Chronic Heart Block and Sinoatrial Disorder

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More than 90 per cent of pacemakers are implanted because of bradycardia associated with heart block or sinoatrial disorder[1]. Sinoatrial disorder is currently the major indication for pacing in the USA[2], and is the indication for an increasing proportion of new implants in other countries, including the UK. Since it was first popularised by Ferrer[3] in 1968 under the title of 'sick sinus syndrome', sinoatrial disorder has been the subject of many papers describing the clinical and electrophysiological features. For want of data on the pathology and natural history of the condition it has been treated as being analogous to heart block. Studies published during the last three years imply that this approach is incorrect.

Definition

The definitions of the various degrees of heart block are generally known and accepted. Unfortunately, this clarity does not apply to sinoatrial disorder, since there is a considerable overlap between the physiological bradycardia seen in athletes training for long distance events and that occurring in patients with pathologically disturbed sinoatrial function. Ironically, ambulatory electrocardiograph monitoring (which it was initially hoped would provide an easy way of diagnosing sinoatrial disorder) has complicated the problem by showing that profound bradycardia and sometimes even sinus arrest may occur in apparently normal young people[4]. In practice, the difficulties are not as great as might be imagined because symptomatic patients suspected of having sinoatrial disorder are usually middle-aged or elderly and rarely in serious training for sports.

A working classification of sinoatrial disorder follows.

Established sinoatrial disorder. A chronic sinus rate below 50 with one or more of the following:

1. Sinus pauses of two seconds or more (sinus arrest or sinoatrial block).
2. Profound bradycardia with an atrial rate below 40, usually associated with junctional rhythm.
3. Paroxysmal supra-ventricular or ventricular tachycardia, such as atrial fibrillation, atrial flutter, or atrial or ventricular tachycardia.

Potential sinoatrial disorder. A chronic unexplained sinus bradycardia in the absence of any of the factors above.

Aetiology

In at least one American hospital[5] drugs contribute to bradycardia in as many as 40 per cent of candidates for pacemakers. This figure may not be generally applicable to the UK, but iatrogenic bradycardia is not rare and in 22 per cent of cases referred to the Devon Heart Block and Bradycardia Survey the bradycardia was considered to be drug-induced.

The disease process most often blamed for bradycardia was coronary artery disease. However, in 1963 Zoob and Shirley Smith[6] reported that only 12 of 51 patients with complete heart block had evidence of ischaemic heart disease. Subsequently, coronary care monitoring confirmed that conduction disturbances were common within a few hours or days of infarction, but, in survivors, conduction usually returned to normal. Sinus function is also often disturbed soon after myocardial infarction involving the inferior heart wall, and usually returns to normal even more quickly than heart block. As with chronic heart block, the blame for chronic sinoatrial disorder has, until very recently, been attributed to vascular disease[7,8]. However, this view is not supported by recent epidemiological and pathological evidence. Table 1 gives data collected from the Devon Heart Block and Bradycardia Survey, which was started in 1968 and involved a direct approach to the GPs (approximately 300) in East and North Devon who looked after a population of from half to three quarters of a million people[9,10]. Only 8 per cent of 436 patients with complete heart block and 16 per cent of 131 patients with sinoatrial disorder gave a past history of myocardial infarction.

Pathological studies have confirmed the clinical impression that chronic bradycardia is not usually the result of cardiac ischaemia. Harris *et al.*[11] studied the heart and conducting system of 65 cases of complete block and found significant coronary artery disease in only ten. Table 2 summarises the result of a recent pathological

Table 1. Possible aetiological factors in past history of patients with bradycardia.

Past History	Complete block %	Chronic sinoatrial disorder %	Potential sinoatrial disorder %
Cardiac infarction	8	16	16
Rheumatic fever	9	11	10
Diphtheria	8	15	9
Diabetes	5	2	2
Thyroid disease	2	6	5
None of these	71	58	67
Total no. in group	436	131	305

Table 2. Pathological findings in 200 cases of chronic heart block collected by Davies at St George's Hospital[12].

	No. of cases	%
Idiopathic bilateral bundle branch fibrosis	76	38.0
Calcific A-V block	22	11.0
Ischaemic (coronary artery disease)	35	17.5
Congestive cardiomyopathy	26	13.0
Other causes	41	20.5
Total	200	100.0

study of 200 cases[12]. In approximately half the cases the abnormalities were confined to the specialised conducting myocardial cells. The most common abnormality was idiopathic bundle branch fibrosis, a condition initially described by Lenègre[13], which was present in almost 40 per cent of the series. In a further 11 per cent the bundle of His was obliterated by calcification extending from the aortic or mitral valve rings[14,15].

