

# SICK SINUS SYNDROME - A PLEA FOR A LONG TERM PROSPECTIVE STUDY

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The Sick Sinus Syndrome (Fig.1), recognized and detailed 10 years ago (1), has become an important part of clinical cardiology (2). It is not a rarity. Physicians throughout the world assert the frequency with which the diagnosis arises at present (3). The problem of documenting its incidence however is a real one since no extensive and numerically large prospective survey is yet available. For example, if one seeks the end stage of the syndrome, where pacing is essential, recent statistics (4) show that there are approximately 400,000 people throughout the world who are living with a pacemaker and nearly 156,000 of these live in the United States - which is 1 in every 1300 Americans. In the U.S. the pacemaker implantation rate is 270 per million and that means 60,000 people a year receive a pacing unit. It is now thought at least half - or perhaps more - of these patients need pacing for a failing SA node and most of the rest for complete heart block. Thus perhaps 30,000 individuals are being diagnosed as having late stage SA node disease. The only prospective incidence attempt comes to us from Devon, England where Shaw and Kekwick (5) surveyed the same community of 600,000 people for 8 years consecutively for the incidence of heart block and sino-atrial disorders. A total of 436 patients with the Sick Sinus Syndrome (SSS) were seen in 8 years (79 or 17 percent of those were paced). The calculated incidence figure of SSS was 108 per million population. Similarly, if one takes the figure of 20 percent to 30 percent for subjects with the Sick Sinus Syndrome who will require a pacemaker, - a reasonable estimate at least in our present state of knowledge - one can extrapolate from this that since the number of those paced annually in the U.S. for a failing SA node is probably around 30,000 per year, there are perhaps 100,000 patients with dysfunction of the primary pacemaker in any one year in the U.S. Thus one realizes that this syndrome represents a sizable problem in cardiovascular disease. However, any such guesses must be moderately inaccurate since they probably do not include pediatric patients (who are being seen more and more often) and cannot hope accurately to measure the whole pool of patients since these move, sometimes very slowly, along the horizontal or time course of this multifaceted disorder. Add to the-

se difficulties in estimation of the total number of sufferers with SSS the fact that the natural history of the disorder in most cases occupies many years. This, of course, makes diagnosis difficult but it also strongly suggests that the deterioration in function of the SA node is not necessarily a rapid one or even a continuous one. Intermittent phases may occur. This introduces the possibility that if one could characterize, in basic cellular fashion, the processes responsible for early changes, chemical or even physiologic interventions could be developed which would interfere with the noxious sequence leading to final and permanent failure of the SA node. It is not inconceivable that the SA nodal tissue transplants could become a feasible consideration in the future. This of course would necessitate even more knowledge of the disorder, which can run a course of 10 to 40 years.

Figure 1

## DIAGNOSTIC FEATURES OF THE SICK SINUS SYNDROME

- A. Depression of the SA Node function, as shown by:
  - 1. Sinus Bradycardia
  - 2. Sinus Exit Block
  - 3. Sinus Arrest - of non-vagal origin.
- B. Cessation of all SA Node Function, as shown by an absence of Sinus Rhythm, with replacement by:
  - 1. Another Supraventricular Rhythm,
  - 2. Usually permanent Atrial Fibrillation (with a slow ventricular rate),
  - 3. Slow Atrial or Junctional Rhythm (55-75/min.),
  - 4. Episodes of Sinus Bradycardia alternating with Atrial Fibrillation or Atrial Flutter with Rapid Ventricular Rates.

