

Etiology and Patho-anatomy of Mitral Regurgitation on Two-Dimensional Echocardiography

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Abstract

Once it is determined that a given case is of severe mitral regurgitation, elucidating the mechanism and etiology of MR is prime concern for the echocardiographer as well as for the surgeon. Exact knowledge of patho-anatomy of mitral regurgitation helps in deciding correct management of severe mitral regurgitation. It helps in deciding that if a patient is a candidate for mitral valve repair or replacement and what are chances of a successful repair. This paper provides details of various mechanisms of mitral regurgitation and patho-anatomy of common etiologies of mitral regurgitation on two-dimensional echocardiogram. Impact of this information on management of severe mitral regurgitation is also discussed in brief.

Keywords: Echocardiography; Mitral Regurgitation; Etiology; Patho-Anatomy; Mechanisms

Abbreviations

MR: Mitral Regurgitation; LV: Left Ventricle; AML: Anterior Mitral Leaflet; PML: Posterior Mitral Leaflet; PLAX: Parasternal Long Axis; LA: Left Atrium; IPMD: Interpapillary Muscle Distance

Once it is determined that a given case is of severe mitral regurgitation (MR), elucidating the mechanism and etiology of MR is prime concern for the echocardiographer as well as for the surgeon. Exact knowledge of patho-anatomy of mitral regurgitation helps in deciding correct management of severe MR. It helps in deciding that if a patient is a candidate for mitral valve repair or replacement and if repair can be performed, then, what are chances of its success.

Anatomy of mitral valve

Anatomically, mitral valve (MV) apparatus can be divided into annulus, leaflets, chordae tendineae, papillary muscles and underlying myocardium. In a normal heart, mitral annulus is connected to ventricular myocardium through papillary muscles via annular chordae. This is largely responsible for stability of mitral valve apparatus in various loading conditions.

Assessment of mitral valve annulus

Normal mitral valve annulus is saddle shape and oval. Annular diameter decreases by ~25% in systole and this corresponds to a decrease in diameter of 14%, assuming a circular shape. Annulus should be examined for size in parasternal long axis (PLAX) view which is

considered to be abnormally large if its diameter is more than 21 mm/m² of body surface area in mid diastolic frames (Figure 1). Ratio of annulus diameter to length of AML of more than 1.3 is also considered abnormal [1]. Annulus is dilated in presence of left ventricle dilatation and left atrial dilatation. Annular dilatation may be idiopathic. Annular dilatation is both a cause and effect of MR. Annulus should also be examined in PLAX, Apical 4 chamber and short axis views for presence and extent of any calcification. Annular calcification interferes in systolic contraction of annulus and thus contributes to MR.

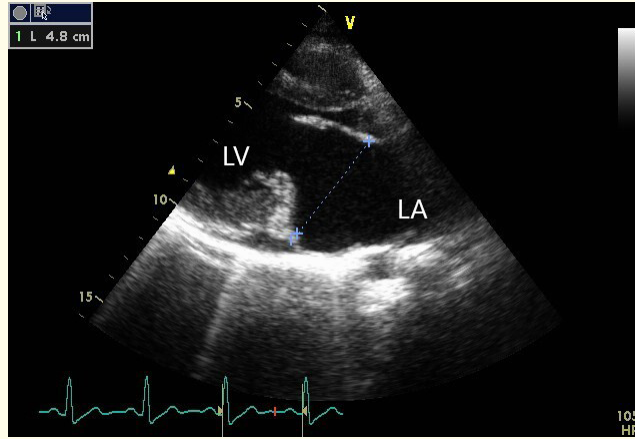


Figure 1: Parasternal long axis view showing dilated mitral valve annulus of size 4.8 cm.

Assessment of leaflets

Mitral valve has two leaflets. Anterior mitral leaflet (AML) and posterior mitral leaflet (PML). Total area of these leaflets is twice the area of annulus. AML is longer than PML and is attached to ~1/3rd of annular circumference. Area of both leaflets is nearly equal. PML is divided into three scallops P1, P2 and P3 by two indentations at the leaflet margin. P1 is towards left atrial appendage. P3 is towards tricuspid valve. Indentations in AML are not clear but are assumed to be there. Classical PLAX view shows P2 and A2 scallop. An anterior tilt of probe towards pulmonic valve shows P1 and A1 scallop while posterior tilt of probe towards tricuspid valve shows P3 and A3 scallop [1].

