Prevalence, Clinical Significance, and Natural History of Left Ventricular Apical Aneurysms in Hypertrophic Cardiomyopathy

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Background—Hypertrophic cardiomyopathy (HCM) is the most common genetic heart disease characterized by a diverse clinical and phenotypic spectrum. This study reports the prevalence, morphology, clinical course, and management of an underrecognized subgroup of HCM patients with left ventricular apical aneurysms.

Methods and Results—Of 1299 HCM patients, 28 (2%) were identified with left ventricular apical aneurysms, including a pair of identical twins. Aneurysms were recognized at a wide age range (26 to 83 years), including 12 patients (43%) who were ≤50 years of age. Apical aneurysms varied considerably in size (maximum dimension, 10 to 66 mm), were dyskinetic/akinetic with thin rims, and were associated with transmural (and often more extensive) myocardial scarring identified by late gadolinium enhancement cardiovascular magnetic resonance. Apical aneurysms were recognized by echocardiography in only 16 of 28 patients (57%) but by cardiovascular magnetic resonance in the 12 patients undetected by echocardiography. Left ventricular chamber morphology varied; however, 19 patients (68%) showed an “hourglass” contour, with midventricular hypertrophy producing muscular narrowing and intracavitary gradients in 9 patients (74±42 mm Hg). Sarcomeric protein missense mutations known to cause other phenotypic expressions of HCM were present in 3 patients. Over 4.1±3.7 years of follow-up, 12 patients (43%) with left ventricular apical aneurysms experienced adverse disease complications (event rate, 10.5%/y), including sudden death, appropriate implantable cardioverter-defibrillator discharges, nonfatal thromboembolic stroke, and progressive heart failure and death.

Conclusions—Patients with left ventricular apical aneurysms represent an underappreciated subset in the heterogeneous HCM disease spectrum with important clinical implications, often requiring a high index of suspicion and cardiovascular magnetic resonance for identification. Apical aneurysms in HCM are associated with substantial cardiovascular morbidity and mortality and raise novel treatment considerations. (Circulation. 2008;118:1541-1549.)

Key Words: aneurysm ■ cardioverter-defibrillators, implantable ■ heart arrest ■ hypertrophic cardiomyopathy ■ magnetic resonance imaging ■ remodeling

Within the diverse clinical and morphological spectrum of hypertrophic cardiomyopathy (HCM),1-5 a subgroup of patients characterized by left ventricular (LV) apical aneurysms has emerged. Previous descriptions of HCM patients with LV apical aneurysms, however, have been confined to case reports6-10 or small patient series.11-16 Consequently, this subgroup of patients has remained underappreciated, and its clinical profile and natural history are largely undefined. In this regard, it has been our impression that LV apical aneurysms in HCM may be associated with important and unrecognized clinical implications. Therefore, we believe it is timely to characterize, within a large HCM patient cohort, the expression of this novel subset of patients with respect to prevalence, clinical course, and management strategies.

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Patient Selection
The study population included 1299 patients with clinically diagnosed HCM who were consecutively enrolled and evaluated from 1993 to 2006 at 2 centers: the Minneapolis Heart Institute Foundation...
obstruction, and mitral regurgitation were assessed with commercially available instruments. Magnitude of LV hyper-
trophy at some time during the patient's clinical course.1,3,4

Diagnosis of HCM was based on echocardiographic documentation of a hypertrophied, nondilated left ventricle in the absence of another cause of LV aneurysm formation by (1) the absence of significant coronary arterial narrowing (>50% stenosis) in the left anterior descending artery by conventional angiography or cardiac computed tomography angiogram (n = 23) and (2) absent history of chest pain, coronary risk factors, and acute coronary syndrome in the other 5 patients (all <50 years of age). Myocardial bridging of the left anterior descending artery was present in 3 (13%) patients.

All study patients signed a statement previously approved by the Internal Review boards of the respective participating institutions, in which patients agreed to the use of their medical information for research purposes.

