

Journal für Kardiologie

Austrian Journal of Cardiology

Österreichische Zeitschrift für Herz-Kreislauserkrankungen

Case report: Shorter-than-normal QT interval and provokable right precordial ST segment elevation in three patients with suspicious arrhythmogenic right ventricular cardiomyopathy

Peters S, Trümmel M, Koehler B
Journal für Kardiologie - Austrian Journal of Cardiology 2011; 18 (9-10), 326-328

Homepage:

www.kup.at/kardiologie

Online-Datenbank
mit Autoren-
und Stichwortsuche



Offizielles
Partnerjournal der ÖKG



Member of the ESC-Editor's Club



Offizielles Organ des
Österreichischen Herzfonds



ACVC
Association for
Acute CardioVascular Care

In Kooperation
mit der ACVC

Indexed in ESCI
part of Web of Science

Indexed in EMBASE

Krause & Pachernegg GmbH • Verlag für Medizin und Wirtschaft • A-3003 Gablitz

P.b.b. 02Z031105M,

Verlagsort: 3003 Gablitz, Linzerstraße 177A/21

Preis: EUR 10,-

Veranstaltungskalender

Hybrid-Veranstaltungen der Herausgeber des **Journals für Kardiologie**

Finden Sie alle laufend aktualisierten Termine
auf einem Blick unter

www.kup.at/images/ads/kongress.pdf

Shorter-than-normal QT interval and provokable right precordial ST segment elevation in three patients with suspicious arrhythmogenic right ventricular cardiomyopathy

S. Peters¹, M. Trümmel², B. Koehler²

¹St. Antonius-Hospital Gronau GmbH, Gronau; ²Cardiology, Klinikum Dorothea Chr. Erxleben gGmbH Quedlinburg, Quedlinburg, Germany

Abstract

According to an American Heart Association Scientific Statement of the contemporary definition and classification of cardiomyopathies, suspicious arrhythmogenic right ventricular cardiomyopathy is demonstrated in 3 cases of typical Brugada syndrome with a shorter-than-normal QTc interval. The history and the findings of electrocardiology, echocardiography, right ventricular angiography and electrophysiology are described in detail.

Introduction

An American Heart Association Scientific Statement of contemporary definition and classification of cardiomyopathies describes arrhythmogenic right ventricular cardiomyopathy, Brugada syndrome and short QT syndrome as classical genetic diseases [1]. Three cases of variants of Brugada syndrome with a shorter-than-normal QTc interval of < 360 ms in males and of < 370 ms in females [2] were associated with suspicious arrhythmogenic right ventricular cardiomyopathy.

Case Reports

Case 1: A 36-year-old male presented as an out-patient because of non-exertional chest pain, no syncope, no history of sudden cardiac death in the family. In the 12-lead ECG (amplitude 10 mV, paper speed 50 mm/s) a prolonged QRS interval in right precordial leads of more than 110 ms and a saddle-back ST-segment elevation in lead V2 with a small epsilon potential was found. QTc interval measured in lead II and lead V5 according to Bazett formula was at mean 340 ms before ajmaline administration (Fig. 1). Echocardiography revealed normal left ventricular function, no valve abnormalities and a right ventricle with normal dimensions, normal global function and contraction abnormalities of the apex and dilation of the right ventricular outflow tract. Coronary artery disease could be excluded by coronary angiography; right ventricular angiography in 30°RAO and 60°LAO projection could confirm echocardiographic findings. Ajmaline challenge disclosed type 1 ST-segment elevation in V1 and V2 with the development of right bundle branch block and negative T

