

Sinus node dysfunction in children and adolescents: treatment by implantation of a permanent pacemaker in 26 patients

Firat Kardelen¹, Alpay Çeliker¹, Sema Özer¹, Şencan Özme¹, Ali Oto²

¹Cardiology Unit, Department of Pediatrics, and ²Department of Internal Medicine, Hacettepe University Faculty of Medicine, Ankara, Turkey

SUMMARY: Kardelen F, Çeliker A, Özer S, Özme Ş, Oto A. Sinus node dysfunction in children and adolescents: treatment by implantation of a permanent pacemaker in 26 patients. Turk J Pediatr 2002; 44: 312-316.

Sinus node dysfunction has been reported rarely in pediatric patients with structurally normal hearts. It has been diagnosed with increasing frequency in children and young adult patients with congenital heart defect, especially in patients who have undergone corrective cardiac surgery related with atrial tissue.

Between 1984-1999, 26 patients who were under 22 years of age underwent implantation of a permanent pacemaker for treatment of sinus node dysfunction at our medical center. This subset of patients represents 18.5% of all patients who required permanent pacemakers during this time. The mean age of the 17 male and 9 female patients at initial implantation was 9.2 ± 6 years (range, 0.5 to 22 years). Of the 26 patients, 18 (69%) had associated cardiovascular disease and in 11 (34.6%) patients, sinus node dysfunction developed after a cardiac operation.

The patients were followed up for a total 1,227 (47 ± 45 , range 2-176, median 34) pacing months. All symptomatic patients noted a resolution of symptoms after pacing had been performed, and they remained free of symptoms at the latest follow-up examination. Mean acute pacing thresholds and mean latest pacing thresholds for the endocardial atrial and ventricular leads, mean acute impedance and mean latest impedance for the endocardial atrial and ventricular leads and mean acute p wave voltage and the latest p wave voltage did not differ significantly.

In this report, we review our experience in children who required implantation of a permanent pacemaker for treatment of sinus node dysfunction during a 15-year period.

Key words: sinus node dysfunction, children, adolescents, permanent pacemaker implantation.

Sinus node dysfunction (SND) that necessitates permanent pacemaker therapy is much less common in children. The diverse clinical and electrocardiographic manifestations of this disorder were first described in adults¹. But, it has been diagnosed with increasing frequency in children and young adult patients with congenital heart defect, especially in patients who have undergone corrective cardiac surgery related with atrial tissue. SND has been reported less frequently in pediatric patients with structurally normal hearts².

In this report, we review our experience in young patients who required implantation of a permanent pacemaker for treatment of SND during a 15-year-period.

Material and Methods

Between 1984-1999, 26 patients who were under 22 years of age underwent implantation of a permanent pacemaker for treatment of SND at our medical center. The total number of patients with permanent cardiac pacemaker implantation performed at our institution during this period was 140. This subset of patients represents 18.5% of all patients who required permanent pacemakers during this time. The mean age of the 17 male and nine female patients at initial implantation was 9.2 ± 6 years (range 0.5 to 22 years).

The diagnosis of SND was based on the following electrocardiographic findings: 1) sinus pause or arrest for more than two seconds, 2)

sinus bradycardia (less than appropriate for age) 3) severe sinus dysrhythmia 4) slow escape rhythm 5) sinoatrial exit block (2° type I and II) 6) the bradycardia-tachycardia syndrome 7) sinus node re-entry tachycardia and 8) atrial muscle re-entry tachycardias.

The initial symptoms or signs of the patients were fatigue in 10, palpitation in four, syncope in four, dizziness in one and breath-holding spell in one. Six of the patients were asymptomatic at the time of pacemaker implantation. Indications for permanent pacing were SND with correlation of symptoms during age-inappropriate bradycardia in 22 patients and bradycardia-tachycardia syndrome with the need for long-term antiarrhythmic treatment other than digitalis in four patients. The 22 patients' Holter results were sinus arrest in six, pause more than 2.5 sec in six, sinus bradycardia + pause in four, sinus bradycardia in three, and atrial flutter/fibrillation in three (Table I).

