Rhythm Abnormalities in Children with Isolated Ventricular Noncompaction

ALPAY ÇELIKER, SÜHEYLA ÖZKUTLU, EMBIYA DILBER, and TEVFIK KARAGÖZ
From the Department of Pediatric Cardiology, Hacettepe University Faculty of Medicine, Ankara, Turkey


Background: Isolated ventricular noncompaction (IVNC) is a serious cardiomyopathy with a generally poor prognosis. It is characterized by the presence of prominent ventricular myocardial trabeculations and deep intertrabecular recesses, in the absence of other structural heart defects. This cardiomyopathy is usually associated with ventricular dysfunction, thromboembolic events, and rhythm problems.

Methods and Results: This article describes 11 children who have rhythm abnormalities associated with IVNC. On admission, eight children had complaints including palpitation, syncope, heart failure, and bradycardia. Ventricular arrhythmias were diagnosed in four children, sinus node and atrioventricular conductance disturbances in six children, and Wolff-Parkinson-White syndrome and associated tachycardia in one child. Three children with ventricular arrhythmias received an automatic implantable cardioverter defibrillator (ICD). Three patients died of cardiac problems during the follow-up period.

Conclusion: Since many of rhythm abnormalities, including life-threatening ventricular arrhythmias, may be seen in patients with IVNC, children with IVNC should be screened for arrhythmias. An ICD may be the best treatment for some of these patients. (PACE 2005; 28:1198–1202)

ventricular noncompaction, rhythm abnormalities, implantable cardioverter defibrillator, children

Introduction

Isolated ventricular noncompaction (IVNC) is a rare cardiomyopathy that results from an abnormal arrest of endomyocardial embryogenesis. The anatomical pattern of IVNC is characterized by prominent ventricular myocardial trabeculations with deep intertrabecular recesses. The long-term prognosis of the disorder is usually poor with a high morbidity and mortality due to heart failure, arrhythmias, and endocardial clot formation with systemic embolization. This study was designed to report cardiac rhythm abnormalities in 11 children with IVNC.

Methods

This study consists of 11 children with IVNC and arrhythmias followed in a pediatric cardiology department. All IVNC patients were screened for rhythm abnormalities, and patients who had arrhythmias were included in this study. This accounts about one-fourth of all children with IVNC diagnosed in our department. Personal and family history of patients and physical examination findings were recorded. Chest roentgenogram, 12-lead electrocardiogram, two-dimensional echocardiogram, 24-hour Holter monitoring, and treadmill exercise testing were evaluated in all cases. Additionally, intracardiac electrophysiological studies, with programmed ventricular stimulation (PES), were performed in four children.

A complete echocardiographic examination was performed in all children. Quantitative evaluation of left ventricular dimension and systolic function was done in accordance with data published elsewhere. Diagnosis of IVNC was based on the presence of numerous, excessively prominent trabeculations associated with deep intertrabecular recesses. The left ventricular segment with the maximal measured wall thickness was used for the analysis of the noncompaction to compaction ratio. For the diagnosis of noncompaction, the noncompacted endocardial segment of the myocardium has to be at least twice as thick as the compacted epicardial segment. Coexisting cardiac abnormalities that sometimes exhibit similar myocardial pattern of sinusoids, such as ventricular outflow tract obstruction, were excluded.

Results

Clinical data, echocardiographic and electrophysiological features, of the children are presented in Table I. There were seven males and four females. Age at presentation ranged from 1 day to 12 years and the duration of follow-up ranged from 1 day to 6 years. Eight children presented with symptoms, including palpitation, syncope, heart failure, and bradycardia. Three children with isolated sinus bradycardia had no complaints. None of the children had any extracardiac malformation. Familial occurrence was documented in a first-degree relative in two children. Three children died during the follow-up period.
### Table I.
Clinical Data, Electrophysiological and Echocardiographic Characteristics of Children with Isolated Ventricular Noncompaction