Thickness of mitral leaflets is measured in diastole at full excursion. Thickest portion of leaflet is measured. Normal thickness is up to 3 mm at age less than 20 years and is up to 5 mm for age more than 40 years. Between 20 - 40 years of age, up to 4 mm thickness is considered normal. It is measured at frequency > 2 MHz and frame rate more than 60 per second. Normal motion of leaflets include complete excursion of both leaflets in diastole with AML moving anteriorly and PML moving posteriorly. In systole, both leaflets join each other at tips with 3 - 5 mm overlap. This 3 - 5 mm part is called zona-coapta [2].

Mechanistically, there can be disruption of leaflet structure (perforation, thickening, shortening, rigidity retraction, calcification or mass lesions), excessive motion or reduced motion of leaflets causing MR.

Disruption of leaflet can be due to perforation caused by infective endocarditis. Post-inflammatory fibrosis and retraction causes malcoaptation leading to MR as in rheumatic heart disease or systemic lupus erythematosus. Carcinoid syndrome also causes fibrous degeneration of valve tissue. Post-radiation valvulopathy also causes fibrosis of leaflets with formation of nodules. Mass lesions like localized thickening, nodules, specks of calcification and vegetation may also cause malcoaptation which leads to MR (Figure 2 and 3).

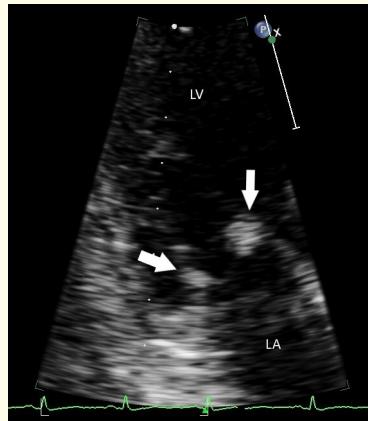


Figure 2: Zoomed view of apical long axis view showing vegetations attached to tips of anterior mitral leaflet and posterior mitral leaflet in case of ovarian carcinoma with liver metastasis, suggestive of non-bacterial thrombotic endocarditis causing moderate mitral regurgitation.

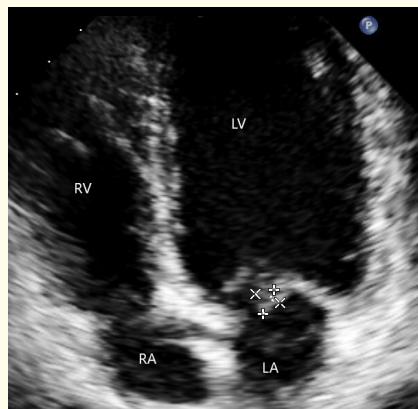


Figure 3: Apical-4-chamber view showing mass lesion on posterior mitral leaflet in a young girl with fever causing mild mitral regurgitation suggesting infective endocarditis.

“Excessive motion” of leaflets manifests as billowing, prolapse or flail leaflets. Prolapse and billowing may be due to disorder of connective tissue or may be due to elongation or redundancy of chordae tendineae. Flail leaflet is usually a result of chordal rupture but may also occur due to excessive chordal elongation. Billowing is defined as superior systolic displacement of body of leaflet, of 2 mm or more, above the annular line without any significant displacement of tip or coaptation point. It is similar to prolapse except for the fact that prolapse by definition should have tip or coaptation point displaced at least 2 mm above the annular line. In case of prolapse, the tip of leaflet faces left ventricular (LV) apex. Prolapse is best seen in PLAX view (Figure 4 and video 1). It can be identified in apical long axis view too. It is forbidden to comment on prolapse in apical-4-chamber view as it may be overestimated in this view. Prolapse not only occurs in myxomatous degeneration of mitral valve or classical Barlow’s disease, it may also occur in ischemic heart disease, rheumatic heart disease or degenerative disease of heart valve. We must try to identify the number of scallops involved in prolapse and extent of leaflet calcification. Involvement of more than 3 scallops (especially when AML is involved), extensive leaflet calcification, large central jet and annular size of more than 50 mm predict unsuccessful repair of MV in cases of prolapse. A flail segment is one which is completely separate from chordae

and its tip faces posterior left atrial (LA) wall (Figure 5 and video 2). Uncommonly, excessive elongation and redundancy of chordae can produce flail leaflet without chordal rupture. Chordal rupture is identified as a linear, usually mildly hyperechoic, free floating structure attached to one of the mitral leaflets which prolapses into LA in systole. A strong differential of ruptured chordae is linear elongated degenerated tissue arising from the leaflets. Presence of severe MR and flail leaflet favor ruptured chordae over degenerated tissue [1].

