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CLASSIC CASE REPORT
Arrhythmias/electrophysiology

A case report of profound atrioventricular block in an endurance athlete: how far do you go?

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Background Athletes presenting with 1st-degree atrioventricular block (AVB) on 12-lead electrocardiogram (ECG) may present a diagnostic conundrum, especially when significantly prolonged and associated with higher degrees of block. A pragmatic stepwise approach to the evaluation of these patients is, therefore, crucial.

Case summary A 19-year-old waterpolo player was referred for assessment of a 1st-degree heart block and one isolated episode of syncope. All other cardiac investigations were within normal limits except for a 24-h ambulatory ECG which showed Mobitz 1 AVB and episodes of 2:1 block occurring in the context of Wenchebach. An electrophysiological study (EPS) was performed which effectively excluded infranodal conductive tissue disease, confirming physiological intranodal block.

Discussion The increase in vagal tone is one of the physiological adaptations to an increased demand in cardiac output in athletes, which explains the presence of 1st-degree AVB in up to 7.5% of athletes. The presence of 2:1 AVB on 24 h ECG raises doubts whether the 1st-degree AVB on resting ECG is pathological or physiological, especially considering this particular patient had suffered an episode of syncope. When this diagnostic uncertainty persists despite non-invasive investigations, including cardiopulmonary exercise testing, invasive EPS may be required to assess the refractoriness of the AV node and at what level within the cardiac conductive system block occurs. The electrophysiological study can effectively rule out infranodal disease by confirming physiological intranodal block using incremental atrial pacing.

Keywords Case report • Athlete • Athlete's heart • Atrioventricular block • Physiological • Cardiomyopathy • Electrophysiological studies

ESC Curriculum 8.1 Sports Cardiology • 8.5 Primary prevention • 2.3 Cardiac magnetic resonance • 5.2 Transient loss of consciousness • 6.5 Cardiomyopathy

Learning points

- The evaluation of profound 1st-degree atrioventricular block (AVB) in athletes requires a stepwise pragmatic approach.
- Invasive assessment with an electrophysiological study to discern between physiological and pathological AVB may be necessary in athletes in the grey zone.

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Introduction

First-degree atrioventricular block (AVB) is a common training-related change, found in up to 7.5% of athletes on a resting electrocardiogram (ECG).[1] The electrophysiological study (EPS) assessment of the atrioventricular (AV) node in endurance athletes favours intrinsic adaptation, independent of vagal tone.[2] Downregulation of the funny channel (HCN4) is one of the most common causes of physiological bradycardia in athletic individuals, leading to a corresponding drop in funny current, an important pacemaker mechanism.[3,4] Nevertheless, further evaluation is warranted in the presence of arrhythmic symptoms, a broad QRS, an abnormal axis, an inappropriate sinus rate response during exercise.[1] The presence of high-grade AVB on prolonged ambulatory ECG monitoring should also raise the suspicion of cardiac pathology. We will be presenting a case of profound AVB in an endurance competitive athlete suffering from syncope.

Timeline

Timeline	Outcome
1 year prior to referral	Syncope during competition. Normal blood work and no arrhythmias during admission at the time. Isolated profound 1st-degree atrioventricular block (AVB) on electrocardiogram (ECG) was the only positive finding.
Referred for assessment	ECG again showed profound 1st-degree AVB. Echocardiography showed borderline left ventricle (LV)/right ventricle (RV) dimensions with low/normal systolic function. Holter showed Mobitz I AVB with periods of 2:1 AV block.
6 weeks	Cardiac magnetic resonance imaging again showed LV/RV volumes to be at the upper limits of normal with low/normal systolic function.
8 weeks	Cardiopulmonary exercise testing demonstrated good AV conduction during exercise with excellent functional capacity.
3 months	Electrophysiological study suggestive of intranodal physiological adaptation. Reassured. Cleared for competitive sport.

