JACC: CASE REPORTS VOL. 4, NO. 23, 2022

CASE REPORT

ADVANCED

CLINICAL CASE

Accelerated Idioventricular Rhythm in a Young Athlete

Physiology or Pathology?

Nick R. Bijsterveld, MD, PhD,[a,b] Saskia N. van der Crabben, PhD,[c] Maarten Groenink, MD, PhD,[a] Arthur Wilde, MD, PhD,[a] Harald Jørstad, MD, PhD[a]

ABSTRACT

An accelerated idioventricular rhythm was seen on a routine preparticipation electrocardiogram of a 19-year-old healthy and symptom-free athlete. Family history was negative for cardiac disease. Additional investigations revealed a hypertrophic cardiomyopathy, confirmed with cardiac magnetic resonance imaging and genetic analysis. Accelerated idioventricular rhythm in young athletes warrants careful clinical evaluation. (**Level of Difficulty: Advanced.**)

HISTORY OF PRESENTATION

Before sports college admittance, a 19-year-old male athlete underwent sports physician-led preparticipation screening. A resting electrocardiogram (ECG) was performed, which revealed accelerated idioventricular rhythm (AIVR) alternating with sinus bradycardia, with normal PR and QRS conduction intervals (**Figure 1**). The athlete reported no history of cardiac disease, no complaints, and denied any (pre) syncopal events or chest discomfort. His sports history reported high levels of physical activity from the age of 8 years, with currently 9 sports-hours (63 MET/h) per week, mainly soccer. He had no other relevant medical history and reported a negative family history for cardiac disease and sudden cardiac death (SCD). The sports physician referred the athlete to our sports cardiology outpatient clinic for further analysis of the abnormal ventricular rhythm.

LEARNING OBJECTIVES

- To understand differential diagnosis and diagnostic approach of an AIVR in an asymptomatic individual without family history or symptoms.
- To understand value of different diagnostic modalities to distinguish between pathology and physiology in a patient with AIVR.

ACCELERATED IDIOVENTRICULAR RHYTHM

AIVR is associated with cardiac pathology and is observed in cardiomyopathies, after myocardial infarction, and during electrolyte disturbances or digoxin intoxication.[1-3] AIVR has also been reported as a benign rhythm variant without evidence of

From the [a]Department of Cardiology, Amsterdam UMC, Heart Centre, Amsterdam, the Netherlands; [b]Department of Cardiology, Flevoziekenhuis, Almere, the Netherlands; and [c]Department of Human Genetics, Amsterdam, Amsterdam, the Netherlands.
Eugene Chung, MD, served as Guest Associate Editor for this paper.
The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the Author Center.

Manuscript received June 27, 2022; revised manuscript received September 15, 2022, accepted September 23, 2022.

ISSN 2666-0849 https://doi.org/10.1016/j.jaccas.2022.09.023

ABBREVIATION AND ACRONYMS

AIVR = accelerated idioventricular rhythm

CMR = cardiac magnetic resonance

ECG = electrocardiogram

HCM = hypertrophic cardiomyopathy

SCD = sudden cardiac death

underlying cardiac pathology in children, adolescents, and athletes.[4-7]

