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CASE REPORT

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Is There a Typical Doppler Pattern in Patients With Apical Hypertrophic Cardiomyopathy With Aneurysm?

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ABSTRACT

Nineteen consecutive patients with apical hypertrophic cardiomyopathy and apical aneurysm underwent a comprehensive echo-Doppler including continuous wave Doppler at midventricular level. Three different flow patterns, pattern A (more frequent), pattern B, and pattern C, and expression of different intracavitary pressure gradients were defined. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2023;14:101836) © 2023 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

It has been reported that about 2%-10% of patients with apical hypertrophic cardiomyopathy (AHCM) have apical regional dysfunction, apical outpouching, or aneurysm in the presence of normal coronary arteries.[1]

Several studies demonstrated the incidence, morphologic variability, and prognosis of AHCM with aneurysm.[2,3] However, despite the relevance of this disease and the known diagnostic role of echo-Doppler and magnetic resonance imaging, Doppler flow patterns at the level of the tunnel that divides the left ventricle (LV) from the aneurysmal cavity have not been adequately studied and classified. These Doppler patterns may explain the pathophysiology of the disease; the paradoxical diastolic flow jet from the apex toward the base implies increased intracavitary pressure in the LV apex and consequently decreased coronary pressure. The LV cavity obstruction may lead to the cessation of systolic flow at the midventricular level, resulting in blood trapping within the apical chamber.

LEARNING OBJECTIVES

- To understand the Doppler flow pathology causing LV obstruction and aneurysm.
- To identify different Doppler patterns in AHCM.

Continuous-wave (CW) Doppler and pulsed-wave (PW) Doppler analysis are useful instruments for the evaluation of blood flow within the LV. We found in the majority of our cases an interruption of blood flow from the LV apex to the base during midsystole (void pattern), at the level of the muscular hypertrophy in the mid-LV, followed by paradoxical jet flow out of the aneurysm in diastole. Other reports[4] described a complete reverse flow (from the LV to the apex in systole and from the aneurysm to the LV in early diastole).

The aims of this case series of patients with AHCM with aneurysm are to evaluate the Doppler flow patterns at the level of the midapical LV obstruction and

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ABBREVIATIONS AND ACRONYMS

AHCM = apical hypertrophic cardiomyopathy

CMR = cardiac magnetic resonance

CW = continuous-wave

LV = left ventricle

PW = pulsed-wave

TTE = transthoracic echo

to correlate these findings with the anatomical and morphologic characteristics of the LV cavity obtained using transthoracic echo (TTE), contrast echo, or cardiac magnetic resonance (CMR).

STUDY POPULATION AND METHODS

Nineteen consecutive ambulatory AHCM patients with aneurysm were enrolled in this case series. All patients had a clinical and instrumental diagnosis of apical aneurysm defined as a discrete, thin-walled, dyskinetic, or akinetic segment of the most distal portion of the chamber.

All underwent a TTE and 13 underwent a CMR. In 6 cases CMR was not performed for technical reasons (5 due to claustrophobia and 1 due to the presence of a pacemaker). Contrast echo was performed in 4 cases. Complete TTE and Doppler examinations were performed in all patients according to the recommendations of the American Society of Echocardiography.[5]

LV volumes and ejection fraction, left atrial volume, thickness of the LV septum and posterior-lateral wall, mitral regurgitation, and patterns of the mitral inflow were measured in accordance with guidelines.

The area and diameter of the aneurysm were measured using manual tracings of the maximal dimension of the cavity in standard or off-axis apical views. The length of the tunnel that was created by the obstruction was measured in systole in the ideal view that allowed us to visualize the entire trajectory of the flow inside this structure, with the additional help of color flow and/or contrast agents. Color Doppler was useful in identifying the flow acceleration and in positioning the Doppler cursor or Doppler sample volume.

From the apical 4-chamber views, CW Doppler and PW Doppler patterns were acquired at the mid-LV level and at different levels of the tunnel. Patterns were annotated and described in detail, including cases with a void pattern, and 2 other different patterns of flows. The presence or absence of a gradient in the midventricular/apical region (late peaking maximal systolic gradient or maximal early diastolic gradient) was noted. An early diastolic gradient was defined as a velocity gradient between the apex and base during isovolumic relaxation or early diastole, associated with a delay of diastole at the apical level and transiently higher pressure at the apex.