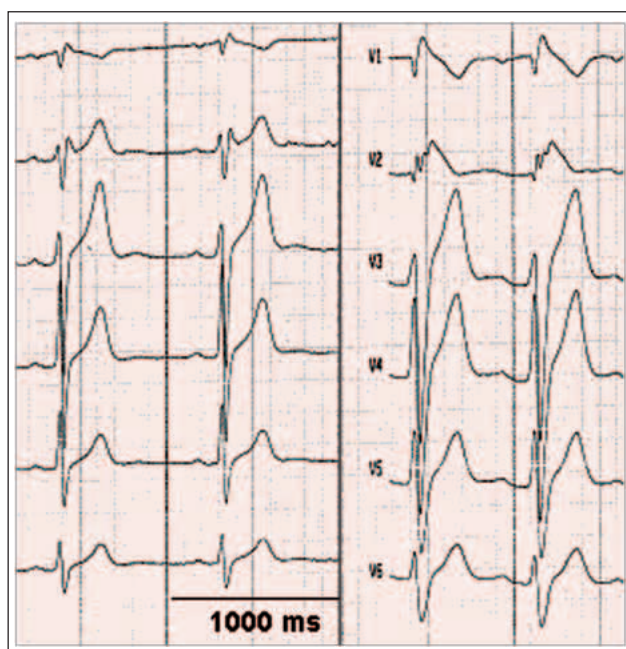


Figure 1: Left: Standard precordial ECG leads V1–V6 (10 mV amplitude, 50 mm/s paper speed) of case no. 1 with prolonged QRS duration in right precordial leads, small epsilon potential in V2 and saddle-back ST-segment elevation in V2. QTc interval 340 ms (NOTICE: horizontal line marking 1000 ms); right: precordial leads of case no. 1 after ajmaline administration with coved-type ST elevation in V1 and V2 and right bundle branch block configuration.

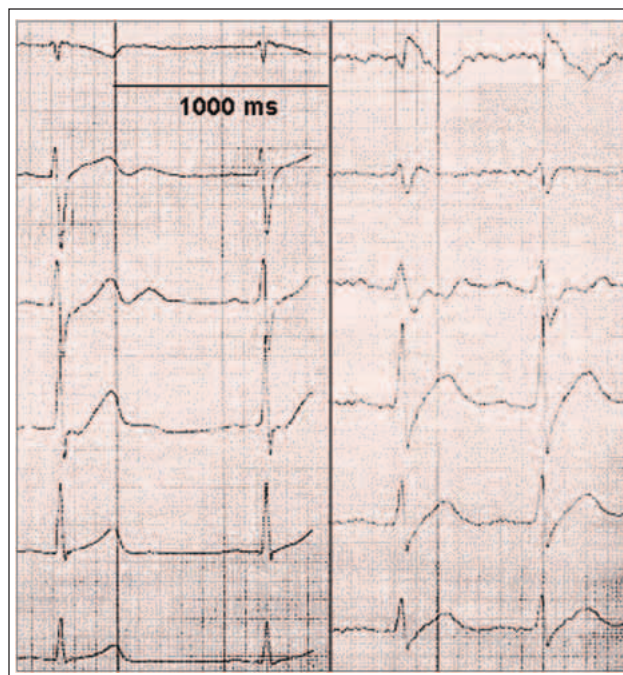


Figure 2: Left: Standard precordial ECG leads V1–V6 (amplitude 10 mV, paper speed 50 mm/s) of case no. 2 with prolonged QRS duration in right precordial leads, small epsilon potential in V1 and V2 and prominent U waves in V2 and V3. QTc 340 ms (NOTICE: horizontal line marking 1000 ms); right: precordial ECG leads of case no. 2 after ajmaline administration with coved-type ST elevation in V1 and right bundle branch block configuration.

waves in V1 (Fig. 1). The patient refused programmed ventricular stimulation and was free of symptoms in a 6 years' follow-up.

Case 2: A 29-year old male with an inborn brain damage and known epileptia in the childhood was admitted to the hospital because of syncope without any medication. There is no history of sudden cardiac death in the family. Standard 12-lead ECG showed a QTc interval measured in lead II and in lead V5 according to Bazett formula of at mean 340 ms before ajmaline administration, prominent U waves in V2 and V3 and right precordial QRS prolongation of 110 ms. Small epsilon potentials were present in V1 and V3 (Fig. 2). Echocardiography revealed a localised apical akinesia of the right ventricle with a prominent moderator band. During the electrophysiological study no sustained ventricular arrhythmias were inducible. Right ventricular angiography confirmed major apical dilatation and akinesia in 30°RAO and 60°LAO projection. Ajmaline challenge demonstrated significant type 1 ST-segment elevation in V1 with typical right bundle branch block and negative T waves in V1 (Fig. 2). Antiepileptic therapy was restarted; syncopes do not appear in a 5 years' follow-up.

Case 3: A 54-year-old female was admitted to hospital because of recurrent syncopes. No history of familial disease. In the 12-lead standard ECG right precordial QRS prolongation of 110 msec with T wave inversions in V1 and V2 and a QTc interval measured in lead II and lead V5 according to Bazett formula of at mean 360 ms before ajmaline administration was striking (Fig. 3). Coronary artery disease could be excluded by coronary angiography. Biplane right ventricular angiography could demonstrate major apical and inferior akinesia. Programmed ventricular stimulation could not induce sustained ventricular arrhythmias. Ajmaline challenge could provoke significant type 1 ST elevation in V1 and V2 with typical right bundle branch block and negative T waves in V1 and V2 (Fig. 3).