Of the 26 patients, 18 (70%) had associated cardiovascular disease (Table I). Transposition of great arteries (TGA) was encountered in four patients. In 11 of 26 (34.6%) patients, SND developed after a cardiac operation. Surgical procedures included Mustard or Senning in four patients, Fontan operation in one, closure of the secundum atrial septal defect in five, and correction of endocardial cushion defect in one. Other surgical procedures, not strongly related with sick sinus syndrome, included repair of coarctation of aorta, valvuloplasty for stenosis of aorta, closure of ventricular septal defect and resection of subaortic ridge, each in one patient. Eight (30%) patients had no cardiovascular abnormality.

Statistical analysis was performed using paired *t* tests. A *p* value <0.05 was considered significant.

Results

The 26 patients were followed for a total of 1,227 months (47 ± 45 , range 2-176,

Table I. Clinical and Pacemaker Data of Patients

Case	Associated cardiac disorders	Age at implantation (y)	Initial symptom or signs	Holter	Follow-up (m)	Mode	Medication
1 (YB)	None	12	Bradycardia	Sinus arrest	124	VVIR	
2 (MB)	None	2	Syncope	Sinoatrial block	176	DDDR	
3 (IK)	None	8	Bradycardia	Sinus bradycardia	110	VVIR	
4 (IT)	None	11	Fatigue	Pause	101	VVIR	sotalol
5 (AHE)	None	12	Syncope	S. Brady+Pause	28	VVIR	
6 (OC)	None	2	Breath-holding	S. Brady+Junc. rhythm	12	VVI	
7 (ZÖ)	None	0.5	None	Sinus bradycardia	37	VVI	
8 (KC)	None	8	None	Brady-Tachy snd	67	VVIR	
9 (MEG)	TGA (Senning)	1	Bradycardia	Sinus arrest+pause	9	AAI	
10 (EK)	TGA-Mustard	21	Bradycardia	S. Brady+Pause	2	AAIR	
11 (AC)	TGA (Senning)	0.75	Bradycardia	Sinus arrest+pause	22	VVIR	
12 (AD)	TGA (Senning)	4	Fatigue	Atrial fibrillation	27	VVIR (Ep)	Quinidine, digoxin
13 (MI)	TA (Fontan)	15	Palpitation	Brady-Tachy snd	49	VVIR (Ep)	
14 (HK)	Closure of ASD+TVR	14	Palpitation	Brady-Tachy snd	58	VVI (Ep)	Amiodarone, digoxin coumadin
15 (SKo)	Closure of ASD+Repair of mitral cleft	11	Palpitation	Atrial Flutter	6	AAIR	
16 (AA)	Closure of ASD	8	Palpitation	Atrial fibrillation	120	VVIR	
17 (AI)	Closure of ASD	16	Syncope	Sinus arrest, J. rhythm	12	AAIR	
18 (HS)	Correction of ECD	8	Dizziness	Pause, Junc. rhythm	12	VVIR	
19 (SKr)	Closure of ASD+MVR	16	None	Pause	44	DDDR	
20 (SG)	VSD+SAS repair	9	None	S. Brady+Pause	28	AAIR	
21 (EO)	Aort coarctation repair	10	Fatigue	Sinus arrest	62	VVIR	
22 (HY)	AS operation	22	Syncope	Pause	2	AAIR	
23 (TK)	Myocarditis	12	Fatigue, CHF	Pause	39	DDDR	Digoxin
24 (SK)	Dilated cardiomyopathy	2	Fatigue, CHF	Brady-Tachy snd	41	VVI	Quinidine, digoxin.
25 (BD)	PFO+PSSVC	6	Fatigue	Pause	31	AAIR	
26 (MU)	Bicuspid Aorta	8	Bradycardia	S. Brady+Pause	8	VVIR	

TGA: transposition of great arteries, TA: tricuspid atresia, ASD: atrial septal defect, TVR: tricuspid valve replacement, ECD: endocardial cushion defect, MVR: mitral valve replacement, Subao: subaortic, PFO: patent foramen ovale, PLSVC: persistent left superior vena cava, CHF: congestive heart failure, S. Brady: sinus bradycardia, Brady-Tachy: bradycardia-tachycardia, Junc: junctional, VVIR: single chamber ventricular rate responsive pacemaker, DDDR: dual chamber rate responsive pacemaker, VVI: single chamber ventricular pacemaker, AAIR: single chamber rate responsive atrial pacemaker, Ep: epicardial, AS: aortic stenosis, Snd. sinus node dysfunction, AAI: single.

median 34). All symptomatic patients noted a resolution of symptoms after pacing had been performed, and they remained free of symptoms at the latest follow-up examination.