Case presentation

A 19-year-old male Caucasian competitive waterpolo player, engaging in 20 h of moderate- to high-intensity physical activity on a weekly basis was referred for cardiovascular assessment follow an isolated episode of syncope 1 year previously during competition. This had occurred soon after the patient came out of the pool at the end of the race. It was preceded with lightheadedness and nausea, lasting ~30 s. He was fully oriented and recovered spontaneously a few minutes later. He was referred to hospital and all investigations including blood tests, telemetry, and a computerized tomography pulmonary angiogram were normal. Physical examination was largely unremarkable, with sinus bradycardia (50 b.p.m.), normal blood pressure, and no added heart sounds. He was never symptomatic before that point. There was no family history of sudden cardiac death. He was not on any regular medications. The patient was eventually referred to the Sports Cardiology Clinic months later after having discussed this with his family doctor during his routine yearly screening appointment. The presence of symptoms in the context AVB prompted referral.

A 12-lead ECG revealed sinus bradycardia with profound 1st-degree AVB (*Figure 1*). The PR interval was measured as 365 ms. The QRS satisfied voltage criteria for left ventricular (LV) hypertrophy. QRS duration and axis were normal, with no evidence of QRS fragmentation. A transthoracic echocardiogram was consistent with an athletic heart. The LV and right ventricular (RV) volumes were at the upper limits of normal. The systolic function of both ventricles was also towards the lower limits of normal, with an LV and RV ejection fraction of 53% and 52%, respectively. Diastolic parameters were normal with the left atrium mildly dilated. Diastolic parameters 24-h ambulatory ECG monitoring revealed prolonged periods of sinus bradycardia, 1st-degree AVB and intermittent 2nd-degree Mobitz I (Wenckebach) AVB (*Figure 2*). Nocturnal episodes of 2:1 AVB in the context of Wenckebach were also recorded (*Figure 3*). He did not report any symptoms during ambulatory ECG monitoring.

Cardiopulmonary exercise testing (CPET) was normal with a VO_{2MAX} of 42.5 mL/kg/min (93% of predicted). Ventilatory efficiency was normal (VE/VCO2 28.4). There was a normal blood pressure response. No arrhythmias were recorded. Atrioventricular conduction was also normal throughout the test, with PQ prolongation again seen in recovery. There was no objective evidence of cardiac or ventilatory limitation to exercise (heart rate recovery at 1 min was 28 b.p.m., O2/Pulse 17.1 mL/beat which was 104% of predicted). Cardiac magnetic resonance (CMR) imaging was also performed because of possible arrhythmic symptoms in the context of conduction abnormalities and ventricular function towards the lower limits of normal. This revealed a low–normal LV and RV ejection fraction (LVEF 52%, RVEF 51%), with normal chamber dimensions. No macroscopic fibrosis was present on late enhancement sequences. Family screening of both his parents and his two siblings was negative. The pros and cons of genetic testing were also discussed. In the absence of a definite clinical phenotype, the team opted against referral.

All these secondary investigations failed to confirm the presence of a definite cardiac phenotype. The presence of symptoms increased the suspicion of conduction disease in the context of early dilated cardiomyopathy. He was referred for an EPS for better risk stratification and phenotypic characterization, a pre-requisite prior to giving full clearance for competitive sports. Baseline sinus node recovery time was measured at 231 ms, baseline AH interval at 307 ms and an HV interval measured at 38 ms. Atrioventricular Wenckebach was achieved by pacing the atrium at 740 ms (81 b.p.m.). This improved to 320 ms (188 b.p.m.) following the administration of isoprenaline.