Definitions
Diagnosis of HCM was based on echocardiographic documentation of a hypertrophied, nondilated left ventricle in the absence of another cardiac or systemic disease capable of producing a similar magnitude of hypertrophy at some time during the patient's clinical course.1,3,4 LV apical aneurysm was defined as a discrete thin-walled dyskinetic or akinetic segment of the most distal portion of the chamber with a relatively wide communication to the LV cavity.17–19

Potentially lethal events in which patients were successfully resuscitated from cardiac arrest or received appropriate therapies for ventricular tachycardia/fibrillation from an implanted defibrillator were regarded as equivalent to sudden death.

Echocardiography
Two-dimensional echocardiograms were performed in each patient with commercially available instruments. Magnitude of LV hypertrophy, outflow obstruction, and mitral regurgitation were assessed as previously described.5,26 Ejection fraction was calculated from echocardiographic images with modified Simpson's rule or the peak instantaneous outflow gradient ≥30 mm Hg by continuous-wave Doppler echocardiography under resting conditions.21

Cardiovascular Magnetic Resonance
Cardiovascular magnetic resonance (CMR) was performed in 22 patients with a Siemens Sonata-Avanto (Erlangen, Germany) or a Philips Gyroscan ACS-NT (Best, the Netherlands) 1.5-T whole-body scanner with dedicated cardiac coils. Breath-hold cine steady-state free precession images were acquired in multiple short-axis and 3 long-axis orientations. Complete ventricular coverage was achieved with contiguous 10-mm-thick slices. A late gadolinium enhancement (LGE) protocol was used 15 minutes after intravenous administration of 0.2 mmol/kg gadolinium-DTPA (Magnevist, Schering, Berlin, Germany) with breath-held segmented inversion-recovery sequence (inversion time, 240 to 300 ms) acquired in the same views as the cine images.

Genetic Testing
Mutational analyses were performed by polymerase chain reaction, denaturing high-performance liquid chromatography (Transgenomic, Omaha, Neb), and direct DNA sequencing for mutations in all translated exons for the 8 most common sarcomeric HCM genes: MYBPC3-encoded myosin binding protein C (MYBPC3), MYH7-encoded β-myosin heavy chain (MYH7), MYL2- and MYL3-encoded regulatory and essential myosin light chains (MYL2 and MYL3), TPM1-encoded troponin I (TPN1I), TNNT2-encoded cardiac troponin T (TNNT2), TPM1-encoded α-tropomyosin (TPM1), and ACTC-encoded cardiac actin (ACTC).

Statistical Analyses
Data are expressed as mean ± SD. Proportions and categorical data were compared by use of Fisher's exact test. CIs for proportions were calculated from the binomial equation. Continuous normally distributed data were compared by use of unpaired and paired Student's t tests. The annual event rate was calculated as the number of adverse clinical events divided by the total observation time of the cohort starting from the time of first evaluation at one of the study centers. Statistical analyses were performed with SAS for Windows version 9.1 (SAS Institute Inc, Cary, NC).

The authors had full access to and take full responsibility for the integrity of the data. All authors have read and agree to the manuscript as written.

Results
LV Apical Aneurysms

Prevalence
LV apical aneurysms were identified in 28 of 1299 study patients (2.2%; 95% CI, 1.4 to 3.1). This proportion was similar (P = 0.5 for difference) for both participating centers (Minneapolis: n = 19 of 947, 2.0%; Boston: n = 9 of 352, 2.6%).

