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CASE REPORT

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Early repolarization electrocardiography pattern with unexplained syncope during training in a young black African non-elite athlete: An accidental finding?

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Abstract

Until recently it was generally thought that early repolarization is benign. But a recent article in the NEJM (Haissaguerre et al.) suggests that some persons with early repolarization may be at risk of life-threatening ventricular tachyarrhythmia. Unexplained syncope or sudden death occurs mostly during sleep. However, some cases of cardiac arrest during exertion have been reported.

We report the case of a 39 year-old black African male with early repolarization pattern on electrocardiogram who regularly experienced dizziness (and one episode of transient loss of consciousness) exclusively while exercising. Detailed examination was normal. Under quinidine therapy, he experienced no further episodes. Increasingly reported cases of cardiac arrest in Africans, and significant prevalence of early repolarization in this population, have to be taken into account since the Haissaguerre et al. report. Further evidence of the lethal consequences of this syndrome are needed, bearing in mind that diagnostic tools for life-threatening arrhythmias are often scarce in sub-Saharan Africa. (Cardiol J 2009; 16, 3: 259–263)

Key words: early repolarization syndrome, ventricular arrhythmias, syncope

Introduction

Transient loss of consciousness (TLOC) is a common disease with a cumulative lifetime incidence of 35% [1]. TLOC is usually due to either seizure or syncope. Syncope during exertion is often serious and of cardiac origins, while syncope immediately after stopping exercise is generally neurally-mediated (reflex syncope) and benign [2].

Known cardiac causes of syncope are structural and non-structural. Potential structural cardiac causes of exercise-related syncope are coronary

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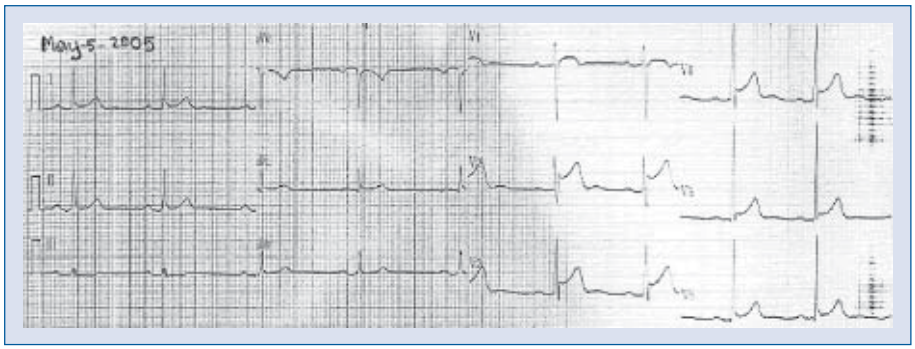

Figure 1. One year before loss of consciousness, ECG displayed prominent J point in anterolateral (V2–V6, I, aVL) and inferior (II, aVF) leads. An elevation of J-STT junction (the so-called Osborn wave) of at least 0.1 mV from baseline manifested as QRS slurring (I, II, aVL, aVF, V6) or notching (V2–V5) with upward concavity. Early R-wave progression in precordial leads with increased voltage in V4–V6 than standard leads. QTc interval is relatively short: 390 ms.

disease, aortic stenosis (AS), hypertrophic cardiomyopathy (HCM) and arrhythmogenic right ventricular dysplasia/cardiomyopathy (ARVD/C). Electrical heart diseases such as long QT syndrome (LQTS), short QT syndrome (SQTS), catecholaminergic polymorphic ventricular tachycardia (CPVT) are common as causes of non-structural exercise syncope. When these pathologies are excluded as potential causes of unexplained syncope or cardiac arrest during exertion, early repolarization pattern in electrocardiogram (ECG) should be taken into account in light of the Haissaguerre et al. report [3]. We report an unusual feature of exertion syncope with early repolarization variant (ERPV).

