MEDICAL GRAND ROUNDS
PARKLAND MEMORIAL HOSPITAL
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SICK SINUS SYNDROME (BRADY-TACHYCARDIA)

Palpation of the pulse as depicted in an illuminated miniature from a medieval Latin translation of Avicenna's writing.

INTRODUCTION

A survey of the patients receiving permanent pacemakers at Parkland Memorial Hospital during the past four years exemplifies the frequency and the magnitude of the sick sinus syndrome in our patient population and prompted this review.

Permanent Pacemakers For Sick Sinus Syndrome

	Number	Per Cent
1970	7	33%
1971	18	41%
1972	14	36%
1973 (2/3)	6	22%

THE SPECTRUM OF HEART BLOCK

Conduction disturbances resulting in heart block may be of sudden or progressive onset and may result in complete or incomplete disruption of conduction through the normal pathways from the sinoatrial (SA) node, through the atrioventricular (AV) node, His bundle, and three fascicles of the ventricular conduction system (right bundle, anterior division of the left bundle, and posterior division of the left bundle). Clinically, these conduction blocks may cause:

1. Bradyarrhythmias resulting in low cardiac output with congestive heart failure, fatigue or specific organ signs of low perfusion as in (a) the central nervous system (CNS), resulting in Adams-Stokes attacks, strokes, or mental confusion; (b) the kidney, resulting in increasing blood urea nitrogen (BUN); or (c) the heart, resulting in angina.

2. Sudden asystole, followed by a fatal arrest or ventricular tachycardia and/or fibrillation. A secondary focus may take over and maintain a cardiac rhythm, but this pacemaker is usually less reliable than the SA node and results in a slower rate (bradyarrhythmias).

The diagnosis of heart block can be made clinically; however, the electrocardiogram gives the definitive diagnosis and is essential prior to institution of therapy. Patients with CNS symptoms suggesting intermittent heart block, such as intermittent dizziness or syncope, may require prolonged electrocardiographic monitoring in a coronary care unit or with a portable battery-operated ECG recorder to verify the presence and type of block.

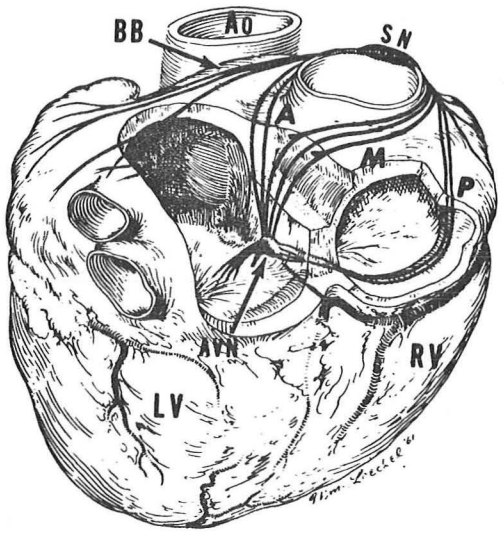
Types of Conduction Block[1]

1. Sinoatrial (SA) block (sinus arrest)
2. Atrioventricular (AV) block
   a. First degree (prolonged P-R interval)
   b. Second degree
      (1) Wenckebach (Mobitz Type I)
      (2) Mobitz Type II
   c. Third degree or complete
3. Ventricular conduction system blocks (fascicular blocks)
   a. Incomplete trifascicular
      (1) Alternating right bundle branch block and left bundle branch block ("bilateral bundle branch block")
      (2) Prolonged P-R interval with left bundle branch block
      (3) Prolonged P-R interval and right bundle branch block and left axis deviation
      (4) Prolonged P-R interval and right bundle branch block and right axis deviation
   b. Complete trifascicular (complete block)

1. Mullins, CB: Heart Block. In: *Current Therapy*, ed. by Howard F. Conn, W. B. Sanders Co., 1972, pp. 159-164.

ATRIAL CONDUCTION SYSTEM

Four specialized atrial pathways have been anatomically and electrophysiologically described which conduct electrical stimuli more rapidly than atrial muscle and propagate the SA impulse throughout the atria and to the AV node. These tracts are composed in part of Purkinje-type cells and provide direct cellular continuity from the sinus node to the AV node.[2]



This drawing depicts the three internodal pathways: anterior (A), middle (M), and posterior (P). Bachman's bundle (BB) contains the major interatrial pathway as well as the initial portion of the anterior internodal pathway. Ao = aorta, LV = left ventricle, RV = right ventricle, sn = sinus node, AVN = AV node.

From James[2]

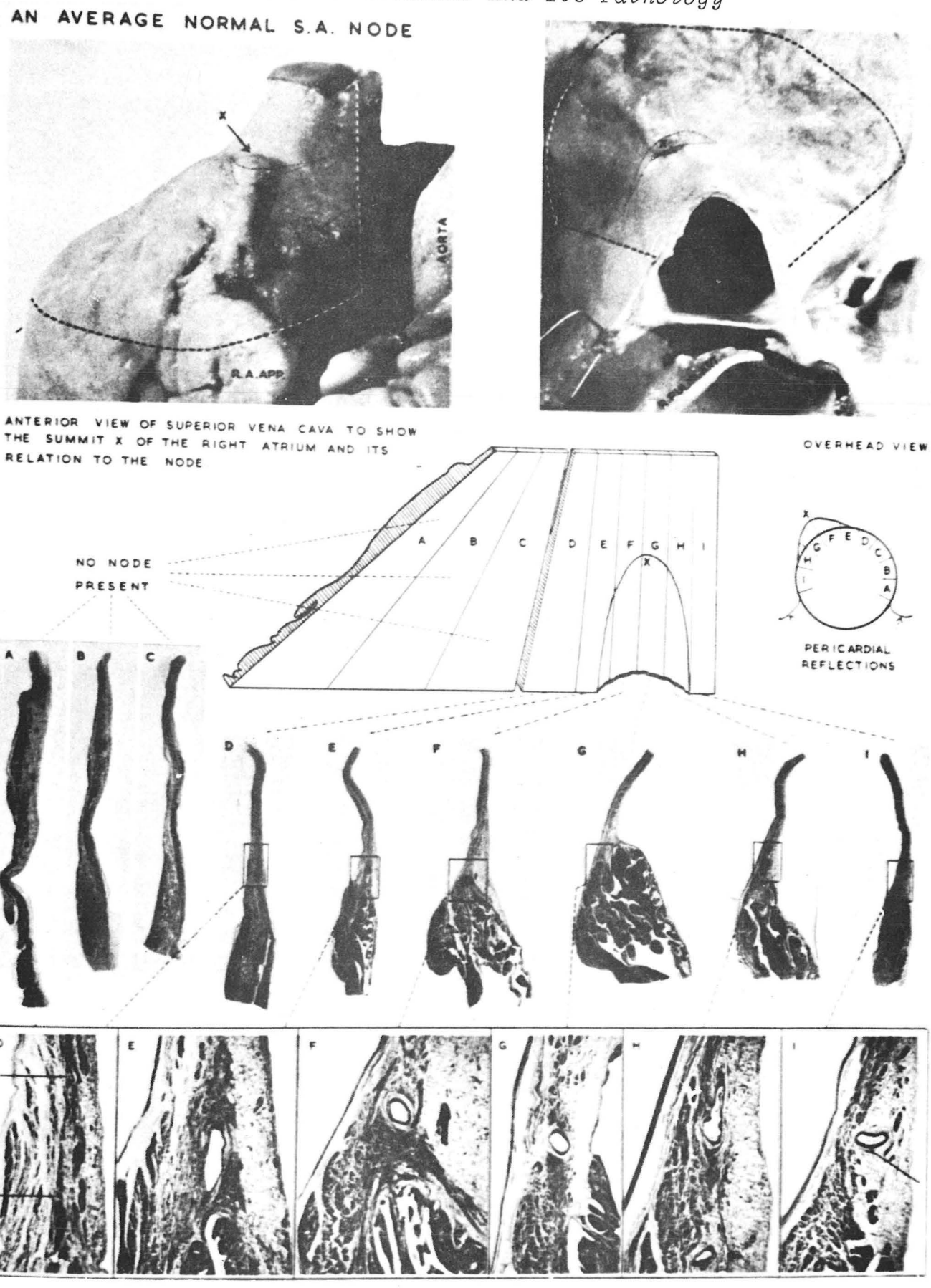
P wave changes may occur due to three possible mechanisms: 1) actual translocation of pacemaker locus; 2) temporary blocks in the atrial preferential pathways, and 3) variable exit sites from the sinus node.