Figure 1. LV long-axis 4-chamber CMR cine steady-state free precession images in systole from 6 HCM patients with thin-walled LV apical aneurysms. A through C, Medium to large aneurysms (arrowheads) and maximal LV wall thickening at midventricular level, with muscular apposition of the septum and LV free wall producing distinct proximal (P) and distal (D) chambers. C, LV apical aneurysm in a 59-year-old man (without a history of atrial fibrillation) contains a sizable intracavitary thrombus attached to the rim of the aneurysm (arrow). D through F, Relatively small apical aneurysms (arrowheads) with hypertrophy located predominantly more distal within the LV chamber (*). LA indicates left atrium; VS, ventricular septum.
LV Morphology and Outflow Obstruction

LV Shape and Pattern of Hypertrophy

Two distinctive LV morphological forms were identified (Figure 1). Most common was an “hourglass” shape (n=19, 68%), with maximal wall thickness present in the midventricle (18±3 mm) and considerably less or no hypertrophy evident in the distal and proximal portions of LV wall (Figure 1A through 1C). In these patients, mid systolic muscular apposition of the septum and LV free wall (length, 21±12 mm; range, 10 to 43 mm) produced distinct proximal and distal chambers, the latter of which included but was not necessarily limited to the aneurysm itself. In the other 9 patients (32%), hypertrophy was predominant in the distal portion of LV chamber (20±4 mm; Figure 1D through 1F).

Obstruction to LV Outflow

Intraventricular pressure gradients were present at rest in 10 apical aneurysm patients (36%). Only one of these patients had dynamic LV outflow tract obstruction caused by typical mitral valve systolic anterior motion and mitral-septal contact (ie, gradient, 50 mm Hg). The other 9 patients had midcavity obstruction (in the absence of systolic anterior motion), which was the consequence of midventricular muscular apposition in an hourglass-shaped chamber (gradients, 74±42 mm Hg; range, 25 to 150 mm Hg). Intraventricular pressure gradients were absent at rest in the other 18 study patients.

Clinical Profile

Demographics and Characteristics

The baseline demographics of the study population are summarized in the Table. The 28 aneurysm patients were 52±13 years of age (range, 26 to 83 years) at study entry; 12 (43%) were <50 years and 3 (11%) were <35 years of age. Eighteen patients (64%) were male. Maximal LV thickness was 19±3 mm (range, 14 to 25 mm), LV ejection fraction was 60±12% and ≤50% in 7 patients (25%). At the most recent evaluation, 18 patients (64%) were asymptomatic (New York Heart Association [NYHA] functional class I), 5 (18%) were mildly symptomatic (NYHA class II), and 5 (18%) were severely symptomatic (NYHA classes III to IV). Three patients (11%) had a history of atrial fibrillation/flutter.

Compared with the 1271 non–LV apical aneurysm HCM patients at study entry, the 28 patients with apical aneurysms were older (52±13 versus 45±20 years; P=0.001), had...
larger LV end-diastolic cavity dimensions (50±6 versus 42±9 mm; P<0.001), and had lower maximal LV wall thickness (19±3 versus 21±8 mm; P=0.001). There were no differences with respect to gender (male, 64% versus 63%; P=0.8), left atrial dimension (40±6 versus 41±8 mm; P=0.5), or NYHA functional class (1.3±0.5 versus 1.6±0.7; P=NS).

**12-Lead ECG**
Among the 28 LV apical aneurysm patients, the initial ECG demonstrated 2 common patterns: convex ST-segment elevation (≥1 mm in ≥2 contiguous leads), usually in V1 through V4 (n=13) and associated with T-wave inversion (n=9), and T-wave inversion without ST-segment elevation, usually in V1 through V4 (n=15). In 5 of these 15 patients, Q or QS waves also were present in leads V1 through V4 or II, III, and AVL.

**Ambulatory (Holter) ECG Monitoring**
A 24-hour Holter ECG was performed in 24 patients within 6 months of the initial evaluation. Of these 24 patients, 10 (42%) had ≥1 run of nonsustained monomorphic ventricular tachycardia (3 to 11 beats at 120 to 215 bpm, including 2 patients with 2 runs of nonsustained monomorphic ventricular tachycardia each, 4 patients with 3 runs, and 1 patient with 7 runs).

