

## IMAGES IN CARDIOLOGY

### Midventricular Obstructive Hypertrophic Cardiomyopathy with Apical Aneurysm: Left Ventricular Hemodynamic and Angiographic Findings

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#### Abstract

A case of a 50-year-old patient with a midventricular obstructive hypertrophic cardiomyopathy is presented and the left ventricular hemodynamic and angiographic findings are depicted and discussed. *Rhythmos 2022; 17(2):36-38.*

**Key Words:** hypertrophic cardiomyopathy; midventricular obstruction; apical aneurysm; “hourglass” left ventricle; left ventriculography; cardiac magnetic resonance imaging; ventricular tachycardia; sudden death

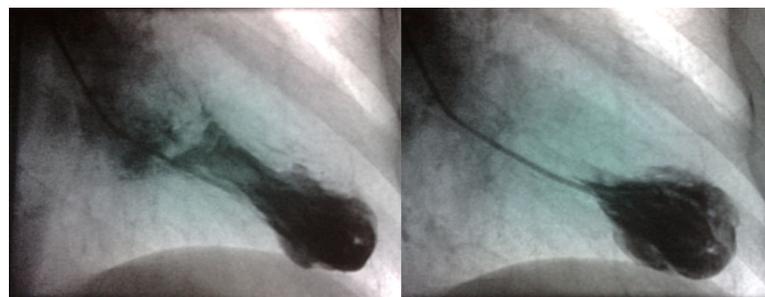
**Abbreviations:** CMR = cardiac magnetic resonance (imaging); HCM = hypertrophic cardiomyopathy; ICD = implantable cardioverter defibrillator; LGE = late gadolinium enhancement; LV = left ventric-le(ular); LVOT = left ventricular outflow tract; MVO = midventricular obstruction; NYHA = New York Heart Association; SCD = sudden cardiac death; VT = ventricular tachycardia

A 50-year-old male presenting with chest pain syndrome and repolarization changes on ECG was referred for cardiac catheterization. Coronary angiography showed normal coronary arteries; however, an impressive pressure gradient of 130 mmHg was recorded during left ventricular (LV) catheterization between the apical (LV) and basal (outflow) part of the LV (LVOT) (**Figure 1**). Subsequent LV angiography showed systolic mid-cavitary obliteration with small tubular LV cavity during diastole (**Figure 2**). The patient was placed on beta-blocker therapy with ensuing remission of symptoms.



**Figure 1.** A left ventricular pressure of 278/7 mmHg was recorded with use of a pigtail catheter inserted into an apical position of the left ventricle (LV). During a pullback recording, the LV pressure dropped to 148/4 at the LV outflow tract

(LVOT) with no further systolic gradient recorded in the aorta (Ao) (148/66 mmHg).



**Figure 2.** A left ventricular angiogram depicted an “hourglass” shaped LV with tubular mid-LV cavity during diastole (left panel), which was completely obliterated during systole (right panel). The apex remained akinetic compatible with an apical LV aneurysm.

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Hypertrophic cardiomyopathy (HCM) is a genetic disease, inherited as an autosomal dominant disorder, characterized by a wide spectrum of morphologies and natural history.<sup>1,2</sup> Mid-ventricular (mid-cavitary) obstructive (MVO) HCM is an uncommon morphologic type/variant of HCM, characterized by the presence of a pressure gradient between the apical and basal chambers of the left ventricle (LV) (**Fig. 1**).<sup>3</sup> It is suggested that the most common HCM morphology associated with MVO is due to the apposition of septum and lateral wall producing a small, hyperdynamic cavity, or less commonly due to the systolic apposition of hypertrophied papillary muscle and lateral wall at the mid-cavitary level, producing two distinct LV chambers in an “hour glass” shaped LV, while an LV apical aneurysm of varying size is also noted (**Fig. 2**).<sup>4</sup> Some investigators have suggested that this aneurysm may be due to fibrotic (as suggested by apical late gadolinium enhancement - LGE on cardiac magnetic resonance imaging-CMR) and thinned apex possibly due to prior infarction, although such patients have been shown to have no concurrent coronary artery disease, albeit their course may be complicated by apical thrombus.<sup>5,6</sup> However, the appearance of “apical aneurysm” may also be due to concurrent apical HCM, proved histologically in one case, leading to akinetic apex.<sup>7,8</sup>

Others have suggested that the apical aneurysm may be secondary to the increased after-load and increased apical pressure arising from significant pressure gradient of the MVO, as shown in the present case, leading to fibrosis/scar at the rim of the aneurysm and the adjacent apical areas.<sup>3</sup> Such fibrosis may also explain the arrhythmogenic substrate harboring the foci for monomorphic ventricular tachycardia (VT) leading to sudden cardiac death (SCD). Importantly, the variants of