2. James, TN, and Sherf, L: P waves, atrial depolarization, and pacemaking site. In: *Advances in Electrocardiography*, ed. by R.T. Schlant and J.W. Hurst, Grune and Stratton, 1972, pp. 37-59.

PATHOLOGY OF THE SA NODE

If one examines the sinu-atrial junction (where the superior vena cava joins the right atrium) in the human heart, the atrium will be seen to climb forwards on the anterolateral aspects of the vena cava to reach a summit, falling away again towards the medial side.

The Human Pacemaker And Its Pathology[3]



Anatomy and histology of sinu-atrial node. From Hudson[3]

I - Node artery distinct. The arrow indicates a few node cells adjacent to the artery

H - A little nodal tissue mainly below the artery

G - Node now elliptical, with the artery above its centre

F and E - Main body of the node

D - The artery has now divided into branches, two of which are indicated by arrows. Stretched out between the two arteries is a broken line of nodal tissue, hardly distinguishable in the picture from nearby atrial muscle.

The main part of the node lies immediately beneath the epicardium, just below the summit X, more commonly towards its right side than its left. The node usually lies immediately adjacent to the fatty epicardium, with no intervening atrial muscle.

Histology of the Normal Node. The average normal adult node is a crescentic vascular neuromyocardial structure, quite distinctly marked off from the neighboring tissues by being embedded in a variable amount of fibro-elastic tissue. At its largest part it is about three or four times the size of a pin's head. From this part, it tapers medially and laterally to a point.

The *muscle* is striated. The fibers are branching and interwoven so that some appear cut transversely and others at various angles. At the periphery, the fibers run more vertically and they are longer and straighter, often forming an incomplete border to the node. The node muscle fibers are usually a little smaller than those of the right atrium.

Nervous tissue is abundant in the node itself, and in the nearby epicardium there are always numerous parasympathetic ganglia and autonomic nerves to be found. The nerves to the pacemaker are derived mainly from the right vagus and sympathetic, the left vagus and sympathetic serving the atrioventricular node.

The nerve bundles in the epicardium are often closely associated with muscle fibers.

3. Hudson, REB: The human pacemaker and its pathology. Brit. Heart J. 22:153, 1960.

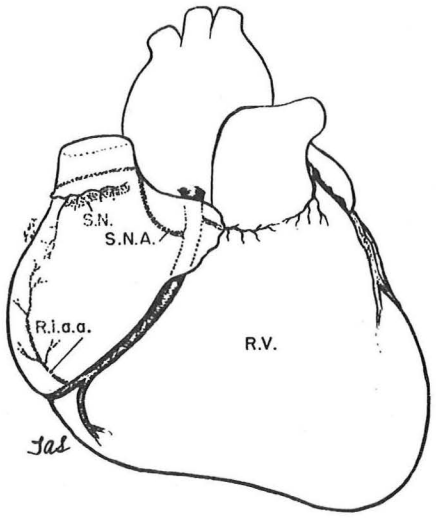
SA NODE ARTERY

The sinus node is almost always supplied by one artery trans-

versing its structure. The presence of the artery in the node is constant; however, its origin and its course in route varies.

In approximately 45% of human hearts the sinus node artery originates from the right coronary artery and in 55% from the left coronary artery. When it arises from the right coronary artery, most often it originates within the first few centimeters and occasionally only a few millimeters distal to its aortic ostium.

From its original origin in the proximal right coronary artery, this vessel, approximately 1 mm in diameter, proceeds medially and cephalad along the anterior wall of the body of the right atrium beneath the atrial appendage. It may then penetrate the anterior margin of the interatrial septum, ascend to the level of the entrance of the superior vena cava and encircle its orifice counter-clockwise, or bifurcate at the anterior margin of the vena cava and send one branch in the above course and the other directly into the crista terminalis and node; the two branches together encircle the base of the vena cava, in the latter case. As a third variation, the artery sometimes terminates as a single vessel circling the base of the vena cava, supplying the node, in a clockwise direction, passing through the crista terminalis and reaching the septal side of the superior vena cava at the end of its course.[4]

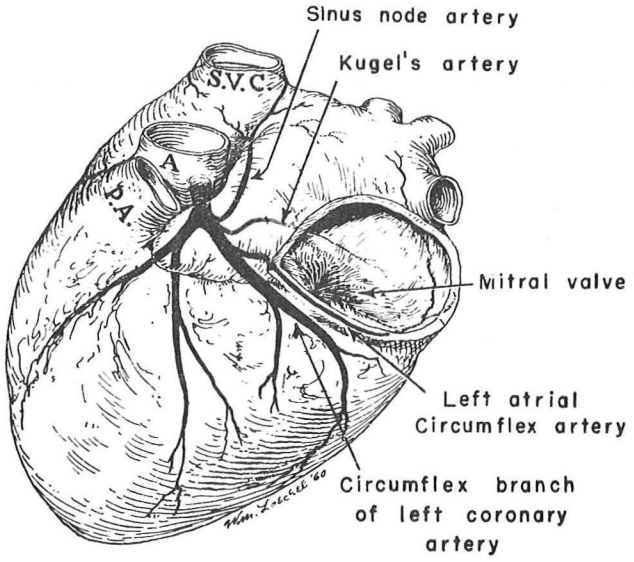


Artist's drawing of the sinus node artery arising from the right side. S.N. is sinus node, S.N.A., sinus node artery, R. i.a.a., right intermediate atrial artery, and R.V., right ventricle.

From James[4]

When the sinus node artery arises from the left side, it originates almost exclusively from the left circumflex artery, just beyond the bifurcation of the main left coronary artery.

From its origin just beyond the bifurcation of the left coronary artery, the sinus node artery ascends a few millimeters and then turns along the body of the anterior left atrium beneath the atrial appendage to reach the anterior margin of the interatrial septum. At this point it may penetrate the interatrial septum a short distance, all the while ascending to the dorsal interatrial septal groove. It then encircles the base of the superior vena cava in the same type of arterial circle as is formed when the sinus node artery originates from the right coronary artery.[4]



Artist's drawing of the sinus node artery arising from the left, also illustrating other important atrial arteries (Chap. 11).

From James[4]

Although small arteries regularly distribute in the region of the interatrial septum from both the right and the left coronary arteries, it is rare for a major artery to supply the sinus node from both sides. When the supply is bilateral, the two rami contribute equally to the formation of an arterial circle nourishing the node. In abnormal hearts (ASHD) the anastomoses of the sinus node artery are a major route of collateral circulation.

