Clinical and electrophysiologic characterization of automatic junctional tachycardia in adults

MICHAEL A. RUDER, M.D., JESSE C. DAVIS, M.D., MICHAEL ELDAR, M.D., JOSEPH A. ABBOTT, M.D., JERRY C. GRIFFIN, M.D., JOHN J. SEGER, M.D., and MELVIN M. SCHEINMAN, M.D.

ABSTRACT  Junctional ectopic tachycardia has been described in infants but not in adults. Five adults with rapid symptomatic paroxysmal junctional tachycardia, distinct from the more common slower nonparoxysmal junctional tachycardia, were recently evaluated. The tachycardia was irregular (rate 120 to 250) and accompanied by periods of atrioventricular dissociation and narrow QRS complexes. A junctional origin was documented during electrophysiologic study in four of the five patients. Analysis of Holter recordings; the response to exercise, isoproterenol, and propranolol; and the effects of atrial and ventricular stimulation appeared to implicate abnormal automaticity of a high junctional focus that was catecholamine sensitive or dependent as the tachycardia mechanism. All patients responded somewhat to β-blockers, although a combination of procainamide and propranolol proved to be the most effective therapy in one patient and another chose electrode catheter ablation of the atrioventricular junction rather than continued drug therapy. Thus, junctional ectopic tachycardia may occur in adults and its mechanism appears to be related to abnormal automaticity that is catecholamine sensitive or dependent. Initial therapy should include β-blockers but selected patients may require more aggressive management.


Rapid junctional ectopic tachycardia has been described in infancy, frequently in association with congenital cardiac defects, and may possibly occur in a familial pattern. Typical features include rapid, irregular heart rates and atrioventricular dissociation. Junctional ectopic tachycardias of this sort generally respond poorly to drug therapy and are associated with a very poor prognosis. To our knowledge, this arrhythmia has not been described in adults. The purpose of this report is to describe five adults with junctional ectopic tachycardia. Detailed analysis of the electrocardiograms, electrophysiologic studies, and response to therapy is provided.

Material and methods

Five patients with symptomatic, rapid, irregular tachycardias were referred for evaluation. Detailed medical histories, physical examinations, chest x-rays, and echocardiograms were obtained in all patients. In addition, multiple 24 hr Holter recordings were available for analysis in all and three of the patients had undergone cardiac catheterization before referral. Four of the five patients underwent invasive electrophysiologic studies for purposes of elucidating the mechanism of the arrhythmia, drug testing, and/or evaluation of nonpharmacologic therapy.

Electrophysiologic studies. Patients were studied in the unsedated, postabsorptive state after informed consent was obtained. All antiarrhythmic drugs were discontinued for at least five half-lives before testing. Three quadripolar electrode catheters were inserted percutaneously into a femoral vein of each patient and positioned across the tricuspid valve, against the high lateral right atrium and the right ventricular apex. Electrocardiographic leads V1, I, and III, and the intracardiac recordings from the high right atrium, His bundle region, and right ventricle were displayed on the Electronics for Medicine VR-12 oscilloscope and recorded simultaneously at a paper speed of 50 or 100 mm/sec. Atrial and ventricular stimulation were performed with a programmable digital stimulator (Bloom, Inc., Redding, PA). The stimuli were 2 msec in duration with current strength twice diastolic threshold for ventricular stimulation or 5 mA for atrial stimulation.

Atrial overdrive pacing at cycle lengths of 600 to 270 msec and programmed atrial stimulation at a basic drive cycle length of 500 msec with the introduction of one and two extrastimuli were performed in all patients. Ventricular overdrive pacing at cycle lengths of 600 to 270 msec was then performed. Programmed ventricular stimulation at basic drive cycle lengths of 500 and 400 msec with the introduction of up to three ventricular extrastimuli was performed; the tachycardia proved virtually incessant in patient 2. In three patients, isoproterenol was infused to increase the sinus rate to at least 130 beats/min. Isoproterenol was not infused in patient 2 because of documented initiation of tachycardia and acceleration with exercise. The effect of the infusion of propranolol (0.1 to 0.2 mg/kg) on
spontaneous tachycardia was studied in all patients who underwent electrophysiologic study.

