Electrocardiographic Recognition of Sinus Node Dysfunction in Children and Young Adults

STEVEN M. YABEK, M.D., RICHARD E. SWENSSON, M.D., AND JAY M. JARMAKANI, M.D.

SUMMARY Twenty-four children and young adults with electrocardiographic evidence of sinus node dysfunction (SND) are described. The patients, whose ages ranged from three days to 25 years, were divided into three groups based on the type of SND. Ten patients had persistent and inappropriate sinus bradycardia (group I); twelve patients had episodes of prolonged sinus arrest (group II); and seven patients had repeated episodes of Mobitz type I or II sinoatrial exit block (group III). Eighteen patients had significant heart disease and in 11 of these the SND followed corrective cardiac surgery. Seven were operations for atrial septal defect and transposition repair. Seven patients with SND were symptomatic, having either syncope (3) or near syncope (2) episodes (4). All three with syncope have received permanent demand pacemakers. These data show that sinoatrial exit block occurs in many young patients with SND (29% in this series). Its recognition, however, requires careful electrocardiographic analysis in order to differentiate sinus bradycardia and sinus arrhythmia. Although SND most commonly occurs in children with congenital or acquired heart disease, particularly following corrective cardiac surgery, it may occur in the absence of other cardiac abnormalities.

SINUS NODE DYSFUNCTION (SND) is a well-recognized clinical entity in adults and implies both depressed automaticity and impaired sinoatrial conduction.1-8 In the past, little attention had been paid to disorders of the sinus node in infants and children. Recent reports in the literature have cited cases of SND in otherwise healthy children,9, 7 but most reports have concentrated on children with congenital heart disease either prior to or following corrective cardiac surgery.9-11 Most of the reported cases in children have presented with inappropriate sinus bradycardia, periods of sinus arrest with junctional or ventricular escape rhythms, ectopic atrial arrhythmias or Bradycardia. There has been little mention of disordered sinoatrial conduction as a manifestation of SND in the pediatric age group.

In this communication, we report 24 young patients both with and without congenital heart disease who have electrocardiographic evidence of SND.

Materials and Methods

Patients came from the pediatric cardiology services of the UCLA School of Medicine and the University of New Mexico School of Medicine. All electrocardiograms taken by these services during the period from October 1973 to June 1976 were reviewed. Twenty-four patients having evidence of SND on two or more 12-lead electrocardiograms taken at least one day apart were identified. There were 15 males and nine females, and their ages ranged from three days to 25 years. Other than the presence of SND, patients were considered to be free of heart disease if there was no suggestion of its presence by physical examination, electrocardiography or chest X-ray. Some of the study patients were taking digitalis glycosides or other antiarrhythmic or sympathetic medications. These patients were included only if they were free of all such medications for a period of at least five days during which time the electrocardiographic evidence for SND persisted.

The 24 patients were subdivided into three groups based on the type of SND as determined electrocardiographically. Patients with persistent or repeated episodes of inappropriate sinus bradycardia, defined as rates less than 50 beats/minute, and an inability to raise the rate by at least 20% with intravenous atropine administration (0.01 mg/kg) or submaximal exercise were included in group I.12-15 Patients having episodes of sinus arrest with no evidence of spontaneous atrial activity for periods greater than two P-P intervals, but not equaling a whole number multiple of the P-P interval, were included in group II. The control P-P interval was determined by averaging the three P-P intervals immediately preceding each pause. This group included patients with or without evidence of A-V nodal or ventricular escape beats. Group III was composed of patients having repeated episodes of Mobitz type I or II second degree sinus node exit block. These patients were identified using criteria established by Schamroth14 and Schamroth and Dove.16 Wenckebach (Mobitz type I) conduction defects at the sinoatrial level were identified by noting progressively shortening P-P intervals followed by a long pause. The sequence then was repeated without a progressive lengthening of the intervening P wave intervals. The progressive shortening of P-P intervals preceding the pause, caused by sinoatrial conduction times which were increasing by decreasing increments, is analogous to the shortening of R-R intervals that occurs in A-V nodal Wenckebach periodicity. To aid in differentiating those patients with sinoatrial Wenckebach periods from those with sinus arrhythmia, each P-P interval was plotted against its succeeding P-P interval as described by Schamroth and Dove18 (fig. 1).

Mobitz type II second degree sinoatrial conduction block was diagnosed when a long P-P interval, due to an apparent episode of "sinus arrest," equalled a whole number multiple of the control P-P interval. We realize that episodes of high
grade Mobitz type II second degree exit block combined with a sinus arrhythmia might go undiagnosed using this criterion. Patients having these latter arrhythmias would be erroneously included in group II, but there was no way to accurately make the distinction clinically.