4. James, TN: *Anatomy of the Coronary Arteries*, Hoeber, 1961

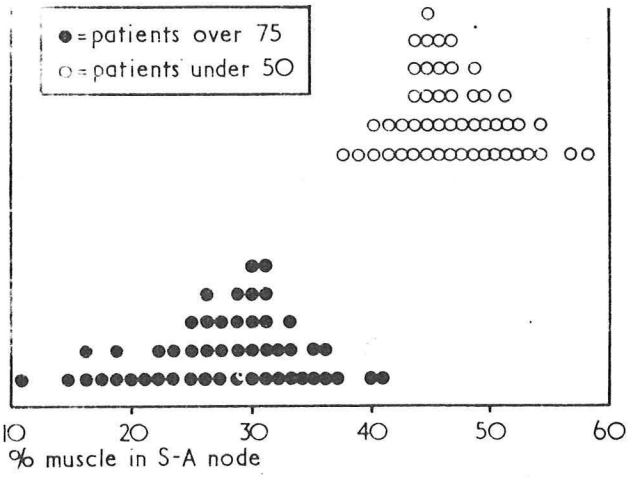
AGING CHANGES IN THE SA NODE

Lev has demonstrated that the sinoatrial node has certain characteristics which differentiate it from the atrial musculature: 1) the muscle fibers of the node after birth are smaller in diameter and length and they are arranged in a plexiform manner; 2) the

nuclei after 2 years of age are less in diameter than those of the atrial musculature; 3) the muscle fibers have a lesser number of myofibrils with less prominent striations; 4) intercalated discs are not found; 5) the reticular network has a finer mesh consisting of more delicate fibrils; and 6) the collagen and elastic fibers are more numerous.[5]

With advancing age the following changes occur: 1) the node grows more slowly than the atrial musculature; 2) there is an increase in collagen up to about 40, and a minimum increase thereafter; 3) there is an increase in elastic fibers throughout life; 4) there is an increase in reticular fibers with an accentuation of the individual fibers; 5) there is an apparent loss of muscle fibers after 40, with focal hypertrophy of individual fibers and focal thickening of the elastic and reticular nets; and 6) there is an infiltration of fat about and in the node.[6]

These findings have been confirmed by Davies and Pomerance in a comparison of per cent muscle in the SA node in patients under 50 years and over 75 years of age.[6]



Comparison of the per cent muscle in the sinoatrial node in young and old subjects.

From Davies and Pomerance[6]

The patients in these studies did not have significant coronary artery disease. In fact, the sinus node artery seemed to increase in size slightly with age.

These aging changes in the SA node could, in many instances, be responsible for abnormal function of the sinus node in the older age group patients with resultant sinus bradycardia-tachycardia and arrest syndromes.

5. Lev, M: Aging changes in the human sinoatrial node. J. Gerontol. 9:1, 1954.
6. Davies, MJ, and Pomerance, A: Quantitative study of ageing changes in the human sinoatrial node and internodal tracts. Brit. Heart J. 34:150, 1972.

PATHOLOGICAL STUDIES OF THE SA NODE CORRELATED WITH ARRHYTHMIAS

A. Chronic Arrhythmias

B. Myocardial Infarction

A. Chronic Arrhythmias

Hudson examined 65 hearts of patients with a variety of cardiac disease and correlated the pathological changes in the SA node with the cardiac rhythm in the patient prior to death and found the following:[7]

Node obviously damaged	15	14 - established arrhythmia, usually atrial fibrillation
		1 - normal rhythm
Node not found with certainty	1	established atrial fibrillation
Node Within Normal Limits	49	1 - foetal heart (rhythm not known
		2 - recent atrial fibrillation
		5 - paroxysmal atrial fibrillation
		41 - normal rhythm

In the damaged nodes, the constant lesion was the depletion and poor staining of nodal muscle with corresponding increase of fibrous, fibro-elastic, or fatty tissue. Sometimes the node was so badly damaged that only the arteries could be identified with certainty. The right atrium nearly always showed muscle damage and increase of interstitium in these cases, and it seems that the node and atrium were affected together. It is probable that in a damaged node, the nervous components will be involved also, since they are an integral part of node structure. Atheroma of the arteries in the node was mild and uncommon.

Sims also examined the SA node histologically in a series of 31 patients with atrial arrhythmias prior to death and compared these with patients dying without arrhythmias and no evidence for heart disease.[8] He found a consistent loss of nodal fibers. These findings suggest that the majority of chronic atrial arrhythmias are secondary *to permanent pathological changes in the SA node and may not be expected to return to normal, i.e.*, patients with chronic atrial fibrillation frequently cannot be expected to maintain a normal sinus rhythm after cardioversion, in fact may on occasion never develop a normal functioning sinus node (normal sinus rhythm) and may require a pacemaker or a return to atrial fibrillation for management.

7. Hudson, REB: St. Cyre's Lecture at the Royal Society of Medicine, 1960.
8. Sims, BA: Pathogenesis of atrial arrhythmias, Brit. Heart J. 34:336, 1972.

B. Myocardial Infarction

Atrial arrhythmias occur in about 10% of acute myocardial infarctions and the commonest of these arrhythmias is atrial fibrillation[9]. When atrial fibrillation is sustained after myocardial infarction, it has been reported to be associated with an 89% mortality[10]. James reported the findings in 11 patients dying of myocardial infarction who developed atrial arrhythmias prior to death[11]. In all 11 cases a coronary occlusion was present proximal to the origins of both the sinus node artery and AV node artery. In 6 of the hearts gross ecchymoses were found in the sulcus terminalis directly over the sinus node.

Microscopically, infarction of the sinus node was found in all 11 cases. Lesser changes were apparent in the AV node in 5 cases. These changes were acute and consisted primarily of hemorrhage and edema; however, collagen deposition and fatty infiltration were present in scattered foci in all the sinus nodes, suggesting that previous focal damage had occurred.

The sites of damage in the sinus node were characteristic in every case, occurring at the junctions of the node with the right atrium and sinus intercavarum. Hemorrhages at these locations involved Purkinje tracts leaving the node and may be presumed to be associated with impairment of normal transmission of the sinus impulse to the rest of the heart. Pathologic changes were less often seen in the central portion of the node or in the more distal atrial muscle, suggesting that the exit junctions of the node may be peculiarly vulnerable to acute hypoxia.

Summary of the Arrhythmias and Nodal Pathology in 11 Cases of Myocardial Infarction[11]

Case	Sinus Node	AV node	Atrial Arrhythmias
1	Infarcted	No pathology found	Multiple atrial arrhythmias
2	Infarcted	No pathology found	Atrial fibrillation
3	Infarcted	No pathology found	Atrial fibrillation
4	Infarcted	Infarcted	Incomplete AV block, then atrial fibrillation
5	Infarcted	No pathology found	Atrial fibrillation
6	Infarcted	Sclerotic AV node artery	Atrial fibrillation
7	Infarcted	No pathology found	Atrial tachycardia (176/ minute)
8	Infarcted	Infarcted	Atrial fibrillation
9	Infarcted	No pathology found	Multiple atrial arrhythmias
10	Infarcted	Infarcted	Intermittent sinus arrest
11	Infarcted	Degeneration	Atrial flutter then fibrillation

James suggests the following factors may influence the onset of atrial arrhythmias during an acute myocardial infarction:

1. Depressed sinus node "dominance"
2. Impaired sinus impulse transmission (SA block)
3. Vagal and vagomimetic reflexes
4. AV node injury
5. Extranodal atrial injury
6. Atrial distension
7. Increased circulating catecholamines

Since the sinus node artery is the largest and most constant atrial artery in man one might expect an occlusion proximal to its origin to produce a large atrial infarct. This did not prove to be

case in the 11 cases studied; only 4 of the 11 having a grossly recognizable atrial infarct, and the largest of these was less than 1 cm2 in size; all 4 were in the right atrium. Thus, extranodal atrial injury does not seem to be a regular accompaniment of sinus node infarction, but is an associated factor contributing to the onset of atrial arrhythmias in less than half of the cases.

Anatomical and electrocardiographic correlations can be made in patients with acute myocardial infarctions and atrial arrhythmias with a high degree of certainty.

If an atrial arrhythmia occurs in a patient with an inferior myocardial infarction, the coronary occlusion is most likely to be proximal to the origin of the sinus node artery. Since true inferior infarctions are in 90% of cases due to right coronary occlusion, and since the sinus node artery from the right (55% of humans) arises most frequently within 2 or 3 cm from the aorta, the coronary occlusion in a posterior myocardial infarction with an atrial arrhythmia is most likely in the first 2 or 3 cm of the right coronary artery.

