Sudden Cardiac Death, RBBB, and Right Precordial ST-Segment Elevation

To the Editor:

We have followed with interest the growing series of patients with the right bundle-branch block (RBBB) and right precordial ST-segment elevation ECG pattern. Beginning in 1992, Brugada and Brugada described 8 patients with this distinctive ECG pattern and a history of aborted sudden death.1 The series was expanded to 47 patients in 1997, including 15 asymptomatic individuals in whom an abnormal ECG was found during routine screening (n=10) or during screening of relatives of an aborted sudden cardiac death victim (n=5).2 The most recent expansion comprises 63 patients, including 22 asymptomatic individuals, 9 of whom were screened for family reasons.3 The incidence of serious ventricular arrhythmias was similar in symptomatic and asymptomatic individuals. Drug treatment proved ineffective, and accordingly, implantation of an automated implantable cardioverter-defibrillator (AICD) was advised as the treatment of choice in all patients identified by means of this ECG, regardless of their history.3

In 1960, the late Professor Dirk Durrer identified a male patient (46 years of age) with a saddle-type ST-segment elevation in leads V1 through V3. This patient, with a negative family history for sudden cardiac death, was followed up for almost 40 years. He never had any complaints. With the exception of an acute anteroseptal myocardial infarction (MI) in 1989, no structural heart disease could ever be demonstrated. The Figure shows some of the ECG recordings from this patient that were made over the years. An unstable elevated ST segment in the right precordial leads was accompanied by a gradual shift of the electrical axis to the left. Q waves appeared in leads V1 and V2 after his MI.

The Brugada brothers will probably agree with us that this patient, now in good shape at the age of 84, should not immediately receive a defibrillator. What about other asymptomatic individuals? To us, it seems that a couple of issues need to be addressed before we accept their rather straightforward advice. First of all, a clear definition of what one really should look for has never been given properly. It is clear that no
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Demonstrable heart disease should be present, but is 1-mm ST-segment elevation sufficient? Should RBBB, which actually might represent a pronounced J wave, be present? Should the ECG pattern be exaggerated by sodium channel-blocking drugs? And what about the family history? With regard to risk stratification, the latter issue seems of particular importance to us. However, no details, which ought to be present, are given. Are the 6 previously asymptomatic individuals who became symptomatic randomly divided between those identified during routine screening and those who were members of a family with a sudden cardiac death victim? The latter, by the way, may also be the case in individuals identified during routine screening.

In light of the patient discussed above (Figure), we firmly believe that before an AICD is implanted in every individual with the “Brugada sign,” at least the issues raised above need to be settled. Some of them may be addressed by the authors directly. Others probably need a discourse at large, involving scientists not predisposed to recognize the syndrome.

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Response

Drs Wilde and Düren address a very important issue concerning the use of implantable defibrillators in asymptomatic patients with what they call the “Brugada sign.” To illustrate their skepticism, they present the single case of a patient with an abnormal ECG followed up over a 40-year period without complaints.

In 1992, we described the syndrome of right bundle-branch block, ST-segment elevation, and sudden death in patients without structural heart disease. Since then, an increasing number of publications about the syndrome are available in the literature. Most of these series refer to symptomatic patients and show a high recurrence rate during follow-up, precluding any discussion about the indication of implantable defibrillators in them. In the article in Circulation, we reported that 6 (27%) of 22 asymptomatic patients developed symptoms (sudden death or aborted ventricular fibrillation) during 34±32 months of follow-up. Age, sex, family history, inducibility of arrhythmias, and previous treatment were not helpful in stratifying the risk of subsequent sudden death. On the basis of these limited data, we recommended the use of implantable defibrillators in asymptomatic patients with the characteristic ECG pattern. We have learned so far that the syndrome is often familial, genetically determined, and due to a mutation in the cardiac sodium channel. We also know that the ECG signs are variable in time and that sodium channel–blocking agents can be used to unmask and modulate the ECG pattern.

Drs Wilde and Düren will have to agree that this represents a lot of information available only 6 years after initial description of the syndrome. New studies are being conducted to clarify the specificity and sensitivity of the ECG pattern, comparing the saddle-back type with the coved pattern, to identify new mutations, to discern the value of antiarrhythmic manipulation of the ECG, to understand the electrophysiological mechanisms of the syndrome, and to stratify patients according to risk. Until these data become available, we have to rely on what we have, and a 27% sudden death rate in young, apparently healthy asymptomatic patients seems more than sufficient to indicate the need for an implantable defibrillator. We cannot definitely say whether the ECG of the patient mentioned by Drs Wilde and Düren is diagnostic of the syndrome, and if so, why he is still alive. However, evidence-based medicine requires more than a single observation to make scientific conclusions, and this is what we try to do at present.