INVESTIGATIONS

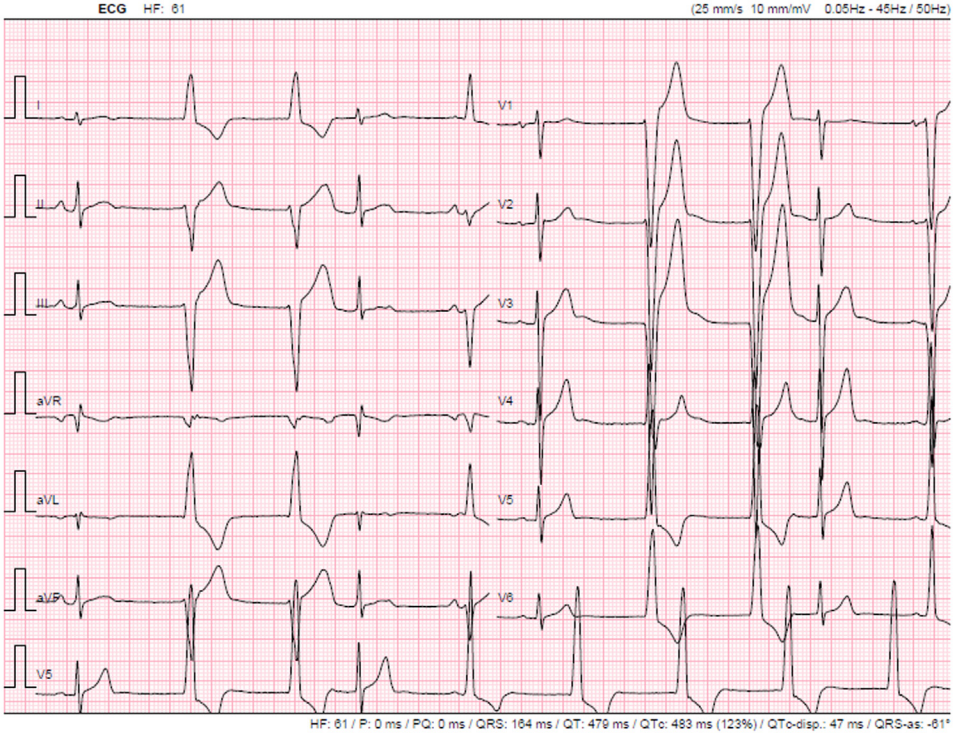
Extensive history-taking focusing on symptoms and family history revealed no additional relevant information. Physical examination was normal with a body mass index of 20 kg/m^2, blood pressure 135/70 mm Hg, and normal cardiac sounds without murmurs. A subsequently performed 24-h Holter analysis revealed sinus rhythm 40-130 beats/min but with AIVR 3.7% of total beats (**Figure 2**). Cardiopulmonary exercise testing demonstrated a maximum oxygen consumption of 3,180 mL/min (57 mL/min/kg), a maximum heart rate of 188 beats/min (104% of predicted maximum), and a normal blood pressure increase. The ECGs during exercise revealed no arrhythmias, and normal sinus rhythm and conduction was seen throughout the entire test. Echocardiography demonstrated biventricular dimensions and ejection fractions within normal limits (Video 1). Septal and posterior wall thicknesses were 10 and 7 mm, respectively. Left ventricular global longitudinal strain was abnormal and showed decreased values ranging from -9% to -16% in the basal segments (**Figure 3**).

DIFFERENTIAL DIAGNOSIS

The patient was discussed at our multidisciplinary sports cardiology team meeting, which includes a broad field of expertise, including electrophysiologists, imaging cardiologists, sports cardiologists, pediatric cardiologists, sports physicians, and clinical geneticists. The sports cardiology team consensus was that AIVR is seen regularly in athletes, both by sports physicians and sports cardiologists. The majority of the team agreed that this young athlete's AIVR, in the absence of complaints and/or a positive family history of cardiac disease, could be a finding within normal limits.

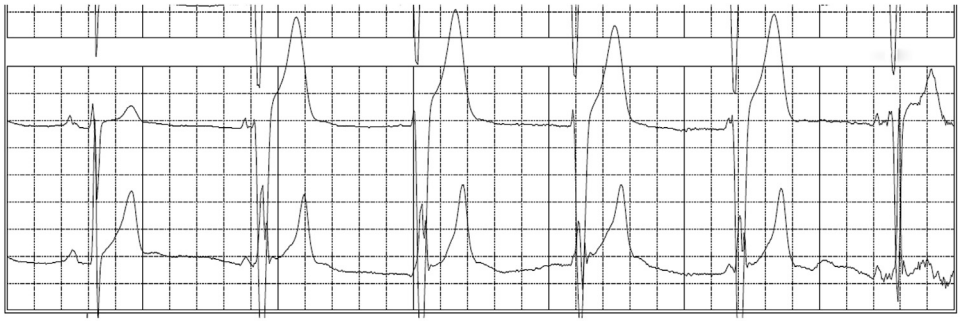
However, consensus could not be reached that pathology was excluded. First, although atrial ectopic and junctional rhythm is considered to be a finding

