MEDICAL PRACTICE

Hospital Topics

Sick Sinus Syndrome: Experience of a Cardiac Pacemaker Clinic

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Summary

Out of a pacemaker clinic population of 182 patients 21 (11.5%) were found to have the sick sinus syndrome. Their ages ranged from 30 to 80 years and averaged 62. Ischaemic heart disease was more commonly an aetiological factor than in patients with chronic atrioventricular heart block. Other aetiologies were familial cardiomyopathy, postcardiac surgery, and dystrophia myotonica.

Cardioversion before pacemaker insertion was a hazardous procedure. After insertion the occurrence of tachycardias and the need for drug therapy were reduced. One patient no longer required a pacemaker once atrial fibrillation became established.

A high incidence of cerebral embolization was observed and the use of anticoagulant drugs therefore merits serious consideration.

Failure of inhibition of demand-type pacemakers occurred in two patients. Two patients who exhibited competition with fixed-rate pacemakers died. Two patients were treated with electrodes surgically implanted on the right atrium. It is suggested that fixed-rate pacemakers may be contraindicated and that sequential atrioventricular demand pacing is theoretically ideal.

Introduction

Growing attention is being focused on the so-called "sick sinus syndrome," a term originally used by Lown,¹ who observed it immediately after cardioversion. He defined it as a defect in elaboration or conduction of sinus impulses characterized by chaotic atrial activity, changing P-wave contour, and bradycardia interspersed with multiple and recurrent ectopic beats and runs of atrial and nodal tachycardia.

The entity has been known for many years. Laslett² in 1909 described the case of a young woman who had fainting attacks associated with the cessation of atrial as well as ventricular activity and Levine³ in 1916 reviewed the literature on sinoatrial heart block and added four cases of his own, one of which was associated with paroxysms of atrial fibrillation. He quoted a similar case described by Schott⁴ in the German literature. This syndrome of alternating bradycardia and tachycardia was redescribed by Short⁵ in 1954.

It has become evident that these conditions are more common than was initially realized and can be associated with significant symptoms. Of importance is sinoatrial syncope, which was reviewed by Easley and Goldstein.⁶ These workers also classified two types of sick sinus syndrome, type I associated with sinus bradycardia and sinoatrial block, and type II with these features together with episodes of tachycardia. With the greater awareness of the syndrome and the availability of modern E.C.G. monitoring equipment for diagnosis a number of detailed descriptive series and reviews have been published.⁷⁻¹⁰

Therapy for the symptoms resulting from the sick sinus syndrome can be difficult since drugs to control the tachyarrhythmias aggravate the bradyarrhythmias. Cardiac pacemakers have been used for the condition since 1966.^{6 11-16}

Theoretically they have overcome the problem of severe bradycardia, leaving only the associated tachycardia to be treated pharmacologically. The solution to the therapeutic

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problem, however, is not entirely simple and additional difficulties arise.

For this reason we review here our experience with the use of permanent implanted cardiac pacemakers in patients with the sick sinus syndrome who have been treated at the Royal Infirmary of Edinburgh.

Methods

The case notes and E.C.G.s of patients with permanent cardiac pacemakers were reviewed to determine the initial cardiac rhythms, the indications for the pacemaker insertions, and the subsequent problems. Patients fulfilling the criteria for the sick sinus syndrome as defined by Lown¹ were studied in detail. For uniformity a postal questionnaire was sent to these patients asking for their observations about their symptoms before and after pacemaker insertion.

A total of 182 patients were reviewed. These included 139 live patients who had their pacemaker inserted between 1966 and January 1974 and 43 pacemaker patients who died between January 1971 and January 1974. We could not trace accurate details of patients who died before 1971.

Altogether 21 patients (11.5%) were found to have the sick sinus syndrome. Not all patients diagnosed as having this syndrome are treated by cardiac pacing at this hospital.

Results

Age and Sex.—Of the 21 patients with the sick sinus syndrome 11 were men and 10 were women. Their average age at the time of pacemaker implantation was 61.8 years and the range was 30 to 80 years. Of the remaining 161 patients 90 were men and 71 were women. Their average age at the time of implantation was 69.2 years.

Mortality.—Six of the 21 patients (28.6%) died. Death occurred one to 19 months after implantation and the average was 5.3 months. The cause of death was myocardial infarction in two patients, cerebrovascular accidents in two, uncontrollable ventricular arrhythmias in one, and acute hepatocellular necrosis attributed to two halothane anaesthetics in one.

Duration of Pacemaker Therapy.—In 14 of the 15 live patients the duration of pacemaker therapy at January 1974 ranged from four to 60 months and averaged 22 months. The other patient had her pacemaker system removed after 19 months because of failure to pace and pressure necrosis of the skin over the box. It was then considered unnecessary to continue pacing as she was in established atrial fibrillation with a good ventricular rate. She subsequently remained asymptomatic.