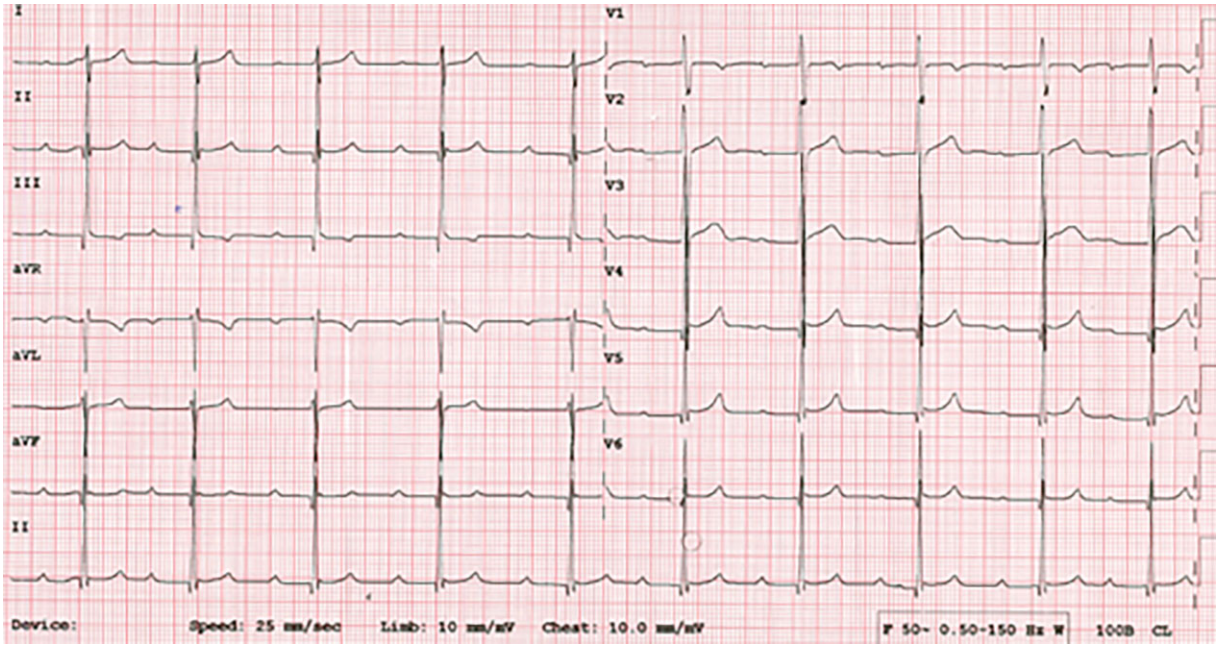


Figure 1 Resting electrocardiogram showing 1st-degree atrioventricular block (PR interval 365 ms).

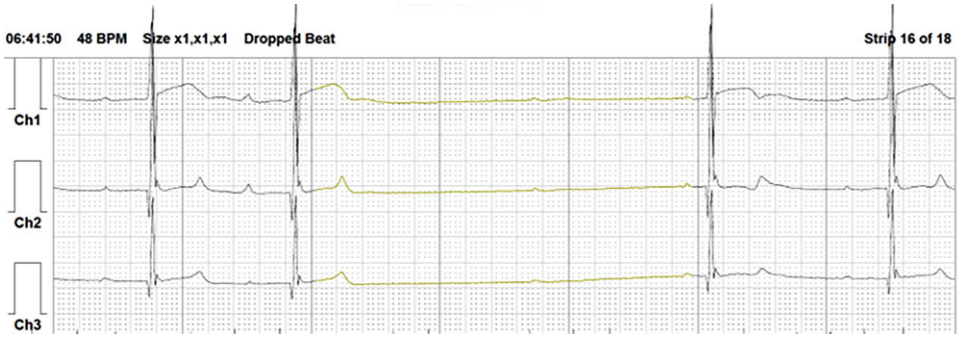


Figure 2 Holter monitoring depicting Mobitz Type 1 atrioventricular block.