FIGURE 1 Resting Electrocardiogram



Resting electrocardiogram performed during routine preparticipation screening showing alternating sinus rhythm and accelerated idioventricular rhythm (AIVR).

FIGURE 2 **24-h Holter Analysis**



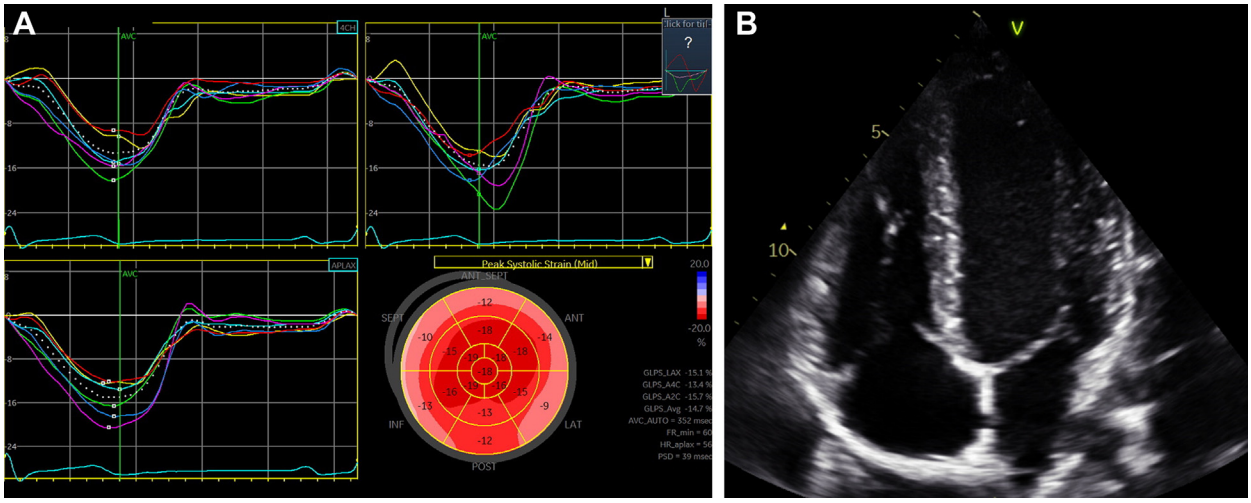
During 24-h Holter analysis frequent episodes of asymptomatic of accelerated idioventricular rhythm (AIVR) were observed.

within normal limits in athletes' resting ECGs, AIVR specifically is not.[8] Second, there was a decreased echocardiographic global longitudinal strain in the basal segments of the left ventricle, which is an abnormal, albeit nonspecific finding. Therefore, the sports cardiology team advised a multimodal approach, recommending in particular a cardiac magnetic resonance imaging (CMR) document or exclusion of (subtle) structural heart disease, based on the possible association between AIVR and underlying heart disease.[1-3]