**Results**

**Clinical features** *(table 1).* The age at onset of tachycardia ranged from 13 to 23 years (mean 19). Incidence of tachycardia varied from relatively infrequent to many times daily. Similarly, duration of tachycardia varied from several seconds to hours. Exercise and stress were frequent precipitating factors in all patients except No. 4. All the patients had significant symp-
toms during tachycardia; syncope occurred in two. Two patients had concomitant heart disease (one an atrial septal defect and one a ventricular septal defect), whereas the others had normal hearts.

**Electrocardiograms.** The rate of the tachycardia varied from 110 to 250. QRS complexes during tachycardia were identical to those during sinus rhythm, except for rate-related aberrancy in two patients. The tachycardia was grossly irregular except in patient 5 in whom it was only slightly irregular. P waves were

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**TABLE 1**

**Junctional ectopic tachycardia in adults**

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Age/sex</th>
<th>Associated heart disease</th>
<th>Duration of symptoms (years)</th>
<th>Arrhythmia pattern</th>
<th>Rate</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>20/F</td>
<td>—</td>
<td>7</td>
<td>Grossly irregular</td>
<td>150–250</td>
<td>His bundle ablation*</td>
</tr>
<tr>
<td>2</td>
<td>22/M</td>
<td>Atrial septal defect</td>
<td>1</td>
<td>Grossly irregular; Wenckebach exit block</td>
<td>110–180</td>
<td>Nadolol</td>
</tr>
<tr>
<td>3</td>
<td>58/M</td>
<td>—</td>
<td>36</td>
<td>Grossly irregular</td>
<td>145–180</td>
<td>Propranolol</td>
</tr>
<tr>
<td>4</td>
<td>24/M</td>
<td>Ventricular septal defect</td>
<td>1</td>
<td>Grossly irregular</td>
<td>150–170</td>
<td>Procainamide, propranolol</td>
</tr>
<tr>
<td>5</td>
<td>31/M</td>
<td>—</td>
<td>15</td>
<td>Slightly irregular; Wenckebach exit block</td>
<td>140–150</td>
<td></td>
</tr>
</tbody>
</table>

*Aβ-Blockers controlled the tachycardia but were not tolerated.*

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**FIGURE 1.** The 12-lead electrocardiogram and rhythm strip (lead II) during tachycardia for patient 1. The tachycardia is irregular (rate averages 160/min), with rate-related aberrancy, atrioventricular dissociation, and captured complexes. Rhythm strips during tachycardia were diagnosed as bouts of atrial fibrillation or multifocal atrial tachycardia.
either not consistently discernible or appeared dissociated from the tachycardia. Initial referral diagnoses included bursts of atrial fibrillation, multifocal atrial tachycardia, or polymorphous ventricular tachycardia.

Representative electrocardiographic strips are shown in figures 1, 2, and 3.

**Electrophysiologic study.** Four of the five patients underwent electrophysiologic study. Spontaneous tachycardia was induced in three patients. In the fourth patient, tachycardia was not inducible, although atrial fibrillation was inducible.

**FIGURE 2.** Electrocardiographic strips for patient 2 showed alternate bradycardia and tachycardia. The 12-lead electrocardiogram shows a slow (35 to 60 beats/min) junctional rhythm. The lead I rhythm strip (bottom) shows an irregular supraventricular tachycardia without discernible P waves.

**FIGURE 3.** Recording from telemetry lead during paroxysms of tachycardia in patient 3. The tachycardia is irregular (rate 145 to 180) with rate-related aberrancy. The patient was originally diagnosed as having polymorphous ventricular tachycardia as well as supraventricular tachycardia.
cardia was seen during the study in all four. A "junctional" origin of the tachycardia was confirmed in that a His deflection preceded each ventricular depolarization with a normal HV interval (figure 4). There were periods of atrioventricular dissociation during tachycardia in all patients. In some patients, retrograde conduction with varying HA intervals occurred at times. During these periods, the atrium could be dissociated from the tachycardia by pacing the high right atrium. The tachycardia was irregular in all patients. During study, this irregularity was not due to intermittent sinus capture, electrotonic interaction between sinus and ectopic tachycardia depolarization (figure 5), or Wenckebach periodicity or exit block from the tachycardia focus.

