

Is Repair to Err? Unique Anatomical Considerations of the Submitral Apparatus in Hypertrophic Cardiomyopathy May Influence Mitral Valve Repair Versus Replacement

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Learning Objective

Analyze the impact of mitral valve and subvalvular abnormalities on surgical decision-making in hypertrophic cardiomyopathy (HCM) using echocardiographic and intraoperative findings.

Case Description

We present a 64-year-old male with hypertrophic cardiomyopathy (HCM), status post alcohol septal ablation nine years prior, who underwent elective mitral valve (MV) surgery for severe mitral regurgitation (MR). MV repair was planned.

Intraoperative Findings

- Pre-bypass transesophageal echocardiography (TEE) revealed elongated mitral leaflets with severe P2 and P3 prolapse.
- Intraoperative findings showed multiple misaligned chordae—some myxomatous and elongated, others abnormally inserted onto incorrect leaflets or the left ventricular wall.
- Successful repair would have required extensive dissection and multiple artificial chordae, with a potential need for a second bypass run if inadequate.

Management

- Given the patient's comorbidities, including COPD and the uncertain long-term durability of repair, the decision was made to proceed with valve replacement.
- A 33 mm bioprosthetic valve was implanted, demonstrating good function and no paravalvular leak on post-bypass TEE.