MANAGEMENT

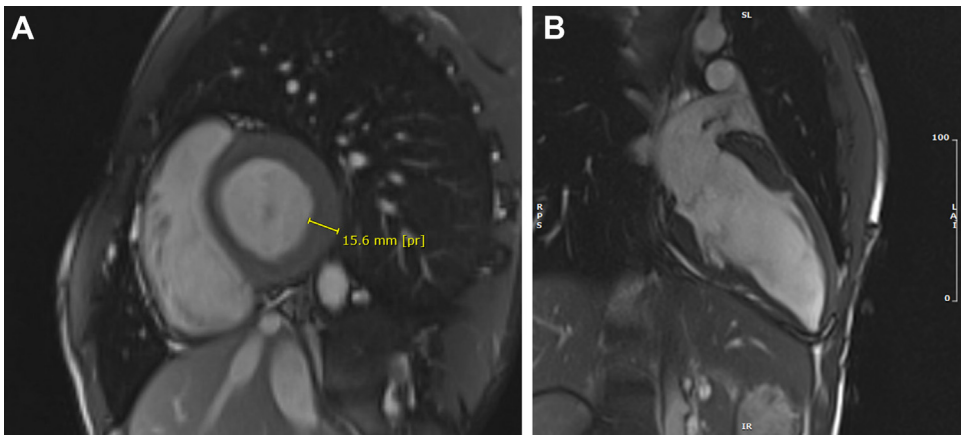
The CMR revealed a structurally abnormal heart, with asymmetric hypertrophy of the basal inferolateral wall, with a maximum myocardial thickness of 15 mm (**Figure 4A,** Video 2). Crypts were also seen in the basal inferior and basal anterior segments (**Figure 4B**, Video 2). No late gadolinium enhancement was seen (Video 2). These phenotypic findings were deemed to be highly suspicious for hypertrophic cardiomyopathy.[9] Subsequent genetic analysis

FIGURE 3 **Echocardiographic Myocardial Strain**



(A) The basal segments of the left ventricle showed decreased myocardial strain values using speckle tracking echocardiography. **(B)** Normal dimensions and wall thickness in the 4-chamber view.

FIGURE 4 CMR



(A) Basal short axis showing asymmetric hypertrophic myocardium with maximum wall thickness of 15.6 millimeter. **(B)** 2-chamber view showing basal myocardial crypt formation **(yellow arrows)**. CMR = cardiac magnetic resonance.

identified a pathogenic (class 5) *MYBPC3* c.2373dup (p.Trp792Valfs*17) variant,[10] a Dutch founder variant,[11] and confirmed the diagnosis of hypertrophic cardiomyopathy (HCM). Cascade family screening revealed hitherto unreported cardiac complaints in the athlete's younger brother, with episodes of exercise-induced palpitations and dizziness. Echocardiographic analysis revealed asymmetric myocardial hypertrophy with a septum thickness of 22 mm. Carrier testing for the pathogenic MYBC3 variant confirmed that the brother had this variant. Further cascade screening also identified their sister and father as carriers. Additional diagnostics are currently being conducted, including CMR and ambulant rhythm monitoring in all identified carriers.

DISCUSSION

With these new results, a repeat evaluation took place in our multidisciplinary sports cardiology team, this time focusing on potential detrimental cardiac effects of (the continuation of) sports in an athlete with de novo diagnosis of hypertrophic cardiomyopathy. Risk factors for exercise-induced SCD in HCM were evaluated.[12] In our patient, the family history was negative for SCD. Although septal wall thickness was modestly increased, cardiac fibrosis and apical aneurysm were absent, and there was no relevant left ventricular pressure gradient, combined with a normal blood pressure response during exercise. There were no nonsustained ventricular tachycardias or premature ventricular beats on repeated ambulant rhythm monitoring and exercise tests. Finally, the patient remained asymptomatic during follow-up. Although not validated in athletes, according to the European Society of Cardiology sports guideline, we calculated the HCM 5-year SCD risk score, which was 1.74%.[10] Based on all of these findings, the sports cardiology expert team concluded that there were only mild phenotypic HCM features and no high-risk markers for SCD.[12-14] Therefore, our sports advice was "symptom limited," without specific restrictions, and continuation of sports-college education, dependent on close clinical monitoring by experts in the field of sports cardiology and cardiomyopathies.