E.C.G. Features.—Three patients had the type I sick sinus syndrome and 18 the type II (Easley and Goldstein's⁶ classification). The E.C.G. features documented specifically are shown in table I.

Presentation.—Most of the patients presented with multiple symptoms, the most prominent ones being syncope and dizziness (18 patients), dizziness without syncope (2), palpitations (15), dyspnoea (13), chest pain (9), and lassitude (3).

TABLE I—E.C.G. Features Documented in 21 Patients with Sick Sinus Syndrome

	No. (%) of Patients		No. (%) of Patients
Sinus bradycardia	16 (76)	First-degree heart block	6 (29)
Sinus arrest	12 (57)	Second-degree heart block	2 (10)
Junctional bradycardia	12 (57)	Third-degree heart block	1 (5)
Wandering atrial pacemaker	9 (43)	Right bundle-branch block	2 (10)
Supraventricular ectopic beats	9 (43)	Left bundle-branch block	4 (19)
Atrial flutter	10 (48)	Intraventricular conduction defect with marked left axis deviation	1 (5)
Supraventricular tachycardia	10 (48)	Myocardial infarction	5 (24)
Atrial fibrillation	9 (43)	Left ventricular hypertrophy	2 (10)
Ventricular tachycardia	8 (38)	Right ventricular hypertrophy	1 (5)

Associated Illnesses.—Some patients had associated clinical conditions which may be of aetiological significance. Five (24%) had previously had documented myocardial infarction and two had rheumatic mitral valve disease. Three patients were siblings and their illness was considered to be a familial cardiomyopathy. One man had previously had a pericardiectomy for tuberculous calcific pericarditis together with a repair of a coincidental atrial septal defect. Another had dystrophia myotonica. Two patients were diabetic and four had hypertension. Another five had no associated medical or surgical conditions. Ten patients (48%) suffered cerebrovascular events. Four, in whom the episodes were considered to be embolic, were left with residual neurological defects; three of these events occurred before pacemaker insertion and one occurred afterwards. One followed a bout of atrial fibrillation. Two of these patients had an associated disease which might have been implicated; one had mitral valve disease and one polyarteritis nodosa. Another four patients had transient cerebral ischaemic episodes with observed neurological signs but no permanent damage, only one of which occurred after pacemaker insertion.

Drug Therapy.—A summary of the drugs given before and after the insertion of a permanent pacemaker is given in table II.

TABLE II—Drug Therapy in Sick Sinus Syndrome Patients before and after Insertion of Permanent Pacemaker

	No. Given Drug before Insertion	No. Given Drug after Insertion
Isoprenaline compounds	9	
Ephedrine	4	
Atropine	11	
Propantheline	5	
Lignocaine	4	
Bretylium	1	
Verapamil	1	
Edrophonium	1	
Procainamide	4	
Quindine	2	4
Beta-blocking drugs	8	7
Digoxin	11	9
Diuretics	8	6
Anticoagulants	5	2
Anticonvulsants	5	2
Antihypertensives	3	2
Hypoglycaemics	2	2
Steroids	3	2
Antituberculosis agents	1	1
Thyroxine	1	1
No cardiac drugs		3

Cardioversion.—Six patients were treated by cardioversion at some stage in their management. One had a total of 14 cardioversions in four years. Bradycardia at times resulted but not of serious degree. A second patient who had two cardioversions also had resultant, untreated bradycardia. A third patient had three electrical conversions, one of which was followed by nodal bradycardia, which was treated with intravenous atropine. A fourth man was slow to recover from a countershock after cardiac surgery, immediately after which he described "absence episodes." The other two patients experienced serious effects; after their initial cardioversion for a tachyarrhythmia they had such severe bradycardia associated with hypotension and poor cardiac output that immediate temporary cardiac pacing had to be established. After the insertion of a permanent pacemaker one of these patients again required treatment by electrical means as this was the only successful therapy for his ventricular tachycardias. One such shock produced ventricular fibrillation which reverted with a further shock. Ventricular arrhythmias were the eventual cause of this patient's death.

Arrhythmias after Pacemaker Insertion.—The 13 live patients with the type II sick sinus syndrome were sent a questionnaire asking about their symptoms. Before pacemaker insertion 11 were aware of palpitations and afterwards seven no longer experienced them. Of those who continued to notice palpitations all said that they occurred less often. All

the patients considered that their pacemakers had helped them.