If an atrial arrhythmia occurs in a patient with an anterolateral myocardial infarction, the occlusion is most likely in the proximal left circumflex or main left coronary artery. These high occlusions result in a large per cent of left ventricular infarctions. It is easy to see why the mortality rate is high in this group of patients.

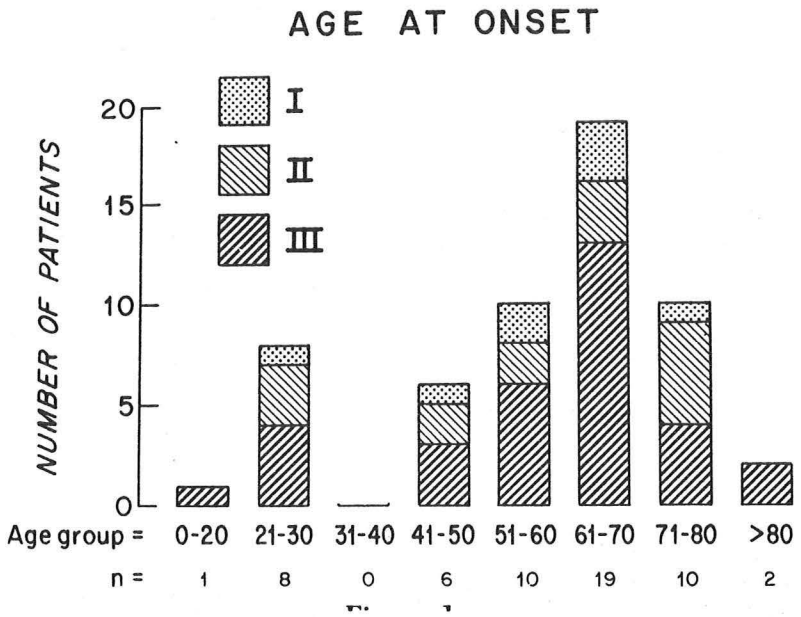
9. Friedberg, CK: *Diseases of the Heart*. Ed. 2, Philadelphia, W. B. Saunders Co., 1956.
10. Askey, JM, and Neurath, JM: The prognostic significance of auricular fibrillation in association with myocardial infarction. Amer. Heart J. 29:575, 1945.
11. James, TN: Myocardial infarction and atrial arrhythmias. Circulation 24:761, 1961.

SICK SINUS SYNDROME

Ferrer[12] has clearly defined the clinical syndrome of a sick sinus node. Disease of the sinus node is of increasing clinical import and over the past few years a variety of clinical states have been recognized as pointing primarily to pathology of the human pacemaker; hence, these conditions have been grouped together as the sick sinus syndrome. This syndrome appears not only in adults, in whom there is a wide etiologic spectrum, but also in children with congenital disorders[13] and indeed probably explains a number of sudden deaths in children and young athletes.[14] The fact that this potentially lethal sequence of events has escaped clinical recognition until recently is due to a number of reasons. Most important of these is the fact that sinus bradycardia has always been presented

as essentially a benign condition, and the sick sinus syndrome usually declares itself with this arrhythmia at its inception. Secondly, the multifaceted clinical espressions of the sick sinus syndrome beclouded the possibility of a single etiology until it was possible to collect continuous rhythm samples in patients with undiagnosed cardiac disorders and thus piece together what had seemed to be unrelated and separate cardiac arrhythmias.

It is usual to find the chronic sick sinus syndrome in subjects of older age; however, it also occurs in the young as well as in every decade of life up to the 80's and involves both females and males. As James has noted[14] in the young athlete who died suddenly at 18 years, vague symptoms had begun at age 13 when a sick sinus syndrome went unappreciated. At age 14 his heart rate was 42 beats/minute and rose to only 70 beats/minute on exercise. At 15 1/2 years, he blacked out several times and at 18 years he died while playing football. The deaf children with lesions in the sinus node and prolonged Q-T intervals, and hence an increased duration of the ventricular vulnerable period[13,14] appear to have syncope or death due to ventricular rather than atrial arrhythmias.



From Rubenstein[15]

The age distribution of 56 patients reported by Rubenstein, *et al.*, is shown above[15]. Note the binodal distribution. The three groups represent subsets -- I - sinus bradycardia; II - sinus arrest or block; III - tachycardia syndrome.

12. Ferrer, MI: The sick sinus syndrome in atrial disease. JAMA 206:645, 1968.
13. James, TN: Congenital deafness and cardiac arrhythmias. Amer. J. Cardiol. 19:627, 1967.
14. James, TN, Froggatt, P, and Marshall, TK: Sudden death in young athletes. Ann. Intern. Med. 67:1013, 1967.
15. Rubenstein, JJ, Schulman, CL, Yurchak, PM, and de Sanctis, RW: Clinical spectrum of the sick sinus syndrome. Circulation 46:5, 1972.

CLINICAL MANIFESTATIONS

The clinical expressions of disordered sinus node activity can be multifaceted, intermittent, and even difficult to elicit. The basic physiologic defects center about hypoperfusion of the vital circulations, especially the brain, the heart, and the kidneys, as a consequence of sinus arrest, severe sinus bradycardia, or the rapid tachyarrhythmias which arise as escape manifestations following sinus pauses. Diminished cerebral arterial blood flow of a mild nature, particularly if periodic, can be cryptic, and one must be alert to slight personality changes such as irritability, fleeting memory losses, and nocturnal wakefulness, as well as to the more obvious changes such as slurred speech, pareses, errors of judgment, dizziness, and syncopal attacks. Often the patient himself has difficulty recalling such "spells" of lightheadedness or momentary lapses. Generalized body fatigue, muscle aching, mild digestive disorders (due to congestive phenomena and low cardiac output during periods of ventricular failure), modest and periodic oliguria, and fleeting pulse irregularities often called premature contractions may seem so vague a constellation of complaints that mild cardiac failure is overlooked. Periodic fulminating and unexplained episodes of acute pulmonary edema are now recognized as secondary to the sick sinus syndrome in some instances. Transitory but repetitive sinus bradycardia sets the stage for mild ventricular failure, especially if there is underlying coronary disease. Then the tachyarrhythmia, usually atrial fibrillation with rapid ventricular response or atrial tachycardia, follows and severe failure with pulmonary edema brings the patient to an emergency room. By this time the tachyarrhythmias may have ceased, and one finds a subject in severe respiratory distress with an inappropriately slow sinus mechanism.

Unless the periodic bradycardia or arrest episode is perceived, it may take considerable time to arrive at a correct diagnosis of a chronically disordered and dying sinus node. A deceptive feature also is the fact that no other signs of heart disease may exist for some time unless one carefully searches the electrocardiogram. Evidences of the existent atrial disease can be seen in the abnormally wide P waves of intraatrial block, or when P wave change their shape, direction, or vectorial orientation, suggesting a new site of automaticity outside the failing SA node. The new atrial rhythm is seldom

rapid and indeed fires at rates close to those of normal sinus rhythm. In these instances the atrial rate is usually slightly faster than that of the sluggish SA node, and the ectopic rhythm may be considered an escape phenomenon. Intraatrial block may precede symptomatic disease by several years.

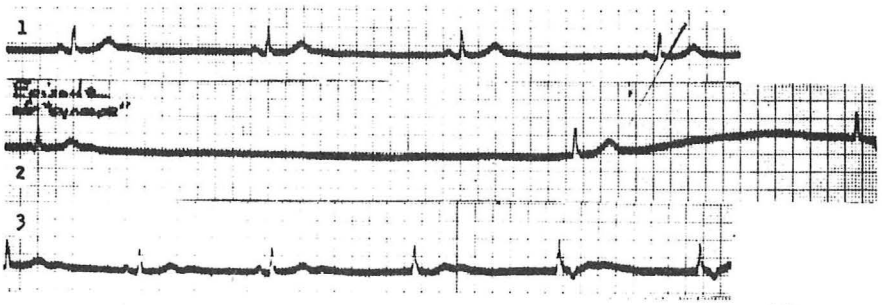
The slow unfolding of the sick sinus syndrome is usually a chronic affair. The sick sinus syndrome can begin acutely, however, especially in myocardial infarction or sudden coronary insufficiency. In that event syncope, shock, pulmonary edema, or simply the symptoms associated with severe hypotension without shock are seen[16].