FOLLOW-UP

We discussed our advice with the patient and his college medical team, and after a process of shared decision-making, the patient continued his sports college education and sports activities. At 1.5 years of follow-up after diagnosis, the patient is performing well, demonstrates no relevant cardiac changes, and remains asymptomatic. Due to the exercise-induced complaints and severe asymmetrical septal hypertrophy of 22 mm, his younger brother was given immediate sports restrictions advice, pending further analysis.

CONCLUSIONS

AIVR is defined as a ventricular ectopic rhythm with a frequency between 50 and 110 beats/min. The mechanism is accelerated phase 4 depolarization of His-Purkinje fibers or ventricular myocardium.[15] AIVR

is associated with cardiac pathology, such as after myocardial infarction, cardiomyopathies, electrolyte disturbances, and with digoxin intoxication,[1-3] but is also seen in patients without any cardiac abnormalities, and has been described in children, adolescents, and athletes.[4-7]

The hypothesis of physiological AIVR constitutes an increased vagal and decreased sympathetic tone combined with enhanced ventricular pacemaker activity (enhanced automaticity). Distinguishing between physiology and pathology requires careful cardiac evaluation, including cardiac imaging and ischemia detection. In our young patient, there was a high probability of benign AIVR, supported by the combination of a negative family history, young age, high level of sports activity, and normal cardiopulmonary exercise testing. Furthermore, this phenomenon is reported by sports physicians who regularly screen (young) athletes. Currently, data on the incidence of AIVR in patients with HCM is limited and mainly consist of case reports. Therefore, although an association between AIVR and HCM in our case is plausible, we cannot exclude that the AIVR was an incidental finding, unrelated to the structural cardiac disease. Studies comparing (hypertrophic) cardiomyopathy patients with controls should be performed to further quantify the relationship between AIVR and underlying pathology, and determine whether there is a certain "dose" of AIVR that could be considered benign. Given the paucity of data, we believe that our case demonstrated that cardiac analysis is warranted to exclude underlying pathology even in the presence of a negative family history. A young athlete with AIVR mandates careful, multidisciplinary evaluation in addition to sensitive imaging studies to distinguish between physiology and pathology. In this patient, echocardiographic longitudinal strain analysis, which has been shown to be able to identify early stages of myocardial dysfunction,[16] and is recommended by the American Heart Association/American College of Cardiology and the European Society of Cardiology[17] to perform in routine clinical practice, was an important tool for discovering the underlying cardiac disease. Whether this measurement is sensitive enough for exclusion of (early stage) myocardial pathology is yet unknown. A multimodal approach that includes CMR has a high sensitivity in diagnosing structural heart disease, but the availability of this modality is resource dependent. Based on our findings, we advise that in athletes with AIVR both high-quality strain analysis performed by an experienced echocardiographist and CMR should be part of a multimodality diagnostic approach.

In our patient, the diagnosis of HCM did not results in a sports restrictions based on extensive evaluation of the multidisciplinary sports cardiology expert team. However, it should be kept in mind that numerous factors in this process are dependent on national and regional factors, such as local legislation, or the role of the university or college team physicians.

FUNDING SUPPORT AND AUTHOR DISCLOSURES

The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

ADDRESS FOR CORRESPONDENCE: Dr Nick R. Bijsterveld, Department of Cardiology, Amsterdam UMC, Heart Centre, location AMC, Meibergdreef 9, 1105 AZ, Amsterdam, the Netherlands. E-mail: n.r.bijsterveld@amsterdamumc.nl.

REFERENCES

1. Delise P, D'Este D, Monico E, et al. Accelerated idioventricular rhythm with isorhythmic A-V dissociation. An electrophysiological study. *G Ital Cardiol*. 1980;10:1588-1593.

2. Wang L, Liu H, Zhu C, et al. Clinical characteristics and therapeutic strategy of frequent accelerated idioventricular rhythm. *BMC Cardiovasc Disord*. 2021;21:425.