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Hemopericardium in a Patient With Systemic Lupus Erythematosus

To the Editor:
The Clinicopathological Conference in Circulation for July 21, 1998, was fascinating and ably discussed, with a useful review of the literature. I think one might supplement observations that might have been made during the patient’s hospital course and might have been acted on. There is no mention of the neck veins, which may have been difficult to “read,” but the echocardiographic description of the pericardial effusion includes “without evidence of hemodynamic compromise.” Quite clearly, this indicates absence of chamber collapses. However, collapses need not be present even in florid tamponade and particularly in some cases with chamber hypertrophy. Similarly, the absence of pulsum paradoxus correlates well with left ventricular hypertrophy. Moreover, the initially high blood pressures (especially in a patient who had been hypertensive) are not incompatible with severe cardiac tamponade. The really ominous finding was the progressive bradycardia. In the absence of
uremia, this should have been a signal that something critical was afoot (we are not told the effect of atropine therapy, but it is likely to have been unsuccessful). The evidence did not permit specific suspicion of dissecting aortic aneurysm, but the patient was quite ill, and there was a pericardial effusion, and therefore further investigation of that effusion might have been prompted by the undue bradycardia. Of course, pericardiocentesis of a hemopericardium due to aortic rupture could precipitate complete collapse by permitting freer bleeding from the aorta. (Operative drainage would have been the appropriate management.) These comments are not to criticize but rather to supplement the presentation of a very interesting case.

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**Response**

We appreciate Dr Spodick’s comments on the Clinicopathological Conference. The general observations that he makes related to potential clinical clues to cardiac tamponade are important points to remember. In this particular case, however, some points of clarification should be noted. The neck veins were originally “flat” and did not change on transfer to the intensive care unit. The pericardial effusion seen on the transthoracic echocardiogram was of moderate size, but it predominantly surrounded the right atrium and was consistent with a localized or loculated fluid collection. A percutaneous attempt at pericardiocentesis given the location was not possible. We do agree that the episodes of bradycardia were troublesome. The clinical service felt it was appropriate to exclude pulmonary embolism or loculated fluid collection. A percutaneous attempt at pericardiocentesis would have been difficult given the location, and therefore further investigation of that effusion might have been prompted by other findings. Of course, pericardiocentesis of a hemopericardium due to aortic rupture could precipitate complete collapse by permitting freer bleeding from the aorta. (Operative drainage would have been the appropriate management.) These comments are not to criticize but rather to supplement the presentation of a very interesting case.

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**Administration of Adenosine in Sinus Rhythm for Diagnosis of Supraventricular Tachycardia**

To the Editor:

We read with interest the article by Belhassen et al describing the usefulness of the administration of ATP during sinus rhythm for the noninvasive diagnosis of dual AV node physiology in patients with AV nodal reentrant tachycardia. We would like to present our experience with the administration of intravenous adenosine during sinus rhythm in patients with supraventricular tachycardia, including AV nodal reentry and AV reentry using a concealed left lateral accessory pathway. Our protocol consisted of the administration of 12 mg of adenosine during sinus rhythm, with simultaneous recording of 2 surface ECG leads and intracardiac electrograms. Standard criteria were used for the electrophysiological diagnosis of the tachycardia mechanism. All tachycardias were successfully ablated afterward. Our results in patients with AV nodal reentry agree with those of Belhassen et al. Three (75%) of 4 patients showed evidence of dual AV nodal physiology, including sudden increases in the PR interval in 2 and echoes in 2. Findings in our 12 patients with concealed accessory pathways (none of them with dual AV node physiology) were interesting. Ten patients (80%) developed AV echo beats (1 to 4 per patient). These echo beats occurred ~10 seconds after adenosine administration, were preceded by a gradual increment in the AH interval (from 76±13 to 114±33 ms), and had a sequential atrial activation identical to that seen during orthodromic AV reentry. In 2 patients, the echoes initiated short runs of AV reentry. When the surface ECG recordings were analyzed by an observer blinded to the intracardiac recordings, retrograde P waves could be identified in 8 of 10 patients with echoes. The RP’ interval for these echo beats was ≥80 ms. In contrast, in most patients with AV nodal reentry, echo beats are buried in the QRS and can only be inferred by a subsequent resetting of the sinus rate.) When lead I had been monitored, the retrograde P’ wave was characteristically negative, suggesting the location of the pathway. We conclude that the administration of adenosine in sinus rhythm with simultaneous recording of multiple surface ECG leads is useful for the noninvasive diagnosis of the mechanism of supraventricular tachycardia. In addition to Belhassen’s criteria favoring AV nodal reentry, the timing and polarity of the retrograde P wave of echo beats can suggest the presence (and location) of a concealed accessory pathway. This will be particularly important in the not insignificant number of patients with AV reentry and concomitant dual AV node physiology.