Case report

A 39 year-old black African male lost consciousness while he was playing football. Eyewitnesses (other players) reported a true absence of consciousness with eyeball revulsion and tongue biting. He spontaneously recovered consciousness after a short time. The eyewitnesses reported no post-event disorientation. The patient remembered that before he lost consciousness he experienced dizziness about 15 minutes into the game and was forced to bend over, as had happened during previous episodes. In the three months before syncope he regularly experienced sudden weakness and dizziness during soccer games. He never had such symptoms at rest and there was no history of sudden death in first degree relatives. He regularly played soccer one hour a week. When he was aged 29, for recruitment purposes he was subject to an ECG showing early repolarization in apicolateral (V3–V6, I, aVL) and inferior (II, aVF) leads; mimicking acute ischemic disease. One year before the cardiac arrest, a systematic check-up with ECG confirmed marked ERPV (Fig. 1). After recovering consciousness, the patient underwent detailed cardiac and neurological evaluation: clinical examination including carotid sinus massage, electroencephalogram, ambulatory ECG recording, brain magnetic resonance (MRI) and coronary angiography were within normal limits. The ECG displayed ERPV. Two-dimensional echocardiography suspected apical left ventricular non-compaction, but cardiac MRI excluded this diagnosis, as well as ARVD. AS and HCM were also excluded. Ventricular ectopic beats did not appear during exercise stress testing, which induced normalization of repolarization (Fig. 2). The tilt-test was not performed and the patient gave up playing soccer. Almost two years later, after publication of Haissaguerre et al. [3] we again contacted the patient and suggested that we study him for arrhythmia vulnerability by programmed ventricular stimulation. Unfortunately he refused. He agreed to take quinidine hydrochloride after counselling. ECG before treatment showed a prominent J point with upward concavity ST segment elevation in apicolateral (V3–V6, I, aVL) and inferior (II, aVF) leads (Fig. 3). Quinidine (300 mg twice daily) consistently reduced the early repolarization pattern after two weeks (Fig. 3). Under quinidine therapy, the patient has recently started playing soccer again once a week, and had no complaints at three months follow up.

Diagnostic criteria

Electrocardiographic features of ERPV have been recently described [4, 5]. Although these features are quite diverse, they have one factor in

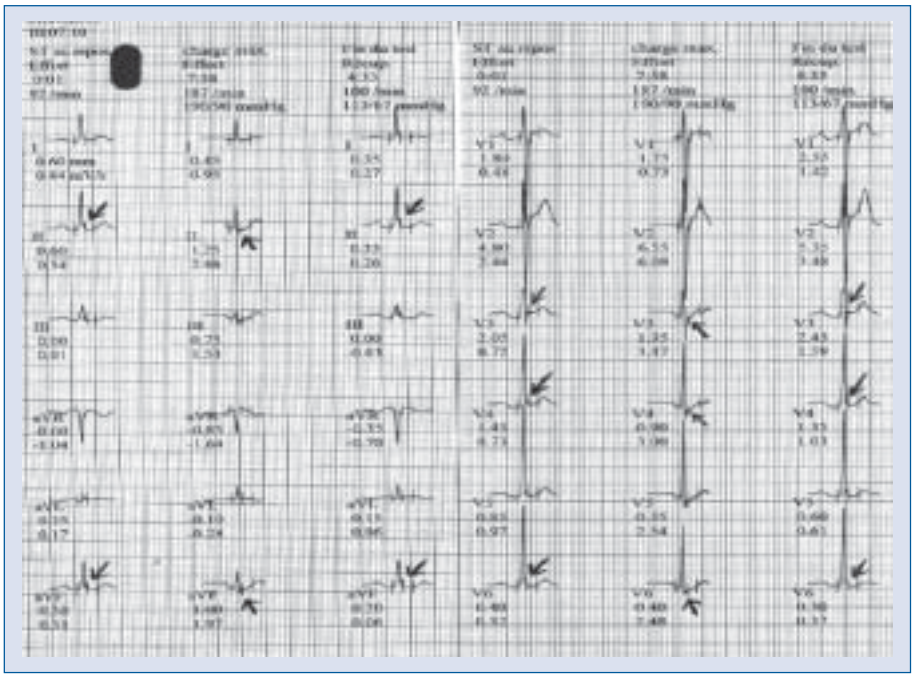

Figure 2. Exercise stress testing (Bruce protocol). The prominent J point with slurring (notching in V3) descending leg of the QRS complex (I, II, V4–V6) at the rest (0:01, HR = 92/min) disappeared during effort (7:58, HR = 187/min). Arrows point out where the slur (notch) appears before the test, during effort and the recovered period; HR — heart rate.