**Clinical Course**
Over the follow-up period of 4.1±3.7 years, 12 patients (43%) experienced an adverse clinical event (Figure 4),
Table. Continued

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including sudden cardiac death (n=2, including 1 patient who also had nonfatal thromboembolic stroke), aborted cardiac arrest (n=2, including 1 patient presenting with ventricular fibrillation), appropriate implantable cardioverter-defibrillator (ICD) intervention for ventricular tachycardia/ventricular fibrillation (n=3, including 1 patient with embolic stroke), and progressive heart failure with increase of ≥1 NYHA class and/or death (n=5). Annual HCM-related adverse cardiovascular event rate was 10.5%/y. Six of these 12 patients also developed systolic dysfunction (ejection fraction ≤50%).

Relation of Aneurysm Size to Clinical Outcome
HCM patients with large LV apical aneurysms were more likely to experience an adverse disease complication (5 of 6, 83%) than patients with medium or small aneurysms (7 of 22, 32%; P=0.02) (Figure 5). Specific disease complications were more common in association with large or medium compared with small aneurysms, ie, sudden death events (3 versus 1, respectively), LV systolic dysfunction (7 versus 0), progressive heart failure symptoms (3 versus 2), and embolic stroke/LV apical thrombus (4 versus 0).

Management Considerations

**Medical Treatment**
Of the 28 patients with LV apical aneurysm, 24 were treated with β-blockers, calcium channel blockers, or both. No patient underwent invasive septal reduction therapy (ie, surgical myectomy or alcohol septal ablation) for relief of heart failure symptoms. Both patients who experienced an embolic stroke were in sinus rhythm at the time of the event, including one with a history of atrial flutter and one who was...
taking warfarin. Two other patients had a large thrombus within the aneurysm identified by CMR (Figures 1C and 2C), but neither patient had a history of atrial fibrillation/flutter.

Implanted Defibrillators

Two patients received an ICD for secondary prevention after cardiac arrest (patients 22 and 23 in the Table), one of whom experienced multiple appropriate shocks for ventricular fibrillation 42 months after implant (patient 22). In 15 other patients, ICDs were implanted prophylactically largely on the basis of the presence of an apical aneurysm associated with transmural scarring, generating an increase in perceived arrhythmic risk. In 8 of these 15 patients, the 5 traditional primary prevention risk factors (ie, maximal LV wall thickness ≥30 mm, syncope, family history of sudden death caused by HCM, nonsustained monomorphic ventricular tachycardia on Holter monitoring, and abnormal blood pressure response to exercise) were absent.

Among the 15 patients implanted for primary prevention, 3 experienced an appropriate ICD discharge for ventricular tachycardia/ventricular fibrillation (at 33, 54, and 64 years of age), and each had ≥1 conventional risk factors. However, of the 4 patients experiencing either sudden death or aborted cardiac arrest, 3 had no risk markers.

Genetics

Of the 28 aneurysm patients, 14 (50%) had a family history of HCM and/or were found to have a disease-causing sarcomere mutation. A history of HCM-related sudden death in ≥1 relative was present in 6 patients, including a set of identical twins. Four disease-causing sarcomere mutations were identified in 3 of 9 LV apical aneurysm patients in whom genotyping was performed, with 1 patient (patient 22 in the Table) harboring 2 MYBPC3 mutations (Q921E and R502W) and the other 2 patients harboring -MYHC7 (R403Q) and TNNT2 (G82R) mutations, respectively (patients 1 and 19 in the Table).