Because of recurrent syncopes and the result of the ajmaline challenge with negative T waves in V1 and V2 an ICD was implanted without documentation of ventricular arrhythmias in the holter of the ICD and recurrent holter monitorings despite recurrent syncopes in a 4 years' follow-up. After a lead fracture of the ICD and because a repeated electrophysiological study was again negative, the ICD was explanted.

■ Molecular Genetics

In patient no. 2 a mutation screening of the laboratory of Prof. C. Antzelevitch in Utica, USA, was conducted. A mutation of SCN5A gene (P2006A) was detected. Patient 1 lived in abroad and missed the screening. In patient no. 3 there are up to now no mutations found.

■ Discussion

These 3 cases reported present for the first time the possible coincidence of shorter-than-normal QTc interval and provokable right precordial ST-segment elevation by ajmaline challenge and right ventricular abnormalities suspicious of arrhythmogenic right ventricular cardiomyopathy [3].

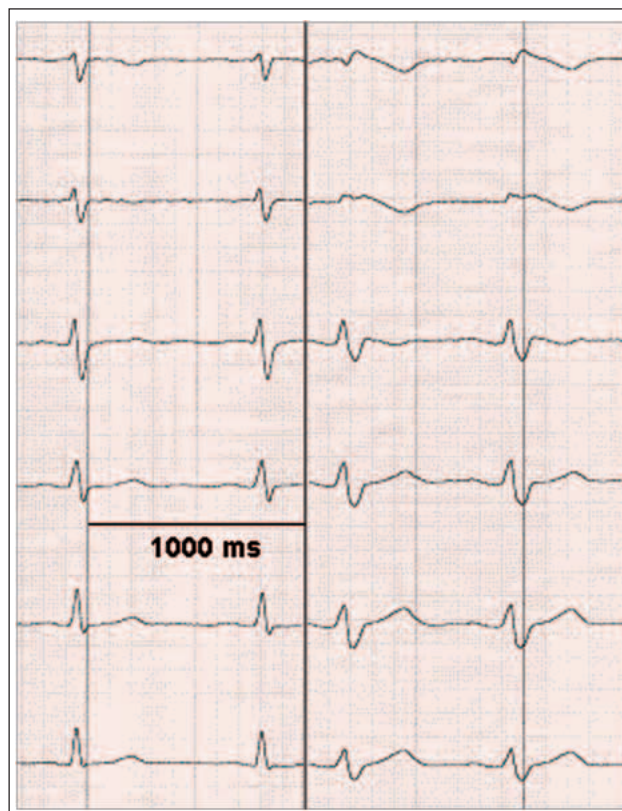


Figure 3: Left: Standard precordial ECG leads (10mV amplitude, paper speed 50 mm/s) of case no. 3 with prolonged QRS duration in right precordial leads and T wave inversions in V1 and V2. QTc interval 360 ms (NOTICE: horizontal line marking 1000 ms); right: precordial leads of case no. 3 after ajmaline administration with significant coved-type ST elevation in V1 and right bundle branch block.

With regard to ECG features the three patients revealed coved-type ST segment elevation after ajmaline administration in two patients in two right precordial leads [4] and in one patient in only one right precordial lead. A similar clinical and arrhythmic profile has been described in typical Brugada patients with type 1 ST-segment elevation in a single right precordial lead [5]. In all three reported cases ajmaline led to typical right bundle branch block. This special ECG feature can be found in a certain percentage of cases [6]. Nevertheless, ECG findings after ajmaline challenge are striking in all three cases with a mixture of depolarisation and repolarization abnormalities discussed in Brugada syndrome [7]. Case no. 3 demonstrated type 1 ST segment elevation and negative T waves in V1 and V2 which is a major risk factor for the development of ventricular fibrillation in typical Brugada patients [8].

Shorter-than-normal QTc interval seen in all three cases is very similar to what has been found in mutations of the alpha1- and beta 2b-subunit of the L-type calcium channel gene in patients with Brugada syndrome [2]. These mutations were tested in two patients and were negative. The difference was that typical ST-segment elevation was only provokable by ajmaline testing.

The finding of a mutation encoding the SCN5A (P2006A) gene in patient 2 is a very rare finding in a case of sudden infant death syndrome [9] and is expressed at the low rate in a general population of less than 0.4%. This polymorphism has

no diagnostic specificity [10]. Recently, P2006A was the only finding in a case of Brugada syndrome and recurrent ventricular fibrillation in case of acute tomb-stone like myocardial infarction (unpublished data Prof. Antzelevitch, Utica, USA).