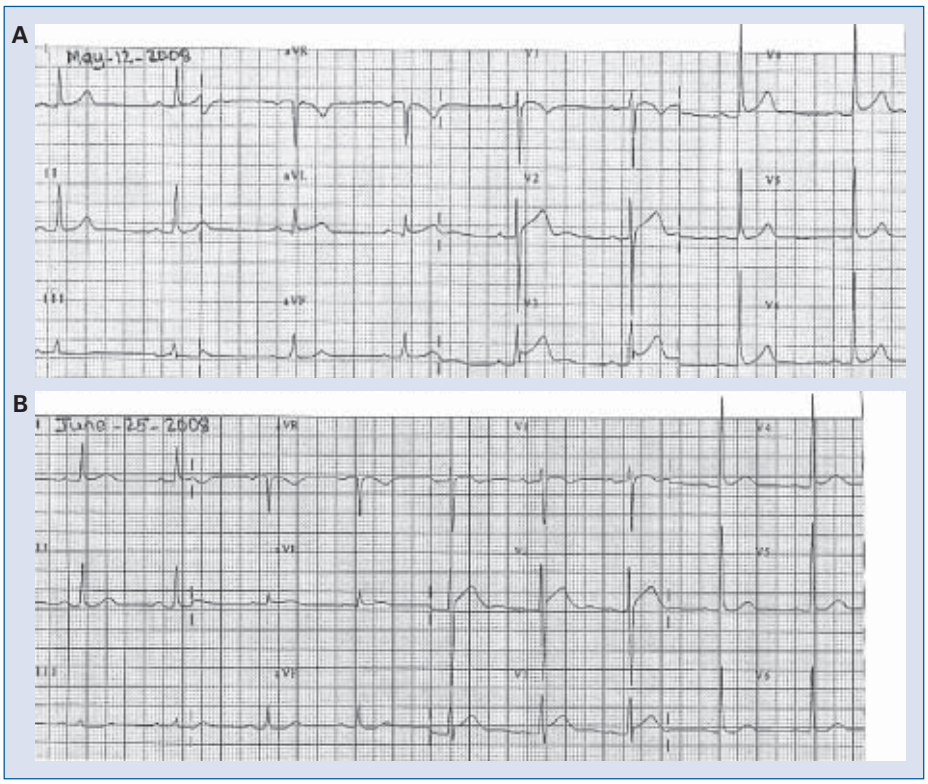

Figure 3. A. Before quinidine treatment — ECG shows early repolarization as QRS slurring in inferolateral (I, II, aVL, V4–V6) and prominent J point followed by notching ST elevation in V3. Lead V4 displays maximal R wave voltage. Intrinsicoid deflection is rapid in leads displaying early repolarization; **B.** Under quinidine therapy — after two weeks (300 mg twice daily): prominent J wave, slurring J-STT elevation and ST elevation with upward concavity were significantly reduced in leads V4–V6 and II. 'Western saddle' pattern was concealed in lead V3.

common: slurring or notching that produces a positive hump in QRS-ST junction: a prominent J wave resembling an Osborn wave. This aspect is found at the junction of the end of the QRS complex and the beginning of the ST segment. The lead with prominent QRS-ST junction of at least 0.1 mV commonly shows ST segment elevation displaying upward concavity. QRS voltage is increased in concerned leads, mostly in V4 and V5. An asymmetric QRS complex with slurring and a reduced slope angle of the ascending positive R wave, an extremely rapid intrinsicoid deflection of descending part of QRS wave and a prominent J wave are common signs. The T wave can be inverted or biphasic. Dynamic changes may occur in the width and height of the wave. The J wave may show circadian changes which may not always be present, and usually disappear during exercise. The common location of early repolarization pattern are leads I, II, III, VF, VL, V4 to V6 but right precordial leads (V2, V3) may also show abnormalities [4]. The ERPV which includes the above mentioned ECG features and a history of unexplained syncope or cardiac arrest due to ventricular fibrillation (VF) seems to have the following properties: the magnitude of J-point elevation ≥ 1.5 mm, the QRS width and the QT interval both may be shorter than normal. Quinidine use, exercise testing, or the infusion of isoproterenol consistently reduces or eliminates early repolarization.