All these considerations make clear that the delineation of the course of the SSS needs widespread and further documentation. Multiple *prospective* studies of the syndrome are urgently needed. Obviously the starting point of such investigations would be the culling of massive amounts of data on sinus bradycardia, limiting the level to rates of 50 per minute or less. Sinus bradycardia usually initiates the SSS. The insurance industry is in an ideal position to assist in such a survey since it is already its current custom to label any insurance applicant with a sinus bradycardia of 50 per minute with an M.I.B. code, and such cases could be culled for study. Insurance medicine, through the M.I.B. coding, could maintain files on its applicants, both adult and juvenile, instead of discarding data after seven years. Long-term data is no longer available in hospital populations where much primary information (such as electrocardiograms and chest x-rays) is destroyed after 5 years because of space limitation. As Harnes has pointed out recently (6), a data-base can and should be set up at the M.I.B. Center for Medico-Actuarial Statistics. The data there should not be destroyed after a certain period but should be transferred to a computer storage system and collected under appropriate headings. Support by life insurance companies for such mortality-morbidity studies would be justified not only because of the production of improved actuarial practices, but also because clinical medicine would benefit greatly from such joint research projects. The addition of the other elements of the SSS besides sinus bradycardia, namely sinus arrest and sinus exit block, could be and should be included in any such data base, but these probably appear later in the dysfunctional state and are much more difficult to diagnose. Intraatrial block, as defined by an abnormally wide P wave and identified by an M.I.B. code (a), is another relatively easy datum to insert into a prospective data base. It adds great weight to the possibility that sinus bradycardia is of the primary and not the secondary (usually vagotonic or drug effect) type. This point is well illustrated by the history of a 55 year old business man who applied for a large amount of life insurance (a million dollars). This level of insurance required a resting and a exercise electrocardiogram during the insurance evaluation. The man had been athletic all his life and so his slow resting heart rate had been previously accepted by his physician as evidence of good conditioning. His resting rate at his insurance examination varied between 32 and 48/minute and on a double Masters Step Test his heart rate initially reached only 105/minute. This was immediately followed by the frequent appearance of periods of sinus arrest (during the exercise). A review of his previous electrocardiograms going back 10 years demonstrated the resting rates of sinus rhythm were 75-85/minute 10 years before, although he was then exercising daily and was in prime shape. Five years before his present evaluation intraatrial block appeared for the first time and resting rates were then 56 to 62/minute. The wide P wave announced the

atrial disease that was already at work on the SA node, and the mild bradycardia became important only because it was measured against a known and previously more rapid rate. It only became severe bradycardia (32-48/minute) 5 years later. Obviously athletic conditioning was not the cause of his current slow heart rate. Rather he had developed the sick sinus syndrome.

Investigative studies for all insurance applicants with sinus bradycardia, with or without the P wave abnormality of intraatrial block, could include examination of all prior insurance or other medical data and prospective follow-up could also be advised. The mechanisms needed to accumulate such a body of data need not be cumbersome when computer storage and analysis is now available in most insurance companies. At present there is real difficulty for insurance underwriters to pick out bad or malignant sinus bradycardia from the physiologic or good sinus bradycardia. More guidelines are needed. Holter monitoring for 24 hours is a very useful requirement to acquire in such evaluations, since markedly slow sinus rates (18-25/minute) may be seen only during sleep. Stress tests can also be used to determine a blunted rise in heart rate.

Electrophysiologic investigations of SA node dysfunction have not as yet provided the clinician with a definitive diagnostic test for a diseased SA node and it appears that it will be some years before this is achieved, although Doctor James Reiffel (7) and his group at the Columbia-Presbyterian Medical Center have some new and very exciting hopes in this direction. Hence we cannot afford to wait any longer before setting into motion a structured attempt at uncovering the beginnings of a disease process the eventual clinical expression of which is often life threatening, and which dooms the subject to an electronic pacemaker for the rest of his life. It is now known that at least half, or perhaps more, of the patients with electronic pacemakers receive them for SA node failure. The proper moment for the insertion of such pacers remains very hard to select in many cases, since we do not know the speed of development of dangerous dysfunction in any one individual. If one could establish points along an individual curve and hence define its slope, this picture might change. A tragic example of this lack of knowledge of the direction and speed of the dysfunction is the story of a 3 month old baby with periods of sinus arrest, and sluggish SA nodal function checked by electrophysiologic studies, but who had no real symptoms. The child was followed by his pediatrician until age 12 years when he was doing so well his parents refused further care. Sudden death occurred at age 15.

Once a subject with the SSS is paced electrically he or she can no longer be followed as a *pure form of the syndrome* in a prospective study of the untreated SSS. Obviously however, as a subgroup of the paced patients must also be surveyed long-term. The coding of such patients, i.e. those with pacemakers, should, if

possible, also include a code for atrial fibrillation, if present. When this is the rhythm in the atria, while the ventricles are independently paced, the possibility of atrial thrombi and embolization exists. There have been case reports of such emboli in SSS patients and this may be one mechanism for sudden death in the paced patient.

### SUMMARY

Clearly then, there is an important need to acquire data on large numbers of subjects with only sinus bradycardia who are or may be at the beginning of a disease process - SA node dysfunction - which can eventually and quite suddenly, threaten life and which, if the syndrome develops slowly, can have considerable morbidity. Insurance medicine - covering life insurance candidates and health insurance candidates as well - can render a large service to clinical cardiology by undertaking such a survey. In turn the information developed would assist the industry in providing fair insurance coverage for a group of applicants whose numbers is likely to be increasing yearly.

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(a) Footnote: It has been suggested to the M.I.B. Center that the code representing P wave abnormality should be subdivided into (a) for atrial enlargement (tall P waves) and (b) for intraatrial block (wide P waves).

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