Right ventricular abnormalities based on echocardiographic and angiographic examinations could be found in all three cases in the so-called “triangle of dysplasia” [11]. Whether these three cases really represent arrhythmogenic right ventricular cardiomyopathy is difficult to decide as all patients only demonstrate with a maximum of one major and one minor diagnostic criteria according to modified Task Force criteria [12] and represent borderline ARVD/C patients. Endomyocardial biopsies were not taken and magnetic resonance imaging was not done. According to right ventricular angiography two patients had major diagnostic criteria with akinesia of the apex and in the other patient akinesia of the inferior and apical region of the modified Task Force criteria.

Two of 3 patients had a history of syncope leading to the question whether ICD implantation was absolutely necessary [13]. In case no. 2 syncope did not appear after initiation of antiepileptic therapy in a five year’s follow-up. In case no. 3 ICD implantation was performed, but a lead fracture made a revision necessary. Multiple holter monitorings showed no ventricular tachycardia and no ventricular fibrillation despite recurrent syncope in a 4 years’ follow-up. A tilt table testing after four years revealed a positive result and electrophysiological testing was again negative. In borderline cases of arrhythmogenic right ventricular cardiomyopathy the value of ICD implantation in cases with syncope is still under debate [14]. In typical Brugada syndrome event rates in asymptomatic patients were low. Inducibility of ventricular tachyarrhythmias and family history of sudden cardiac death were not predictors of cardiac events [15].

The report of these three cases demonstrate a mixture of puzzling electrocardiographic and angio- and echocardiographic findings suggestive of ion channelopathies and cardiomyopathies supporting the attempt of the American Heart Association to define ion channelopathies as primary cardiomyopathies [1].

The dilemma is that all cases do not represent up to now a relevant phenotype of arrhythmogenic right ventricular cardiomyopathy, short QT syndrome and Brugada syndrome. The fact that in all these patients no arrhythmias appeared in a 4 to 6 years’ follow-up is striking; a longer follow-up is needed.

References:

1. Maron BJ, Towbin JA, Thiene G, Antzelevitch C, Corrado D, Arnett D, Moss AJ, Seidman CE, Young JB. Contemporary definitions and classification of the cardiomyopathies: an American Heart Association Scientific Statement from the Council on Clinical Cardiology, Heart Failure and Transplantation Committee; Quality of Care and Outcomes Research and Functional Genomics and Translational Biology Interdisciplinary Working Groups; and Council on Epidemiology and Prevention. *Circulation* 2006; 113: 1807–16.
2. Antzelevitch C, Pollevick GD, Cordeiro JM, et al. Loss-of-function mutations in the cardiac calcium channel underlie a new clinical entity characterized by ST-segment elevation, short QT intervals, and sudden cardiac death. *Circulation* 2007; 115: 442–9.
3. Peters S. Advances in the diagnostic management of arrhythmogenic right ventricular dysplasia-cardiomyopathy. *Int J Cardiol* 2006; 113: 4–11.
4. Antzelevitch C, Brugada P, Borggrefe M, Brugada J, Brugada R, Corrado D, Gussak J, LeMarec H, Nademanee K, Perez Riera AR, Shimizu W, Schulze-Bahr E, Tan H, Wilde A. Brugada syndrome: report of the second consensus conference: endorsed by the Heart Rhythm Society and the European Heart Rhythm Association. *Circulation* 2005; 111: 659–70.
5. Richter S, Sarkozy A, Paparella G, Henkens S, Boussy T, Chierchia GB, Brugada R, Brugada J, Brugada P. Number of electrocardiogram leads displaying the diagnostic coved-type pattern in Brugada syndrome: a diagnostic consensus criterion to be revised. *Eur Heart J* 2010; 31: 1357–64.
6. Antzelevitch C, Brugada P, Borggrefe M, Brugada J, Brugada R, Corrado D, Gussak J, LeMarec H, Nademanee K, Perez Riera AR, Shimizu W, Schulze-Bahr E, Tan H, Wilde A. Brugada syndrome: report of the second consensus conference. *Heart Rhythm* 2005; 2: 429–40.
7. Tukkie R, Sogaard P, Vleugels J, de Groot IK, Wilde AA, Tan HL. Delay in right ventricular activation contributes to Brugada syndrome. *Circulation* 2004; 109: 1272–7.
8. Atarashi H, Ogarawa S. New ECG criteria for high-risk Brugada syndrome. *Circ J* 2003; 67: 8–10.
9. Wang DW, Desai RR, Crotti L, Arnestad M, Insolia R, Pedrazzini M, Ferrandi C, Vege A, Rognum T, Schwartz PJ, George AL Jr. Cardiac sodium channel dysfunction in sudden infant death syndrome. *Circulation* 2007; 115: 368–76.
10. Shinlapawittayatorn K, Du XX, Liu H, Ficker E, Kaufman ES, Deschenes I. A common SCN5A polymorphism modulates the biophysical defects of SCN5A mutations. *Heart Rhythm* 2011; 8: 455–62.
11. Marcus FI, Fontaine GH, Guiraudon G, Frank R, Laurenceau JL, Malergue C, Grosgeat Y. Right ventricular dysplasia: a report of 24 adult cases. *Circulation* 1982; 65: 384–98.
12. Marcus FI, McKenna WJ, Sherrill D, Basso C, Bauce B, Bluemke DA, Calkins H, Corrado D, Cox MG, Daubert JP, Fontaine G, Gear K, Hauer R, Nava A, Picard MH, Protonotarios N, Saffitz JE, Sanborn DM, Steinberg JS, Tandri H, Thiene G, Towbin JA, Tsatsopoulou A, Wichter T, Zareba W. Diagnosis of arrhythmogenic right ventricular cardiomyopathy/dysplasia: proposed modification of the Task Force Criteria. *Eur Heart J* 2010; 31: 806–14.
13. Jung W, Andresen D, Block M, Böcker D, Hohnloser SH, Kuck KH, Sperzel J. Leitlinien zur Implantation von Defibrillatoren. *Clin Res Cardiol* 2006; 95: 696–708.
14. Corrado D, Calkins H, Link MS, Leoni L, Favale S, Bevilacqua M, Basso C, Ward D, Boriani G, Ricci R, Peccini JP, Dalal D, Santini M, Buja G, Iliceto S, Estes NA 3rd, Wichter T, McKenna WJ, Thiene G, Marcus FI. Prophylactic implantable defibrillator in patients with arrhythmogenic right ventricular cardiomyopathy/dysplasia and no prior ventricular fibrillation or sustained ventricular tachycardia. *Circulation* 2010; 122: 1144–52.
15. Probst V, Veltmann C, Eckardt L, Meregalli PG, Gaita F, Tan HL, Babuty D, Sacher F, Giustetto C, Schulze-Bahr E, Borggrefe M, Haissaguerre M, Mabo P, Le Marec H, Wolpert C, Wilde AAM. Long-term prognosis of patients diagnosed with Brugada syndrome: Results from the FINGER Brugada Syndrome Registry. *Circulation* 2010; 121: 635–43.

Correspondence:

Prof. Dr. med. Stefan Peters
St. Antonius-Hospital Gronau GmbH
D-48599 Gronau
Möllenweg 22
e-mail: speters@st-antoniushospital-gronau.de

Mitteilungen aus der Redaktion

Besuchen Sie unsere Rubrik

☒ Medizintechnik-Produkte



Neues CRT-D Implantat
Intica 7 HF-T QP von Biotronik



Artis pheno
Siemens Healthcare Diagnostics GmbH



Philips Azurion:
Innovative Bildgebungslösung

Aspirator 3
Labotect GmbH



InControl 1050
Labotect GmbH

e-Journal-Abo

Beziehen Sie die elektronischen Ausgaben dieser Zeitschrift hier.

Die Lieferung umfasst 4–5 Ausgaben pro Jahr zzgl. allfälliger Sonderhefte.

Unsere e-Journale stehen als PDF-Datei zur Verfügung und sind auf den meisten der marktüblichen e-Book-Readern, Tablets sowie auf iPad funktionsfähig.

☒ Bestellung e-Journal-Abo

Haftungsausschluss

Die in unseren Webseiten publizierten Informationen richten sich **ausschließlich an geprüfte und autorisierte medizinische Berufsgruppen** und entbinden nicht von der ärztlichen Sorgfaltspflicht sowie von einer ausführlichen Patientenaufklärung über therapeutische Optionen und deren Wirkungen bzw. Nebenwirkungen. Die entsprechenden Angaben werden von den Autoren mit der größten Sorgfalt recherchiert und zusammengestellt. Die angegebenen Dosierungen sind im Einzelfall anhand der Fachinformationen zu überprüfen. Weder die Autoren, noch die tragenden Gesellschaften noch der Verlag übernehmen irgendwelche Haftungsansprüche.

Bitte beachten Sie auch diese Seiten:

[Impressum](#)

[Disclaimers & Copyright](#)

[Datenschutzerklärung](#)