Twenty-three patients received the following transvenous pacing system: ventricular demand (VVI, VVIR) 12 patients (46%); atrial demand (AAIR) 8 patients (30%); and dual chamber (DDDR) 3 patients (11%) (Table I). Eight of 11 atrial leads (72%) and six of fifteen ventricular leads (40%) had screw-in mechanism. The remaining leads had tined fixation mechanism. There was no malfunction of leads.

Five patients initially received ventricular demand epicardial system. Two of them had high myocardial stimulation threshold and high lead impedance that necessitated replacement via transvenous approach. The remaining epicardial systems were good.

There was no atrial sensing or capture problem in atrial and dual chamber pacing systems. Atrioventricular synchronization was good in both groups.

Mean acute pacing thresholds during implantation and mean latest pacing thresholds for the endocardial atrial leads were 1.17 ± 0.45 V and 1.65 ± 0.75 V, respectively ($p > 0.05$). Mean acute pacing thresholds and mean latest pacing thresholds for the endocardial ventricular leads were 0.98 ± 0.45 V and 0.97 ± 1.3 V, respectively ($p > 0.05$). Mean acute impedance and mean latest impedance for the endocardial atrial leads were 560 ± 177 Ohm and 591 ± 165 Ohm, respectively ($p > 0.05$). Mean acute impedance and mean latest impedance for the endocardial ventricular leads were 552 ± 178 Ohm and 827 ± 937 Ohm, respectively ($p > 0.05$). Mean acute p wave voltage was 1.9 ± 1.3 mV and the latest p wave voltage was 1.2 ± 0.6 mV ($p > 0.05$) (Table II).

Table II. Acute and Latest Pacemaker Measurement with Telemetry

	Implantation	Latest
Threshold		
Atrial leads	1.17 ± 0.45 V	1.65 ± 0.75 V
Ventricular leads	0.98 ± 0.45 V	0.97 ± 1.3 V
Impedance		
Atrial leads	560 ± 177 Ohm	591 ± 165 Ohm
Ventricular	552 ± 178 Ohm	827 ± 937 Ohm
P wave voltage	1.9 ± 1.3 mV	1.2 ± 0.6 mV

All but two of our patients were alive and asymptomatic. The deaths were not thought to be pacemaker related. The first patient died suddenly at home. He had a transvenous pacemaker system after operation for atrial septal defect. The second patient was operated for TGA and epicardial pacemaker system was removed four years after operation. He had taken quinidine and digoxin for atrial fibrillation with rapid ventricular response. The cause of death could have been either medication or life-threatening dysrhythmias.

Discussion

Sinus node disease has many names, all of which describe the same set of syndromes. Sinoatrial node disease is probably the most accurate, whereas sick sinus syndrome is possibly the most memorable. This condition is defined as an affliction of the sinoatrial node that either prevents impulse generation or prevents or delays the conduction of sinoatrial impulses to the surrounding atrial tissue. This affliction may be a pathologic process in or around the sinoatrial node, or it may be a pathophysiologic phenomenon of abnormal function of the autonomic nervous system that adversely influences impulse generation within the node or conduction out of it³.

The clinical manifestations of SND are related directly to age, the function of the remaining conduction system and the underlying hemodynamic state. Poor feeding, lethargy, or signs of congestive heart failure may be associated with severe bradycardia in infants. In older children, bradycardia may manifest as general fatigue, the inability to maintain the same level of activity as peers, or increased sleep requirement with or without change in activity. Fatigue was the most common sign in our patients. Syncope and palpitation were found in eight patients. Dizziness and syncope are difficult to detect in infants and young children. Also, it is imperative to evaluate the patient's rhythm when a child presents with unexplained seizures⁴.

Although several types of classification of SND have been offered, the causes of SND in children are best classified as either nonsurgical or surgical⁵. When no other cause is found, SND is named as idiopathic. Thirteen of our patients were idiopathic. Five of 13 patients had associated cardiac disease not strongly related

with sick sinus syndrome. Familial occurrences have been reported but are probably uncommon⁶. Two of our patients were siblings. Acquired or familial myocardial diseases such as cardiomyopathies and inflammatory or ischemic diseases encompass a wide range of possible causes of SND⁴. One of our patients had dilated cardiomyopathy and another had viral myocarditis. Medications, particularly antiarrhythmic drugs, are an important cause of SND in children. Two of our patients with brady-tachycardia syndrome received antiarrhythmic medication.