16. Ferrer, MI: The sick sinus syndrome. Circulation 47:635, 1973.

ELECTROCARDIOGRAPHIC MECHANISMS

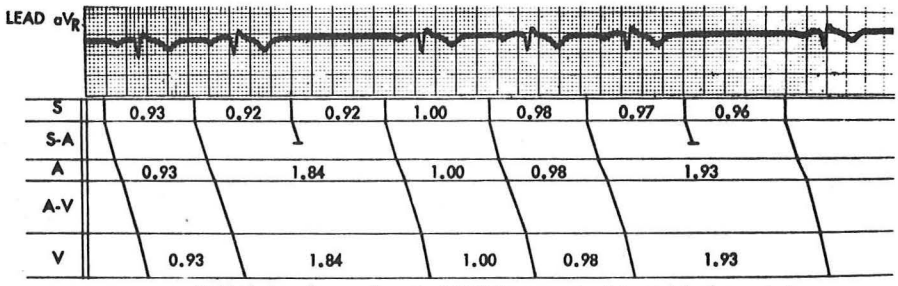
The electrocardiographic mechanisms which have been considered as part of the sick sinus syndrome are:

1. Sinus arrest - appearing for short intervals with no other (escape) rhythm arising or longer periods with replacement of sinus rhythm by another atrial or nodal rhythm.



From Myerburg[17]

2. SA block[17].



SA block. In each pause the entire P-QRS-T sequence is missing and the long cycle is approximately equal to two of the sinus cycles.

3. Chronic or recurrent atrial fibrillation due to sinus node failure. May be associated with a slow ventricular rate due to concomitant AV block.

4. Sinus node failure following cardioversion for atrial fibrillation.

5. Marked sinus bradycardia with or without PAC's.

6. Drug-induced SA block or bradycardia.

7. Brady-tachycardia syndrome[18].

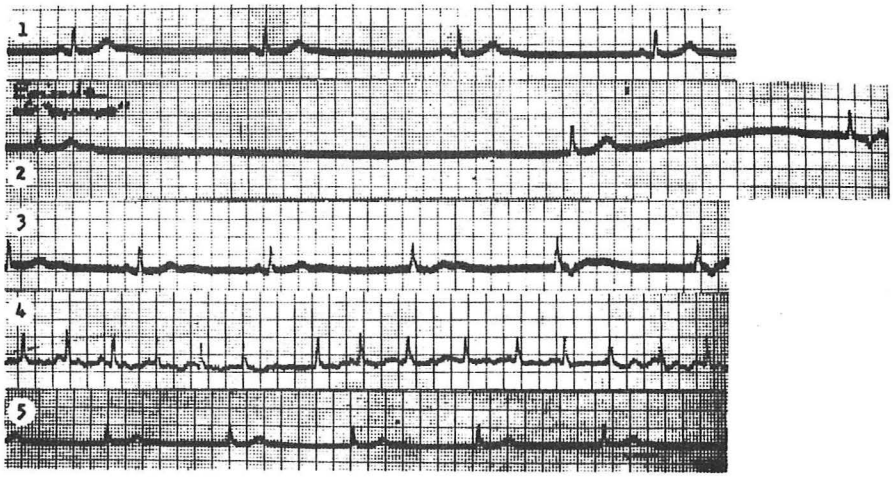
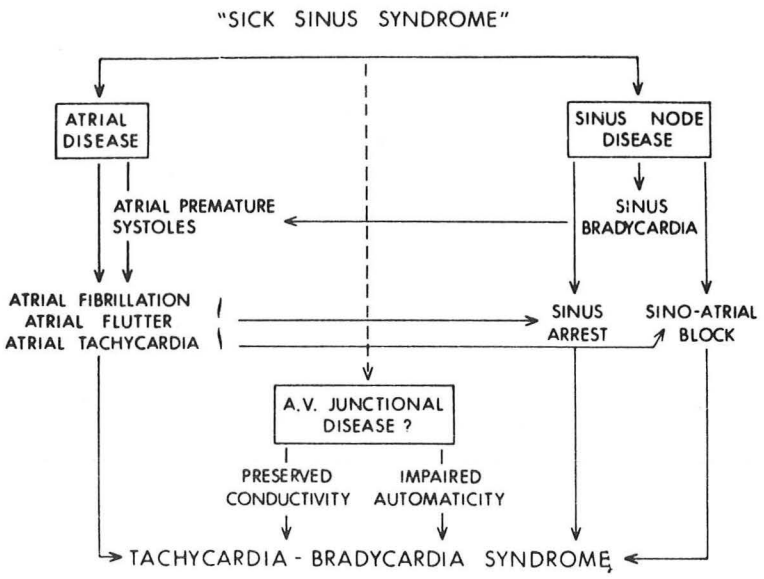


FIG. 1. Control electrocardiogram, sinus bradycardia at 26 per minute.
FIG. 2. Asystole of 6 seconds' duration resulting in syncope. Note recovery by a nodal beat.
FIG. 3. Conversion to a low nodal rhythm at 32 per minute, 90 seconds after atropine, 1.2 mg. intr nously.
FIG. 4. Paroxysmal atrial fibrillation at 110 per minute.
FIG. 5. Mid-nodal rhythm at 40 per minute after atropine, 2.0 mg. intravenously.

From Birchfield[18]

These conditions are likely to be interrelated, and more than one may be present at different times in the same patient. Because of these similarities and coexistence, confusion has resulted from mass grouping of multiple arrhythmias into the *sick sinus syndrome*. A proposed interrelation of the various functional disorders of the atria, sinus node and AV junctional tissues resulting in the tachycardia-bradycardia syndrome is schematized by Kaplan, *et al.*[19].



From Kaplan[19]

Whatever the underlying diseases, it may be said that sick sinus syndrome results either from direct structural damage to the sinoatrial node whether ischemic in origin or otherwise; or from sympathetic-parasympathetic imbalance as a result of (1) disturbances from circulating hormones or drug agents, or (2) neurovegetative derangement of the ganglionic fibers to the sinoatrial node by the disease process.

Mechanisms of Sick Sinus Syndrome

1. Parasympathetic or sympathetic imbalance in the SA node
2. Poor function of the SA node
3. SA node exit block
4. Direct drug suppression

17. Myerburg, RJ: Electrocardiographic diagnosis of sinus node rhythm variations and SA block. *In:* Advances in Electrocardiography (Eds.) Schlant and Hurst, Grune and Stratton, 1972.
18. Birchfield, RI, Menefee, EE, Bryant, GDN: Disease of the sinoatrial node associated with bradycardia, asystole, syncope and paroxysmal atrial fibrillation. Circulation 46:20, 1957.
19. Kaplan, BM, Langendorf, R, Lev, M, and Pick, A: Tachycardia-bradycardia syndrome (So-called "sick sinus syndrome"). Amer. J. Cardiol. 31:497, 1973.
20. Hurst, JW, and Logue, RB: *The Heart*. 2nd Ed., McGraw-Hill, 1970, pp. 48-49.

ETIOLOGY

A multitude of basic anatomic lesions may be responsible for the dysfunction of the sinus node: (1) ischemia; (2) sclerotic; (3) rheumatic and inflammatory conditions; (4) pericarditis; (5) cardiomyopathies; (6) collagen disease; (7) surgical and traumatic injury; (8) metastatic disease; (9) infiltrative disease, amyloid, hemochromatosis, sarcoidosis; (10) thyrotoxicosis and (11) idiopathic.