Discussion

Syncope while exercising, but not on cessation of exercise, is very often cardiogenic and three mechanisms are often involved: mechanical obstruction when AS or obstructive HCM is present; tachyarrhythmias due to coronary disease, HCM, ARVD, electrical heart diseases such as LQTS, SQTS, CPVT and bradyarrhythmias due to paroxysmal heart block. Some authors report anecdotal data of syncope and sudden cardiac death with prominent J-waves [6–11]. These isolated case reports have mentioned such changes in the QRS-ST junction in men with idiopathic VF. However, the lack of information from large multicenter studies has remained a drawback. Haissaguerre et al. [3] were the first to demonstrate with high accuracy the possible link between early repolarization pattern and unexplained syncope or sudden cardiac death due to VF. In their work, the link between this ECG pattern and malignant arrhythmias has been supported by both the accentuation of early repolarization (increase of a prominent J point) before the onset of arrhythmia, and the origin of triggering beats from the region of early repolarization. Syncope or cardiac arrest occurred mostly at rest, but some patients experienced it during exertion. Quinidine diminished the ECG pattern and eliminated the recurrence of arrhythmia in 100% (4/4) of patients [3].

In fact, quinidine has been shown to restore transmural electrical homogeneity and abort arrhythmic activity in this condition. Gussak and Antzelevitch [12] and Yan and Antzelevitch [13] have demonstrated the role of quinidine in restoring transmural dispersion of repolarization by reducing outward potassium Ito current, leading to normalize the J point (the so-called Osborn wave). Syncope during exertion is often serious, and of cardiac origins, while syncope immediately after stopping exercise is generally benign and of vasovagal (common faint) or situational causes. Although we have not proved the arrhythmia susceptibility of this patient, many of the facts could point to it: clinical event while exercising, normal carotid sinus massage, exclusion of other causes of exercise-related syncope as AS, obstructive HCM, coronary disease, ARVD, LQTS, SQTS and CPVT. However, we assumed that tilt-testing would definitively help to exclude neurally-mediated syncope. Unexplained syncope during exertion in this young adult without structural heart disease, negative extra-cardiac evaluation, early repolarization features in the ECG, consistent reduction of early repolarization during exercise stress testing and a possible efficacy on junctional changes of quinidine after three months of follow up are likely features of the syndrome described by Haissaguerre et al. [3]. Moreover, syncope in a patient with the prominent J wave has already been described [6].

Our patient underwent a detailed cardiac, neurological and psychiatric evaluation without any finding apart from early ERPV in ECG (Fig. 1): slurring and notching of J-STT elevation with upward concavity, increased QRS voltage in V4 and V5, asymmetric QRS complex with a reduced slope angle of the ascending R wave, an extremely fast intrinsicoid deflection of descending R wave, a prominent J wave and different morphologic forms and spatial distributions of ST elevation. This pattern of repolarization has been overlooked until recent findings suggesting a possible link between ERPV and idiopathic ventricular arrhythmia were reported [3], leading to the proposition of an electrophysiological (EP) study and quinidine treatment to our patient almost two years later. This case report is very unusual in so far as he combined both features

of cardiac and neutrally-mediated mechanisms. Uncompleted diagnostic evaluation including tilt-testing and EP study look like the situation which is common in sub-Saharan Africa. Cardiac arrest is increasingly reported in black Africans [14] and epidemiological studies have shown that ERPV is more prevalent in dark-skinned persons [15]. For these reasons, in the future, clinicians will be faced with the crucial issue of deciding whether unexplained syncope in a patient with ERPV was an arrhythmic event or not. This decision is even more difficult in sub-Saharan Africa where diagnostic tools for arrhythmic sudden death and syncope such as Holter monitoring, external or internal loop recorders, tilt-testing, EP study and cardiac imaging techniques are limited.