RESULTS

Table 1 shows the main clinical data of the study population. Mean age was 65 years, 42% were female, all patients were in sinus rhythm during the echo examination (3 had a history of paroxysmal atrial fibrillation), 52% had hypertension, and 32% had significant ventricular arrhythmias. None of the patients had significant LV outflow tract gradient.

All patients except 1 were on pharmacologic therapy, 14 were on beta-blockers alone, 3 were on beta-blockers plus calcium channel blockers, and 1 was on calcium channel blockers alone.

Table 2 shows the main echocardiographic data. The left ventricular ejection fraction was normal in all patients except one in whom mild-moderate hypertrophy was present in the basal and medium segments.

Gradients and length of the tunnel as well as the dimension of the aneurysm were variable. A thrombus in the apical aneurysm was found in 2 cases at the TTE evaluation.

CMR (**Table 3**) confirmed a thrombus in the 2 cases and identified a thrombus in another 1, moreover, 13 cases had fibrosis of the apical wall of the aneurysm.

Figure 1 illustrates the 3 different Doppler patterns identified in our series: 12 had pattern A (more common); 6 had pattern B, characterized by a blunted early systolic flow and a paradoxical early diastolic flow; and 1 had pattern C characterized by complete paradoxical flow in systole and diastole.

The dimensions of the aneurysm were not correlated to the pressure gradient. The median length of the tunnel was 17 mm, the median diameter was 4 mm, clearly defining the obstruction shape.

Figure 2 shows an example of a multimodality approach in the evaluation of AHCM with an aneurysm with a TTE, contrast-echo, CMR, and Doppler pattern.

DISCUSSION

In this case series of patients with AHCM with aneurysm, we identified 3 different flow patterns in the LV. Pattern A is characterized by early systolic flow from the LV apex to the base, interrupted during systole (void) and followed by early diastolic emptying; pattern B is identified as blunted early systolic flow and paradoxical early diastolic flow; pattern C is defined as paradoxical flow both in systole and diastole. These data are in agreement with the data from Po et al[6] that showed midsystolic Doppler signal voids in patients with akinetic apical aneurysms and paradoxical jet in 20 of 21 of their case series.

TABLE 1 Clinical Characteristics of the Study Population

Case No.	Gender	Age (y)	BSA (m^2)	HTN	NYHA Functional Class	NSVT	Therapy
1	F	75	1.69	No	II	No	BB
2	M	63	1.99	No	I	Yes	BB
3	F	78	2.06	Yes	III	No	BB + CCB
4	M	67	1.96	Yes	I	Yes	BB
5	M	40	1.85	No	I	No	None
6	F	75	1.84	Yes	I	Yes	BB
7	M	50	2.05	No	I	No	BB
8	M	63	2.21	Yes	II	Yes	CCB
9	F	78	1.68	Yes	I	No	BB
10	F	75	1.71	Yes	I	No	BB + CCB
11	F	36	1.58	No	I	No	BB
12	M	55	1.90	Yes	I	No	BB
13	M	41	1.82	No	I	No	BB
14	M	69	2.10	Yes	II	Yes	BB
15	M	73	1.89	Yes	II	No	BB
16	F	83	1.86	No	I	No	BB
17	M	46	2.05	No	I	No	BB
18	M	72	1.94	No	I	No	BB
19	F	89	1.41	Yes	II	Yes	BB + CCB
Overall population	M (57.9%) F (42.1%)	65 ± 15	1.82 ± 0.24	No (47.4%) Yes (52.6%)	I (68.4%) II (26.3%) III (5.3%)	No (68%) Yes (32%)	BB (89.5%) CCB (21.1%)

BB = beta-blockers; BSA = body surface area; CCB = calcium-antagonist; F = female; HTN = hypertension; M = male; NSVT = nonsustained ventricular tachycardia; NYHA = New York Heart Association.