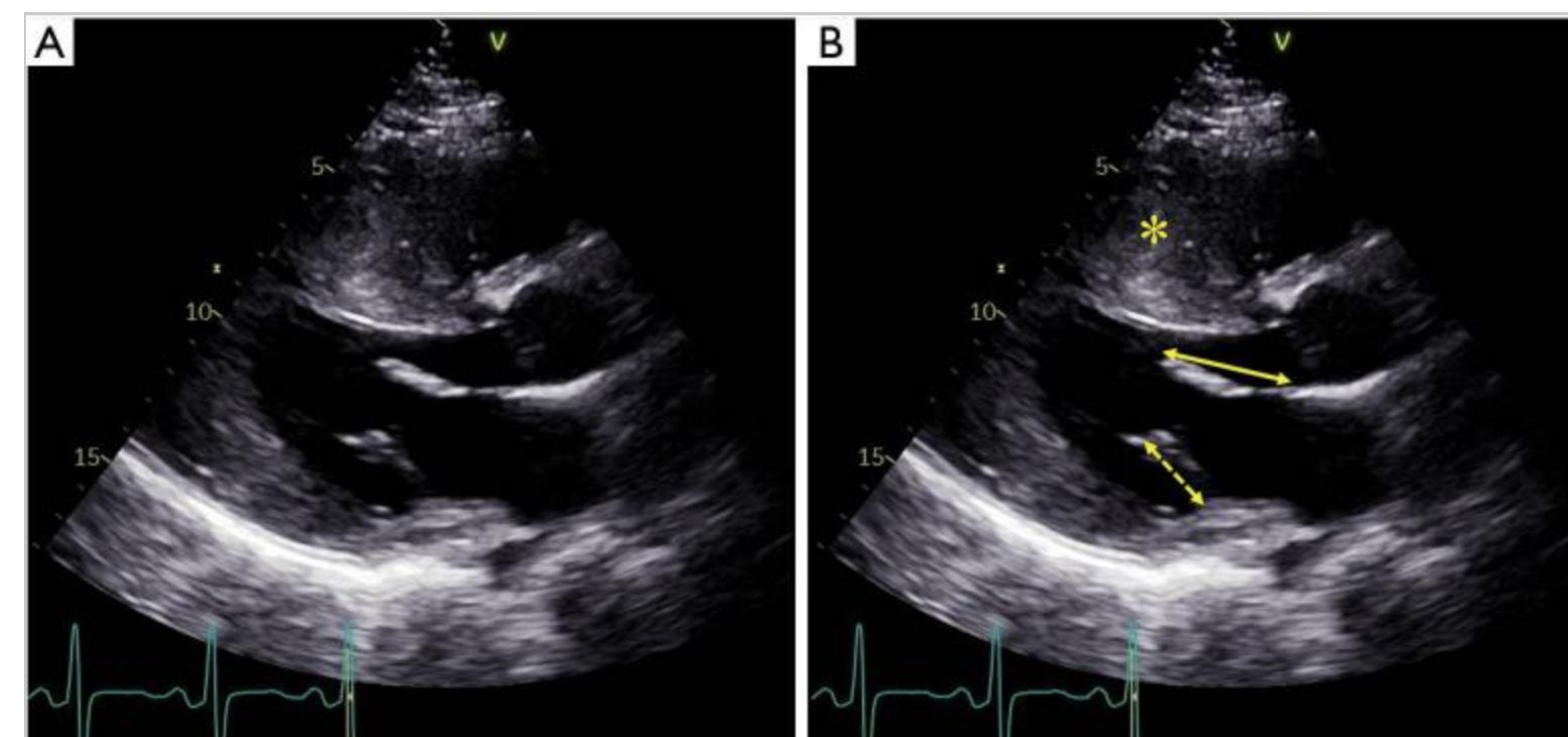


Figure 1. Elongated mitral valve leaflets in hypertrophic cardiomyopathy in mid-diastolic parasternal long-axis view [2].

↔ Elongated anterior mitral leaflet, measuring 28 mm
↔ Elongated posterior mitral valve leaflet, measuring 16 mm
★ Thickened interventricular septum measuring 30 mm with a reverse curve morphology