3. Shah JA, Naz F, Kumar R, et al. Incidence of cardiac arrhythmias in acute myocardial infarction patients undergoing primary percutaneous coronary intervention and associated outcomes during the first 24 hours. *Cureus*. 2021;13:e12599.

4. Bernard Y, Dambrine P, Ravault M, Pernot C. Accelerated idioventricular rhythm in children. Apropos of a case. *Arch Mal Coeur Vaiss*. 1981;74:623.

5. Riva UR, Budriesi N, Fancinelli M, Labriola E. An accelerated idioventricular rhythm and sports activity. Comments on a clinical case and a characterization of the arrhythmia. *Cardiologia*. 1994;39:591-596.

6. Thomas JE, Katapadi M. Accelerated idioventricular rhythm in a healthy teenager post circumcision. *Conn Med*. 2004;68:555-557.

7. Hingorani P, Karnad DR, Rohekar P, Kerkar V, Lokhandwala YY, Kothari S. Arrhythmias seen in baseline 24-hour Holter ECG recordings in healthy normal volunteers during phase 1 clinical trials. *J Clin Pharmacol*. 2016;56:885-893.

8. Sharma S, Drezner JA, Baggish A, et al. International recommendations for electrocardiographic interpretation in athletes. *J Am Coll Cardiol*. 2017;69:1057-1075.

9. Germans T, Wilde AAM, Dijkmans PA, et al. Structural abnormalities of the inferoseptal left ventricular wall detected by cardiac magnetic resonance imaging in carriers of hypertrophic cardiomyopathy mutations cardiac imaging. *J Am Coll Cardiol*. 2006;48:2518-2523.

10. The Task Force for the Diagnosis and Management of Hypertrophic Cardiomyopathy of the European Society of Cardiology (ESC). 2014 ESC Guidelines on diagnosis and management of hypertrophic cardiomyopathy. *Eur Heart J*. 2014;35:2733-2779.

11. Alders M, Jongbloed R, Deelen W, et al. The 2373insG mutation in the MYBPC3 gene is a founder mutation, which account for nearly one-fourth of the HCM cases in the Netherlands. *Eur Heart J*. 2003;24:1848-1853.

12. Hong Y, Su W, Li X. Risk factors of sudden cardiac death in hypertrophic cardiomyopathy. *Curr Opin Cardiol*. 2022;37:15-21.

13. The Task Force on sports cardiology and exercise in patients with cardiovascular disease of the European Society of Cardiology (ESC). 2020 ESC Guidelines on sports cardiology and exercise in patients with cardiovascular disease. *Eur Heart J*. 2021;42:17-96.

14. Ommen S, Mital S, Burke M, et al. 2020 AHA/ACC guideline for the diagnosis and treatment of patients with hypertrophic cardiomyopathy: executive summary: a report of the American College of Cardiology/American Heart Association Joint Committee on Clinical Practice Guidelines. *J Am Coll Cardiol*. 2020;22:3022-3055.

15. Riera AR, Barros RB, de Sousa FD, Baranchuk A. Accelerated idioventricular rhythm: history and chronology of the main discoveries. *Indian Pacing Electrophysiol J*. 2010;10:40-48.

16. Singh R, Sozzi F, Fedacko J, et al. Pre-heart failure at 2D- and 3D-speckle tracking echocardiography: a comprehensive review. *Echocardiography*. 2022;39:302-309.

17. Lang R, Badano L, Mor-Avi V, et al. Recommendations for cardiac chamber quantification by echocardiography in adults: an update from the American Society of Echocardiography and the European Association of Cardiovascular Imaging. *J Am Soc Echocardiogr*. 2015;28:1-39.

KEY WORDS **accelerated idioventricular rhythm, athlete, hypertrophic cardiomyopathy, sports cardiology, ventricular rhythm**

APPENDIX **For supplemental videos, please see the online version of this paper.**