Although many patients with active atrial and junctional tachyarrhythmias undoubtedly have SND, such patients were not included in this study unless they showed evidence of SND as defined by one of the above groups. Patients exhibiting symptomatology related to apparent SND were likewise included only if they satisfied the electrocardiographic criteria described above.

Results

Of the 24 patients with electrocardiographic evidence of SND, 18 had cardiac disease (table 1). Eleven of these 18 patients first developed SND following corrective cardiac surgery, and in seven cases, this surgery was for correction of a secundum atrial septal defect (5) or transposition of the great vessels (2). Each patient had at least three 12-lead electrocardiograms prior to surgery to document the absence of SND. Sinus node dysfunction developed from one day to seven weeks following surgery with a mean onset of eight days. Over an average postoperative follow-up period of 3.5 years, two of these patients have required permanent pacemakers because of recurrent syncopal episodes.

Severe SND occurred in two patients following episodes of acute viral myocarditis. Both patients experienced symptoms secondary to repeated bouts of transient sinus arrest (fig. 2) or Mobitz type II sinoatrial exit block. One patient (patient 19) ultimately required permanent pacing because of syncopal.

Five patients with unoperated congenital heart disease and six patients with no evidence of cardiac disease had SND (table 1). The latter patients were diagnosed while hospitalized for noncardiac disorders.

There were ten patients in group I. Four of these also showed electrocardiographic criteria for inclusion in other groups. Eight patients (3–10) had persistent sinus bradycardia with rates less than 50 beats/minute. In all instances

![Figure 1](image1.png)

**Figure 1. Illustration showing the effect of plotting each P-P interval against its succeeding interval as originally described by Schamroth and Dove.**

*Note: The distributional pattern as seen in sinus arrhythmia. b) The distributional pattern of Mobitz type I second degree sinoatrial exit block plotted from a rhythm strip obtained from patient 23.*

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age</th>
<th>Cardiac diagnosis</th>
<th>Symptoms; treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Group I</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1*</td>
<td>1½</td>
<td>TGV and PA</td>
<td></td>
</tr>
<tr>
<td>2*</td>
<td>1½</td>
<td>TGV and PS</td>
<td></td>
</tr>
<tr>
<td>3†</td>
<td>6</td>
<td>Postop TF</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>6</td>
<td>Normal</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>9</td>
<td>Normal</td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>11</td>
<td>VSD</td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>13</td>
<td>PS</td>
<td></td>
</tr>
<tr>
<td>8†</td>
<td>17</td>
<td>Post myocarditis</td>
<td>Near syncope</td>
</tr>
<tr>
<td>9</td>
<td>19</td>
<td>Normal</td>
<td>Near syncope</td>
</tr>
<tr>
<td>10</td>
<td>25</td>
<td>Postop ASD</td>
<td></td>
</tr>
<tr>
<td>Group II</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>11</td>
<td>2</td>
<td>Postop ASD,</td>
<td>Syncope; pacemaker</td>
</tr>
<tr>
<td></td>
<td></td>
<td>PAPVR</td>
<td>&amp; digoxin</td>
</tr>
<tr>
<td>12</td>
<td>4</td>
<td>Postop TGV</td>
<td></td>
</tr>
<tr>
<td>13</td>
<td>4</td>
<td>Normal</td>
<td></td>
</tr>
<tr>
<td>14</td>
<td>5</td>
<td>PDA</td>
<td></td>
</tr>
<tr>
<td>15</td>
<td>6</td>
<td>Postop TGV</td>
<td>Syncope; pacemaker</td>
</tr>
<tr>
<td>16</td>
<td>15</td>
<td>Postop ASD</td>
<td>Near syncope</td>
</tr>
<tr>
<td>17</td>
<td>16</td>
<td>Postop ASD</td>
<td>Syncope; pacemaker</td>
</tr>
<tr>
<td>18</td>
<td>18</td>
<td>Postop PDA;</td>
<td>Near syncope</td>
</tr>
<tr>
<td></td>
<td></td>
<td>levocardia</td>
<td></td>
</tr>
<tr>
<td>19</td>
<td>20</td>
<td>Post myocarditis</td>
<td>Syncope; pacemaker</td>
</tr>
<tr>
<td>Group III</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>20</td>
<td>3</td>
<td>Normal</td>
<td></td>
</tr>
<tr>
<td>21</td>
<td>3</td>
<td>Normal</td>
<td></td>
</tr>
<tr>
<td>22</td>
<td>1</td>
<td>Postop PDA</td>
<td></td>
</tr>
<tr>
<td>23</td>
<td>3</td>
<td>Postop VSD</td>
<td></td>
</tr>
<tr>
<td>24</td>
<td>8</td>
<td>Postop ASD</td>
<td></td>
</tr>
</tbody>
</table>