In our series of 19 patients with AHCM with aneurysm, we have encountered 3 main patterns distributed as follows: pattern A was identified in 12 (63%), whereas the other 2 patterns were identified in 6 (32%) and 1 (5%), respectively.

From a physiological point of view, systolic blood trapping in the apex escapes from the apex only in late systole-early diastole, when the obstructing neck relaxes (paradoxically because it flows from the apex toward the base in early diastole). Mid-ventricular obstruction leads to the cessation of systolic flow at the midventricular level resulting in blood trapping in the apical chamber, and, when a midventricular obstruction is relieved, blood is allowed to flow from apex to base in late systole or early diastole constituting the basis for paradoxical physiology. In the typical pattern, the severe obstruction and the apical dysfunction cause a virtual absence of flow at the level of obstructing neck; this explains the midsystolic Doppler signal void.

This phenomenon is not present in cases with AHCM without an aneurysm. Several series also showed this void pattern as typical.[6] However, Barbier et al[4] first described another pattern with a paradoxical bidirectional pressure gradient between the base of the LV and the apical cavity, which

TABLE 2 Transthoracic Echocardiography Data

Case No.	LV EDV (mL)	LV EF (%)	IVS (mm)	Posterior Wall (mm)	LAVi (mL/m^2)	E/A	MR Grade[a]
1	86	75	16	8	48	1.0	1
2	114	64	18	14	56	0.4	2
3	81	76	14	10	47	0.2	1
4	93	61	17	9	51	1.5	1
5	99	75	23	12	52	0.3	1
6	64	61	19	9	64		1
7	84	74	21	12	55	1.3	0
8	123	66	20	12	127	1.5	0
9	108	66	12	10	69		1
10	60	66	23	12	55	0.6	1
11	71	69	18	16	80	1.0	0
12	102	63	14	8		0.7	0
13	90	55	16	11	41	1.4	1
14	59	71	12	10	55		0
15	95	71	10	10	28	0.4	0
16	52	79	7	7	42		0
17	104	69	11	9	57	1.2	0
18	118	61	13	12	88	0.8	0
19	56	38	13	12	54	1.4	0
Overall population	86 ± 31	66 ± 9	16 ± 4	11 ± 2	59 ± 21	0.9 ± 0.4	0 (52.6%) 1 (42.1%) 2 (5.3%)

[a]0 = absent/trivial MR; 1 = mild MR; 2 = moderate MR.

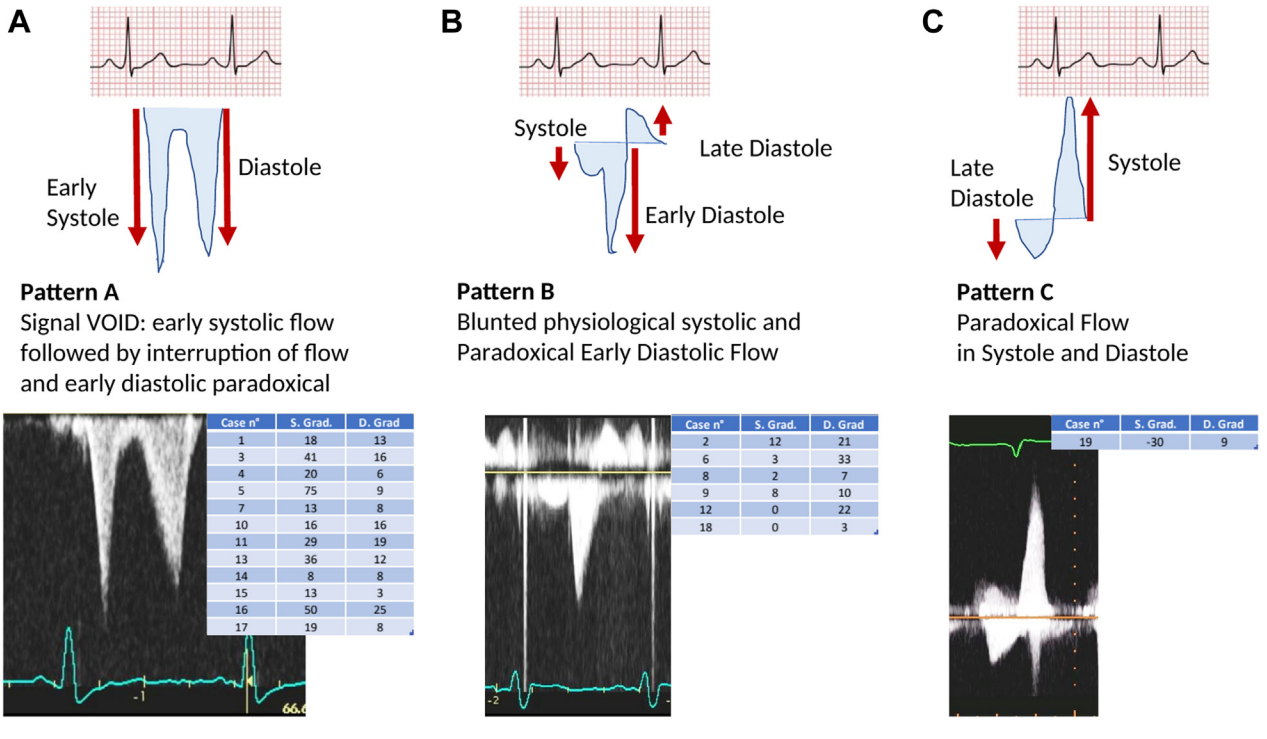
A = mitral inflow A-wave velocity; E = mitral inflow E-wave velocity; EDV= end-diastolic volume; EF = ejection fraction; IVS = interventricular septum; LAVi = left atrium volume index; LV = left ventricular; MR = mitral regurgitation.