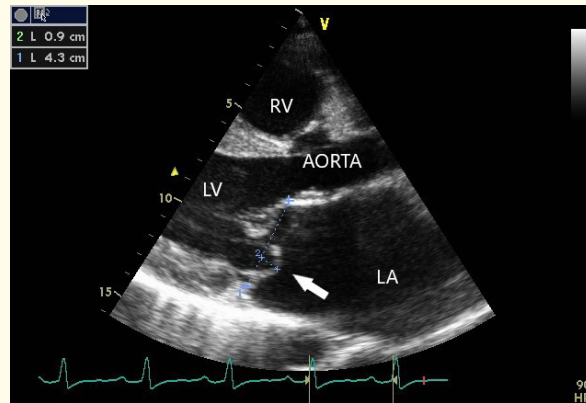


Figure 4: Parasternal long axis view showing prolapse of posterior mitral leaflet (white arrow) 9 mm below annular line.

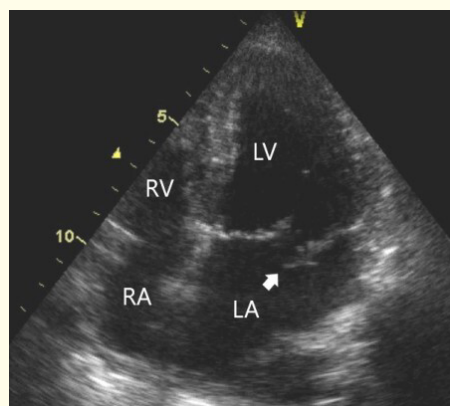
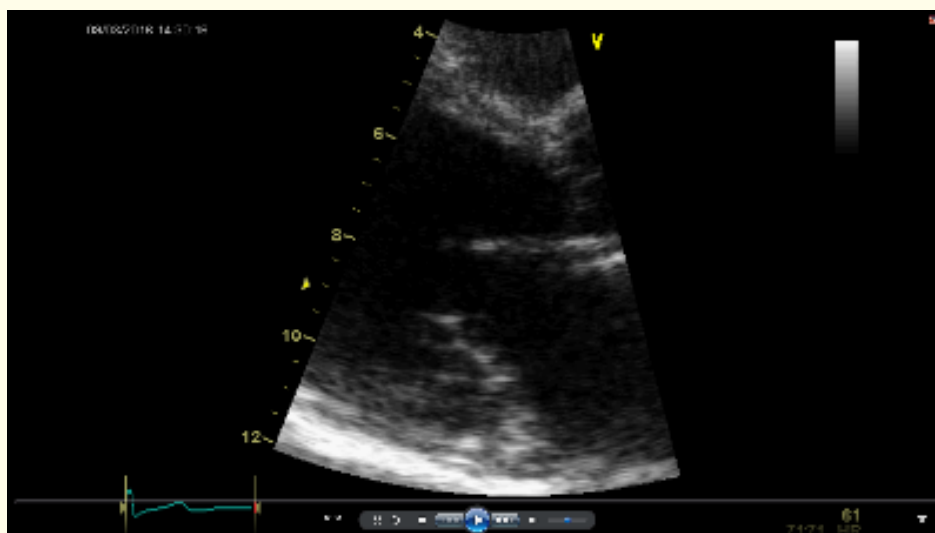
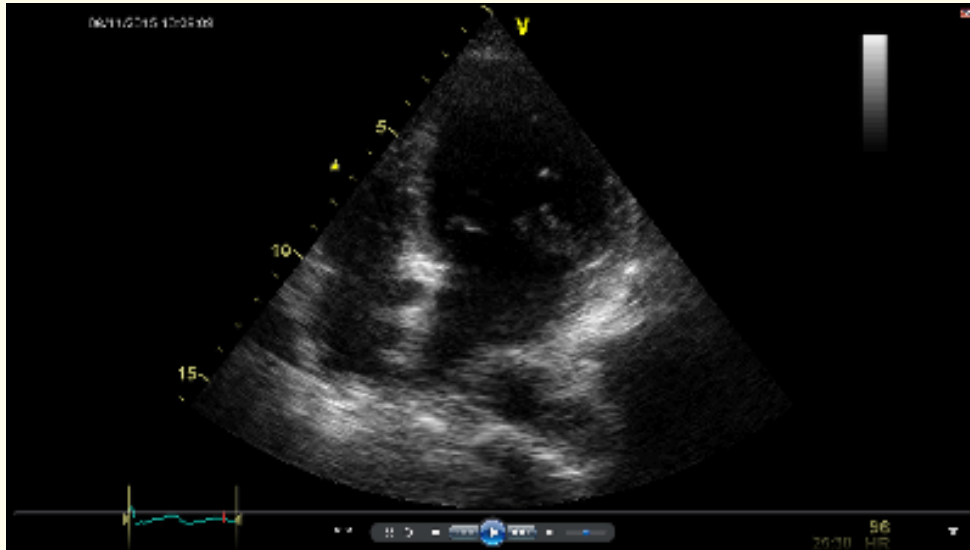


