving durability of MV repair.⁸ However, there are obvious technical challenges to achieving successful MV repairs in patients with absent posterior leaflets.

A literature review of published cases of ULMV surgery showed that clinically significant MR (moderate to severe) was reported in 12 of the 21 published cases. Described mechanisms leading to regurgitation include annular dilation, leaflet prolapse, and chordal rupture.⁵ Five of these patients underwent MV replacement, and a further five underwent repair.⁹⁻¹⁴ Tables 1 and 2 highlight surgical

Table 1 Unileaflet/hypoplastic posterior mitral leaflet cases undergoing surgery

	<i>n</i>	Age, y/sex	Presenting symptom	TTE/TEE	Repair/replacement	Outcome
de Agustin <i>et al.</i> ¹⁵	1	73/F	Dyspnea, 4/6 pansystolic murmur	Elongated and mobile AMVL and mild degree of prolapse, with a small PMVL	The patient was successfully treated using MV replacement, and no complications occurred.	Annual follow-up with TTE and examination
Bacich <i>et al.</i> ¹⁶	1	69/F	Exertional dyspnea	Severe MR, complete prolapse of AMVL	Surgical inspection confirmed a rudimentary PMVL adherent to the left ventricle. The valve was successfully replaced with a 29-mm Hancock biologic prosthetic valve, with preservation of anterior and posterior tensor apparatus.	Reduction of MR
Joshi <i>et al.</i> ¹⁷	1	66/F	Exertional dyspnea, known murmur since childhood	Severe MR	Surgical finding of congenital atresia of PMVL. Choice of valve replacement was made over valve repair given age and likelihood of needing further repair. Anterior leaflet was excised, and a 29-mm St. Jude Medical mechanical valve was inserted.	Functional MR at 6 wk
Cacioli <i>et al.</i> ¹⁸	1	14/M	Progressive worsening of dyspnea	Severe MR, hypoplasia of PMVL	Surgical inspection confirmed PMVL hypoplasia. The AMVL was large and thin. Decision for MV repair by restrictive annuloplasty with a 28-mm saddle ring (St. Jude Medical), with satisfactory valvular continence	TTE at 11 mo showed no MR
Kalangos <i>et al.</i> ¹⁹	1	10/F	Progressive worsening of dyspnea	Large mobile AMVL, virtually absent PMVL	Surgical evaluation revealed dilated mitral annulus and mobile AMVL, along with two groups of chordae tendineae arising from the AMVL inserted into small conical papillary muscle in the posterior ventricular wall. Mitral valve repair was performed by restrictive annuloplasty with a 32-mm Carpentier Edwards ring.	TTE at 12 mo showed mobile anterior leaflet without any residual leak or stenosis
Yazdan-Ashoori <i>et al.</i> ²⁰	1	76/M	Acute pulmonary edema requiring mechanical ventilation	Elongated AMVL, with hypoplastic PMVL	The patient underwent MV repair with ring annuloplasty and chordal transfer from posterior annulus to A2.	ND

(Continued)

Table 1 (Continued)

	<i>n</i>	Age, y/sex	Presenting symptom	TTE/TEE	Repair/replacement	Outcome
Stojanovic <i>et al.</i> ⁹	1	29/F	Marked LV dysfunction and hammock MV	3/4 MR from PMVL immobility	The patient was found at surgery to have an almost immobile PMVL, with severe hypoplasia of the P2 area. The remnants of P2 were excised and remaining posterior leaflet detached from annulus. Two triangular pericardial patches were used to enhance P1 and P3, and a saddle ring (no. 30; St. Jude Medical) was used for annuloplasty.	TTE at 6 mo showed no MR
Saura <i>et al.</i> ¹⁰	1	51/M	Acute pulmonary edema	Severe MR and AR, with bicuspid AV, virtual absence of PMVL	MV repair was performed with annuloplasty, with failed aortic repair, leading to biological prosthesis.	ND
Zhang <i>et al.</i> ¹¹	1	5/M	Severe dyspnea	ULMV with severe mitral stenosis and mild insufficiency,	Intraoperative views showed that the mitral leaflets were not separated as AMVL and PMVL and developed as a membrane-like structure instead, without any trace of commissures. MV repair was deemed inappropriate, and the patient received successful replacement (17AGFN-756; St. Jude Medical).	ND
Ozkan <i>et al.</i> ¹²	1	62/M	Dyspnea and acute pulmonary congestion	ULMV with severe MR	Surgical evaluation confirmed agenesis of PMVL with absent chordae tendineae and papillary muscle, with severe degeneration of the AMVL and subvalvular apparatus. Excision of the AMVL was performed, with replacement with a no. 31 St. Jude Medical mechanical prosthetic valve.	ND