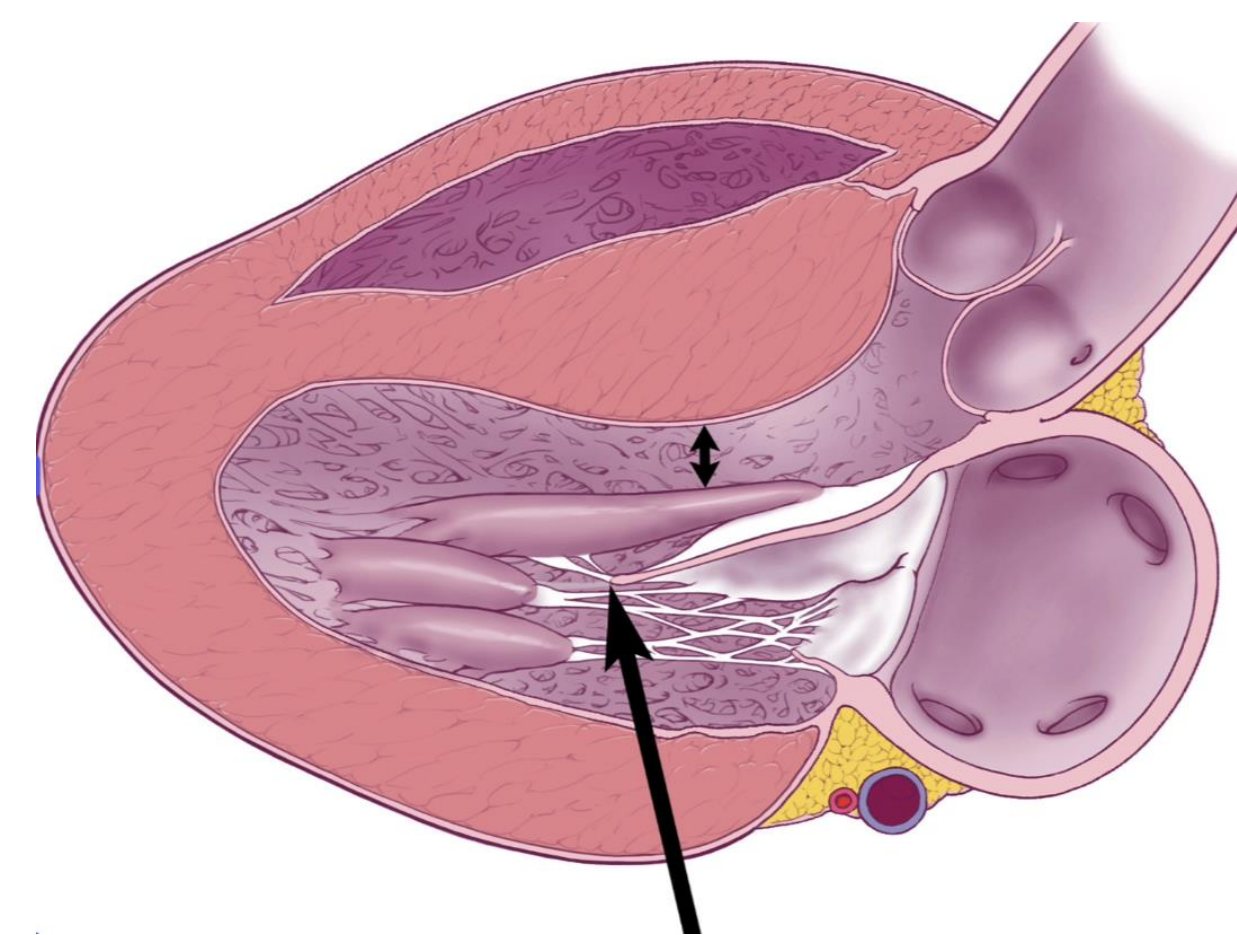


Figure 2. Anomalous PM with aberrant chordal attachments to the free edge of the anterior MV leaflet. *The Journal of Thoracic and Cardiovascular Surgery*.