Sinus node dysfunction occurs after several types of surgical procedure for congenital heart disease. Pathologic and electrophysiologic correlations have revealed that incisions, sutures, and progressive fibrosis in the area of the sinus node and sinus node artery are definite causes of SND in children who have undergone surgery for congenital heart disease. But, SND has also been found before operation in patients with congenital heart disease. The incidence is highest in patients who have undergone atrial repair (Mustard or Senning) for transposition of the great arteries⁷. Five of our patients who had undergone atrial repair (Mustard or Senning) for transposition of the great arteries received permanent pacemaker. Although patients with secundum atrial septal defect may have pre-existing SND, postoperative SND may also be found⁸. Five of our patients with secundum atrial septal defect had postoperative SND.

The true incidence of sudden death relative to SND is unknown. The problem of documentation relates to the distinct possibility of other life-threatening arrhythmias in the same patient. Flinn et al.⁹ showed that the incidence of sudden death was 2.5% in 372 patients who had undergone Mustard repair for TGA; many of them had SND. Gelatt et al.¹⁰ reported that sinus rhythm was present in 77% at five years and in 40% at 20 years from records of 534 children who underwent the Mustard operation. Kirjavainen et al.¹¹ showed that the probability of staying in sinus rhythm was 34% in patients with simple TGA and 7% in patients with complex TGA after Senning operation.

The diagnosis of SND can be suspected on the basis of a careful history and examination of the ECG. Yabek et al.¹² reported 74% of 30

children with various underlying heart diseases who were asymptomatic from SND. Thus, neither the history alone nor the ECG findings is reliable in making an accurate diagnosis of SND. In recent years, several non-invasive and invasive tests have been described in the evaluation of patients with suspected SND. A 24-hour ambulatory Holter monitor remains an important test in the diagnosis of suspected SND¹³. Transtelephonic recorders that are carried or worn by the patient are most useful in patients with intermittent symptoms, especially if the symptoms are not associated with abrupt syncope or if they occur with a brief prodrome. Electrophysiologic tests were not performed for the evaluation of sinus node function in all patients¹⁴.

Bradycardia-tachycardia syndrome (sinus bradycardia alternating with atrial flutter or reentrant atrial tachycardia) is an increasingly frequent problem in young patients following surgery for congenital heart disease. It is clear that long-term drug therapy deemed essential for the control of atrial flutter may result in symptomatic bradycardia in some patients, whereas in others the use of antiarrhythmic agents may potentially increase the risk of ventricular arrhythmias or sudden death in the presence of profound bradycardia. Thus, in young patients with recurrent arrhythmias associated with the bradycardia-tachycardia syndrome, permanent pacing should be considered as an adjunctive form of therapy¹⁵. Four of our patients who had bradycardia-tachycardia syndrome are symptom-free at follow-up after permanent pacemaker therapy. Two of four patients also received antiarrhythmic medication.

Sinus node dysfunction is not itself an indication for pacemaker implantation. Symptomatic bradycardia is considered an indication for pacemaker implantation, provided that other causes of the symptoms have been excluded. Alternative causes to be considered include seizures, breath holding, apnea, or neurally mediated mechanisms⁴.

Single-chamber atrial pacemakers, with rate-responsive capability if appropriate, have been advocated for patients with SND but no evidence of atrioventricular (AV) block. Available data suggest that the incidence of atrial fibrillation in patients receiving atrial or

dual-chamber pacemakers may be lower than in patients receiving ventricular pacemakers¹⁶. Some studies showed a lower mortality in atrial-based pacemaker patients and others showed no significant difference. Short-term crossover studies in patients with SND have shown improved quality of life in dual-chamber versus ventricular pacing¹⁷. Most patients at our hospital who required pacemakers received ventricular demand epicardial or transvenous systems in previous years. When the screw-in leads were developed, we performed atrial or dual-chamber demand transvenous systems with rate-responsive capability.

In conclusion, SND has been diagnosed with increasing frequency in children with congenital heart defect, especially in patients who have undergone corrective cardiac surgery related with atrial tissue. The incidence is highest in patients who have undergone Mustard or Senning procedure for TGA. There is also a significant number of patients who are considered idiopathic because of no associated cardiovascular disease. The diagnosis of SND is difficult because of documentation. When pacemaker therapy is indicated, atrial demand endocardial systems are preferred.

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