Lambda-like ST segment elevation in acute myocardial infarction – a new risk marker for ventricular fibrillation? Three case reports

Uniesienie odcinka ST o kształcie litery lambda w ostrej fazie zawału serca – nowy wskaźnik ryzyka wystąpienia migotania komór? Opis trzech przypadków

Piotr Kukla[1], Marek Jastrzębski[2], Jerzy Sacha[3], Leszek Bryniarski[2]

[1] Department of Internal Medicine, H. Klimontowicz Specialistic Hospital, Gorlice
[2] 1st Departament of Cardiology, *Collegium Medicum*, Jagiellonian University, Kraków
[3] Department of Cardiology, Medical Center, Opole

Abstract

Sudden cardiac death (SCD) is responsible for almost 50% of all cardiac deaths in the U.S. The most common underlying cause of SCD is coronary artery disease, especially acute myocardial infarction (AMI). There are no publications concerning the shape of ST segment elevation in AMI and the risk of ventricular fibrilation (VF) or SCD. We present three cases with AMI and atypical ST segment elevation – 'lambda-wave-like' pattern, complicated with episodes of VF. This ECG pattern resembles the ST segment elevation shape in the type 1C Brugada syndrome. The 'lambda-like' ST segment elevation in AMI may identify patients with increased risk of VF or SCD.

Key words: ventricular fibrillation, ECG, lambda wave, acute myocardial infarction

Kardiol Pol 2008; 66: 873-877

Introduction

Sudden cardiac death (SCD) is responsible for almost 50% of all cardiac deaths in the U.S. [1]. Epidemiological studies have shown that SCD occurs in 0.36 to 1.28 per 1000 persons yearly [2]. The most common cause of SCD is coronary artery disease (CAD) [3].

Recently, Hu et al. showed that some mutations in the gene SCN5A (mutation G400A) could be associated with ventricular fibrillation (VF) and electrical storm during AMI [4]. However, data concerning the shape of ST segment elevation in AMI and the risk of VF or SCD are lacking. It is possible that some patients who suffer from AMI also have genetic abnormalities predisposing to dangerous ventricular arrhythmia, called channelopathies, and that these patients are at particularly high risk of developing VF during acute ischaemia. It may be speculated that the shape of the ST segment elevation or other ECG parameters may be different from that observed in AMI patients without genetic abnormalities.

One of the ST segment abnormalities described in the literature is a lambda-like ST segment pattern. It was first described by Riera et al. [5] and further characterised by Gussak and Bjerregaard in their editorial [6]. Riera et al. described a young man with characteristic ST segment elevation in the inferior (II, III and aVF) and lateral (V6) leads, resembling the Greek letter lambda, called then by Gussak 'action potential-like' shape. This specific ST segment elevation was accompanied by an up-sloping ST segment depression in I, aVL, aVR and V_1-V_5 leads. This patient had a positive SCD family history with 2 victims of SCD occurring at the age of 31 and 39 years. Unfortunately, he died suddenly during Holter ECG monitoring, which showed a short run of polymorphic ventricular tachycardia, rapidly degenerating into fatal asystole. Riera at al. called this phenomenon atypical Brugada syndrome [5], and named the lambda-like ST segment elevation the Gussak wave [7]. Of note, this ECG pattern resembles acute inferior AMI; however, there were no clinical or other signs indicating AMI in the described patient.

Address for correspondence:
Piotr Kukla MD, Klinika Chorób Wewnętrznych, Szpital Specjalistyczny im. H. Klimontowicza, ul. Węgierska 21, 38-300 Gorlice, tel.: +48 18 355 32 00, e-mail: kukla_piotr@poczta.onet.pl

Received: 04 May 2008. **Accepted:** 21 May 2008.

Other interesting data were presented by Yan et al. [8, 9], who proposed the new concept, based on an experimental model, that ST segment elevation in AMI and Brugada syndrome share similar underlying mechanisms. Namely, the loss of the characteristic shape of the 'spike and dome' in the action potential in the epicardium leads to a transmural voltage gradient between the epicardium and the endocardium, which gives rise to a J wave. The main role is played by the Ito current, which is responsible for the prominent notch in the epicardial action potential [10]. The more prominent the notch which occurs between repolarisation phase 1 and phase 2 of the action potential, the greater the predisposition to complete loss of the dome-shape in the epicardium. This in turn causes a transmural voltage gradient throughout the whole of phase 2 of repolarisation, presenting as ST segment elevation on the surface ECG [11].

