

## LEARNING CORNER



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## LOOKING DEEPER INTO MITRAL REGURGITATION

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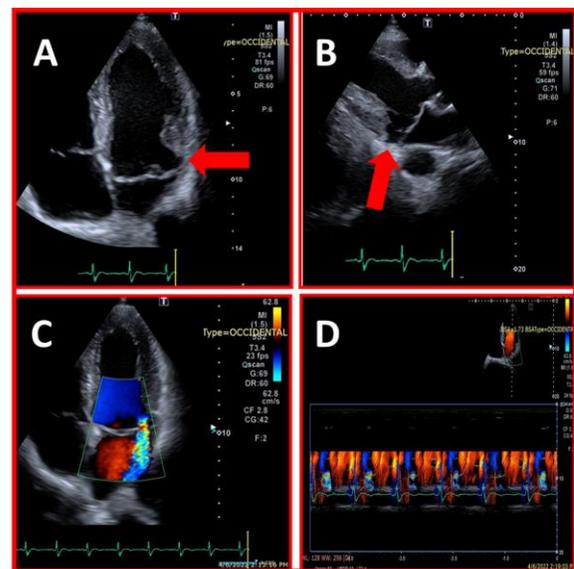
## SUMMARY

A 50 years old male patient previously treated for pulmonary Kochs presented with shortness of breath (functional class II). A non-ejection click and pan-systolic murmur radiating to axilla was heard at apex. Transthoracic echocardiography revealed thickened and markedly prolapsing both leaflets of mitral valve with mild to moderate regurgitation. A clear space was noted between the termination of LV myocardium and atrio-mitral junction (7.7mm) indicating Mitro-annular disjunction (MAD). 24 hours Holter monitoring showed 160 isolated premature ventricular contractions, maximum 20 in one hour. No other arrhythmia was recorded. Beta blocker and an ACE inhibitor were initiated with advice for follow-up as presently no intervention was needed.

## CLINICAL VIGNETTE

A 50 years old male patient previously treated for pulmonary Kochs presented with shortness of breath (functional class II). He was anemic with normal vitals. Precordial examination showed a normally placed forceful apex beat with a non-ejection click and pan-systolic murmur (grade III) at apex radiating to axilla getting prolonged in standing position. ECG was un-remarkable, CXR revealed right upper lobe fibrosis indicative of previously healed tuberculosis, and normal sized heart. Transthoracic Echocardiography revealed normal heart chambers for size, volume and function. Mitral valve leaflets were thickened with marked prolapse of both leaflets and mild to moderate mid to late systolic central regurgitation jet. A deeper look into mitral valve anatomy revealed that the atrio-mitral junction has been displaced superiorly with clear distance between the termination of LV myocardium and hinge point of atrio-mitral junction (7.7mm) indicating Mitro-annular disjunction, easily appreciated in parasternal long axis and apical 4 chamber views, Figure 1A and 1B (also see clips 1 and 2). Mitral regurgitation jet was directed parallel to posterolateral left atrial walls (video clip 3 and 4). 24 hours Holter monitoring showed 160 isolated premature ventricular contractions, maximum 20 in one hour.

No other arrhythmia was recorded. Beta blocker and an ACE inhibitor were initiated with advice for follow-up as presently no intervention was needed.



**Figure 1: Panel A and B, Trans-thoracic apical 4 chamber and parasternal long axis view showing Mitral annular disjunction (7.7 mm). Panel C, mitral regurgitation jet. Panel D color M-mode showing mid to late mitral regurgitation**