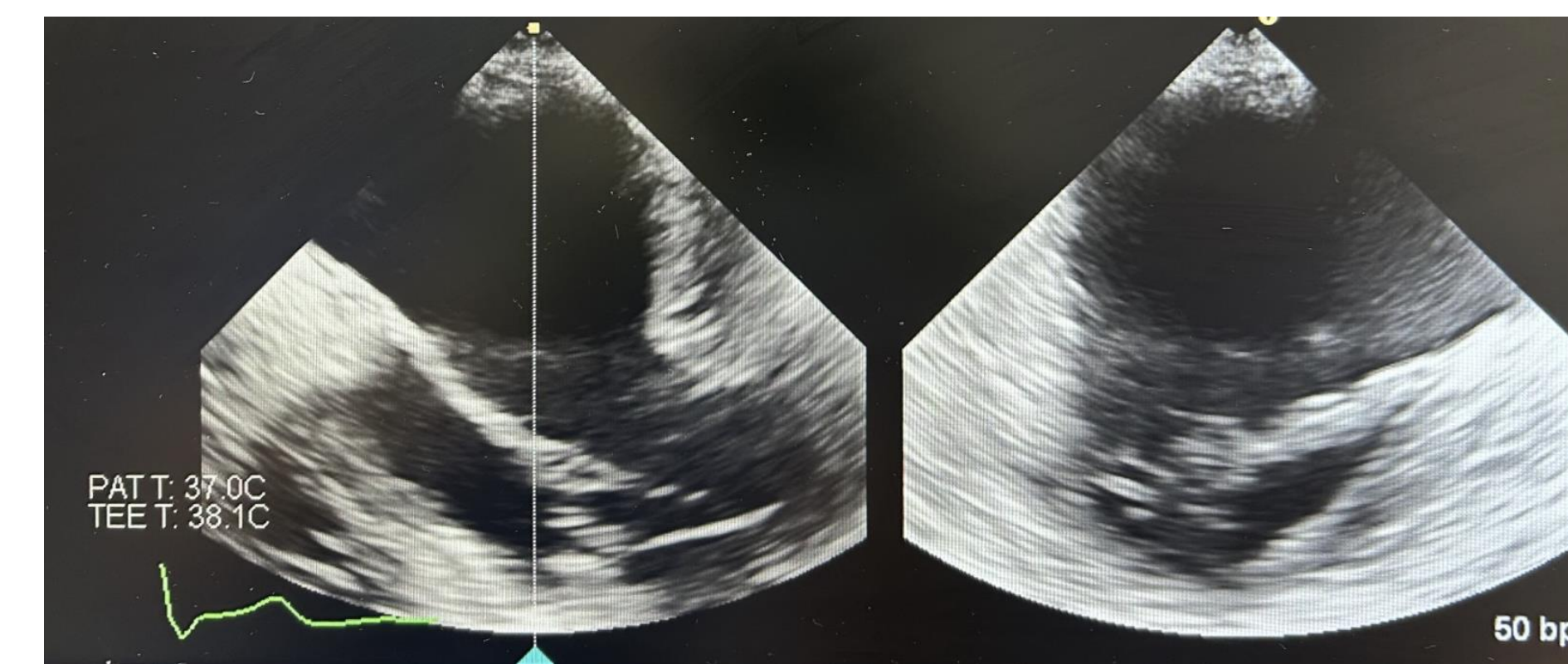


Figure 3. TEE images of the case patient.