Pathological studies of specialised tissue in sinoatrial disorder have been scarce but recently four small series have been reported[16-19]. In none of these studies was major coronary artery disease shown to be an important aetiological factor. The most common pathological finding was an absolute reduction in the number of pacemaker cells in the sinoatrial node. In some instances the node was atrophic or hypoplastic, while in others the pacemaker cells were replaced by fibrous tissue. In some instances a relatively normal node appeared to be isolated from the myocardium of the right atrium by adipose tissue or the deposition of amyloid. In a recent report[20] on two adolescents the atrial preferential pathways were found to be affected by fibrosis and fatty infiltration.

Natural History and Treatment

Except for drug-induced bradycardia, the causes of chronic heart block or sinoatrial disorder are very seldom reversible. Treatment, therefore, must be palliative and aimed at the particular problem the condition presents in the individual patient.

Symptoms

Blackouts (Stokes-Adams attacks) are the most common and troublesome symptoms associated with both heart block and sinoatrial disorder. The disturbance of consciousness is usually the result of a prolonged pause in ventricular activity, caused by either complete heart block with failure of the idio-ventricular pacemaker, or prolonged sinus arrest with gross delay in the escape rhythm. Long-term pacemaker therapy is currently the most reliable method of preventing Stokes-Adams attacks. Drugs such as isoprenaline may increase both sinoatrial node and ventricular myocardial automaticity and will usually increase the heart rate in sinoatrial disorder and in heart block. However, they tend to predispose to extrasystoles or more serious arrhythmias such as ventricular tachycardia or fibrillation. Furthermore, if a dose of the drug is missed or delayed, profound and dangerous bradycardia may recur within a few hours. Unfortunately, no current drug therapy can be regarded as a reliable protection against the risk of attacks of ventricular asystole.

Cardiac failure associated with bradycardia usually responds to standard treatment with diuretics. Unless the heart rate is very slow, severe cardiac failure implies significant deterioration in heart muscle or valve function in addition to disease of the specialised cardiac tissues. Pacemaker therapy is also likely to lead to short-term improvement, but the long-term results are often disappointing, as cardiac failure is likely to recur and be fatal.

Breathlessness or occasional chest pain may result from the failure of the heart rate to increase during exercise; sometimes, in second degree block, the ventricular rate may decrease on effort. A standard ventricular pacemaker may help in these instances (particularly if effort precipitates dizziness), but it has two disadvantages: it cannot increase the ventricular rate above its resting level (usually about 70 a minute), and the priming action of the normal atrial contraction is lost. An atrial synchronised pacemaker theoretically overcomes these disadvantages and might be expected to be particularly helpful in this group of patients.

Survival

Heart Block

The vogue for long-term cardiac pacing was given great impetus in the mid-1960s by the very high mortality reported in unpaced patients. It was suggested that only 50 per cent would live beyond the first 12 months[21,22]. Subsequent studies (Fig. 1) have reported a better outlook in unpaced patients[23,24], but, at least in symptomatic cases referred to major hospital centres, complete block grossly shortens life span. Pacing dramatically improves the outlook of these people[25], although survival is not restored to normal in all age groups[26]. There is certainly a good case for considering long-term pacemaker therapy in most patients referred to hospital with chronic complete block. The position is less clear in instances of asymptomatic block found coincidentally by routine

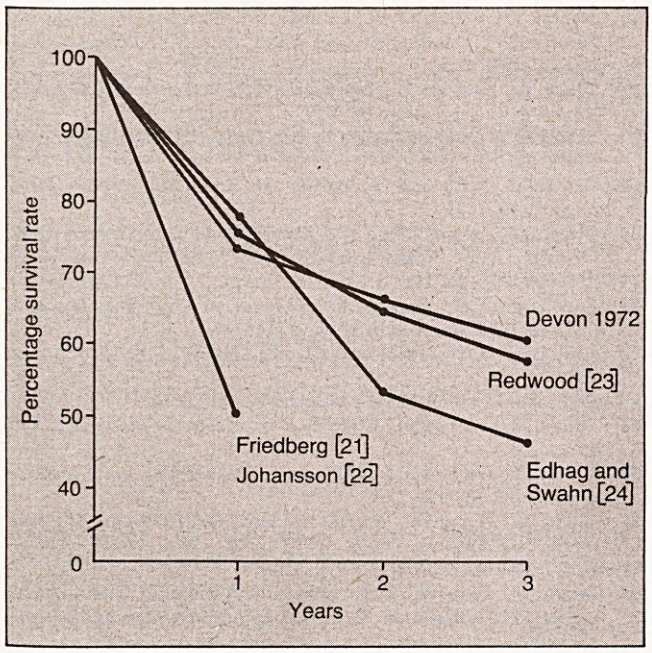

Fig. 1. *The rate of survival in unpaced patients is plotted against the time in years since the diagnosis of complete heart block. Figures in brackets refer to references in the bibliography. (The figures for the Devon survey (1972) are from unpublished data.)*