HCM with apical aneurysm are the ones producing monomorphic VT, rather than polymorphic VT which characterizes the common type of HCM.<sup>7</sup>

Midventricular obstruction (MVO) constitutes ~10% of patients presenting with HCM. A study of 490 HCM patients showed that 46 (9.4%) had MVO.<sup>9</sup> Another study of 572 patients presenting with HCM indicated that 76 (13.3%) of these patients were diagnosed with MVO.<sup>10</sup> A Greek single-center cohort study of 423 patients with HCM (age 49±17 years; 66% male) indicated that MVO was identified in 34 patients (8%).<sup>11</sup> Patients with MVO tended to be more symptomatic (>90% with NYHA class ≥II symptoms) compared with the other HCM patients. Apical aneurysm was identified in 26.5%. On multivariate analysis, presence of MVO strongly predicted progression to end-stage, burned out HCM and related heart failure deaths (hazard ratio-HR 2.62; P=0.047).

The presence of MVO in patients with HCM has important pathophysiological and prognostic implications. As mentioned, in a study comprising 490 HCM patients, MVO, diagnosed when the peak midcavitary gradient was estimated to be ≥30 mm Hg, was identified in 46 patients (9.4%).<sup>9</sup> Patients with MVO were more likely to be symptomatic than those without. MVO predicted death (HR 2.23, P=0.016), particularly sudden death and potentially lethal arrhythmic events (HR: 3.19, P<0.001). Apical aneurysm formation was identified in 28.3% of patients with MVO and strongly predicted HCM-related death (HR: 3.47, P=0.008) and the combined endpoint of SCD and potentially lethal arrhythmic events (HR: 5.08, P<0.001). The presence and severity of MVO has been associated with higher incidence of non-sustained VT in HCM patients.<sup>10</sup>

LV apical aneurysms have rarely been reported in patients with HCM. In a large study of 1299 HCM patients, 28 (2%) were identified with LV apical aneurysms (age range, 26-83 years).<sup>12</sup> Apical aneurysms varied considerably in size (10-66 mm), and were associated with transmural myocardial scarring as shown by LGE on CMR. Left ventricular morphology varied, but the majority (68%) showed an "hourglass" contour, with midventricular hypertrophy producing muscular narrowing and intracavitary gradients in 9 (32%) patients (74±42 mm Hg). During follow-up (4.1±3.7 years), 12 patients (43%) with LV apical aneurysms developed complications (10.5%/y), including SCD, appropriate implantable cardioverter defibrillator (ICD) discharges, nonfatal thromboembolic stroke, and progressive heart failure and death.

A large cohort study of 1332 patients with apical HCM confirmed by CMR showed LV apical aneurysms in 31 (2.3%) patients (aged 53.8±15.1 years).<sup>13</sup> The majority (90%) of these patients had clinical symptoms, and 10% had a family history of HCM. Compared with apical HCM

patients without LV aneurysm, the proportion of systolic MVO and late gadolinium enhancement (LGE) presence, and the LGE extent in apical HCM patients with aneurysms were significantly higher (all P<0.05). The event-free survival rate in apical HCM patients with aneurysm was significantly lower than that in patients without aneurysm (log rank, P=0.01).

A retrospective study indicated that among 1,940 HCM patients, 93 (5%) were identified with LV apical aneurysms (mean age 56 ± 13 years, 69% male).<sup>4</sup> Over 4.4±3.2 years, 3 of the 93 patients with LV apical aneurysms (3%) died suddenly or of heart failure, 22 (24%) had successful interventions including appropriate ICD discharges (n=18), heart transplants (n=2), and successful resuscitation after cardiac arrest (n=2). Five non-anticoagulated patients had nonfatal thromboembolic events (1.1%/year), whereas apical clots were detected in 13 anticoagulated patients, albeit without embolic events. The authors concluded that HCM patients with LV apical aneurysms are at high risk for arrhythmic sudden death and thromboembolic events.

In the subgroup of patients with HCM and MVO, LV apical aneurysms have been reported in over 20% of such patients and have been considered an independent predictor of potentially lethal ventricular arrhythmic events, including ventricular tachycardia (VT)/ventricular fibrillation (VF) leading to SCD.<sup>9</sup> A systematic review of 39 observational studies reporting on 94 patients with HCM with MVO and LV aneurysm (mean age 58.05±11.76 years, 59.6% males) indicated that the most common electrocardiographic finding was T wave inversion occurring in 13.8% of the cases followed by ST elevation (9.5%).<sup>14</sup> Maximal LV wall thickness was 18.89 ± 5.19 mm and paradoxical jet flow was detected in 29.8% of patients. Beta-blockers (58.5%) were the most common drug therapy; amiodarone (10.6%) was employed for VT, which was the most common complication, observed in 39.3% of cases. All-cause mortality was 13.8 % over 16±20.1 months of follow-up. Implantable cardioverter defibrillator (ICD) was implanted in 37% of patients, of whom 26% received appropriate shock therapy.