Familial sinus node disease has been reported.[21,22]

Drug-induced bradycardia may result in this syndrome, especially prone to induce bradycardia are the following: (1) digitalis excess; (2) propranolol; (3) Quinidine; (4) Pronestyl; (5) Aldomet; (6) Guanethidine; (7) Reserpine; (8) Melloril; (9) K+ excess; (10) nicotine; and (11) aerosol propellants with fluorinated hydrocarbons.

Vagotonia certainly may induce this syndrome and plays an intermittent role in the anatomic lesions as they develop.

21. Spellberg, RD: Familial sinus node disease. Chest 60:246, 1971.
22. Allensworth, DC, Rice, GJ, Lowe, GW: Persistent atrial standstill in a family with myocardial disease. Amer. J. Med. 47:775, 1969.

TRIGEMINAL AND GLOSSOPHARYNGEAL NEURALGIA PRODUCING SINUS BRADYCARDIA, ASYSTOLE AND SYNCOPE

Trigeminal and glossopharyngeal neuralgia associated with syncope is uncommon but are clear etiologies for asystole. The syndromes are characterized by intense paroxysmal pain felt in the ear, throat, posterior part of the tongue, soft palate, and lower lateral and posterior parts of the pharynx. The pain is accompanied by bradycardia or asystole and, at times, syncope with associated convulsions.[23-27]

Glossopharyngeal neuralgia is uncommon, compared to trigeminal neuralgia. The condition was first described by Weisenburg in 1910 in a patient with a tumor of the cerebellopontine angle.[28] Riley and associates in 1942 were the first to report cardiac arrest associated with the neuralgia.[23]

Although in most cases glossopharyngeal neuralgia occurs as an isolated condition, it may be associated with trigeminal neuralgia, cardiac arrhythmias, syncope, hypersecretion from the parotid gland on the affected side, and convulsions and unconsciousness in the presence of a normal heart rate and blood pressure.

Paroxysms of pain are precipitated by mechanical touch or by swallowing of foods, particularly cold, salty, bitter, or acid. The most common trigger zone is the tonsillar fossa on the affected side. Bradycardia, hypotension, and syncope followed severe paroxysms of pain, while massage of the carotid sinuses does not reproduce the symptoms. Sinus arrest, sinus bradycardia, and wandering atrial pacemaker are the most common arrhythmias reported in the syndrome. The cause of the arrhythmias is not completely understood, but it is likely that vagal reflexes initiated by the pain are involved.

The treatment of glossopharyngeal neuralgia is directed toward control of pain and abolition of the reflex bradycardia and syncope. Atropine sulfate and isoproterenol (Isuprel) hydrochloride are used as temporary emergency measures to abolish the bradycardia, but have no effect on the pain and cannot be used for long-term therapy. Spraying of the pharyngeal and oral mucosa with local anesthetics or nerve block are of temporary benefit. Anticonvulsant therapy has been suggested as a treatment of trigeminal neuralgia and results have sometimes been satisfactory. Diphenylhydantoin has been advocated since, in addition to its central action, it also stabilizes the peripheral nerve fibers against hyperexcitability.

Temporary demand pacing can be used to control the cardiac arrhythmia and syncope.[29]

Surgical resection of the glossopharyngeal nerve is the most effective permanent form of treatment.

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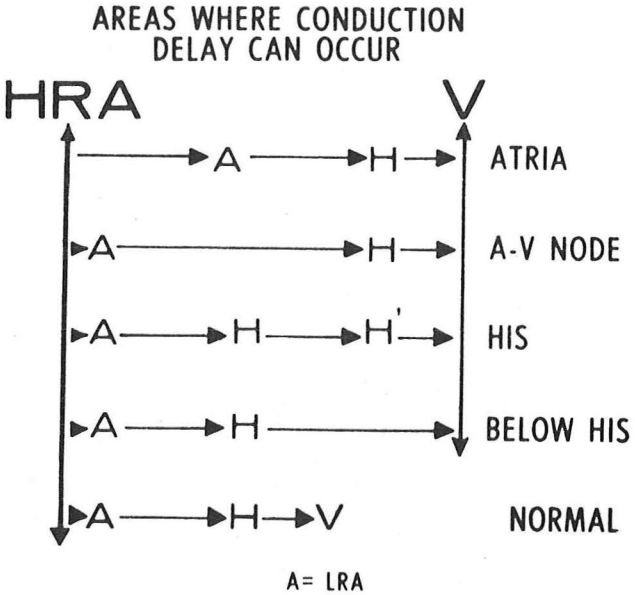
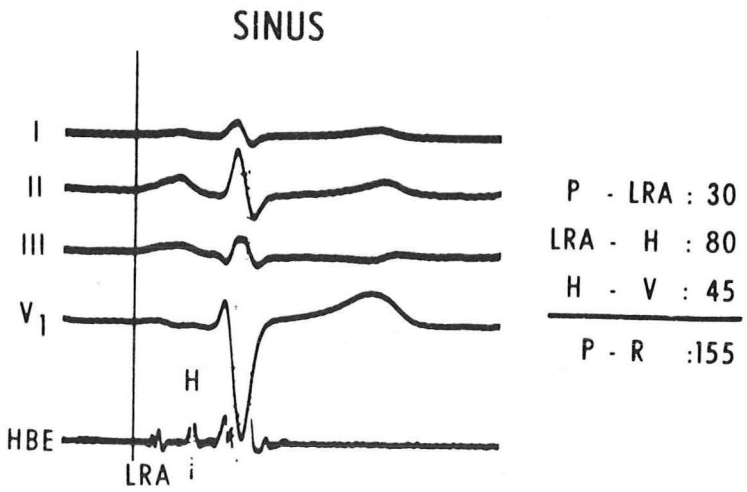
AV CONDUCTION DEFECTS

The sick sinus syndrome is clearly associated with a variety of conduction defects. These defects may occur at several different levels in the conduction system. For complete evaluation His bundle recordings must be made to determine sites of conduction delay.

His Bundle Recording



From Marriott[30]

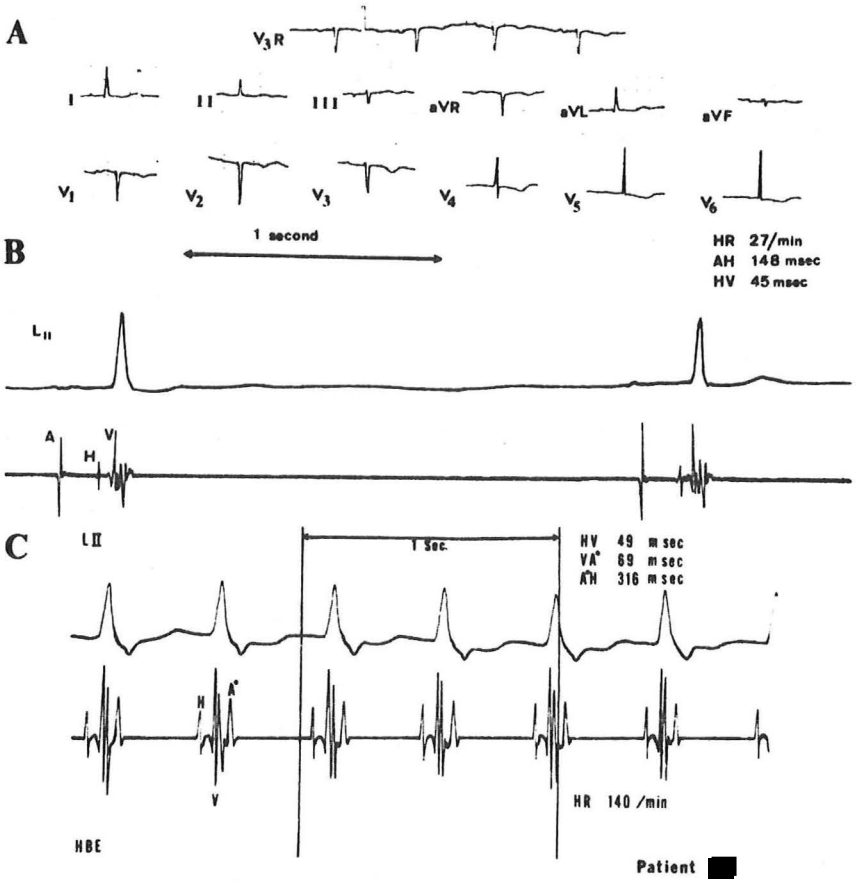


From Marriott[30]