screening procedures[27]. Second degree block commonly progresses to third degree and then presumably has a similar prognosis. However, on occasion, 2:1 atrioventricular block may persist as an apparently stable rhythm for a number of years. Little data are available on this rather uncommon group.

Controversy continues concerning the correct management of bifascicular block following acute myocardial infarction. It is generally agreed that patients with this condition have an increased mortality when compared with those having an uncomplicated infarction, but it is not clear whether death results from progress of the conduction fault or if long-term pacing can reduce the mortality[28-30]. Symptomless bundle branch block or bifascicular block arising *de novo* are not generally regarded as indications for pacing (Table 3).

Table 3. Heart block: prognosis with and without pacing.

Degree/Type of Block	Prognosis	
	Unpaced	Paced
Third degree	Reduced	Improved
Second degree	Reduced in some cases	Improved in some cases
Bifascicular—after acute myocardial infarction	Reduced	?
Bifascicular—incidental	Normal?	No change?
First degree	Normal in most cases	No change in most cases

Sinoatrial Disorder

Chronic sinoatrial disorder has only gained general recognition as a common entity in the last ten years. Relatively few follow-up studies are available. Initial reports are contradictory; some implied poor prognosis even in paced patients[31,32]; others commented upon the long history in some forms of sinoatrial dysfunction[33-35]. The latter view is supported by very recent studies[36-38] (Fig. 2).

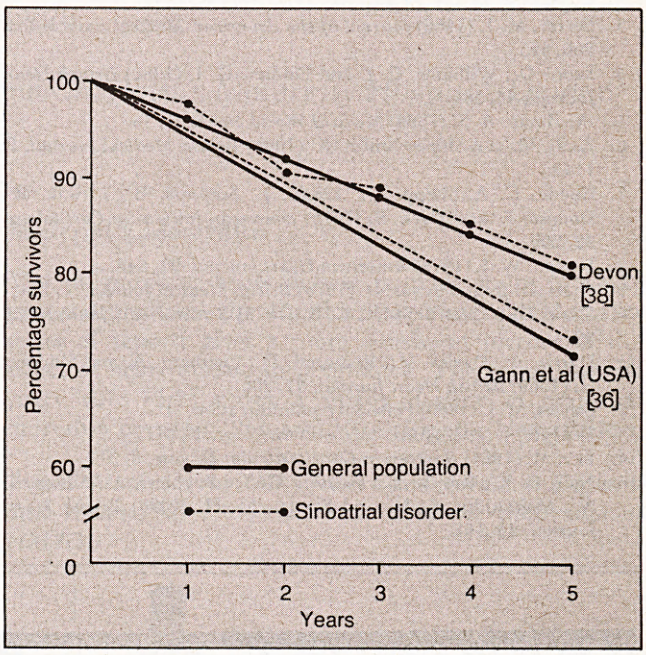

Fig. 2. *The survival curves for patients with sinoatrial disorder are compared with those estimated for normal populations of similar age and sex distribution. (Figures in brackets refer to references in the bibliography.)*

Conclusions

Despite some important recent data on the pathology and natural history of conditions associated with bradycardia, many questions remain unanswered. Autopsy studies show the blood supply to the conducting system and sinoatrial node to be well maintained in most instances, and the disease processes to be primarily located in the specialised cardiac tissues. The cause of these changes remains unknown.

The mortality of patients referred to hospital with complete heart block is abnormally high and the outlook can be improved considerably by pacemaker therapy. However, these findings may not necessarily apply to patients with asymptomatic block found coincidentally.

Current results tend to disprove the poor prognosis previously attached to chronic sinoatrial disorder. Indeed, unless associated with some other serious underlying cardiac disease, it appears to be a relatively benign condition in which survival is unlikely to be prolonged to any major extent by pacemaker implantation. In this condition, then, the main indication for pacemaking should be incapacitating symptoms.

Irrespective of the mechanism producing bradycardia, the most rewarding results of pacemaker therapy are seen in patients with Stokes-Adams attacks.

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