TABLE 3 Characteristics of Apical Aneurysms as Assessed Using Multimodality Imaging

Case No.	Apical Fibrosis by CMR	Tunnel L (mm)	Tunnel D (mm)	Aneurysm Area (mm^2)	Systolic G (mm Hg)	Diastolic G (mm Hg)	Thrombus
1	NP	20	4	10	18	13	No
2	Yes	18	3	8	12	21	No
3	Yes	10	4	10	41	16	No
4	Yes	10	2	8	20	6	No
5	Yes	16	3	19	75	9	No
6	Yes	19	6	27	3	33	No
7	Yes	14	3	19	13	8	Yes
8	Yes	23	3	24	2	7	No
9	Yes	9	2	18	8	10	No
10	NP	23	6	31	16	16	No
11	Yes	16	4	11	29	19	Yes
12	Yes	11	7	36	0	22	Yes
13	Yes	16	8	15	36	12	No
14	NP	20	6	10	8	8	No
15	Yes	14	3	8	13	3	No
16	NP	19	3	11	50	25	No
17	Yes	19	5	12	19	8	No
18	NP	16	5	18	0	3	No
19	NP	22	4	30	-30	9	No
Overall population	13 (68.4%)	17.0 ± 4.2	4.0 ± 1.5	1.25 ± 0.65	17.5 ± 21.0	13.0 ± 7.7	No (84.2%) Yes (15.8%)