<table>
<thead>
<tr>
<th>Patient (Admission)</th>
<th>Sex</th>
<th>Clinical Presentation</th>
<th>Rhythm Abnormality (ECG, Holter Monitorization)</th>
<th>EF/FS (%) (Last Exam.)</th>
<th>EPS</th>
<th>Treatment</th>
<th>Follow-Up (Years)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>F*</td>
<td>Palpitation, heart failure</td>
<td>Sinus tachycardia LAD, multiiform VES, couplet, sinus bradycardia, LAD, deep Q waves, flattened T waves, ST-depression</td>
<td>38/15</td>
<td>VT, 2S</td>
<td>Amiodarone, digoxin</td>
<td>8, 5 (died)</td>
</tr>
<tr>
<td>2</td>
<td>M*</td>
<td>Palpitation</td>
<td>Sinus bradycardia, LAD, deep Q waves</td>
<td>49/25</td>
<td>VT, 1S</td>
<td>ICD implantation, amiodarone</td>
<td>3</td>
</tr>
<tr>
<td>3</td>
<td>F</td>
<td>Syncope</td>
<td>Normal</td>
<td>58/30</td>
<td>VF, AP</td>
<td>ICD implantation, amiodarone</td>
<td>4</td>
</tr>
<tr>
<td>4</td>
<td>M</td>
<td>Syncope, heart failure</td>
<td>Sinus bradycardia, deep Q waves, negative T waves</td>
<td>46/22</td>
<td>VF, 1S</td>
<td>ICD implantation, amiodarone</td>
<td>2, 5</td>
</tr>
<tr>
<td>5</td>
<td>F</td>
<td>Bradycardia, hydrops</td>
<td>Bradycardia (55bpm)</td>
<td>57/29</td>
<td></td>
<td>Temporary PM implantation</td>
<td>8 hours (died)</td>
</tr>
<tr>
<td>6</td>
<td>M</td>
<td>Asymptomatic</td>
<td>Sinus bradycardia (52 bpm)</td>
<td>66/36</td>
<td></td>
<td></td>
<td>4</td>
</tr>
<tr>
<td>7</td>
<td>M</td>
<td>Asymptomatic</td>
<td>Sinus bradycardia (54 bpm)</td>
<td>69/39</td>
<td></td>
<td></td>
<td>3, 5</td>
</tr>
<tr>
<td>8</td>
<td>M</td>
<td>Asymptomatic</td>
<td>Sinus bradycardia (58 bpm)</td>
<td>64/35</td>
<td></td>
<td></td>
<td>2</td>
</tr>
<tr>
<td>9</td>
<td>M</td>
<td>Heart failure, bradycardia</td>
<td>LAD, Mobitz type II AV block, negative T waves</td>
<td>36/14</td>
<td></td>
<td>Digoxin, PM implantation</td>
<td>5 (died)</td>
</tr>
<tr>
<td>10</td>
<td>F</td>
<td>Heart failure, bradycardia</td>
<td>Complete AV block, 50 bpm (junctional)</td>
<td>52/25</td>
<td></td>
<td>PM implantation</td>
<td>2</td>
</tr>
<tr>
<td>11</td>
<td>M</td>
<td>Palpitation</td>
<td>WPW syndrome, SVT attack</td>
<td>35/13</td>
<td></td>
<td>Sotalol</td>
<td>6</td>
</tr>
</tbody>
</table>

F = female; M = male; EF = ejection fraction; FS = fractional shortening; LAD = left axis deviation; WPW = Wolff-Parkinson-White; AV = atrioventricular; bpm = beats/min; VT = ventricular tachycardia; 1S = one extrastimulus from right ventricle; 2S = two extras from right ventricle; VF = ventricular fibrillation; AP = atrial pacing; ICD = implantable cardioverter defibrillator; PM = pacemaker; SVT = supraventricular tachycardia.

*Familial recurrence.
All children had a markedly increased left ventricular trabeculation. The lateral, inferior, and apical segments were most commonly involved. Eight children had decreased left ventricular systolic function. It was normal in other three children who had sinus bradycardia and had no other complaints.

Ventricular arrhythmias were diagnosed in four children, sinus node and atrioventricular conductance disturbance in six children, and Wolff-Parkinson-White (WPW) syndrome and associated tachycardia in one child.