Figure 5: Apical 4 chamber view showing flail posterior mitral leaflet (white arrow).

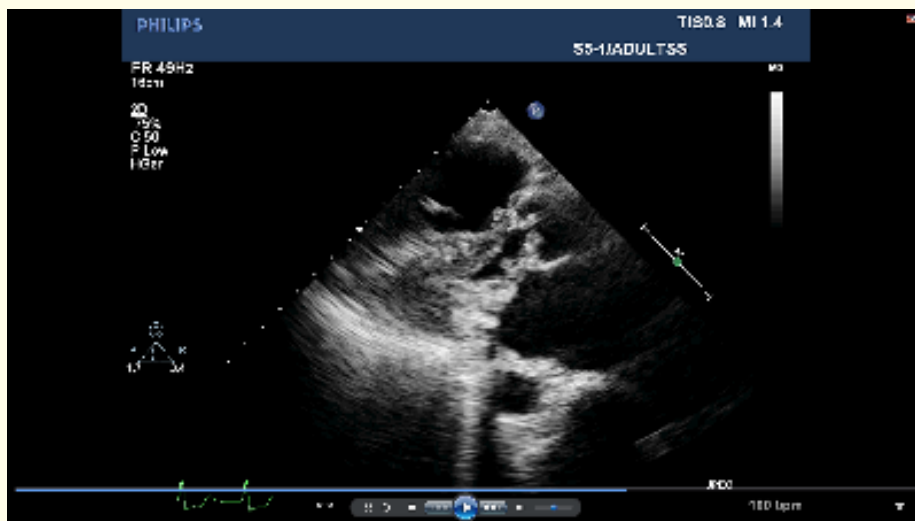


Video 1: Parasternal long axis view showing prolapse of posterior mitral leaflet.

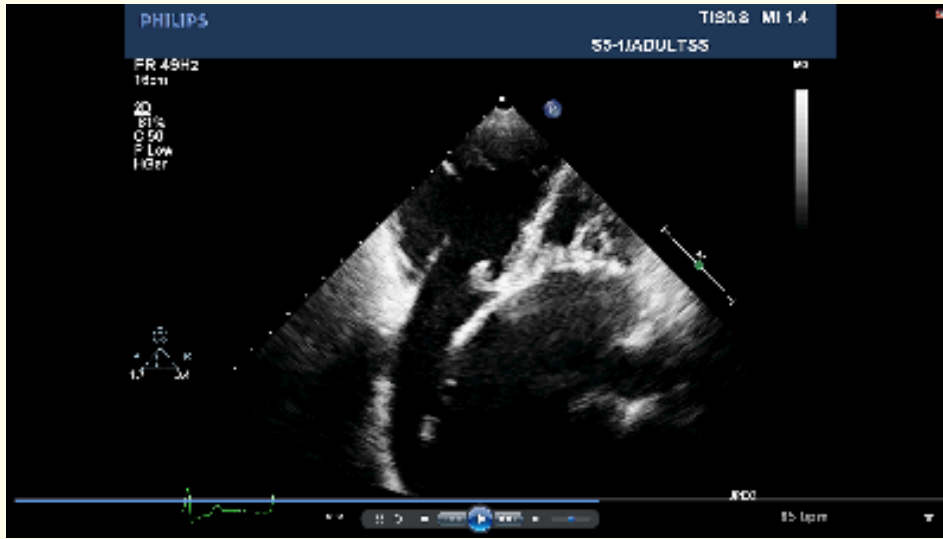


Video 2: Apical 4 chamber view showing flail posterior mitral leaflet.