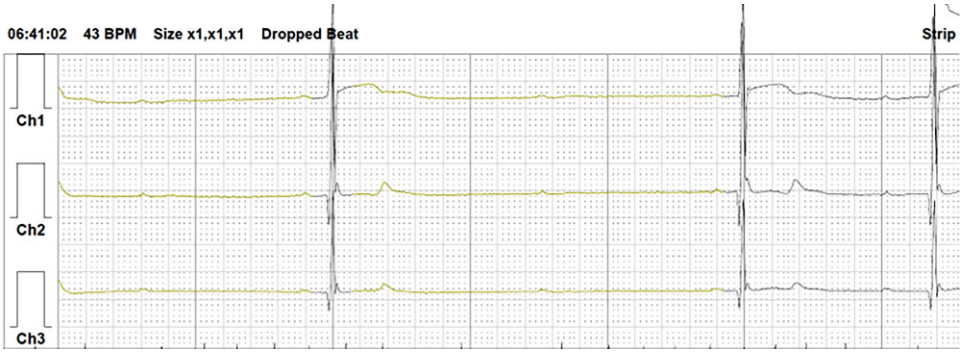


Figure 3 Holter monitoring showing 2:1 atrioventricular block.

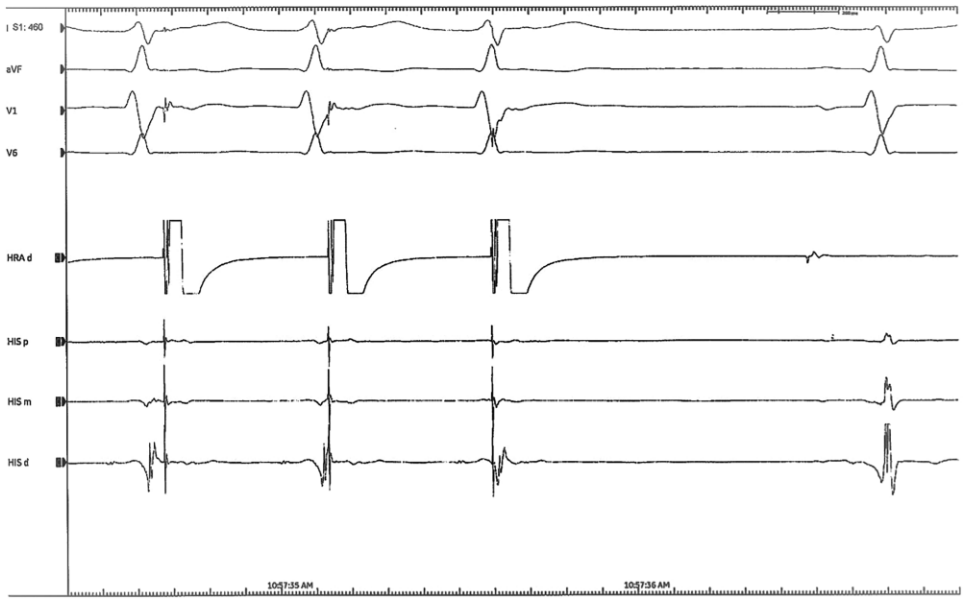


Figure 4 Incremental atrial pacing with atrioventricular block without a His signal, consistent with intranodal block.