*Patients also satisfying criteria for group II.
†Patients also satisfying criteria for group III.
Abbreviations: TGV = transposition of the great vessels; PA = pulmonary atresia; PS = pulmonary stenosis; TF = tetralogy of Fallot; VSD = ventricular septal defect; ASD = secundum atrial septal defect; PAPVR = partial anomalous pulmonary venous return; PDA = patent ductus arteriosus.

![Figure 2](image2.png)

**Figure 2. Electrocadiographic leads II (top) and III (bottom) from patient number 19 demonstrating periods of transient sinus arrest with no escape beats. Asystolic period in upper strip lasted over six seconds and was accompanied by a near-syncope episode.**
intravenous atropine (0.01 mg/kg) or submaximal exercise failed to increase the rate by more than 20% and in no case did the rate increase to more than 58 beats/minute. Two patients, both aged 1½ years, had frequent, episodic sinus bradycardia which was unresponsive to intravenous atropine. Two patients from group I were symptomatic, both experiencing near syncopal episodes.

Twelve patients, including three in group I, satisfied criteria for group II. In 10 patients, each episode of sinus arrest was accompanied by a junctional or ventricular escape rhythm at a rate considerably lower than the intrinsic sinus rate. The tracing in figure 3A was taken from an 18-month-old child with transposition of the great vessels and pulmonary atresia. She had frequent hypercyanotic spells and evidence of SND disappeared. Another infant with severe systemic hypoxemia due to transposition of the great vessels and left ventricular outflow obstruction had SND with episodes of sinus bradycardia progressing to complete sinus arrest with junctional escape rhythms (fig. 3B). This 1½-year-old infant died during attempted surgical correction.

Two patients in group II had periods of sinus arrest unaccompanied by any escape rhythm. One patient was a 20-year-old male (patient 19) with myocarditis (fig. 2) and the other was a 16-year-old female (patient 17) following the repair of a secundum atrial septal defect. Both patients experienced repeated syncopal episodes eventually requiring treatment with permanent ventricular demand pacemakers.

A six-year-old female in group II (patient 15) required recurrent hospitalizations following repair of transposition of the great vessels because of tachy-bradyarrhythmias and frequent episodes of sinus arrest leading to syncope. She was the only patient in the present series having tachyarrhythmias, and her symptoms were not adequately controlled until

![Figure 3](http://circ.ahajournals.org/)

**Figure 3.** Continuous lead II rhythm strips from two patients with transposition of the great vessels and decreased pulmonary blood flow demonstrating periods of sinus arrest. *A*] Sinus rhythm with sudden, transient sinus arrest and junctional escape rhythm. B] Sinus rhythm gradually progressing to sinus bradycardia and sinus arrest with junctional escape rhythm.
the management included both digoxin and the insertion of a ventricular demand pacemaker.

Seven patients, including two from group I, satisfied the electrocardiographic criteria for inclusion in group III. Mobitz type I and II sinoatrial exit block were recognized electrocardiographically in four patients and three patients, respectively. The distributional pattern of points representing succeeding P-P intervals obtained from multiple plots showed little day to day variation and clearly distinguished patients with sinoatrial Wenckebach from those with sinus arrhythmia (fig. 1). The only symptomatic patient in this group was a 17-year-old male (patient 8) who, following acute myocarditis, also developed sinus bradycardia and periods of sinus arrest.

Seven patients had symptoms directly related to the SND. All three patients with syncopal episodes have received permanent ventricular pacemakers. The four patients having at least one episode of near syncope continue to be followed. An additional patient with hypercyanotic spells and marked SND was treated successfully with a systemic to pulmonary shunt.

Discussion

Sinus node dysfunction has been increasingly noted to occur in children and young adults. In most reports, its occurrence has followed major cardiac surgery, especially procedures involving the atria. This is consistent with our present findings. In one recent report, SND was present in 50% of children following repair of sinus venous atrial septal defects. The reported incidence following secundum atrial septal defect and transposition of the great vessels repair is also high. Histologic damage to the sinus node following such surgery has included hemorrhage and necrosis of the node and thrombosis of the sinus node artery. Damage directly related to suture material in and about the node has also been noted. Invasive electrophysiological investigations in some of these patients have suggested SND by showing prolongation of the sinus node recovery time following overdrive atrial stimulation.