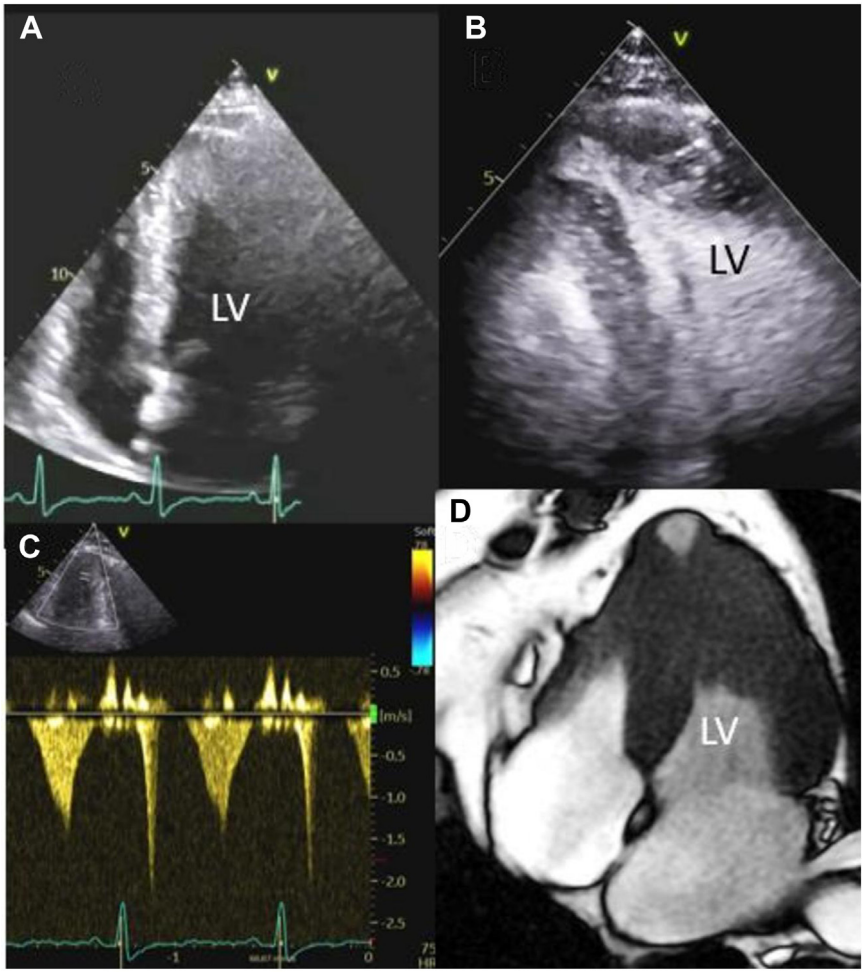
CMR = cardiac magnetic resonance; D = diameter; G = gradient; L = length; NP = not performed.

FIGURE 1 Schematic Representation of the Doppler Pattern



(A) Pattern A (more frequent). **(B)** Pattern B. **(C)** Pattern C. S. Grad = systolic gradient (mm Hg); D. G. = diastolic gradient (mm Hg).

FIGURE 2 **Multimodality Imaging of AHCM With an Aneurysm**



(A) TTE view of left ventricular AHCM. **(B)** Contrast-echo of apical hypertrophy with an aneurysm. **(C)** CW Doppler pattern. **(D)** CMR showing apical hypertrophy with aneurysm. Contrast echocardiography and CMR better identify this small apical aneurysm in comparison with the standard 2-dimensional echocardiography. ACHM = apical hypertrophic cardiomyopathy; CMR = cardiac magnetic resonance; CW = continuous-wave; TTE = transthoracic echo.

in our series was found only in 1 case. We demonstrated an intermediate pattern in 32% of patients, characterized by blunted early systolic flow and paradoxical early diastolic flow (Figure 1).

Our patients had similar clinical presentations as reported in the larger series. The mean age was 65 years, hypertension was present in 52%, paroxysmal atrial fibrillation in 16%, and nonsustained ventricular tachycardia in 32%. Moreover, LV showed normal systolic function (left ventricular ejection fraction: 66%), normal diastolic volumes, and mild-to-moderate hypertrophy of basal and medium walls (mean septum thickness: 16 mm). Tunnel length and area/diameter of the aneurysm were variable, but the mean values were in the range of the majority of other studies.

Lee et al[7] showed a linear relationship between intracavitary gradients and aneurysm size, and observed that a 10-mm Hg increase in intracavitary gradient was associated with a 1.6-mm increase in aneurysm size. There was no significant relationship

between intracavitary gradient and adverse clinical events, including stroke.[7]

The mean systolic and diastolic velocities in our study were largely individually different and not clearly related to aneurysm size.

CONCLUSIONS

In our case series of 19 patients with AHCM and apical aneurysm, we identified 3 different flow patterns at the midventricular level using CW Doppler. The most frequent was pattern A, followed by pattern B and pattern C. All are an expression of increased intracavitary pressure and consequently decreased coronary pressure that may contribute to the pathophysiology of aneurysm formation.

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KEY WORDS apical aneurysm, Doppler pattern of left ventricle intracavitary gradients, hypertrophic cardiomyopathy