Normal Values

			MSEC
P	→	LRA	25 - 45
LRA	→	H	50 - 120
H	→	V	35 - 45

This is a representative His bundle recording from a patient with sick sinus syndrome.[31]



From Mandel[40]

Bradycardia-tachycardia syndrome. (A) Twelve-lead electrocardiogram. (B) His bundle recording obtained during an episode of marked sinus bradycardia. The upper trace is a lead II electrocardiogram. The lower trace is the His bundle electrogram with the A, H, and V depolarizations labeled. A 1-sec time mark is shown at the top of the figure. Heart rate (HR), AH, and HV conduction times are shown to the right of the figure. (C) His bundle electrograms recorded during an episode of tachycardia with figure legends as in (B). Note the short VA conduction time, long AH time, and retrograde P waves (lead II).

AV conduction defects in patients with the sick sinus syndrome are common. Narula analyzed AV conduction in 75 patients with sinus bradycardia (SB) by His bundle (BH) recordings. Forty-five of these patients had normal QRS complexes and 30 had abnormal QRS complexes ($\geq$ 0.12 sec). The conduction times through the atrium (P-A), AV node (A-H), and His-Purkinje system (H-V) were measured during normal sinus rhythm and transient right atrial pacing up to rates of

150/minute. Over-all AV conduction was completely normal only in 25 (33%) of patients. Of these 25 patients, 20 had a narrow QRS complex and five had an abnormal QRS complex. The remaining 50 patients showed abnormal AV conduction in one or more regions (P-A in 10, A-H in 5, BH in 6, H-V in 16, and mixed in 19). In some patients despite normal ECG's, the H-V time was abnormal and 1:1 AV conduction was present during atrial pacing up to 150/minute. Fourteen patients were restudied at intervals of 6 months to 2 years. Three of these 14 patients showed prolongation of conduction times over 1 1/2 to 2 years.

Sinus Bradycardia[32-33]

QRS Complex	No. of Patients*	His Bundle Study			
		Normal		Abnormal	
Normal (≤ 0.10 sec)	45	20	25 (56%)	P-A	8
				A-H	2
				H-V	6
				Mixed	9
Abnormal (≥ 0.12 sec)	30	5	25 (83%)	P-A	2
				A-H	3
				H-V	10
				Mixed	10
Total	78*	25	50 (67%)		

*Mean age = 73 years.

The data demonstrate that SB is often (67%) associated with AV conduction abnormalities. AV conduction was abnormal in 56% of patients with normal and 83% with abnormal QRS complexes. The ECG and atrial pacing are of limited value in assessing AV conduction. The Bundle of His recordings in symptomatic patients with sinus bradycardia may be of clinical significance in the selection of site for pacemaker implantation, that is, atrial or ventricular.

Normal His bundle recordings are no assurance of continued normal conduction since Kulbertus, *et al.*[34] reported a patient with sick sinus syndrome who developed complete heart block 5 days following a normal His bundle recording.

Narula has emphasized that the patients' symptoms with sick sinus syndrome were not correlated with AV conduction defects despite the high association of AV conduction defects with the sick sinus syndrome.[33] They reported that the history of dizziness,

syncope and Adams-Stokes syndrome was elicited from 28 of their patients. These symptoms could not be correlated with AV conduction defects. Thirty-six per cent (18 of 50) of the patients with abnormal AV conduction and 40% (10 of 25) of the patients with normal AV conduction were symptomatic. These symptoms were probably not due to AV conduction abnormalities because 1:1 AV conduction was present during sinus rhythm (or supraventricular impulses). This suggests that these symptoms were primarily dependent on the manifestations of the sick sinus node and, in addition, on the escape interval and automaticity of the subsidiary escape pacemaker during sinus arrest or block.

Patients With Sick Sinus Syndrome

	Total No.	No. Symptomatic
Abnormal AV conduction	50	18 (36%)
Normal AV conduction	25	10 (40%)

30. Marriott, JHL: *Practical Electrocardiography*, 4th Ed. William and Wilkins Company, 1973.
31. Mandel, W, Hayakawa, H, Danzig, R, and Marcus, HS: Evaluation of sinoatrial node function in many by overdrive suppression. Circulation 44:59, 1971.
32. Narula, OS: Atrioventricular conduction defects in patients with sinus bradycardia. Circulation 44:1096, 1971.
33. Narula, OS, Samet, P, Javiere, RP: Significance of the sinus-node recovery time. Circulation 45:140, 1972.
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AUTOMATICITY OF SA NODE IN SSS AND SNRT

Depressed AV Junctional Automaticity (Escape Mechanism)

It is clear that AV junctional (nodal) and ventricular escape rhythms are depressed in the sick sinus syndrome indicating impaired impulse formation by lower automatic centers. Ordinarily, pacemaker cells in the AV junction provide escape beats at rates of 40-50

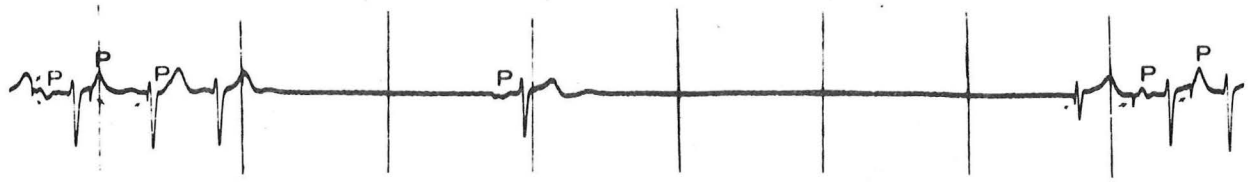
beats/minute when the SA node defaults. However, sinus bradycardias < 30/minute, as well as periods of sinus arrest for 3-5 seconds, are often found in the sick sinus syndrome with no escape rhythm.

This finding suggests that automaticity of the conduction system is independent of the conductivity of the system and occasionally either function may be affected without the other.

Depressed SA Automaticity and Sinus Node Recovery Time

The depressed automaticity in the sick sinus syndrome can be evaluated by atrial pacing and measuring the sinus node recovery time (SNRT or SART).

The normal response to cessation of rapid pacing is the development of overdrive depression with a variable asystolic period and transient bradycardia. After cessation of atrial pacing at 100-160 beats/minute in normal patients, the asystolic period is less than 1.4 sec (sinus node recovery time). In patients with sick sinus syndrome the asystolic period often exceeds 2.0 sec, occasionally requiring pacing for development of any rhythm at all! The sinus node recovery time (SNRT) has been found to occasionally vary from > to < 1.4 sec at different times in the same patient. This may make diagnosis more difficult but fortunately this is not usually the case, *i.e.*, once SNRT is prolonged it remains so.

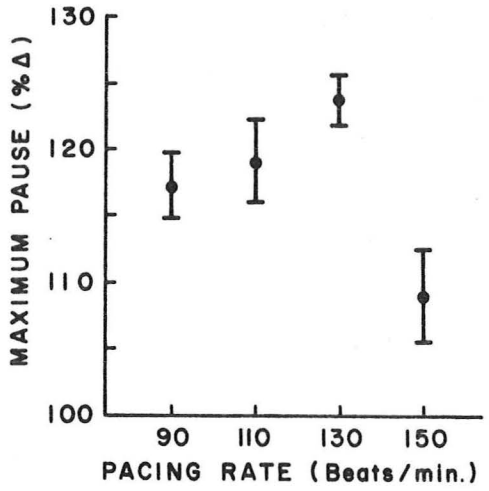


From Rosen[40]

Depressed automaticity shown after cessation of atrial pacing at a rate of 130 beats/minute, the rate producing Wenckebach periods. Paced P waves are labeled P and are preceded by atrial pacing spikes (arrows). After cessation of pacing, there is a 2-sec asystole followed by an ectopic atrial escape beat and then a 4-sec asystolic period. To terminate the second episode of asystole, pacing was resumed at the same time a junctional escape occurred. Time lines are at 1 sec. Paper speed is 50 mm/sec.