## DISCUSSION

Mitral annular disjunction is a relatively unknown entity till now although has been described some 31 years ago by Bharati et al.<sup>1</sup> The condition is usually, identified on trans-thoracic echocardiography (TTE), in parasternal long axis view as a clear space (at least 2 mm), between left ventricular myocardial wall and the hinge point of mitral valve and posterior left atrial wall junction during systole. The annulus shows a curling movement during systole and obliterates during diastole. This usually involves the inferior, posterior and lateral part of mitral annulus, as at the anterior part the posterior aortic leaflet is continuous with anterior mitral leaflet and the two fibrous trigones also intervene. Disjunction may not be completely seen all along the annulus and pathologically areas of normal annular tissue can be seen interspersed. The curling movement of annulus produces traction on papillary muscles (especially posterior) which gets fibrosed and acts as a nidus for ventricular arrhythmias. Usually noted in cases of degenerative type of mitral regurgitation (mitral valve prolapse). A study of 185 cases of severe MR due to mitral valve prolapse by Konda et al showed that MAD was noted in 24% of cases (in Barlow's disease 90% and in fibro-elastic deficiency 20.6%).<sup>2</sup> A large histopathological study of 900 patients by Hutchins et al showed that about 6 % of normal individuals also show MAD.<sup>3</sup> Identification of MAD improves as more sophisticated imaging modality is used for its detection. Mantegazza et al in their study of 131 patients showed the detection rate of 17% by TTE, 25% by Transesophageal echocardiography and 42% by cardiac magnetic resonance imaging.<sup>4</sup> Mantegazza et al in another study of 159 patients of MAD undergoing surgical repair for severe MR showed that P1 and P2 scallops (69.2%) are most commonly involved as compared to P3 scallop (30.8%).<sup>5</sup> Complications of ventricular arrhythmias with sudden cardiac death have been reported with MAD. Dejgaard et al in their study found that MAD distance (especially if greater than 8.5 mm), gadolinium enhancement in papillary muscles, lowered ejection fraction and young age are predictors of serious ventricular arrhythmias.<sup>6</sup>

## LEARNING POINTS

1. The superior displacement of the hinge point of mitral valve leaflet and left atrial wall from left ventricular myocardium is known as mitral annular disjunction.
2. The minimum separation should be greater than 2 mm to classify as MAD. Whole circumference of annulus may not be involved in disjunction process with spared areas seen in pathological specimen for individual cases.

3. MAD is seen in cases of degenerative mitral regurgitation (mitral valve prolapse) especially in Barlow's syndrome and fibro-elastic deficiency.
4. MAD is associated with ventricular arrhythmic events and sudden cardiac death. This propensity increases with fibrosis of papillary muscles especially posterior, increasing length of MAD (greater than 8.5 mm), lower ejection fraction and young age of presentation.
5. In cases of MAD with severe mitral regurgitation the treatment modalities both interventional and surgical are same as for MR due to other causes.

### QUESTION 1

The gold standard test for the diagnosis of MAD is:

- A. Transthoracic echocardiography
- B. Transesophageal echocardiography
- C. Cardiac magnetic resonance imaging

### QUESTION 2

On transthoacic echocardiography Mitral annular disjunction is most commonly seen in;

- A. Parasternal short axis view at mitral valve level
- B. Parasternal long axis view
- C. Apical 4 chamber view

### QUESTION 3

Highest prevalence of MAD is seen in cases of:

- A. Barlow's syndrome
- B. Fibro-elastic deficiency
- C. Secondary mitral regurgitation

### QUESTION 4

Most commonly involved scallop of mitral valve leaflet in MAD is:

- A. A1
- B. A2
- C. P2

## CLIPS

- Parasternal long axis view  
<https://youtu.be/Rh5nbbiRBJY>
- Apical 4 chamber view  
[https://youtu.be/ZaymUDNF\\_BI](https://youtu.be/ZaymUDNF_BI)
- Mitral regurgitation jet in parasternal long axis view  
<https://youtu.be/1ql0wDHJCbq>
- Mitral regurgitation jet in apical 4 chamber view  
<https://youtu.be/MYB0hhFUFjc>

## REFERENCES

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### ANSWERS

- *Question 1:* C
- *Question 2:* B
- *Question 3:* A
- *Question 3:* C

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