Case Reports

Ventricular Arrhythmias

Patient 1 was referred with the diagnosis of dilated cardiomyopathy. On admission, she was 12 years old and had palpitations. Surface electrocardiogram revealed sinus tachycardia, left axis deviation, multiform isolated ventricular extrasystoles, and negative T waves in left precordial leads. Couplets were seen on Holter monitoring. On echocardiography, she had IVNC and left ventricular systolic dysfunction. Intracardiac electrophysiological study induced ventricular fibrillation during incremental atrial pacing to determine the AV conduction that was terminated with transthoracic shock. She continued to have syncope on amiodarone and an ICD was implanted. During follow-up, she had multiple episodes of ventricular fibrillation that were terminated by ICD shock (Fig. 1).

Patient 2 was the first-degree relative of patient 1. He was first seen at the age of 11 years and had palpitations. Surface electrocardiogram revealed sinus bradycardia of 50 beats/min, deep Q waves and negative T waves in left precordial leads. Echocardiography showed IVNC and left ventricular systolic dysfunction. Intracardiac electrophysiological study induced ventricular tachycardia with one extrastimulus, and he was given amiodarone. His complaints continued on amiodarone and an implantable cardioverter defibrillator (ICD) was implanted. Four months after ICD implantation, a right atrial mural thrombus was identified on echocardiography. It completely resolved with low molecular weight heparin. He continues to be anticoagulated with warfarin.

Patient 3 was admitted at the age of 6 years with complaints of nine syncopal episodes within the previous 2 months. Surface electrocardiogram and Holter monitoring were normal. On echocardiography, she had IVNC and decreased left ventricular systolic function. Intracardiac electrophysiological study induced ventricular fibrillation during incremental atrial pacing to determine the AV conduction that was terminated with transthoracic shock. She continued to have syncope on amiodarone and an ICD was implanted. During follow-up, she had multiple episodes of ventricular fibrillation that were terminated by ICD shock.

Sinus Node and Atrioventricular Conduction Disturbance

Four patients had bradycardia inappropriate for their age, which suggested sinus node abnormalities. Patient 5 was seen for the first time at 32 weeks’ gestation because of two siblings who died within the first postnatal day. On fetal echocardiography, the fetus had moderate hydrops. Heart rate was in the 60s and regular. She was delivered by cesarean at the 34th gestational week, weighting 2,600 g. There was no evidence of dysmophia and the karyotype was normal, 46,XX. The electrocardiogram showed bradycardia of about 54 beats/min that was originating from a low atrial region (Fig. 2). Severe spongiform cardiomyopathy was noted on transthoracic echocardiography (Fig. 3). She had no electrolyte and pH...
abnormalities. Pacing was begun promptly with a right ventricular electrode with highest output, but it was not possible to stimulate the ventricular myocardium. Bradycardia and poor ventricular function led to death within the first day. Three patients (patients 6–8) were referred because of sinus bradycardia. They had no other rhythm abnormalities. Echocardiography confirmed the diagnosis of IVNC. Left ventricular systolic function was normal. On Holter monitoring, mean heart rates were 52, 54, and 58 beats/min, and in exercise testing, maximum heart rates were 107, 112, and 117 beats/min, respectively. Holter monitoring did not detect any other rhythm abnormalities. There were also no complaints during the 2- to 4-year follow-up.

Atrioventricular block was diagnosed in two patients. Patient 9 had heart failure on admission. The surface electrocardiogram showed left axis deviation and negative T waves in the left precordial leads. Holter monitoring revealed intermittent Mobitz type I and II second-degree heart block. Heart rate ranged from 45 to 55 beats/min. A right ventricular pacemaker was implanted. He died due to heart failure while awaiting heart transplantation. Patient 10 was a 9-month-old girl, referred because of heart failure and bradycardia. There was cardiomegaly with a cardiothoracic ratio of 0.66 on chest roentgenogram. Surface electrocardiogram showed complete A-V block and junctional rhythm with a ventricular rate of 50 beats/min. Transthoracic echocardiogram showed IVNC and left ventricular systolic dysfunction. An epicardial pacemaker was implanted with a favorable 2-year follow-up.

**WPW Syndrome**

A 7-year-old boy (patient 11) admitted with palpitations and supraventricular tachycardia secondary to manifest WPW syndrome. He also had frequent ventricular ectopy. Transthoracic echocardiography revealed IVNC and left ventricular systolic dysfunction. In this patient, tachycardia attacks were controlled with sotalol.