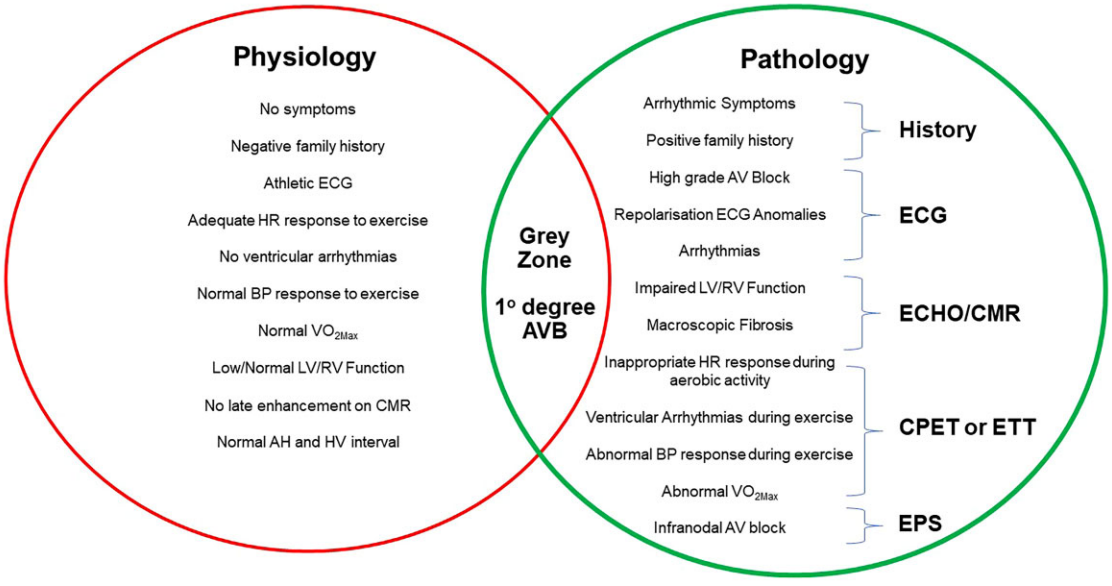


Figure 5 Useful characteristics to differentiate physiology from pathology in athletes with profound 1st-degree atrioventricular block. AV, atrioventricular; AVB, atrioventricular block; BP, blood pressure; CMR, cardiac magnetic resonance; ECG, electrocardiogram; EPS, electrophysiological study; LV, left ventricle; HR, heart rate; RV, right ventricle.

AH interval shortened to 88 ms. No His signal was recorded when in AVB, confirming the presence of intranodal physiological AVB (*Figure 4*). Atrioventricular block improved with rapid atrial pacing and after administering isoprenaline infusion, effectively ruling out infranodal block. He was reassured and was advised to undergo biannual surveillance with echocardiography and ambulatory ECG monitoring. His initial symptoms were attributed to vasovagal syncope in the context of overtraining. This tallied with the acute increase in training volume in preparation for the competition at the time. Since his evaluation, he has not had any recurrence of his symptoms and is training normally. Based on this comprehensive evaluation, no medical treatment was necessary so far.

Discussion

Individuals who regularly engage in at least 4 h of physical activity per week undergo structural, functional, and electrical adaptations in the heart collectively known as the athlete's heart.[5] Increased vagal tone and enlarged cardiac chambers help accommodate the increased demand in cardiac output.[6,7] The type and degree of cardiac adaptation is influenced by age, gender, ethnicity, and sporting discipline. Extreme athletic adaptation may at times overlap with cardiac pathology, traditionally known as the grey zone.[8]

First-degree AVB is a typical training-related change, found in up to 7.5% of athletes on a resting ECG.[1] Electrophysiological study assessment of the AV node in endurance athletes favours intrinsic adaptation, independent of vagal tone.[2] Biomechanical and mechanical effects induced by dilatation and hypertrophy have been proposed as possible mechanisms. Further evaluation is normally only warranted in the presence of arrhythmic symptoms, a broad QRS, an abnormal axis, an inappropriate sinus rate response during exercise.[5] The presence of high-grade AVB on prolonged ambulatory ECG monitoring should also raise the suspicion of cardiac pathology. Pathological AVB in young athletic individuals may be a manifestation of an inherited disorder (Lamin A/C dilated cardiomyopathy, SCN5A Arrhythmogenic Cardiomyopathy or Brugada Syndrome, Myotonic Dystrophy, PRKAG2 syndrome).[1]

Various factors may, however, help differentiate physiological adaptation from pathology (*Figure 5*). The case presented discusses several important factors in the diagnostic work up of an athlete in the grey zone. Further evaluation should be targeted at phenotypic characterization and risk stratification.[5]

Symptoms and family history are extremely important. Symptoms strongly suggestive of cardiac arrhythmias should be evaluated comprehensively. A relevant family history of premature conduction disease, cardiomyopathy or sudden cardiac death should also raise suspicion for a familial disorder. The absence of symptoms and a relevant family history would favour physiological remodelling.