Discussion

HCM is characterized by particularly heterogeneous clinical and morphological expression. An underrecognized and incompletely described subgroup within this broad spectrum comprises patients with thin-walled, akinetic/dyskinetic LV apical aneurysms of various sizes. To clarify the presentation, clinical profile, and prognosis of this patient subset and to provide insights into appropriate management strategies, we have assembled the most substantial series of HCM patients with LV apical aneurysms to date. Despite the unusual morphology expressed by the LV apical aneurysm...
patients, there is evidence that this subgroup in fact constitutes part of the broad HCM disease spectrum, given that one half of our study patients had either a confirmed family history of HCM and/or one of the disease-causing sarcomeric protein mutations, which have been reported to cause more typical phenotypic expressions of HCM.24,25

In the present cohort of almost 1300 HCM patients assembled from 2 specialty centers, the prevalence of LV apical aneurysms was 2%. However, it is likely that our data underestimate the true prevalence of apical aneurysms in the overall HCM population, given that 2-dimensional echocardiography proved unreliable in detecting smaller apical aneurysms in contrast to the higher spatial resolution and detection capability of CMR. This observation is similar to that in which CMR has demonstrated enhanced diagnostic accuracy for the identification of hypertrophy confined to the LV apex26 or anterolateral free wall27 and further underscores the emerging and important role of CMR in the contemporary evaluation of HCM patients.26–30

The clinical course of our HCM patients with LV apical aneurysms was variable but overall proved to be largely unfavorable. Over a relatively short follow-up period, almost one half of the study patients either had died of their disease or survived with severe progressive heart failure symptoms, embolic stroke, or appropriate ICD shocks. The overall rate of adverse disease consequences was 10.5%/y, significantly higher than reported in the general HCM population.1,3,37–39

Adverse clinical outcome was due largely to specific pathophysiological features related to the LV apical aneurysms, the recognition of which also raises several management considerations. In this context, we regard the scarred rim of the aneurysm and the associated extensive areas of LV myocardial fibrosis (identified by LGE CMR as hyperenhancement) to likely represent an arrhythmogenic substrate for the generation of malignant ventricular tachyarrhythmias.18,40 In addition, we found bursts of nonsustained monomorphic ventricular tachycardia on ambulatory (Holter) ECGs to be present in a large proportion of our LV apical aneurysm patients (ie, >40%). Nonsustained monomorphic ventricular tachycardia has been reported to be a determinant of increased sudden death risk in HCM41,42 and more recently has been found to occur more commonly in HCM patients with LGE28 (probably indicative of myocardial fibrosis). It also is possible that the patient subgroup with apical aneurysms could account for some HCM-related sudden deaths that would otherwise be unexplained.23 Indeed, >40% of our study patients with cardiac arrest, sudden cardiac death, or appropriate ICD interventions showed 0 of the 5 conventional primary prevention risk markers used in HCM,23 suggesting that aneurysm formation (and associated transmural myocardial scarring) may itself represent a novel risk marker for some HCM patients. These observations support the importance of contrast-enhanced CMR in defining myocardial fibrosis and scarring in this subset of HCM patients and selectively recommending the option of a prophylactic ICD.22

The dyskinetic/akinetc apical aneurysm in HCM also can provide the structural basis for intracavitary thrombus formation. Two of our patients incurred disabling thromboembolic events and 2 additional patients were unexpectedly found (by CMR) to have a thrombus within their aneurysm. Each of these 4 patients had large aneurysms (≥3 cm in the transverse dimension) and were in sinus rhythm at the time their stroke occurred or when the thrombus was identified fortuitously. These observations, although present in only a small number of patients, nevertheless suggest a potential role on a case-by-case basis for prophylactic warfarin anticoagulation to protect against embolic stroke in HCM patients with sizable apical aneurysms.

In addition, although only a minority of our patients with LV aneurysms (∼25%) developed global systolic dysfunction (ie, ejection fraction ≤50%), this prevalence of the end stage was significantly higher than in the general HCM population (ie, 3%).45 This finding suggests that a similar process of adverse LV remodeling, responsible for myocardial fibrosis and impaired systolic function, may be common to patients with apical aneurysm and those in the end stage. Therefore, these observations emphasize the importance of close interval surveillance among HCM patients with LV apical aneurysms to identify deterioration in LV function, which may prompt implementation of appropriate, targeted management strategies such as afterload-reducing agents, aldosterone inhibitors, ICD implantation,22 and possibly heart transplantation.46

