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Ace-of-Spades Morphology in Apical Hypertrophic Cardiomyopathy With Exertional Syncope and Nonsustained Ventricular Tachycardia: A Case Report

Maria Isabel Sanchez Sanchez [1], Cecilia Andrea Baquerizo Suárez [1, 2], Jaime Patricio Lema Masalema [1], Juliana Patricia Aguilar Moncayo [3], Leslie De Loor Zapata [3]

1. Department of Cardiology, Hospital de Especialidades Guayaquil Dr. Abel Gilbert Ponton, Guayaquil, ECU 2. Graduate School of Health, Universidad Espíritu Santo, Samborondón, ECU 3. School of Medicine, University of Guayaquil, Guayaquil, ECU

Corresponding author: Maria Isabel Sanchez Sanchez, missdosanc@gmail.com

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Abstract

Apical hypertrophic cardiomyopathy (ApHCM), also known as Yamaguchi syndrome, is an uncommon phenotypic variant of hypertrophic cardiomyopathy characterized by predominant hypertrophy of the left ventricular apex and a distinctive ace-of-spades ventricular configuration. A 41-year-old man with a family history of sudden cardiac death presented after an episode of exertional syncope while playing soccer. Electrocardiography showed sinus rhythm with marked T-wave inversion across the precordial leads. Transthoracic echocardiography demonstrated preserved left ventricular systolic function with mid-apical hypertrophy, while cardiac magnetic resonance imaging confirmed the apical hypertrophic phenotype. Twenty-four-hour ambulatory electrocardiographic monitoring documented four episodes of nonsustained ventricular tachycardia (NSVT), with a maximum ventricular rate of 198 beats/minute and a longest episode of approximately 26 seconds. Coronary angiography showed no significant obstructive coronary artery disease. Following individualized sudden cardiac death risk assessment, a dual-chamber transvenous implantable cardioverter-defibrillator was placed for primary prevention, and the patient remained asymptomatic during follow-up while receiving beta-blocker therapy. This case highlights the value of integrating electrocardiographic findings, multimodality cardiac imaging, ambulatory rhythm monitoring, and individualized arrhythmic risk assessment in patients with ApHCM presenting with exertional syncope and NSVT.

Categories: Cardiology
Keywords: ace-of-spades morphology, apical hypertrophic cardiomyopathy, implantable cardioverter-defibrillator, sudden cardiac death, yamaguchi syndrome

Introduction

Apical hypertrophic cardiomyopathy (ApHCM), also known as Yamaguchi syndrome, is a phenotypic variant of hypertrophic cardiomyopathy characterized by predominant hypertrophy of the left ventricular apex. First described in 1976, ApHCM is less common in European and North American HCM cohorts, in which it accounts for approximately 8% of cases, whereas substantially higher proportions have been reported in Asian cohorts, reaching up to 40% in some series [1]. Although hypertrophic cardiomyopathy is commonly inherited in an autosomal dominant pattern, a pathogenic genetic variant is not identified in every patient, reflecting the genetic and phenotypic heterogeneity of the disease [1].

Clinical expression is variable. Some patients remain asymptomatic, whereas others may present with chest pain, exertional dyspnea, palpitations, or syncope [1]. Electrocardiographic abnormalities, particularly deep T-wave inversion in the precordial leads, may provide an important diagnostic clue [1,2]. Transthoracic echocardiography remains the initial imaging modality; however, apical hypertrophy may be underestimated or missed when endocardial visualization is suboptimal or apical views are foreshortened. In these circumstances, cardiac magnetic resonance imaging provides a more complete assessment of apical morphology and may identify associated findings such as apical aneurysm or myocardial fibrosis [1-3].

Although ApHCM was historically regarded as a relatively favorable hypertrophic cardiomyopathy phenotype, it is now recognized that its clinical course is heterogeneous and not uniformly benign. Clinically significant complications may occur, including atrial and ventricular arrhythmias, heart failure, thromboembolic events, apical aneurysm formation, and sudden cardiac death [4-6]. Recognition of these features is therefore important not only for diagnosis but also for individualized risk assessment and long-term surveillance [3,7].

This report describes a patient with ApHCM presenting with exertional syncope, marked precordial T-wave inversion, mid-apical hypertrophy, and nonsustained ventricular tachycardia (NSVT) documented on

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ambulatory monitoring. The case illustrates the complementary role of electrocardiography, echocardiography, cardiac magnetic resonance imaging, and arrhythmic risk assessment in the evaluation of ApHCM.