Electrocardiographic evidence of SND following ventricular surgery, although uncommon, has been reported previously. In the present series, two patients developed evidence of SND following tetralogy of Fallot and ventricular septal defect repair. This may be explained on the basis of damage to the sinus node arising from atriotomy or vena caval cannulation as part of the operative procedure. The occurrence of sinus arrest and sinoatrial exit block in two patients following patent ductus arteriosus ligation cannot be explained as easily. The association of SND with unoperated cardiac defects might imply a congenital defect of the sinus node or its blood supply as part of the spectrum of congenital heart disease.

Sinus node dysfunction has recently been reported in children who show no evidence of congenital or acquired heart disease. The young patients with clinical and electrocardiographic SND described by Scott et al. are, in many respects, similar to the two healthy, young males described by James et al. who died suddenly while engaged in strenuous physical activity. Careful postmortem examination of these latter cases showed hemorrhagic and fibrotic foci within the sinus node and narrowing of the sinus node artery. In this report, we have described six children having electrocardiographic evidence of SND who were hospitalized for noncardiac disorders. In each case, the SND persisted following discharge from the hospital.

Previous reports have virtually ignored the presence of sinoatrial exit block as a cause of SND in children. This contrasts with recent literature from adult patients with SND in whom sinoatrial exit block constitutes a major contribution. In our group of 24 patients, seven (29%) had electrocardiographic evidence of Mobitz type I or II second degree sinoatrial exit block. Since first degree and third degree sinoatrial exit block cannot be diagnosed electrocardiographically, the true incidence of sinoatrial exit block as a cause of SND in this population probably remains underestimated.

First degree sinoatrial block, resulting from a fixed delay in conduction from the sinus node to the surrounding atrial musculature, produces no abnormalities in the P wave morphology or in the atrial rate or rhythm. Third degree sinoatrial block and high grade second degree block with sinus arrhythmia are similarly impossible to diagnose with certainty from the electrocardiogram since neither will result in pauses which are an exact multiple of the normal sinus interval. We believe that many of our cases of sinus arrest which are included within group II are probably patients experiencing transient episodes of sinoatrial exit block which, by the methodology of this study, cannot be diagnosed. Although not used in the present study, measurement of sinoatrial conduction times following multiple premature atrial stimuli, represents a very useful means of uncovering otherwise occult sinoatrial conduction abnormalities.

Adult patients with documented SND frequently have associated conduction system abnormalities and generalized depression of cardiac automaticity involving all subsidiary pacemakers. This depressed automaticity may contribute to the symptomatology experienced by these patients. Of the 12 patients in our series having periods of sinus arrest (group II), two had repeated asystolic periods of three seconds or longer without evidence of escape beats from subsidiary pacemakers. Both patients had syncopal episodes requiring pacemaker therapy. Although sufficient evidence is not available to suggest that SND in children is anatomically as widespread a disease as is seen in adults, the occurrence of prolonged asystolic periods warrants consideration of this possibility.

The sinus node dysfunction observed in two patients with transposition of the great arteries and decreased pulmonary blood flow probably was not related to structural sinus node disease. The sinus bradycardia and periods of sinus arrest seen in patient 1 were reversed following a palliative systemic to pulmonary shunt, suggesting that hypoxemia was responsible for the preoperative SND.

Since the method by which patients were included in this study involved screening of electrocardiograms from pediatric cardiology departments, the patient population must be considered highly selected. Accordingly, no attempt can be made to estimate the frequency of SND among normal children and young adults. We have shown, however, that significant SND does occur as a sequela of cardiac surgery, especially procedures involving the atrium. We have
also demonstrated that sinoatrial exit block is a major, but previously unstressed, manifestation of SND in children and young adults. Its recognition, however, requires careful and patient electrocardiographic analysis so that sinus bradycardia, sinus arrhythmia and sinus arrest can be differentiated.

Acknowledgment

We gratefully acknowledge and thank Mrs. Bobbie FitzGerald for her excellent typing and proofreading of this manuscript.

References


Correction

Electrocardiographic recognition of sinus node dysfunction in children and young adults.
S M Yabek, R E Swensson and J M Jarmakani

Circulation. 1977;56:235-239
doi: 10.1161/01.CIR.56.2.235
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1977 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/56/2/235

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation is online at:
http://circ.ahajournals.org//subscriptions/