Furthermore, no patient in our series experienced a catastrophic ventricular rupture despite the marked thinning of the aneurysm wall, and therefore, our data do not support prophylactic surgical resection for this purpose. Nevertheless, it is possible that in some patients with severe advanced heart failure, operative removal of the aneurysm may be a consideration. Finally, none of the 9 study patients with midventricular hypertrophy and intracavitary gradients underwent either surgical myectomy or alcohol septal ablation (nor dual-chamber pacing) to relieve LV outflow obstruction. Five of these patients were asymptomatic or only mildly symptomatic (and therefore not candidates for a septal reduction procedure), whereas the others had severe heart failure symptoms but were of advanced age or had significant comorbidities.

On the basis of our case series, none of several potential mechanisms11–13,15,16 appears to explain apical aneurysm formation in all HCM patients. First, the hypothesis that LV apical aneurysms and the associated regional myocardial scarring develop secondary to increased LV wall stress as a result of midcavity LV obstruction and elevated intracavitary systolic pressures is supported by only about one third of our patients. Second, a genetic predisposition is suggested by the virtually identical aneurysmal formation identified in a set of 48-year-old asymptomatic monozygotic twins,10 although we have found no apparent clustering of aneurysms in other families. Finally, the possibility that myocardial bridging of the left anterior descending coronary artery promotes apical aneurysm formation seems unlikely because this anomaly was identified in only 3 patients. Therefore, the basic mechanism responsible for the formation of apical aneurysms in HCM patients remains unresolved, and multiple causes are most likely.
Conclusions
HCM patients with LV apical aneurysm represent a previously underrecognized but clinically important subset within the broad HCM disease spectrum. Clinical recognition of this phenotype often requires a high index of suspicion, with CMR most effective for characterization of both the aneurysm and contiguous myocardial fibrosis. LV apical aneurysms in HCM patients are commonly associated with adverse clinical course, including sudden death events, embolic stroke, and progressive heart failure. Therefore, timely recognition of HCM patients with LV apical aneurysms may alter clinical practice by triggering consideration for primary prevention of sudden death with an ICD and possibly prophylactic anticoagulation with warfarin in selected patients with larger aneurysms.

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Disclosures
None.

References
33. Moon JC, McKenna WJ, McCrohon JA, Elliott PM, Smith GC, Pennell DJ. Toward clinical risk assessment in hypertrophic cardiomyopathy with...
The present study reports the prevalence, morphology, clinical course, and management of an underrecognized subgroup of hypertrophic cardiomyopathy (HCM) patients with left ventricular apical aneurysm. Among a large cohort of HCM patients, 2% were identified with a thin-rim apical aneurysm, which varied considerably in size (transverse dimension, 10 to 66 mm), were dyskinetic/akinetic, and were associated with transmural (and often more extensive) myocardial scarring identified by late gadolinium enhancement cardiovascular magnetic resonance. Sarcomere protein mutations known to cause phenotypic expression of HCM were identified in 3 of 9 genotyped patients, suggesting that these patients are part of the broad HCM disease spectrum. In most patients, the left ventricular chamber had an “hourglass” contour, with midventricular hypertrophy producing muscular narrowing and intracavitary gradients in some patients. Left ventricular apical aneurysms were identified by 2-dimensional echocardiography in only one half of the patients but were detected in all those patients imaged by cardiovascular magnetic resonance. Over a mean follow-up of 4 years, HCM patients with left ventricular apical aneurysms experienced a substantial adverse event rate (10.5%/y), including sudden death, appropriate ICD discharges, nonfatal thromboembolic stroke, and progressive heart failure and death. Therefore, identification of HCM patients with left ventricular apical aneurysm requires a high index of suspicion, often relying on cardiovascular magnetic resonance for both diagnosis and detection of myocardial scarring. These data also raise important management considerations in this subset of HCM patients, including consideration for prophylactic implantable cardioverter-defibrillator therapy and anticoagulation.
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