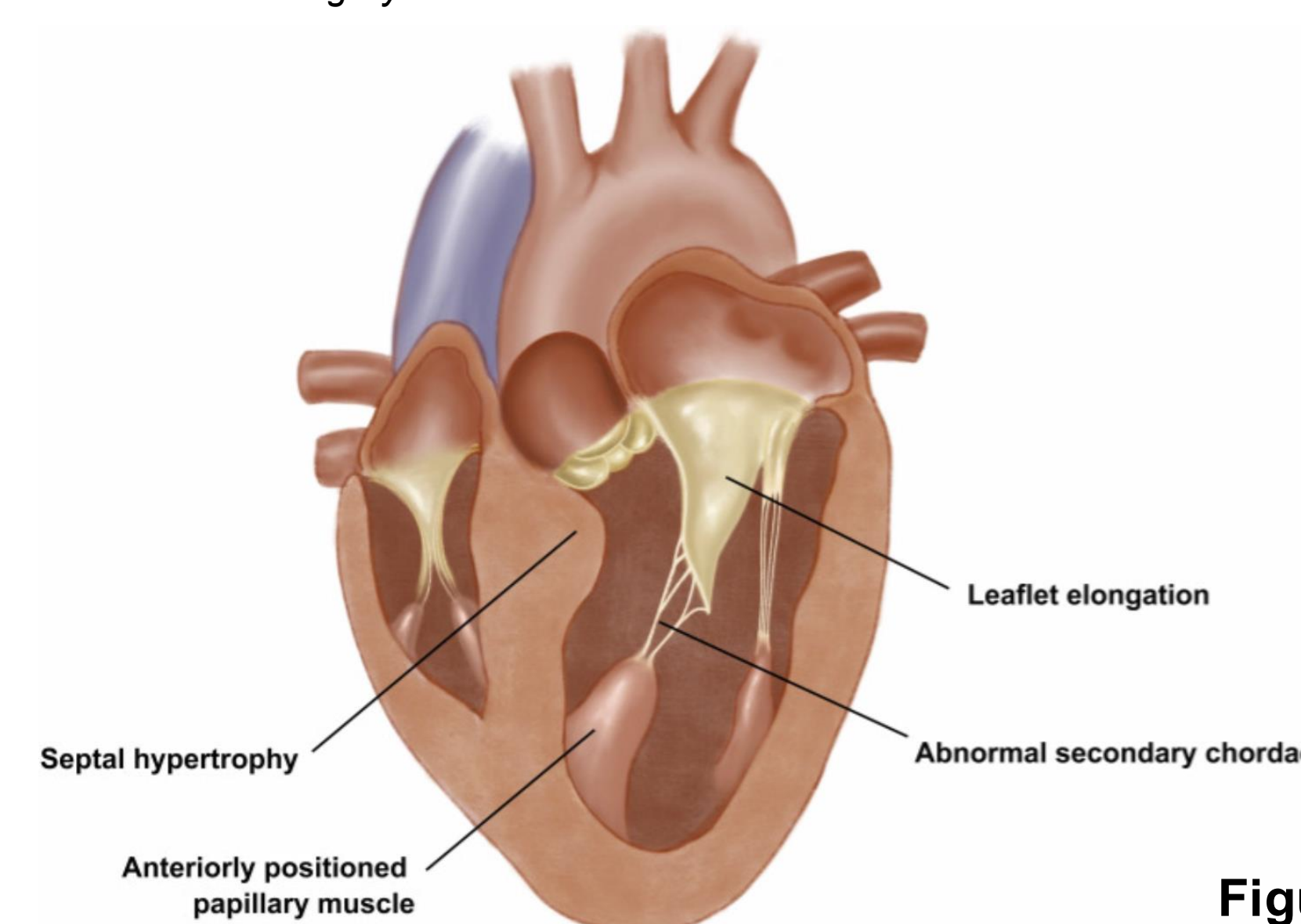


Figure 4. MV leaflet elongation in HCM. *Reviews in cardiovascular medicine*.