Case Presentation

A 41-year-old man with no previously documented cardiovascular disease presented after a transient loss of consciousness while playing soccer. The episode occurred during exertion and lasted approximately 30 seconds, followed by spontaneous recovery. His family history was notable for sudden cardiac death in his maternal grandfather at 52 years of age.

A 12-lead electrocardiogram showed sinus rhythm with marked T-wave inversion across the precordial leads V1-V6 (Figure *1*). Serum troponin I was mildly elevated at 0.15 ng/mL (reference value <0.04 ng/mL). Given the clinical presentation and electrocardiographic abnormalities, further cardiovascular evaluation was undertaken.

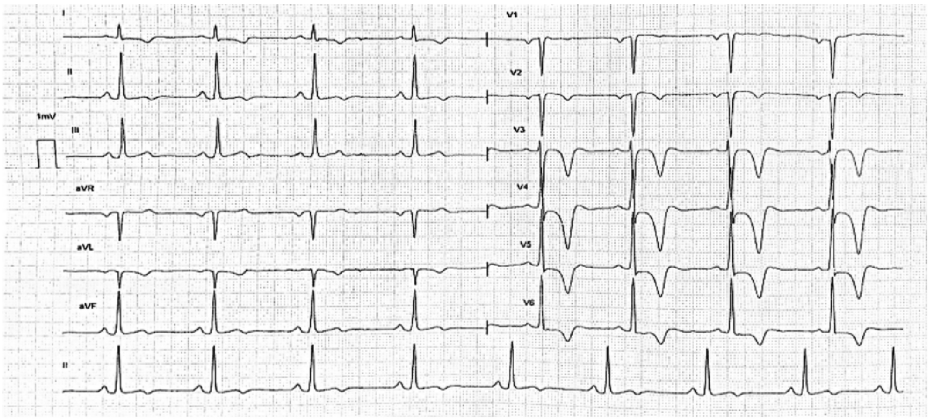


FIGURE 1: Twelve-lead electrocardiogram showing sinus rhythm with marked T-wave inversion in the precordial leads V1-V6.

Transthoracic echocardiography demonstrated predominant mid-apical hypertrophy, with a maximum left ventricular wall thickness of 15 mm, a left atrial diameter of 28 mm, and preserved left ventricular systolic function with a left ventricular ejection fraction of 70% by the biplane Simpson method (Figures *2*-*3*). The resting maximal left ventricular outflow tract gradient was 5 mmHg. A provoked gradient with the Valsalva maneuver was not documented in the available echocardiographic study.

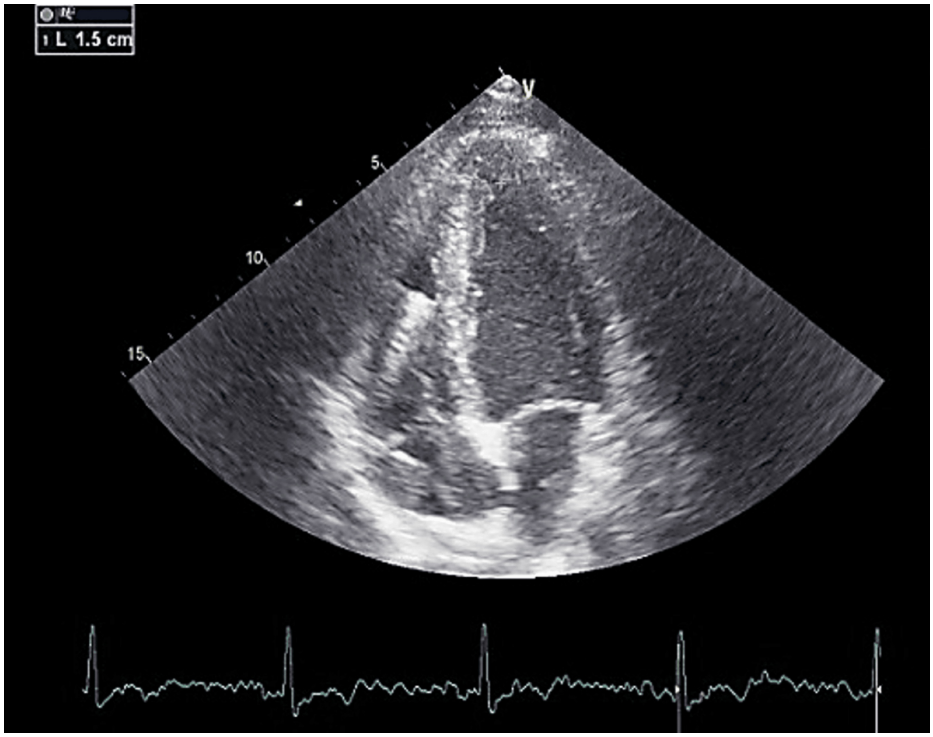


FIGURE 2: Transthoracic echocardiography, apical four-chamber view, demonstrating predominant mid-apical left ventricular hypertrophy with a maximal wall thickness of 15 mm and an ace-of-spades configuration.