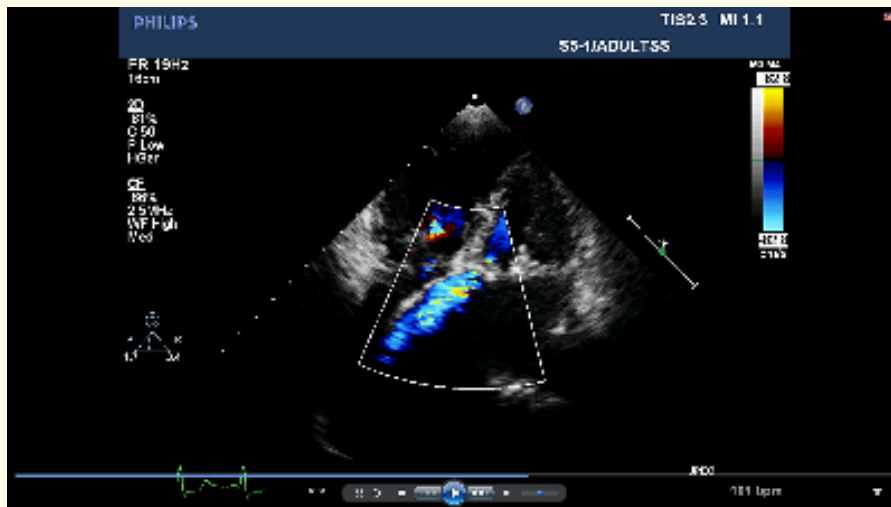
Restriction of motion of leaflets may be due to leaflet pathology or non-leaflet pathology. Leaflet restriction due to leaflet pathology occurs in rheumatic heart disease and carcinoid heart disease. There is thickening of leaflet which may or may not be associated with calcification. Degenerative heart disease may also cause leaflet restriction but it will usually occur in presence of moderate to severe calcification. Characteristically, thickening and calcification start from base of leaflet in degenerative process and it starts from tip of leaflet in rheumatic heart disease. In these disorders, leaflet movement will be restricted in systole as well as diastole (Video 3-5).



Video 3: Parasternal long axis view showing a case of rheumatic heart disease with leaflet thickening, calcification and restricted mobility. Chordal thickening, calcification and fusion are also seen.



Video 4: Apical 4 chamber of view of same patient as in video 3. Showing severely thickened and calcified mitral valve with involvement of sub-valvular apparatus. Note the thickening and restricted mobility of septal tricuspid leaflet.



Video 5: Apical 4 Chamber view of same patient as in video 4 with color Doppler showing highly eccentric jet of mitral regurgitation towards inter-atrial septum.

Non-leaflet pathologies causing restriction of leaflet mobility are ischemic heart disease and dilated cardiomyopathy. This type of MR is known as secondary MR. The leaflet movement is restricted only in systole. Primary mechanism of restriction of movement in these etiologies is tethering of leaflets. Apical and lateral displacement of papillary muscles lead to pulling of chordae away from line of coaptation. Decrease in LV contractility contributes to MR in this condition as the force opposing tethering decreases. Restriction of PML mobility causes decrease in zone of coaptation or at times causes coaptation of AML tip with body of PML what is known as pseudoprolapse of AML

(Figure 6). Tethering of PML leads to displacement of coaptation point posteriorly in LV or may cause frank non-coaptation. It causes a posteriorly directed jet of eccentric MR (Figure 7 and 8). Restriction of both leaflets lead to symmetric tenting of mitral valve. This pushes coaptation point toward apex of LV. It usually causes a central jet of MR. [3-5].