Finally, a recent meta-analysis of 6 studies comprising 2382 HCM patients indicated that the presence of LV apical aneurysm in these patients was associated with an increased risk for SCD events (pooled OR: 4.67; I<sup>2</sup>: 38%) and thromboembolism (pooled OR: 6.30; I<sup>2</sup>: 66%).<sup>15</sup>

## Conclusion

The HCM variant with MVO featured in the present case is encountered in ~10% of patients with HCM. LV apical aneurysms have rarely been reported in patients with HCM (2-5%). However, in the subgroup of HCM with MVO, apical aneurysms are found in over 20% of such

patients and when typical (fibrotic), such aneurysms are associated with an arrhythmogenic substrate of VT/VF and SCD, while they can also harbor intracavitary thrombi with the attendant risk of thromboembolism.

## References

1. Ommen SR, Mital S, Burke MA, et al. 2020 AHA/ACC Guideline for the Diagnosis and Treatment of Patients With Hypertrophic Cardiomyopathy: A Report of the American College of Cardiology/American Heart Association Joint Committee on Clinical Practice Guidelines. *Circulation* 2020;142:e558-e631.
2. Maron BJ, Desai MY, Nishimura RA, et al. Diagnosis and Evaluation of Hypertrophic Cardiomyopathy: JACC State-of-the-Art Review. *J Am Coll Cardiol* 2022;79:372-89.
3. Cui L, Tse G, Zhao Z, et al. Mid-ventricular obstructive hypertrophic cardiomyopathy with apical aneurysm: An important subtype of arrhythmogenic cardiomyopathy. *Ann Noninvasive Electrocardiol* 2019;24:e12638.
4. Rowin EJ, Maron BJ, Haas TS, et al. Hypertrophic Cardiomyopathy With Left Ventricular Apical Aneurysm: Implications for Risk Stratification and Management. *J Am Coll Cardiol* 2017;69:761-73.
5. Martinez MW, Sorajja P, Ommen SR. Mid-cavitary obstruction in hypertrophic cardiomyopathy. *Heart* 2008;94:1287.
6. Mavilakandy A, Ahamed H. Mutation of the MYL3 gene in a patient with mid-ventricular obstructive hypertrophic cardiomyopathy. *BMJ Case Rep* 2022;15.
7. Wilson P, Marks A, Rastegar H, Manolis AS, Estes NA, 3rd. Apical hypertrophic cardiomyopathy presenting with sustained monomorphic ventricular tachycardia and electrocardiographic changes simulating coronary artery disease and left ventricular aneurysm. *Clin Cardiol* 1990;13:885-7.
8. Fath AR, Jokerst CE, Aglan A, Mihiyawi N, Mookadam F. Multimodality Imaging Demonstrating an Apical Variant Hypertrophic Cardiomyopathy in an Uncommon Pentad. *J Investig Med High Impact Case Rep* 2020;8:2324709620934324.
9. Minami Y, Kajimoto K, Terajima Y, et al. Clinical implications of midventricular obstruction in patients with hypertrophic cardiomyopathy. *J Am Coll Cardiol* 2011;57:2346-55.
10. Nie C, Zhu C, Xiao M, et al. Mid-ventricular obstruction is associated with non-sustained ventricular tachycardia in patients with hypertrophic obstructive cardiomyopathy. *Clin Cardiol* 2021;44:555-62.
11. Efthimiadis GK, Pagourelis ED, Parcharidou D, et al. Clinical characteristics and natural history of hypertrophic cardiomyopathy with midventricular obstruction. *Circ J* 2013;77:2366-74.
12. Maron MS, Finley JJ, Bos JM, et al. Prevalence, clinical significance, and natural history of left ventricular apical aneurysms in hypertrophic cardiomyopathy. *Circulation* 2008;118:1541-9.
13. Yang K, Song YY, Chen XY, et al. Apical hypertrophic cardiomyopathy with left ventricular apical aneurysm: prevalence, cardiac magnetic resonance characteristics, and prognosis. *Eur Heart J Cardiovasc Imaging* 2020;21:1341-50.
14. Elsheshtawy MO, Mahmoud AN, Abdelghany M, et al. Left ventricular aneurysms in hypertrophic cardiomyopathy with midventricular obstruction: A systematic review of literature. *Pacing Clin Electrophysiol* 2018;41:854-65.
15. Papanastasiou CA, Zegkos T, Karamitsos TD, et al. Prognostic role of left ventricular apical aneurysm in hypertrophic cardiomyopathy: A systematic review and meta-analysis. *Int J Cardiol* 2021;332:127-32.