We present three patients with AMI and lambda-like atypical ST segment elevation, complicated with episodes of VF. In all these patients recurrent polymorphic VT and VF were observed, responding to a high dose of beta blockers but resistant to intravenous amiodarone.

Case 1

This male patient, aged 74 years old, was admitted to our institution because of acute coronary syndrome with ST segment elevation in V_4-V_6, I and aVL. Coronary angiography showed that the infarct-related artery was the first diagonal branch. We observed 7 episodes of VF within four hours. Amiodarone was ineffective, but metoprolol administered in bolus doses i.v. up to 30 mg diminished the arrhythmia and VF did not recur again. The characteristic pattern of ST segment elevation is shown in Figures 1 and 2.

Case 2

A 41-year-old man suffered from 3 episodes of VF. His admission ECG showed ST segment elevation in leads I, aVL and V_2-V_6. Coronary angiography did not reveal any significant coronary artery stenosis. Echocardiography showed left ventricular ejection fraction (LVEF) of 45%, increased LV diastolic diameter of 66 mm, hypo- and akinetic anterior wall and anterior part of the interventricular septum as well as mitral insufficiency grade II/III. Ventriculography showed hypokinesia of the antero-

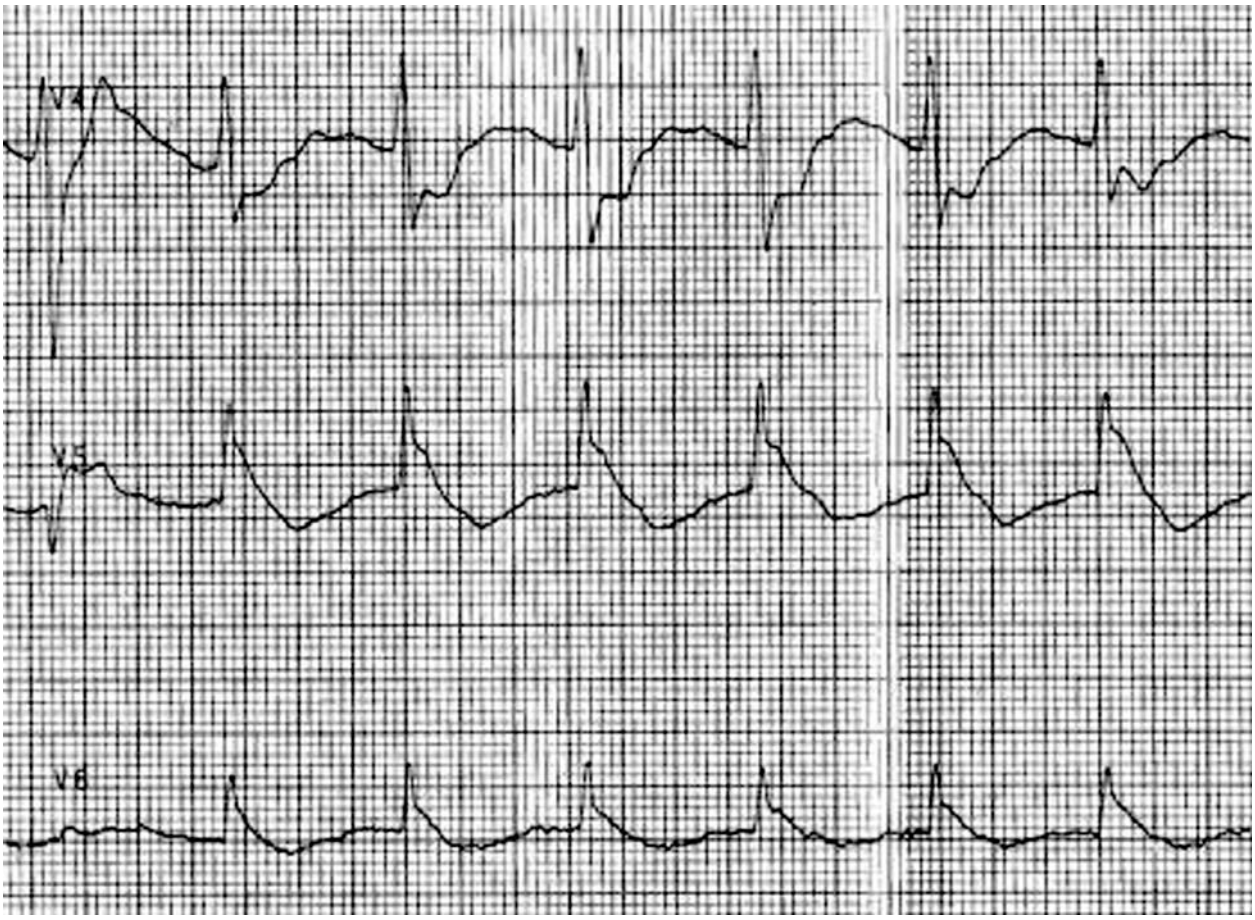

Figure 1. Three left precordial leads with 'lambda-like' ST segment elevation in patient 1

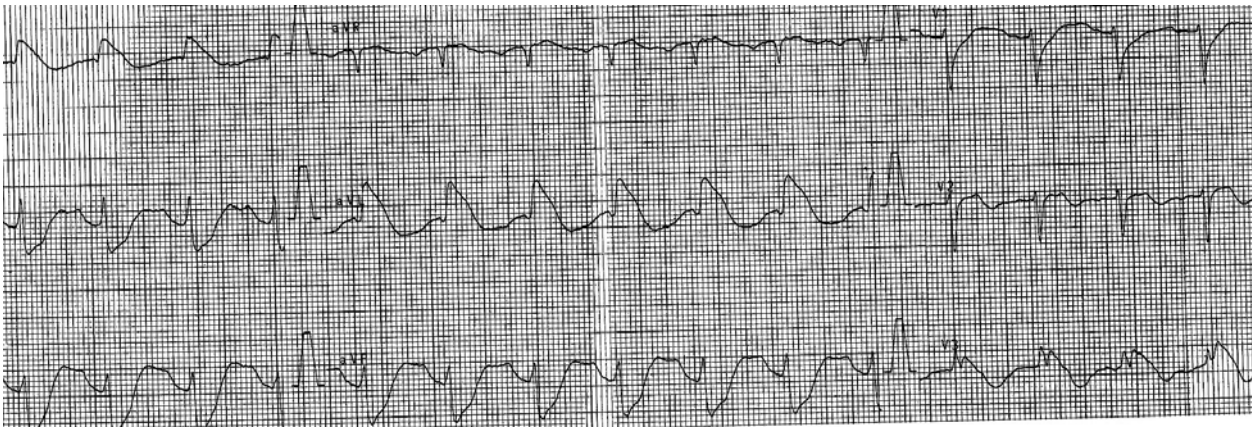

Figure 2. The remaining ECG leads in patient 1

lateral LV and apical segment. The levels of necrotic enzymes were elevated; however, three DC shocks have to be taken into account when interpreting enzyme concentrations. The maximal creatine phosphate (CK) level was 11042 U/l, CK-MB max. was 407 U/l, and troponin I max. was 38.58 ng/ml. The ECG (precordial V_1-V_6 leads) recorded on admission is presented in Figure 3.

Case 3

An 82-year-old man was transferred to our institution from another hospital because of acute coronary syndrome with ST segment elevation, complicated by 4 episodes of VF. Coronary angiography showed subtotal occlusion of the proximal part of the left descending artery. A bare metal stent was implanted. Echocardiography showed LVEF of 33%, with akinesia of the mid and apical segments of the anterior LV wall and anterior part of the interventricular septum. Maximum values of necrosis markers were: CK – 3567, CK-MB – 666 U/l and troponin I >30 ng/ml. The ECG recorded on admission is presented in Figure 4, and the ECG obtained on the next day after PCI in Figure 5.