A resting 12-lead ECG is undoubtedly very important in the diagnostic work up. The absence of pathological ECG patterns including high-grade AVB, a wide QRS, repolarization anomalies (pathological T wave inversion, Q waves, ST-segment depression, QT prolongation) strongly favours athletic adaptation.[1] The presence of a pathological ECG pattern should always prompt referral for a comprehensive diagnostic work up, especially in symptomatic patients. Prolonged ambulatory ECG monitoring may help record high-grade AVB and/or malignant ventricular arrhythmias throughout the day. The presence of either one will again favour cardiac pathology.

Echocardiography is regarded as the 1st line imaging modality in ruling out significant structural heart disease. The presence of dilated and/or hypocontractile chambers may raise suspicion for a cardiomyopathic process. The absence of structural heart disease would undoubtedly favour athletic adaptation. The presence of chamber dilatation, systolic dysfunction, and/or fibrosis on CMR will all help ascertain the likelihood of pathology in athletes presenting in the grey zone.[9,10] Using athlete-specific reference ranges may also help decrease the false positive rates, potentially resulting in a misdiagnosis in an otherwise young healthy athletic individual.[11]

As discussed previously, normal AV conduction during exercise favours physiological adaptation. An inappropriate sinus rate response is traditionally present in athletes who present with high-grade AVB.[1] A cardiopulmonary exercise assessment may give more information on cardiorespiratory fitness and evidence of cardiac limitation to exercise. Normal VO_{2MAX} and a normal physiological response will strongly favour athletic adaptation. Standard exercise tolerance testing is a reasonable alternative when CPET is not routinely available.

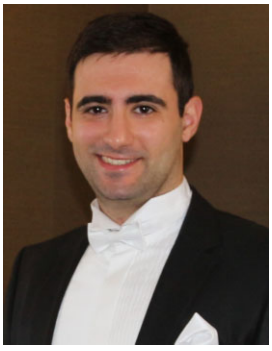
Current guidelines also encourage referral for invasive EPS when diagnostic uncertainty persists despite non-invasive evaluation.[12] This may help differentiate intranodal from infranodal AVB.

PR interval in the case presented was within normal ranges for an athletic individual.[1] The presence of syncope prompted further evaluation as per current guidelines,[12] using a systematic stepwise approach.[13] 2:1 AVB raised the suspicion for significant conduction disease. The presence of low/normal systolic function of both ventricles also leads to a cardiomyopathy suspicion. Both these may raise the suspicion of an early Lamin A/C cardiomyopathy phenotype.

Several factors elicited during the evaluation were, however, reassuring. Aquatic athletes often show significant cardiac remodelling in response to the high physiological demand. Normal sinus rate response during exercise, normal cardiorespiratory fitness, absence of ventricular arrhythmias on ambulatory monitoring, and no fibrosis on CMR all favoured athletic adaptation. Negative family screening also helped rule out a familial inherited disorder.

The presence of syncope in the context of 2:1 AVB may traditionally have been a reasonable indication for permanently pacing this individual. Profound electrical remodelling in athletes may, however, occasionally overlap with cardiac pathology, hence why a comprehensive secondary evaluation is warranted in borderline cases. He would have been falsely mislabelled with an inexistent cardiac phenotype, itself carrying important lifelong implications (sporting career, family planning, life insurance policy). Implanting a pacemaker in an aquatic athlete would undoubtedly have also led to a higher incidence of lead fracture due to repetitive ipsilateral arm movements. Now more than ever, the latest European Society of Cardiology sports cardiology guidelines encourages a shared decision-making approach, respecting the athlete's autonomy after providing all the relevant information about the impact of sport and the potential adverse events which may occur.[14] A meticulous comprehensive assessment in a tertiary centre is strongly advised in such difficult cases. Electrophysiological studies may help rule out cardiac pathology in athletes presenting with profound athletic remodelling.

Lead author biography



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Supplementary material

Supplementary material is available at *European Heart Journal—Case Reports* online.

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Slide sets: A fully edited slide set detailing these cases and suitable for local presentation is available online as Supplementary data.

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

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