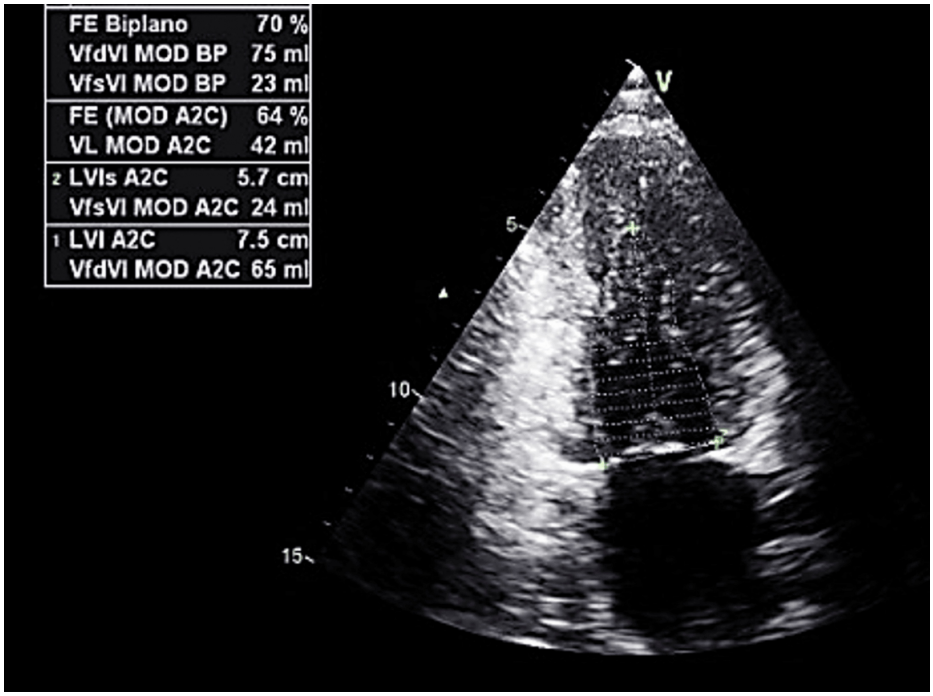


FIGURE 3: Transthoracic echocardiography, apical two-chamber view, demonstrating predominant mid-apical left ventricular hypertrophy with preserved left ventricular systolic function.

Twenty-four-hour ambulatory electrocardiographic monitoring showed sinus rhythm with occasional premature ventricular complexes and supraventricular ectopic beats. Four episodes of NSVT were recorded,

with a maximum ventricular rate of 198 beats/minute. The longest episode lasted approximately 26 seconds. The Holter report did not document symptom correlation with the NSVT episodes or specify whether the ventricular tachycardia morphology was monomorphic or polymorphic. No pathological pauses were reported.

Coronary angiography demonstrated no significant obstructive coronary artery disease. Cardiac magnetic resonance imaging confirmed predominant hypertrophy of the mid-apical and apical regions of the left ventricle, supporting the diagnosis of ApHCM (Figures *4*-*5*).



FIGURE 4: Cardiac magnetic resonance imaging demonstrating predominant hypertrophy of the mid-apical and apical regions of the left ventricle.