Sinus node function was normal except in patient 2 in whom there was profound sinus bradycardia and prolonged sinus node recovery times. Antegrade and retrograde atrioventricular conduction was normal in all patients and there was no evidence for dual atrioventricular nodal pathways. The tachycardia began and terminated spontaneously and could not be in-

**FIGURE 4.** Spontaneous tachycardia in patient 1. Recordings include surface leads V1, I, and III and bipolar electrograms from the right atrium (HRA) and His bundle (HBE). The atrial (A-A intervals) and ventricular cycle length (R-R intervals) are listed. The first three junctional complexes are aberrantly conducted and atrioventricular dissociation is present. The high to low orientation of the atrial electrograms suggest a sinus origin. The presence of atrioventricular dissociation excludes atrial or atrioventricular reentrant tachycardia and is strong evidence against atrioventricular nodal reentry.

**FIGURE 5.** Phase response curve of a typical tachycardia episode in relation to sinus discharges in patient 1. The x axis (ED to SND) represents the time after the junctional ectopic discharge (ED) at which a single spontaneous sinus node discharge (SND) occurs. The y axis (delta ETCL) represents the prolongation (positive values) or abbreviation (negative values) of the ectopic tachycardia cycle length (ETCL). There is no consistent relationship between the timing of the sinus impulse and its effect on tachycardia cycle length, as would be expected to occur if the irregularity of the tachycardia were due to electronic modulation of the ectopic tachycardia focus by sinus node discharges.
duced by atrial or ventricular overdrive pacing with multiple drive rates or by programmed atrial or ventricular stimulation (figure 6). During spontaneous tachycardia, neither atrial or ventricular pacing nor programmed stimulation caused acceleration or termination of the junctional tachycardia (figure 7).

The infusion of isoproterenol increased the frequency of episodes of spontaneous tachycardia and acceler-

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**FIGURE 6.** Atrial overdrive pacing during sinus rhythm (patient 2) at a cycle length of 350 msec fails to initiate tachycardia. The postpacing pause is terminated by a junctional escape complex. Inability to initiate tachycardia with either atrial or ventricular overdrive or by programmed stimulation is further evidence against a reentrant or triggered mechanism. Abbreviations as in figure 4.

**FIGURE 7.** Atrial overdrive pacing during an episode of sustained junctional tachycardia does not appear to produce perceptible change in the tachycardia cycle length. The baseline irregularity of the tachycardia rate is noted. The tachycardia could not be terminated by atrial, ventricular overdrive, or programmed pacing. Inability to terminate the tachycardia with pacing supports an automatic mechanism.
FIGURE 8. Control tracings from patient 2 during tachycardia showing retrograde atrial activation (low septal atrial to high atrial sequence) from the junction. Spontaneous (or induced) premature depolarizations did not consistently affect the tachycardia cycle length.

FIGURE 9. Tracings from patient 2 after administration of propranolol. The low septal to high right atrial sequence persists but episodic antegrade block from the junctional focus is present. The response to propranolol is very suggestive of a high junctional focus with antegrade block occurring within the atrioventricular node.

ated the rate of tachycardia in patients 1 and 3 but not in patient 4. It was not used in patient 2 because of the clear association of tachycardia with exercise. The infusion of propranolol either abolished spontaneous tachycardia or markedly reduced its incidence. In patient 2, infusion of propranolol (0.20 mg/kg) produced antegrade block from the tachycardia focus (figures 8 and 9) before termination of the tachycardia.

Treatment. Patients 1, 2, and 3 had been treated with various antiarrhythmics, including procainamide, lidocaine, quinidine, verapamil, and disopyramide, without effect. All patients responded in part to β-
blockade. The incidence of tachycardia was reduced but not abolished by oral propranolol in patient 4 and procainamide eliminated junctional ectopy. Patient 5 refused drug therapy.

Various β-blockers were uniformly effective in patient 1 but caused intolerable side effects. This patient opted for catheter ablation of the atrioventricular junction as a means of controlling her arrhythmia. This was accomplished by use of two direct-current shocks of 200 J each. She subsequently underwent insertion of a DDD pacemaker and is symptom free.

Discussion

Clinical features. While rapid, irregular junctional rhythms have been previously described in infants and young children, this is, to our knowledge, the first report of this entity in adults. The clinical manifestations of junctional ectopic tachycardia in adults share many features previously described for infants but appear to differ in other respects. Gillette and his colleagues, for example, have stressed the malignant course of junctional ectopic tachycardia in infants whose deaths were attributed to either congestive heart failure or uncontrollable arrhythmias. Conceivably, the more benign course in our patients was related to the fact that the tachycardia was paroxysmal rather than incessant in nature. Previous reports in children have emphasized that these arrhythmias are often unresponsive to drug therapy, although rate reduction with propranolol, amiodarone, or more recently propafenone, has been observed. In contrast, the response to β-blockers in our patients was good to excellent and one patient responded best to combined procainamide and β-blocker therapy.