As the prevalence of ECG features of early repolarization is fairly high in the general population without affecting life expectancy for a significant proportion, the challenge is to distinguish both groups, with and without risk markers of sudden cardiac death [15]. Although our understanding of atypical patterns of ECG is steadily improving, we need to document the accuracy of non-invasive tests and signs of junctional changes in inferolateral leads for which ventricular ectopic activity would be likely. Thus, countries with low incomes, insufficient medical facilities and high prevalence of ERPV will have to address the problem of loss of consciousness in subjects having early repolarisation ECG pattern.

Limitations of the study

Although this report did not show the occurrence of ventricular tachyarrhythmia leading to syncope, detailed history taking, physical examination and additional investigations were consistent with Haissaguerre's description of this syndrome. However, we are aware that being event-free for three months of follow-up on quinidine may be random; ECG repolarization changes in transitional chest lead (V3) could be due to electrode placement and generally the pattern of J-STT can also be dynamic, changing in degree from one recording to the next.

Conclusions

This study looks at ERPV and unexplained syncope that could be caused by a self-terminating idiopathic VF and which should alert the attention of clinicians to this entity. Increasingly reported cases of cardiac arrest in Africans and a significant prevalence of ERPV in this population have to be taken into account.

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References

1. Ganzeboom KS, Mairuhu G, Reitsma JB, Linzer M, Wieling W, van Dijk N. Lifetime cumulative incidence of syncope in the general population: a study of 549 Dutch subjects aged 35–60 years. J Cardiovasc Electrophysiol, 2006; 17: 1172–1176.
2. Brignole M, Alboni P, Benditt DG et al. Task force on syncope, European Society of Cardiology. Guidelines on management (diagnosis and treatment) of syncope: Update 2004. Europace, 2004; 6: 467–537.
3. Haïssaguerre M, Derval N, Sacher F et al. Sudden cardiac arrest associated with early repolarization. N Engl J Med, 2008; 358: 2063–2065.
4. Boineau JP. The early repolarization variant: An electrocardiographic enigma with both QRS and J-STT anomalies. J Electrocardiol, 2007; 40: 3 (e1–e10).
5. Riera Pérez RA, Uchida Hiroshi A, Schapachnik E et al. Early repolarization variant: Epidemiological aspects, mechanism, and differential diagnosis. Cardiol J, 2008; 1: 4–16.
6. Bjerregaard P, Gussak I, Kotar SL, Gessler JE, Janosik D. Recurrent syncope in a patient with prominent J-wave. Am Heart J, 1994; 127: 1426–1430.
7. Klasky AL, Oehm R, Cooper RA, Udalstova N, Armstrong MA. The early repolarization normal variant electrocardiogram: Correlates and consequences. Am J Med, 2003; 115: 171–177.
8. Mehta M, Jain AC, Mehta A. Early repolarization. Clin Cardiol, 1999; 22: 59–65.
9. Letsas KP, Efremidis M, Pappas LK et al. Early repolarization syndrome: Is It always benign? Int J Cardiol, 2007; 114: 390––392.
10. Garg A, Finneran W, Feld KF. Familial sudden death associated with terminal QRS abnormality on surface 12-lead electrocardiogram in the index case. J Cardiovasc Electrophysiol, 1998; 9: 642–647.
11. Nam GB, Kim YH, Antzelevitch C. Augmentation of J waves and electrical storms in patients with early repolarization. N Engl J Med, 2008; 358: 2078–2079.
12. Gussak I, Antzelevitch C. Early repolarization syndrome: Clinical characteristics and possible cellular and ionic mechanisms. J Electrocardiol, 2000; 33: 299–309.
13. Yan GX, Antzelevitch C. Cellular basis for the electrocardiographic J wave. Circulation, 1996; 93: 372–379.
14. Bonny A, Tonet J, Fontaine G et al. Brugada syndrome in pure black Africans. J Cardiovasc Electrophysiol, 2008; 19: 421–426.
15. Wellens HJ. Early repolarization revisited. N Engl J Med, 2008; 358: 2063–2065.