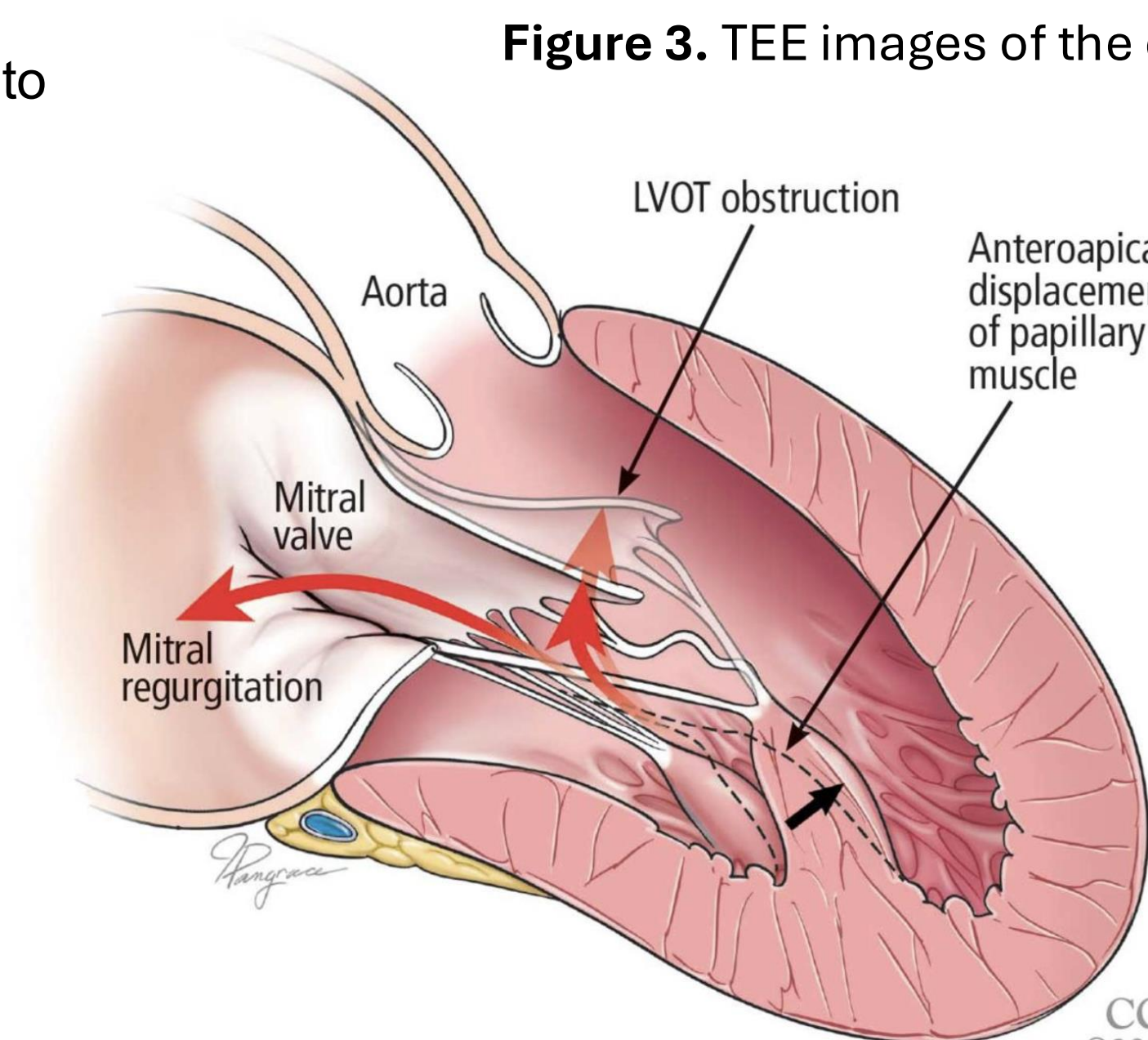


Figure 5. Bifid PM, anterior displacement of PM, and subsequent slack in chordae lead to SAM of the mitral valve, causing LVOT obstruction and simultaneous mitral regurgitation. *Cleveland Clinic Journal of Medicine*.

Discussion

- HCM is characterized by an asymmetrically hypertrophied left ventricle with risk of LVOT obstruction during systole.
- While traditionally considered a myocardial disease, congenital malformations of the MV contribute to LVOT obstruction in a substantial subset of patients [1]. These can involve:
 - mitral leaflets
 - chordae tendineae
 - ventricular wall
 - papillary muscles
- In a study of 94 patients with HCM, 62 (66%) demonstrated malformations, including leaflet elongation and anomalous papillary muscle insertion directly onto anterior mitral leaflets- highlighting that the disease process is not confined to myocardium [1].
- TEE-based studies quantified elongation of mitral leaflets in HCM versus controls: anterior leaflet length averaged 31 ± 4 mm in HCM versus 22 ± 3 mm in controls; the posterior averaged 20 ± 2 mm versus 15 ± 3 mm, respectively ($P < 0.00001$) [2]. These elongated leaflets can be problematic to repair during bypass.
- Subvalvular abnormalities, such as anterior displacement of papillary muscles and chordal misalignment, also promote systolic anterior motion by creating slack in the chordae tendineae [3].

Conclusions

Recognition of mitral apparatus abnormalities is crucial before and during open-heart mitral valve surgery. They can significantly alter intraoperative decision-making and may necessitate valve replacement over repair. Patient-specific bypass risks must be considered. This underscores the importance of multidisciplinary approaches when managing mitral pathology in HCM.

References

- [1] Klues, H. G., Maron, B. J., Dollar, A. L., & Roberts, W. C. (1992). Diversity of structural mitral valve alterations in hypertrophic cardiomyopathy. *Circulation*, 85(5), 1651–1660.
 - [2] Guigui, S. A., Torres, C., Escobar, E., & Mihos, C. G. (2022). Systolic anterior motion of the mitral valve in hypertrophic cardiomyopathy: a narrative review. *Journal of thoracic disease*, 14(6), 2309–2325.
 - [3] Maron, B. J., Nishimura, R. A., & Danielson, G. K. (1998). Pitfalls in clinical recognition and a novel operative approach for hypertrophic cardiomyopathy with severe outflow obstruction due to anomalous papillary muscle. *Circulation*, 98(23), 2505–2508.
- Further references available upon request. Please email Lana.Shteynman@stonybrookmedicine.edu