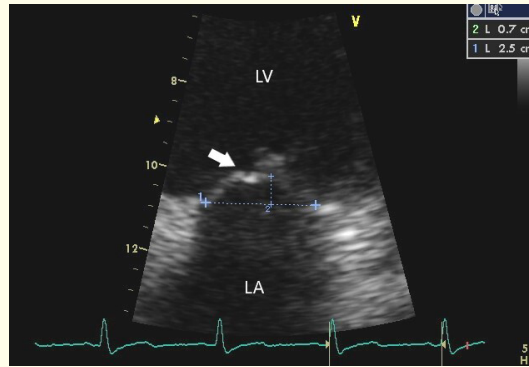


Figure 6: Zoomed section of apical-4-chamber view showing anterior mitral leaflet coapting with posterior mitral leaflet proximal to tip of posterior leaflet (pseudoprolapse).

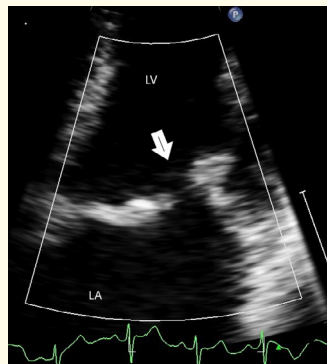


Figure 7: Zoomed section of apical-4-chamber view showing non-coaptation of leaflets with restricted posterior leaflet mobility in a case of dilated cardiomyopathy.

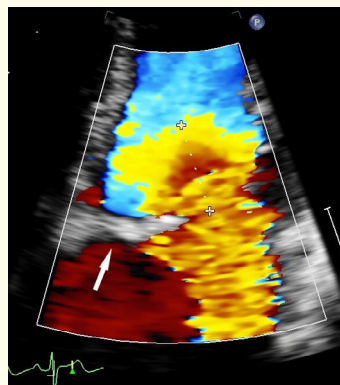


Figure 8: Zoomed section of apical-4-chamber view showing aliasing radius 1.3 cm at aliasing velocity of 42 cm/s by "Proximal isovelocity surface area (PISA)" method suggestive of severe mitral regurgitation (same patient as in figure 7).

Restriction of leaflet can be identified visually but it has to be quantified by parameters of global or regional remodeling. Regional remodeling is assessed by lateral and posterior displacement of leaflets, which in turn is measured by tenting area, tenting height (coaptation distance), postero-lateral angle, inter-papillary muscle distance and posterior papillary-fibrosa distance.

Tenting of leaflets occurs as coaptation point moves away from the annular line. Tenting of leaflets can be symmetric or asymmetric. Asymmetric tenting occurs mostly due to moderate to severe tethering of PML. This leads to decrease in zona-coapta or may cause pseudoprolapse. Symmetric tenting occurs due to tethering of both leaflets leading primarily to malcoaptation. Tenting area, tenting height and postero-lateral angle are measured in apical-4-chamber view in mid-systole. Coaptation distance (tenting height) is the vertical distance between an imaginary line connecting lateral and medial annulus and the point of coaptation of AML and PML (Figure 9). Coaptation distance of up to 7 mm is considered normal. Tenting area is the area of triangle formed between the above said imaginary line with AML and PML (Figure 10). Postero-lateral angle is the angle between this imaginary line and PML. Higher the postero-lateral angle more is the tethering of PML. Coaptation distance of 1 cm or more is usually associated with significant MR and has high likelihood of failure of MV repair. Similarly, tenting area of more than 2.5 cm² or postero-lateral angle of more than 45 degrees is associated with low success rate of MV repair. Interpapillary muscle distance (IPMD) is measured in short axis at papillary muscle level at end-diastole (Figure 11). Distance between posterior papillary muscle and anterior edge of annulus (posterior papillary fibrosa distance) is measured in apical-2-chamber view at end-diastole (Figure 12). Posterior-papillary fibrosa distance of more than 40 mm and IPMD of more than 20 mm is associated with higher likelihood of failure of mitral valve repair. Global remodeling is assessed by left ventricular volume, dimensions and sphericity index. End-diastolic LV volume of more than 140 ml, end-diastolic LV dimension of > 65 mm and end-systolic LV dimension of > 51 mm predict poor success of mitral valve repair. Sphericity index is measured as ratio of long axis to short axis of left ventricle at end-diastole. Normal value is more than 1.5. However, with severe LV remodeling it approaches 1. All these parameters assess the extent of displacement of papillary muscles away from their normal location [3-5]. Annular dilatation is a common accompaniment of restricted leaflet mobility in secondary MR. In presence of severe tethering, if one performs only ring annuloplasty, severe MR is likely to persist post-surgery.