Types of Pacemaker.—Six types of pacemaker had been implanted in the 21 patients—Devices demand, endocardial (15), Medtronic demand, endocardial (1), Stanicor demand, endocardial (1), Devices fixed rate, endocardial (1), Devices fixed rate, epicardial (1), and Devices fixed rate, atrial-epicardial (2).

Pacemaker Complications.—The complications noted with the pacemakers are listed in table III.

TABLE III—Complications of Pacemaker Therapy noted in the 21 Cases

	No. of Cases
Failure of pacemaker inhibition (on demand units)	2
Pacemaker competition with patient's rhythm	2
Pacemaker endocardial electrode displacement	3 (2 in 1 patient)
Electrode fracture	1
"Exit block" with failure to capture	1
High fixed-rate firing immediately after insertion	1
Diaphragmatic stimulation by pacemaker impulse	1
Generation-box extrusion through skin	1
Infection { Local at site of box	1
{ Systemic (with staphylococci)	1
Acute hepatocellular jaundice attributed to halothane anaesthetic	1
Axillary vein thrombosis	1
No complications	10 (48%)

Discussion

INCIDENCE

The incidence of the sick sinus syndrome in the general population is not known. Vohra *et al.*¹⁷ noted that 6-7% of pacemakers are implanted for this condition. Conde *et al.*¹⁸ found 31 such patients in a clinic population of 500, giving an incidence of 6.2%. Our incidence of the sick sinus syndrome within the pacemaker clinic population was 11.5%. This obviously depends on the aggressiveness of the pacemaker policy. At present in the south-eastern region of Scotland 38 per million of the population have permanent cardiac pacemakers implanted every year.

AGE

Our patients were younger than those analysed in recent large series. Of the 52 patients with sinoatrial rhythm disorders treated by cardiac pacing and reviewed by Chokshi *et al.*¹⁹ most were in their eighth decade, the age range being 54 to 89 years. In the series of Conde *et al.*¹⁸ the patients had an average age of 71 years and a range of 55 to 86 years. Fruehan *et al.*¹⁵ reported on a group with an average age of 75 years. In our series six patients (29%) were aged 55 years or less at the time of the pacemaker implantation. The average age was 62 years.

AETIOLOGY

Multiple aetiologies for the sick sinus syndrome have been cited. As with chronic complete heart block many patients have no associated illnesses and a degenerative process is implicated. It is thought, however, that the syndrome is more commonly associated with ischaemic heart disease than is chronic atrioventricular block.²⁰ This appears to be the case in the present series, in which previous definite myocardial infarction had been documented in five patients with the sick sinus syndrome (24%) compared with 14 out of 104 patients (13.5%) who had pacemakers inserted during 1971 to 1973 for complete heart block.²¹ Precise correlations have previously been made in patients with recent myocardial infarction having sinus arrest, junctional rhythms, and syncope and subsequently being shown at necropsy to have involvement of the right coronary artery and artery to the sinus node with infarction of this node.²²

Three siblings are included in the present series and have been described in greater detail elsewhere.²³ Familial cases have been documented previously.^{24 25}

There are other reports of the sick sinus syndrome developing after repair of an atrial septal defect.^{14 26} One of our patients had an atrial septal defect repaired together with a pericardiectomy for calcified tuberculous pericarditis. The latter procedure could easily be associated with damage to the artery to the sinus node. Histological examples of surgical injury to the sinoatrial node were described by Hudson.²⁷

Cardiomyopathy and atrial arrhythmias are recognized in dystrophia myotonica.²⁸ A high proportion of E.C.G. abnormalities have been found in this condition²⁹ and conduction disturbances confirmed by His-bundle studies.³⁰ Patients who almost certainly had the sick sinus syndrome have previously been documented.^{29 31-34}

No patient had thyrotoxicosis, which in the series of Wan *et al.*³⁵ topped the list of aetiological factors. Though many of our patients were treated with digoxin at various times, in none could drug-induced arrhythmias be implicated as the sole factor.

TREATMENT

The extensive range of drugs used in our patients gives an indication of the difficulties experienced in the control of arrhythmias. This is not unexpected as more studies throw light on the nature of the condition; a poor response of the heart rate to the trial use of atropine and isoprenaline is a stress test of use in establishing the diagnosis.¹⁷ After pacemaker insertion drug therapy was reduced but most patients required at least one pharmacological agent.

Our incidence of symptomatic tachyarrhythmias after pacemaker insertion was reduced. Cheng,¹² Zipes *et al.*,¹⁴ and Rokseth *et al.*¹⁶ described similar improvement with pacing but Rubenstein *et al.*⁷ found the degree of prevention of tachyarrhythmias disappointing. Cohen *et al.*³⁶ suggested that a mechanism of retrograde ventricular to atrial conduction was involved in the prevention of supraventricular tachycardias when ventricular pacing was established.