AMVL, Anterior MV leaflet; AR, aortic regurgitation; AV, aortic valve; LV, left ventricular; ND, not documented; PMVL, posterior MV leaflet; TEE, transesophageal echocardiography; TTE, transthoracic echocardiography.

and nonsurgical cases, respectively. Stojanovic *et al.*⁹ used pericardial patches to enlarge the posterior leaflet area in a patient with a hammock MV, in addition to performing an annuloplasty. Their post-operative results were reported to be successful, but follow-up of only 6 months was documented. Some cases of repair involved restrictive annuloplasty rings without intervention on the posterior leaflet itself.¹⁰⁻¹⁴

In summary, we present a rare case of a successful ULMV repair with several unique aspects: (1) the repair was an Australasian first, which introduced a novel approach to MV surgery in our region; (2) multimodality cardiac imaging was used extensively to plan and predict a successful repair, and operative findings were predicted to a high degree by preoperative imaging, allowing the surgeon to plan a successful repair; (3) this is the first reported case to use a tissue-

Table 2 Unileaflet/hypoplastic posterior mitral leaflet cases not undergoing surgery

	<i>n</i>	Age, y/sex	Case description	TTE/TEE	Repair/replacement	Outcome
Kanagala <i>et al.</i> ¹³	3	18/F 17/F 46/F	Family of three, asymptomatic ULMV	Elongated AMVL with small PMVL in all three cases	Not attempted, medical management	Annual follow-up with TTE and examination
Bezgin <i>et al.</i> ¹⁴	1	45/F	Chest discomfort, dyspnea	MV with single leaflet significant LVOT	Metoprolol was used to relieve obstruction and symptoms	Symptom alleviation, annual follow-up
Pourafkari <i>et al.</i> ³	1	45/F	Dyspnea, severe MR	Elongated AMVL, hypoplastic posterior leaflet	Not attempted, patient refused	Annual follow-up
Shah <i>et al.</i> ²¹	1	22/M	Palpitations, incidental finding of ULMV	Elongated AMVL	Not attempted, medical management	Annual follow-up
Candan <i>et al.</i> ²²	1	47/M	Asymptomatic	ULMV	Not attempted, medical management	ND
Bar <i>et al.</i> ⁴	3	62/F 62/M 72/F	Asymptomatic, known pansystolic murmur Routine follow-up after aortic valve replacement Routine follow-up for known aortic regurgitation	Elongated AMVL, hypoplastic PMVL, mild to moderate MR Elongated AMVL, markedly hypoplastic PMVL, minimal mitral insufficiency Moderate aortic insufficiency, AMVL was thickened and prolapsed, PMVL hypoplastic, mild late systolic mitral insufficiency	Not attempted, all three cases medically managed	Regular TTE and examination for all three patients
Heper <i>et al.</i> ²³	1	54/F	Central cyanosis, exertional dyspnea	Absence of PMVL and ASD	Considered inoperative given central cyanosis and bidirectional shunt	Medically managed and routine clinic follow-up
Fazlinezhad <i>et al.</i> ²⁴	1	24/M	Atypical chest pain	Elongated AMVL and hypoplastic PMVL, mild MR, and moderate aortic stenosis	Not attempted	ND

AMVL, Anterior MV leaflet; AR, aortic regurgitation; ASD, atrial septal defect; AV, aortic valve; ND, not documented; PMVL, posterior MV leaflet; TEE, transesophageal echocardiography; TTE, transthoracic echocardiography.

engineered bovine patch that provides a thin but strong and pliable material that may facilitate surgical repair over more traditional techniques in this scenario; (5) durable repair was documented at long-term follow-up, which may advocate for MV repair over replacement in future cases; and (5) this case provides insights into the embryologic basis of ULMV on the basis of imaging and operative findings.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at <https://doi.org/10.1016/j.case.2020.04.009>.

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