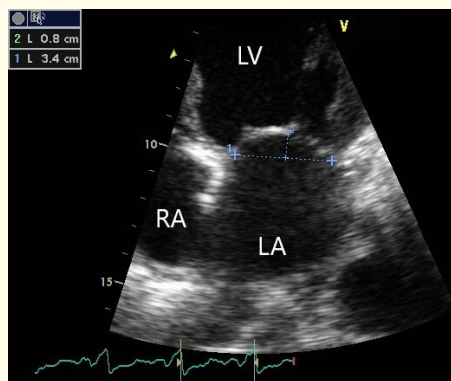


Figure 9: Zoomed section of apical-4-chamber view showing symmetrical tethering with tenting height of 8 mm.

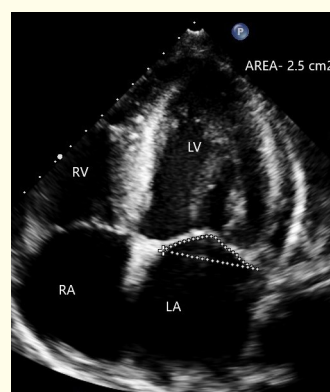


Figure 10: Apical-4-Chamber view showing mitral valve tenting area of 2.5 cm².

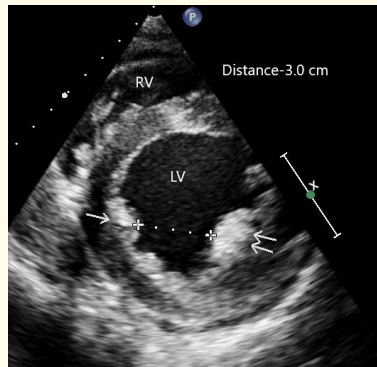


Figure 11: Parasternal short axis view at papillary muscle level showing inter-papillary muscle distance of 3.0 cm. (Single arrow- Postero-medial papillary muscle; Double arrow- Antero-lateral papillary muscle).

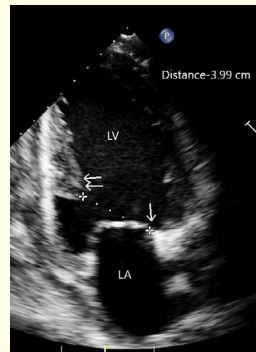


Figure 12: Apical-2-Chamber view showing distance of 3.99 cm between postero-medial papillary muscle (double arrow) and anterior edge of mitral annulus (single arrow).

Chordae tendineae

There are multiple generations of chordae from tip of papillary muscle to mitral valve leaflets/annulus. Chordae from each papillary muscle are attached to both leaflets. They are attached to leaflet margins, annulus and body of leaflets. Their thickening, shortening, fusion, rupture, elongation, calcification or redundancy leads to mitral regurgitation (Video 3-5).

Papillary muscles and underlying myocardium

Papillary muscle rupture is rare and is usually a complication of acute myocardial infarction. It gives rise to acute severe MR and is mostly fatal, unless dealt with promptly. Papillary muscle dysfunction is usually because of ischemia. This involves the underlying myocardial wall too. It leads to restriction of leaflet motion in systole and contributes to MR.

Characteristic morphological features in various common etiologies

- **Rheumatic heart disease:** Restricted PML mobility. Thickening and retraction of AML. Annular dilatation secondary to LA enlargement/atrial fibrillation. Reduced mobility of AML in systole as well as diastole due to chordal fusion. Prolapse of AML.

- **Myxomatous degeneration of mitral valve:** Thickened leaflets. Redundant tissue attached to leaflets. Chordal elongation. Prolapse of leaflets.
- **Dilated/Ischemic cardiomyopathy** - Restricted PML mobility in systole. Increased tenting height, increased tenting are Pseudoprolapse of AML. Dilated mitral annulus. Increased inter-papillary muscle distance. Increased posterior papillary-fibrosa distance. Global or regional wall motion abnormality. Flail leaflet in ischemic heart disease.
- **Sclero-degenerative valvular heart disease:** Basal and mid part of leaflets is thickened. Hyperechoic and calcific leaflets. Restricted mobility. May have flail or prolapsing leaflet.

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