**Discussion**

Patients with IVNC usually have depressed left ventricular systolic function and commonly present with heart failure. Other cardiac abnormalities are endocardial clot formation with systemic embolization and arrhythmias. The high incidence of arrhythmias in IVNC has not been explained by a well-defined morphological abnormality. However, it has been suggested that myocardial ischemia may play a crucial role in the development of impaired systolic function and ventricular arrhythmias in these patients, similar to ischemic heart disease. Different rhythm abnormalities have been reported in patients with IVNC, and ventricular arrhythmias were the most common form. In some cases, it may cause palpitations and syncope as the presenting symptoms. In a previous report, ventricular arrhythmias were documented in five of the eight patients and were the presenting symptoms in two. In another report, ventricular arrhythmias were observed in 14 of the 34 adult patients; 11 had nonsustained ventricular tachycardia and 3 patients had sustained ventricular tachycardia. We documented life-threatening
ventricular arrhythmias in 4 of the 11 patients. Ventricular arrhythmias in IVNC may be resistant to medical treatment and may follow a fatal course.\textsuperscript{1,3,12} In these patients, ICD was tried as a life-saving mode of treatment.\textsuperscript{4,13} Oechslin et al.\textsuperscript{4} implanted an ICD in four patients who had sustained ventricular tachycardia or a presyncopal event and inducible ventricular tachycardia by electrophysiological study. Despite this treatment, one of these patients died from recurrent and refractory ventricular tachycardia. In our cases, ventricular arrhythmias were resistant to medical treatment. An ICD was implanted in three patients as an adjunctive treatment. It seems to be an effective mode of life-saving treatment, since the fibrillation attacks were terminated by the ICD in two patients.

IVNC may also be diagnosed prenatally.\textsuperscript{14,15} Guntheroth et al.\textsuperscript{14} reported fetal hydrops and intrauterine bradycardia in three such cases. Intrauterine demise was noted in two of these cases and the other died in the early postnatal period. They proposed that IVNC might be suspected on fetal echocardiography by the appearance of restrictive cardiomyopathy. Fetal cases generally have poor prognosis, especially if hydrops is present.\textsuperscript{14} One of our patients had fetal hydrops and marked bradycardia with a fatal outcome. Sinus bradycardia is another finding in IVNC.\textsuperscript{1} Three of our patients had sinus bradycardia. They had well-preserved left ventricular systolic function and had no complaints. Effort testing revealed decreased maximum heart rate response. During a follow-up of up to 4 years, no deterioration was seen on left ventricular systolic function.

Only a few cases of heart block have been reported in patients with IVNC.\textsuperscript{1,3,14} These include second-degree and complete A-V block, and some received an implanted transvenous pacemaker. In our series, one child had intermittent first degree and Mobitz type II, and other had complete A-V block. Transvenous pacemakers were implanted in these children with variable results.

WPW syndrome and associated tachycardia were also reported in association with IVNC.\textsuperscript{1,3,10,12} Ichida et al.\textsuperscript{3} reported WPW syndrome in 4 of 27 patients with IVNC; being manifested in three patients and concealed in one patient. In their report, this association is explained as the WPW syndrome is thought to arise from a failed regression of developmental embryologic atioventricular anatomical and electrical continuity which can also be seen in the failing regression of noncompacted myocardium in IVNC.\textsuperscript{3} Supraventricular tachycardia with WPW syndrome may lead to fatal ventricular tachycardia and fibrillation in IVNC patients.\textsuperscript{1,12} One child in our report had paroxysmal supraventricular tachycardia secondary to WPW syndrome. It was controlled with medical treatment and no attack of life-threatening ventricular fibrillation was recorded.

In conclusion, all kinds of rhythm abnormalities including life-threatening ventricular arrhythmias may be seen in patients with IVNC. Therefore, all children with IVNC should be screened for arrhythmias. Those with palpitations or syncope should undergo electrophysiologic testing unless a cause for the symptoms can be documented. An ICD may be the life-saving mode of treatment in patients with refractory ventricular arrhythmias.

References