Discussion

The search for ECG markers of SCD and susceptibility to VF continues. In this report we present three patients with AMI and VF in whom ECG showed a peculiar lambda-like

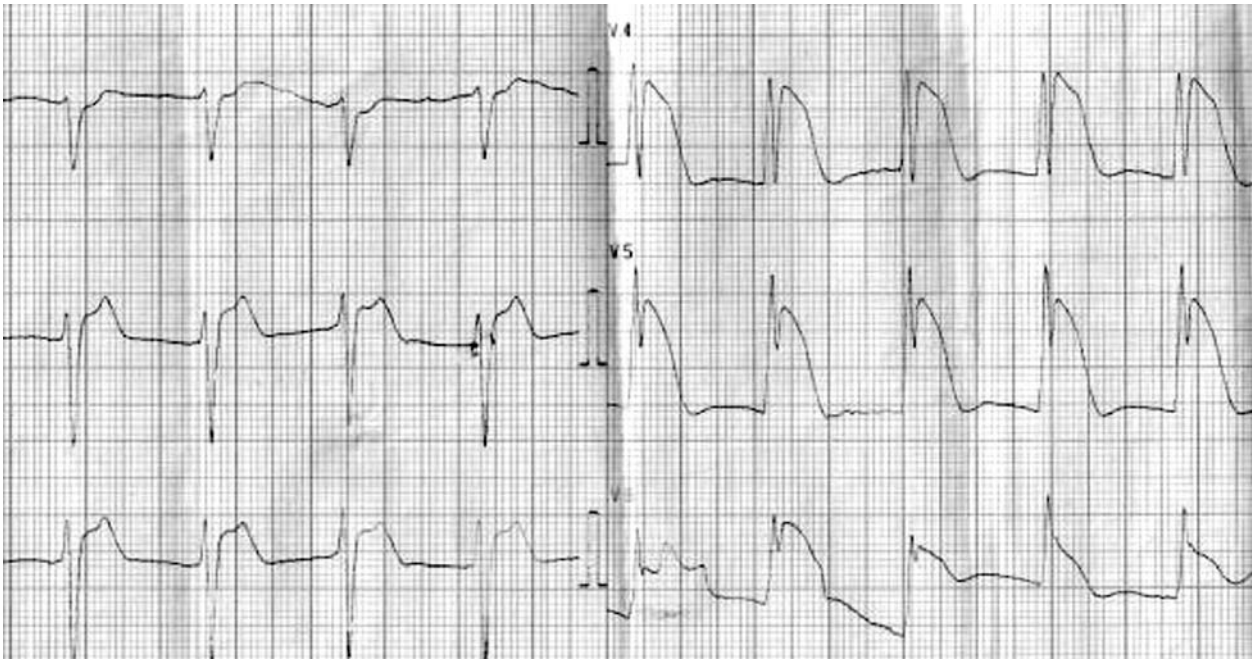

Figure 3. Precordial ECG leds in patient 2

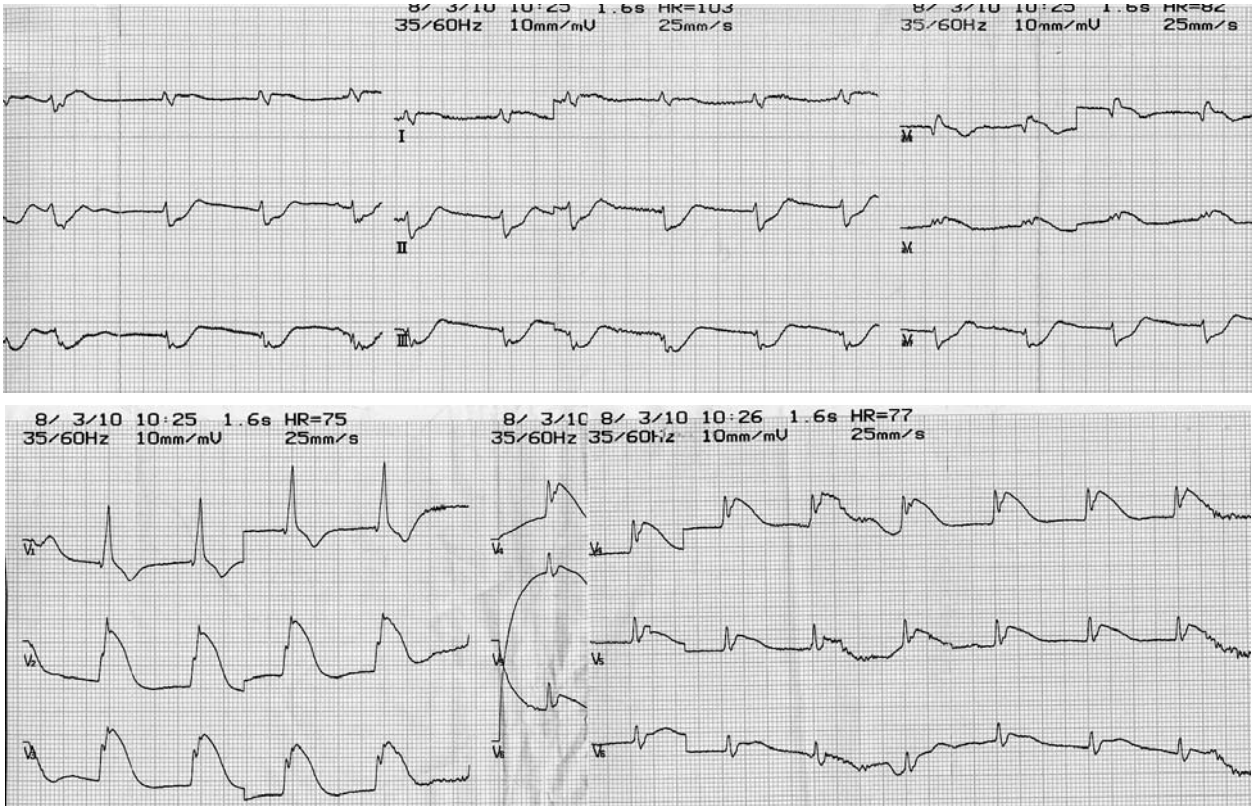

Figure 4. EKG recorded on adminission in patient 3

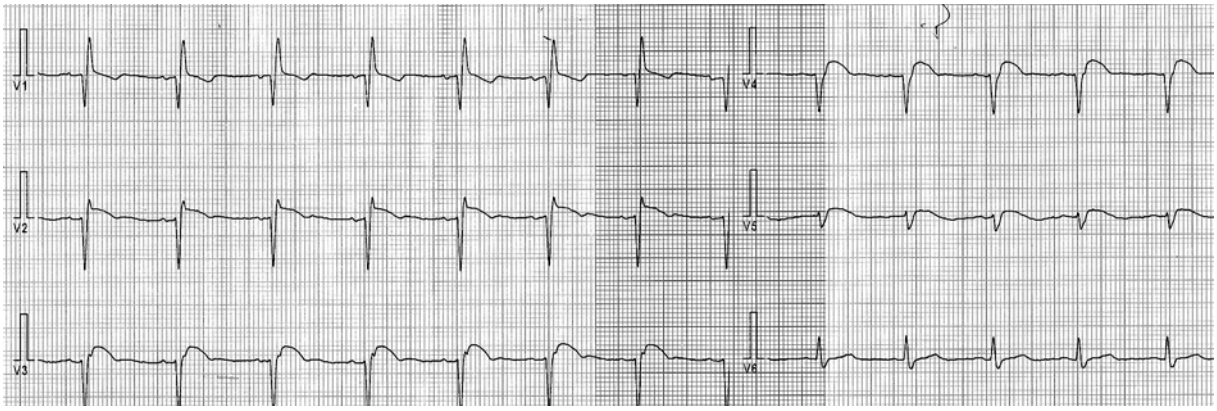

Figure 5. Precordial EKG leads recorded in patient 3 one day ofter admission

ST segment elevation. Further studies, including more patients and genetic assessment, are needed to clarify the possible value of this ECG abnormality in identification of patients at increased risk of VF during acute ischaemia.

This pattern of ST segment elevation was first described by Riera [5] and then named by Gussak 'the Lambda wave' [6]. These ECG peculiarities were associated with sudden cardiac death and cardiac asystole followed by short episodes of ventricular fibrillation.

Recent publications reported some similarities between mechanisms of ST segment elevation and the initiation of VF in AMI and the Brugada syndrome. Because the lambda wave was observed in a patient with VF and SCD, who had atypical Brugada syndrome, one can speculate that a similar ST pattern could be associated with VF in AMI.

Study limitations

No genetic studies were performed in our patients, so the possibility that they had channelopathies remains speculative.