Mechanism of tachycardia. Although it is difficult to precisely define the mechanism of junctional ectopic tachycardia, the weight of evidence would suggest the presence of abnormal automaticity. The presence of atrioventricular dissociation excludes an atrial tachycardia or tachycardia incorporating an accessory atrioventricular nodal bypass tract. Persistent atrioventricular dissociation is very rare in patients with atrioventricular nodal reentry tachycardia. Other features that argue against an atrioventricular nodal reentrant mechanism include inability to initiate or terminate the tachycardia with pacing, spontaneous onset of tachycardia without preceding changes in the spontaneous cycle length, and irregularly irregular tachycardia rate and the absence of either antegrade or retrograde dual atrioventricular nodal conduction patterns in any patient. The response to cardiac pacing did not meet any of the proposed criteria for triggered rhythms. Similarly, the lack of response to calcium-channel antagonists suggests that triggered activity was not the tachycardia mechanism. Dangman and Hoffman have studied the response of cells with abnormal automaticity to overdrive pacing, but the grossly irregular tachycardia rate observed in our study precluded firm conclusions regarding overdrive suppression or acceleration.

The tachycardia in our patients appeared to originate within the atrioventricular junction. The fact that antegrade block above the bundle of His was induced with propranolol in patient 2 (figure 8) and the rapid and enhanced response to exercise and/or isoproterenol seen in all of the patients implicate a focus high in the junction, possibly involving nodal tissue. Administration of atropine may have also been useful in clarifying origin of the tachycardia, but it was not done.

The irregularity of the junctional response observed in these patients remains unexplained. Irregular junctional rhythms have been described in infants with junctional ectopic tachycardia, but detailed analyses of possible causes for the irregular rhythm have not been analyzed. In the present report, analysis of multiple Holter examinations together with the electrophysiological studies conclusively excluded the following possible causes of the irregular rhythm: (1) sinus capture, (2) emergence of at least two foci with different rates, and (3) changes in the junctional rate owing to electronic interaction with sinus impulses (figure 5). In only two subjects could the irregular rates at times be explained by Wenckebach exit block from the junctional focus.

Differential diagnosis. Junctional ectopic tachyarrhythmias can be clearly differentiated from the more common nonparoxysmal junctional tachycardia. The latter occurs predominantly in the setting of acute myocardial ischemia, digitalis excess, chronic obstructive lung disease, rheumatic carditis, metabolic derangements, or after cardiac surgery. These arrhythmias are slower (70 to 120/min) and usually regular, although Wenckebach exit block has been described in the presence of digitalis excess. The evidence to date suggests that nonparoxysmal junctional tachycardia appears to be related to delayed afterdepolarizations.

Care must be taken to differentiate junctional ectopic tachycardia from other arrhythmias. The typical appearance of a rapid, irregularly irregular tachycardia with atrioventricular dissociation excludes an atrial or atrioventricular reentrant tachycardia and is strong evidence against a reentrant supraventricular arrhythmia involving the atrioventricular node. However, in those
cases in which P waves are not consistently obvious, the arrhythmia may be mistaken for atrial fibrillation or multifocal atrial tachycardia. Similarly, irregular, abnormally conducted junctional complexes may simulate bursts of polymorphous ventricular tachycardia. Because it may resemble these other arrhythmias, we believe that junctional ectopic tachycardia may be more common than previously realized.

Therapeutic implications. All patients responded somewhat to β-blockers and these drugs would appear to be the drugs of choice for initial therapy. Care should be taken to assess the heart rate after β-blockade to avoid bradycardia. In patients with alternate junctional tachycardia and bradycardia (patient 2), initial assessment of drug therapy would be best undertaken after insertion of a temporary ventricular pacemaker. The response to type I agents appear to be variable. Three patients were clearly unresponsive to a variety of conventional and experimental type I drugs, while patient 4 responded best to a combination of procainamide and β-blockers. Finally, catheter ablation of the atrioventricular junction is an acceptable alternative treatment when drug therapy is either ineffective or associated with unacceptable side effects.4,5

References
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