In our experience cardioversion in six patients was a rather dangerous procedure. In two patients the ensuing bradycardia was the initial indication of the diagnosis. This was also the experience of Lown,¹ who made the original description. Rubenstein *et al.*⁷ generally avoided the procedure but experienced no problems on five occasions when it was used.

Hazards in the investigative procedures of cardiac catheterization and coronary angiography have also been described.¹⁰ We have no details of problems experienced with preoperative investigations in our two patients who had cardiac surgery as both assessments were done elsewhere.

One of our patients had her pacemaker removed after 19 months. She had previously had sinus bradycardia, sinus arrest, and episodic atrial tachycardias with symptoms of syncope, dizziness, and palpitations. When the pacemaker was removed her cardiac rhythm was infixed atrial fibrillation with an adequate ventricular rate. She remained in this rhythm for four years and there was no recurrence of her symptoms. Two similar cases were recorded by Stock.³⁷ As the natural history of the sick sinus syndrome is still being determined this is an important observation. Pacemakers may not be indicated permanently.

COMPLICATIONS

An impressive incidence of cerebrovascular events of a probable embolic nature was observed in this clinic. Ten patients (48%) suffered such episodes, which were well docu-

mented. Most occurred before pacemaker implantation though one patient developed aphasia and repeated transient cerebral ischaemic attacks after the insertion. Rapidly changing rhythms are implicated in producing emboli. A significant incidence of cerebral embolization (14.3%) was also the experience of Rubenstein *et al.*⁷ Thus we cannot agree with Eraut and Shaw,³⁸ who advised against the widespread use of anticoagulant therapy from their experience of the absence of embolization in 46 patients with the "lazy sinus syndrome." With the serious complications we have observed we now consider the use of anticoagulants when the diagnosis of sick sinus syndrome is made.

The most frequent pacemaker complication was electrode displacement, which is common to all pacemaker insertions. Other features evolved in the sick sinus group, however, which are worthy of note but may not be specific because of small numbers.

Sutton³⁹ described features of inhibition of demand pacemakers due to endocardial voltages which were too low to be sensed by the demand mechanism. This has been observed particularly in acute myocardial infarction, in acute pulmonary embolism, and after cardiac surgery. The phenomenon was noted in two of our sick sinus syndrome patients, neither of whom had those described clinical conditions at the time though one died one month later reportedly of myocardial infarction. The other patient had dystrophia myotonica and was alive and coping adequately. A total of seven of the whole pacemaker clinic population of 182 patients exhibited the phenomenon,²¹ but the other five two had had recent cardiac surgery, one developed the feature terminally with pulmonary embolism, and one had a faulty pacemaker; in only one case was there no previous explanation.

Competition of the pacemaker stimulus with a patient's own heart beat is an occasional feature when a fixed-rate pacemaker unit is implanted in a patient with apparently established complete heart block but who subsequently reverts to episodes of normal sinus rhythm. The generally accepted view of this phenomenon is that with standard pacemakers and a normal myocardium without ischaemia, hypoxia, or postsurgical inflammatory reactions and unassociated with the use of sympathomimetic drugs or anaesthetic agents there is no risk of pacemaker-induced ventricular fibrillation.⁴⁰

We prescribe practolol in such cases to reduce the patient's own heart rate.

PACEMAKER TYPES

Two of our sick sinus syndrome patients were given fixed-rate pacemakers. One had complete heart block associated with the atrial arrhythmias, and the other, though originally given a demand unit, was unintentionally changed to a fixed-rate pulse generator when she underwent thoracotomy for the placement of epicardial wires after recurrent endocardial electrode displacement. Both patients exhibited competition and both died. One died one month after insertion, reportedly of myocardial infarction, and the other died five months after insertion from a cerebrovascular accident. Fixed-rate pacemakers may be contraindicated in the sick sinus syndrome.

A high proportion of patients exhibited ventricular tachycardia in the present series (38%), and this ventricular

irritability may be a factor of importance for intolerance of pacemaker competition.

Two of our patients were treated with surgically implanted electrodes sutured to the right atrium, a technique originally described by Silverman *et al.*⁴¹ It gives the added advantage of useful atrial contraction which improves the cardiac output.⁴² It does, however, entail a thoracotomy. Sowton⁴³ observed that patients with the sick sinus syndrome are usually sensitive to the effect of atrial contraction, and ventricular pacing alone produces low cardiac outputs with poor exercise tolerance. Symptomatic sinoatrial disease is commonly associated with atrioventricular conduction disturbances.^{9, 44} Because of these observations the theoretically ideal method of pacing is with a sequential atrioventricular demand system. This method has been used effectively.^{45, 46}

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