References

1. Huikuri HV, Castellanos A, Myeburg RJ. Sudden death due to cardiac arrhythmias. *N Eng J Med* 2001; 345: 1473-82.
2. Becker LB, Smith DW, Rhodes KV. Incidence of cardiac arrest: a neglected factor in evaluating survival rates. *Ann Emerg Med* 1993; 22: 86-91.
3. Centers for Disease Control and Prevention. State-specific mortality from sudden cardiac death – United States, 1999. *MMWR* 2002; 51: 123-6.
4. Hu D, Viskin S, Oliva A, et al. Novel mutation in the SCN5A gene associated with arrhythmic storm development during acute myocardial infarction. *Heart Rhythm* 2007; 4: 1072-80.
5. Riera AR, Ferreira C, Schapachnik E, et al. Brugada syndrome with atypical ECG: downsloping ST-segment elevation in inferior leads. *J Electrocardiol* 2004; 37: 105-7.
6. Gussak I, Bjerregaard P, Kostis J. Electrocardiographic 'Lambda' wave and primary idiopathic cardiac asystole: a new clinical syndrome? *J Electrocardiol* 2004; 37: 105-7.
7. Riera AR. Proposal of classification of type I Brugada ECG pattern. *Personal communication* 2008.
8. Yan GX, Joshi A, Guo D, et al. Phase 2 reentry as a trigger to initiate ventricular fibrillation during early acute myocardial ischemia. *Circulation* 2004; 110: 1036-41.
9. Yan GX, Antzelevitch C. Cellular basis for the Brugada syndrome and other mechanism of arrythmogenesis associated with ST-segment elevation. *Circulation* 1999; 100: 1660-6.
10. Yan GX, Antzelevitch C. Cellular basis for the electrocardiographic J wave. *Circulation* 1996; 93: 372-9.
11. Shu J, Zhu T, Yang L, et al. ST-segment elevation in the early repolarization syndrome, idiopathic ventricular fibrillation, and the Brugada syndrome: cellular and clinical linkage. *J Electrocardiol* 2005; 38: 26-32.

Electrical storm in acute myocardial infarction

Ihor Gussak, MD, PhD, FACC[1,2], Preben Bjerregaat, MD, DMSc, FACC[3]

[1] New Cardio, Inc, Santa Clara, CA USA

[2] University of Medicine and Dentistry of New Jersey, Robert Wood Johnson Medical School, New Brunswick, NJ USA

[3] Saint Louis University Hospital, St Louis, MO USA

In the USA, the annual rate of acute myocardial infarction (AMI) is close to one million, and almost one quarter of patients with AMI will die suddenly due to the development of fatal ventricular tachyarrhythmias, such as ventricular tachycardia (VT) and ventricular fibrillation (VF) [1]. Although in more than half of such cases sudden cardiac death (SCD) occurs as the first symptom of coronary artery disease [2], conventional cardiovascular risk factors are not predictive of 'coronarogenic' SCD [3]. The search for identification of patients at risk for SCD, including those with AMI, has intensified in recent years. Promising results from experimental and clinical studies have emphasized the pivotal role of family history and subclinical mutation in cardiac channels in the development of repetitive life-threatening arrhythmias during acute ischemia, commonly described as 'electrical storm'.

The term 'electrical storm (ES)' is commonly defined as a state of transient critically impaired electrical stability of the heart culminating in a sequence of life-threatening ventricular tachyarrhythmias (either self-terminating or requiring multiple electrical defibrillations) within a short time (typically during a 24-hour period), although there has been no consistency in the definition of this term. ES is highly resistant to prevention and treatment and is associated with pure clinical outcome, even in patients with implantable cardioverter-defibrillators [4]. Most frequently ES is observed in patients with: (a) primary electrical diseases (PED) of the heart, (b) acute ischemic ('coronarogenic') event, (c) hypothermia during aggressive rewarming, and (d) drug-associated cardiac toxicity. Neither the mechanism/s nor the precipitating factors for ES are well defined, although genetic mutation of cardiac ion channels or gap junctions are considered as a highly likely predisposing factor for ES in acute coronary syndrome (ACS). For instance, Dr. Dan Hu et al. [5] have identified ES in 1 out of 19 consecutive patients who developing VF during AMI. The patient with ES was the only one carrying the SCN5A mutation. Interestingly, this patient developed his first VF at age 70 years and only in the setting of AMI.