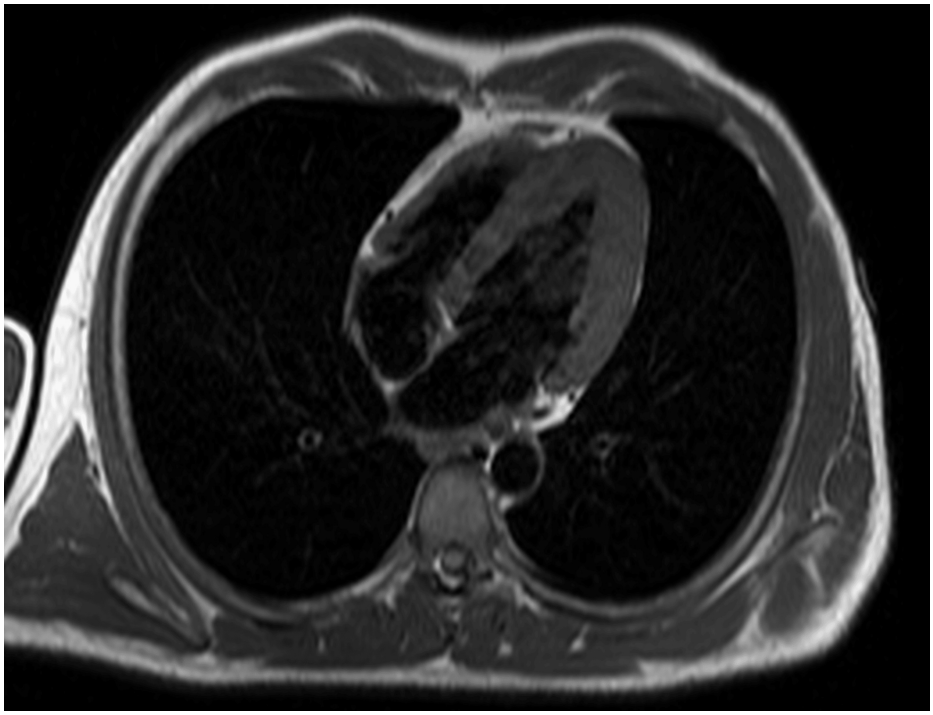


FIGURE 5: Cardiac magnetic resonance imaging, axial view, demonstrating predominant apical left ventricular hypertrophy.

Using the documented clinical variables available for the HCM Risk-SCD model [8], including the resting left ventricular outflow tract gradient because a provoked gradient was not documented, the estimated five-year sudden cardiac death risk was approximately 5.2%. The maternal grandfather's history of sudden cardiac death was not entered as a positive family-history variable because it did not meet the model-specific definition. Given the patient's unexplained exertional syncope and documented NSVT, implantable cardioverter-defibrillator (ICD) candidacy was assessed within the overall clinical risk profile, and a dual-chamber transvenous ICD was placed for primary prevention. The specific rationale for selecting a transvenous system rather than a subcutaneous or extravascular ICD was not documented in the available clinical record.

Genetic counseling and genetic testing were not documented during the available evaluation or follow-up. The patient subsequently remained on beta-blocker therapy and remained asymptomatic during cardiology follow-up.

Discussion

ApHCM, historically known as Yamaguchi syndrome, is a distinctive phenotypic expression of hypertrophic cardiomyopathy characterized by predominant hypertrophy of the left ventricular apex. Its recognition can be challenging because clinical manifestations are heterogeneous and electrocardiographic abnormalities may be striking despite preserved global systolic function. The present case illustrates this pattern, with exertional syncope, marked precordial T-wave inversion, preserved left ventricular ejection fraction, and predominant mid-apical hypertrophy with an ace-of-spades configuration [1,4].

Marked T-wave inversion in the precordial leads is a characteristic electrocardiographic finding in ApHCM and may initially raise concern for myocardial ischemia, particularly when accompanied by symptoms or biomarker elevation [1,2]. In this patient, serum troponin I was mildly elevated; however, coronary angiography demonstrated no significant obstructive coronary artery disease. Transthoracic echocardiography identified mid-apical hypertrophy with preserved systolic function, while cardiac magnetic resonance imaging further characterized the apical distribution of hypertrophy. This complementary use of echocardiography and cardiac magnetic resonance imaging is particularly relevant in ApHCM because the apex may be incompletely visualized or foreshortened on conventional echocardiographic views. Previous reviews have similarly emphasized that careful apical imaging and multimodality assessment are important for recognizing the characteristic morphology and associated structural abnormalities of ApHCM [1-3,9].

Although ApHCM was historically considered to have a relatively favorable prognosis compared with other hypertrophic cardiomyopathy phenotypes, subsequent studies have demonstrated substantial clinical

heterogeneity. Ventricular and atrial arrhythmias, heart failure, thromboembolic events, apical aneurysm formation, and sudden cardiac death may occur during follow-up [4-6]. Accordingly, the presence of an apical phenotype should not, by itself, be interpreted as indicating low arrhythmic risk. Risk assessment should instead integrate the clinical presentation, ambulatory electrocardiographic findings, ventricular morphology, systolic function, and cardiac magnetic resonance findings when available [3,7].