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1. Belhassen B, Fish R, Glikson M, Glick A, Eldar M, Laniado S, Viskin S. Tachycardia mechanism. All tachycardias were successfully ablated afterward. Our results in patients with AV nodal reentry agree with those of Belhassen et al. Three (75%) of 4 patients showed evidence of dual AV nodal physiology, including sudden increases in the PR interval in 2 and echoes in 2. Findings in our 12 patients with concealed accessory pathways (none of them with dual AV node physiology) were interesting. Ten patients (80%) developed AV echo beats (1 to 4 per patient). These echo beats occurred ~10 seconds after adenosine administration, were preceded by a gradual increment in the AH interval (from 76±13 to 114±33 ms), and had a sequential atrial activation identical to that seen during orthodromic AV reentry. In 2 patients, the echoes initiated short runs of AV reentry. When the surface ECG recordings were analyzed by an observer blinded to the intracardiac recordings, retrograde P waves could be identified in 8 of 10 patients with echoes. The RP’ interval for these echo beats was ≥80 ms. In contrast, in most patients with AV nodal reentry, echo beats are buried in the QRS and can only be inferred by a subsequent resetting of the sinus rate.) When lead I had been monitored, the retrograde P’ wave was characteristically negative, suggesting the location of the pathway. We conclude that the administration of adenosine in sinus rhythm with simultaneous recording of multiple surface ECG leads is useful for the noninvasive diagnosis of the mechanism of supraventricular tachycardia. In addition to Belhassen’s criteria favoring AV nodal reentry, the timing and polarity of the retrograde P wave of echo beats can suggest the presence (and location) of a concealed accessory pathway. This will be particularly important in the not insignificant number of patients with AV reentry and concomitant dual AV node physiology.

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Response

We thank Dr Labadet and colleagues for their interest in our work.1 We are pleased to note they observed results similar to ours using adenosine in their group of patients with AV nodal reentry tachycardia. The results they observed in patients with AV reentrant tachycardia are very interesting. We have made similar observations using ATP. Therefore, it seems that administration of adenosine or ATP during sinus rhythm may be of some diagnostic value for most (>90%) regular, paroxysmal supraventricular tachycardia. When using adenosine, however, we recommend that the initial tested dose be 6 mg and not 12 mg. In our study,1 about one third of the patients exhibited signs of dual AV node physiology with 10 mg of ATP (which corresponds approximately to a dose of 5.3 mg of adenosine, taking into account the different molecular weights of ATP and adenosine). The use of an initial dose of 12 mg of adenosine may prevent the unmasking of dual AV node physiology in some patients owing to simultaneous block in both slow and fast pathways. In patients with AV reentry tachycardia, the use of a high initial dose of adenosine may result in complete AV nodal block that prevents the initiation of AV reentrant echos.

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Evaluation of Regional Differences in Right Ventricular Systolic Function

To the Editor

In their recent study on right ventricular function, Geva and colleagues1 indicate that most previous studies have focused on the chamber as a unitary structure. They then present data supporting its traditional division into “sinus” and “infundibular” components. It is disingenuous of these investigators, however, not also to cite the considerable body of evidence that supports the concept that, morphologically, the right ventricle can be analyzed in terms of 3 components, namely, the inlet, the apical trabecular component, and the outlet. The ring of anatomic landmarks that they cite as the infundibular boundary is, in fact, far from uniformly present in the normal heart. The moderator band, extending from the septal band to the anterior papillary muscle of the tricuspid valve, is but one of the many coarse trabeculations to be found in the apical component. Equally important are the septoparietal bands, identified by Goor and Lillehei2 when they proposed the important tripartite approach to right ventricular structure.

There is just as much, if not more, anatomic and embryological evidence to support this tripartite approach as that cited by Geva et al1 in substantiating their bipartite concept. Thus, division of the ventricle is equally well explained in tripartite fashion. The rudimentary right ventricle in double-inlet left ventricle and tricuspid atresia is separated from the dominant left ventricle by the remnant of the apical trabecular septum rather than the infundibular septum. It is this apical muscular septum that carries the atrioventricular conduction axis,3 conduction tissue never being found in the muscular outlet septum. Immunostaining has shown that the conduction axis develops astride this apical muscular septum from the outflow.4 The ventricular outflow tracts are divided by the outflow ridges and then give rise to the muscular outlet septum in malformed hearts, but with no relationship to the conduction tissues.5 In the normal heart, however, these ridges are muscularized to form the free-standing subpulmonary infundibulum, the anatomic feature that makes it possible for the surgeon to remove the pulmonary valve as an autograft and use it in the Ross procedure. This would not be possible if the “infundibular septum” was positioned as shown by Geva et al.1 To have presented their results without any discussion of a potentially tripartite configuration presents an unduly biased account of right ventricular structure.

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Response

We thank Dr Anderson for his interest in our article. His views regarding the anatomic partition of the right ventricle have been published1 and are controversial.2,3 Our article, however, is about regional differences in right ventricular systolic function. A discussion about the different views regarding the anatomic partition of the right ventricle is important but is beyond the scope of our article.

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Evaluation of Regional Differences in Right Ventricular Systolic Function
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