In the article by Dr. Piotr Kukla [6] published in this issue of the Journal, the authors present three patients

Address for correspondence:

Ihor Gussak MD, PhD, FACC, One Robert Wood Johnson Place, P.O. Box 19, New Brunswick, NJ USA 08903-0019, e-mail: igussak@hotmail.com

with AMI associated with multiple episodes of polymorphic VT and VF that were observed during a short period of time (ES). These episodes of ES responded to a high dose of beta-blockers but not to intravenous amiodarone. The authors should be congratulated for their meticulous attention to the clinical and ECG aspects of such events in AMI patients and their potential link to a peculiar ECG abnormality, which they call a 'lambda-like wave'. In the author's opinion, this ECG phenomenon deserves special attention as an ECG marker to identify AMI patients with an increased arrhythmogenic risk.

The term 'lambda' wave was introduced in 2004 [7] and was originally described by the authors as (a) having a peculiar ECG pattern of the QRS-ST complex generated by a remarkably gentle slope of its ascending and descending portions followed by negative T waves in the same leads; (b) being associated with primary cardiac asystole; and (c) having non-ischemic origin. By analogy with the 'lambda wave', we believe that the term 'lambda-like' wave, introduced by Dr. Kukla et al. [6], might be an appropriate way to describe a 'triangulated' pattern of ischemic ST elevation that is associated with ES in AMI settings.

Elevation of the ST-segment is the most ominous ECG sign of acute myocardial injury, especially in AMI. It is postulated that the underlying mechanism of ST-segment elevation during acute occlusion of a coronary artery is due to a heterogeneous loss of the action potential dome that also has been shown to give rise to a transmural dispersion of repolarization culminating in phase 2 reentry [8-10]. It appears that the magnitude of ST-segment deviation in AMI is an independent risk factor for primary VF. Furthermore, Dr. Lukas R.C. Dekker et al. [3] have demonstrated that cumulative ST deviation is independent of time to PTCA and of enzyme-estimated infarct size. For each 10-mm increase in cumulative ST deviation, the odds ratio of VF was 1.59 [3].

We believe that it is too early to insist that the primary cardiac channelopathies are responsible for all cases of ES, in general, and the 'lambda-like' ST-segment elevation for ES associated with AMI. However, it is very likely that genetically determined defects in the cardiac electrophysiological matrix could largely and significantly contribute to ischemia-induced electrical instability and fatal arrhythmias. In addition, one should keep in mind that many antiarrhythmic drugs used to prevent and treat ES could be ineffective and even highly proarrhythmic. In such cases, beta blockers might be considered as the most suitable treatment modality.

References

1. Solomon SD, Zelenkofske S, McMurray JJ, et al. Sudden death in patients with myocardial infarction and left ventricular dysfunction, heart failure, or both. *N Engl J Med* 2005; 352: 2581-8.
2. Davies MJ. Anatomic features in victims of sudden coronary death. Coronary artery pathology. *Circulation* 1992; 85: I19-24.
3. Dekker LR, Bezzina CR, Henriques JP, et al. Familial sudden death is an important risk factor for primary ventricular fibrillation: a case-control study in acute myocardial infarction patients. *Circulation* 2006; 114: 1140-5.
4. Credner SC, Klingenheben T, Mauss O, et al. Electrical storm in patients with transvenous implantable cardioverter-defibrillators: incidence, management and prognostic implications. *J Am Coll Cardiol* 1998; 33: 1909-15.
5. Hu D, Viskin S, Oliva A, et al. Novel mutation in the SCN5A gene associated with arrhythmic storm development during acute myocardial infarction. *Heart Rhythm* 2007; 4: 1072-80.
6. Kukla P, Jastrzębski M, Sacha J, Bryniarski L. Lambda-like ST segment elevation in acute myocardial infarction – a new risk marker for ventricular fibrillation? Three case reports. *Kardiol Pol* 2008; 66: 873-7.
7. Gussak I, Bjerregaard P, Kostis J. Electrocardiographic 'lambda' wave and primary idiopathic cardiac asystole: a new clinical syndrome? *J Electrocardiol* 2004; 37: 105-7.
8. Lukas A, Antzelevitch C. Phase 2 reentry as a mechanism of initiation of circus movement reentry in canine epicardium exposed to simulated ischemia. *Cardiovasc Res* 1996; 32: 593-603.
9. Yan GX, Joshi A, Guo D, Hlaing, et al. Phase 2 reentry as a trigger to initiate ventricular fibrillation during early acute myocardial ischemia. *Circulation* 2004; 110: 1036-41.
10. Di Diego JM, Antzelevitch C. Cellular basis for ST-segment changes observed during ischemia. *J Electrocardiol* 2003; 36: 1-5.