Risk stratification was particularly relevant in this patient because he experienced unexplained exertional syncope and ambulatory monitoring documented NSVT. Re-review of the original 24-hour Holter recording showed four NSVT episodes, with a maximum ventricular rate of 198 beats/min and a longest episode of approximately 26 seconds. No symptoms temporally associated with the NSVT episodes were documented in the available Holter report, and the ventricular tachycardia morphology was not explicitly classified as monomorphic or polymorphic. Using the documented clinical variables, the HCM Risk-SCD model [8] yielded an estimated five-year sudden cardiac death risk of approximately 5.2%. This estimate should not be interpreted as an isolated indication for ICD therapy. Contemporary guidelines emphasize individualized risk assessment rather than reliance on a single numerical threshold. In adults with hypertrophic cardiomyopathy, recent unexplained syncope considered potentially arrhythmic represents an important marker of sudden cardiac death risk, while NSVT detected on ambulatory monitoring provides additional information when considering primary-prevention ICD therapy [7].

The maximum left ventricular wall thickness of 15 mm in this patient did not meet the criterion for massive left ventricular hypertrophy, defined as a wall thickness of 30 mm or greater [7]. Therefore, the decision to implant an ICD should be understood within the context of the overall arrhythmic risk profile, particularly the unexplained exertional syncope and documented NSVT, rather than being attributed solely to the degree of hypertrophy or to the HCM Risk-SCD estimate. A dual-chamber transvenous ICD was implanted for primary prevention. The available clinical record did not document the specific rationale for selecting a transvenous system over a subcutaneous or extravascular device; therefore, no retrospective assumption regarding device selection was made.

Cardiac magnetic resonance imaging supported the mid-apical and apical hypertrophic phenotype. However, detailed late gadolinium enhancement burden and scar distribution were not available in the archived documentation. This limits assessment of the potential contribution of myocardial fibrosis to arrhythmic risk in this case. Similarly, genetic counseling and genetic testing were not documented during the available evaluation or follow-up, limiting genotype-based cascade assessment of potentially affected relatives.

Following ICD implantation, the patient remained under cardiology and electrophysiology follow-up while receiving beta-blocker therapy and was asymptomatic at the most recent documented evaluation. Long-term surveillance remains important in ApHCM because the phenotype may evolve over time and clinically relevant complications may develop despite preserved left ventricular systolic function. Serial clinical assessment, ambulatory rhythm monitoring, device follow-up, and multimodality cardiac imaging therefore remain appropriate components of ongoing care [3,7].

Conclusions

ApHCM is a distinct hypertrophic cardiomyopathy phenotype that may present with exertional syncope, marked precordial T-wave inversion, and ventricular arrhythmias despite preserved left ventricular systolic function. In this case, the integration of electrocardiography, transthoracic echocardiography, ambulatory rhythm monitoring, and cardiac magnetic resonance imaging allowed accurate characterization of the apical phenotype and supported individualized sudden cardiac death risk assessment. The presence of unexplained exertional syncope and documented NSVT emphasized the importance of integrating multiple clinical and imaging variables rather than relying on a single numerical risk estimate. Long-term clinical follow-up, rhythm surveillance, device monitoring, and periodic multimodality imaging remain important for detecting disease progression and potential complications.

Additional Information

Author Contributions

All authors have reviewed the final version to be published and agreed to be accountable for all aspects of the work.

Concept and design: Cecilia Andrea Baquerizo Suárez, Maria Isabel Sanchez Sanchez

Drafting of the manuscript: Cecilia Andrea Baquerizo Suárez

Critical review of the manuscript for important intellectual content: Cecilia Andrea Baquerizo Suárez, Maria Isabel Sanchez Sanchez, Jaime Patricio Lema Masalema, Leslie De Loor Zapata, Juliana Patricia Aguilar Moncayo

Supervision: Cecilia Andrea Baquerizo Suárez, Maria Isabel Sanchez Sanchez

Acquisition, analysis, or interpretation of data: Maria Isabel Sanchez Sanchez, Jaime Patricio Lema Masalema, Leslie De Loor Zapata, Juliana Patricia Aguilar Moncayo

Disclosures

Human subjects: Informed consent for treatment and open access publication was obtained or waived by all participants in this study. **Conflicts of interest:** In compliance with the ICMJE uniform disclosure form, all authors declare the following: **Payment/services info:** All authors have declared that no financial support was received from any organization for the submitted work. **Financial relationships:** All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work. **Other relationships:** All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

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Maria Isabel Sanchez Sanchez and Cecilia Baquerizo Suarez contributed equally to the work and should be considered co-first authors.

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