Narula evaluated the SNRT in normal patients and a group with sinus bradycardia.[39] In 27 of 28 normal patients, the SNRT with atrial pacing ranged from 110 to 525 msec and was independent of the rate and duration of pacing. In 12 of 28 patients with sinus bradycardia, the SNRT was in the same normal range. In 16 of the patients with sinus bradycardia, the SNRT ranged from 560 to 3740 msec and was usually directly proportional to the rate of atrial pacing.

EFFECT OF PACING RATE ON PAUSE



From Mandel[40]

Mandel reported the effect of pacing rate on sinus node recovery time in normal subjects.[40] The pause was directly proportional to the rate until pacing rates of 150 were reached and then the pause suddenly decreased for unexplained reasons. SNRT was reported after Atropine in several of the patients with little change.

Rosen studied 15 patients with symptomatic bradycardia and found 4 patients with SNRT > 2 sec.[41]

It is apparent that atrial pacing and measurement of the sinus node recovery time is a useful clinical tool in assessing sinus node function and thereby aids in the diagnosis of the *sick sinus syndrome*. However, many patients with symptomatic sinus bradycardia may have normal SNRT. This suggests that patients with sinus bradycardia do not comprise a homogeneous group and may or may not show an abnormal response. Some of these patients may have the brady-tachycardia syndrome with a normal sinus node function except in the posttachycardia period when they may have an abnormally prolonged sinus node recovery time but normal function following atrial pacing. Others may have transient defects in sinus node function or rarely may have increased vagal tone resulting in bradycardia.

Increased vagal tone usually does not seem to play a primary role in suppression of the sinus node in patients with the sick sinus syndrome since Atropine often has little effect on the SNRT in these patients. Furthermore, sinus rates rarely exceed 90 beats/minute after Atropine in patients with sick sinus syndrome, whereas normal patients routinely exceed 90 beats/minute postatropine. Atropine can be used as a provocative test for sick sinus syndrome.

Provocative Tests For Sinus Node Function

Maneuver	Normal	Sick Sinus Syndrome
ECG	NSR	Symptomatic bradycardia
Holter 24-hr monitor		Symptomatic bradycardia-tachycardia
		Sinus arrest or sinus block
Carotid sinus massage	Transient bradycardia	Sinus arrest > 3 sec
(Pacer in Place)	(-42% rate)	(Carotid hypersensitivity)
Exercise	Rate ↑	Rate ↑ or no response
Isoproterenol infusion		
Atropine	Rate ↑ 64%	Rate ↑ 25%
1-2.5 mg I.V.	> 90 beats/minute	< 90 beats/minute
Atrial overdrive pacing	SNRT < 1.4 sec	SNRT > 1.4 sec
	Usually < 1 sec	Often > 2 sec

The sick sinus syndrome is composed of a varied group of pathological entities and therefore, as one might expect, these provocative tests are not always positive and neither will all the tests be positive in the same patient.

Perhaps the depressed automaticity and conductivity has a pathophysiologic explanation. The reluctance of atrio-ventricular junctional pacemakers to emerge might be related to the pathological

changes of the intrinsic nerve plexus since it has recently been demonstrated that normal sympathetic neural input is essential to atrio-ventricular junctional automaticity.[42]

Anatomical data also lead to the idea that scattered areas of disturbed conduction might reasonably be expected in various sites of the heart and more particularly around the sinus node and in the supraventricular structures. These obstacles to conduction, embedded in the thin atrial wall, should offer ideal paths for circus movement and re-entrant excitation. They should favor the development of various types of tachyarrhythmias, especially those for which perpetuated re-entrant excitation has been suggested as a possible mechanism, *i.e.*, atrial flutter[43], atrial fibrillation[44,45] and paroxysmal atrial tachycardia.[46]

39. Narula, OS, Samet, P, Javiere, RP: Significance of the sinus node recovery time. Circulation 45:140, 1972.
40. Mandel, W, Hayakawa, H, Danzig, R, and Marcus, HS: Evaluation of sinoatrial node function in many by overdrive suppression. Circulation 44:59, 1971.
41. Rosen, KM, Loeb, HS, Sinno, MZ, Rahimtoola, SH, and Gunnar, RM: Cardiac conduction in patients with symptomatic sinus node disease. Circulation 48:836, 1971.
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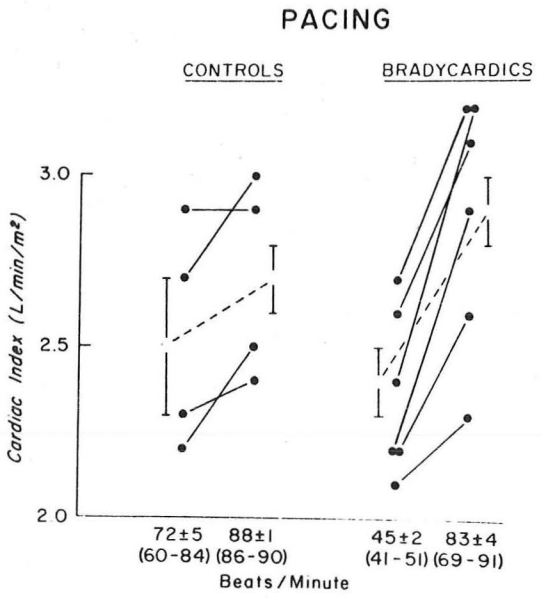
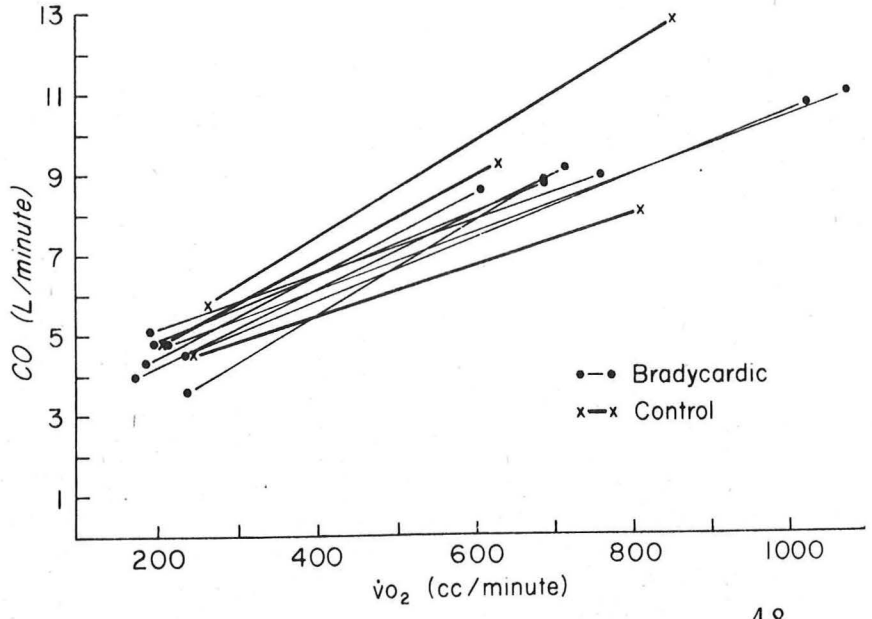
ASYMPTOMATIC BRADYCARDIA

Sinus bradycardia may not represent a pathological entity but may be physiologic as in athletes. Resting heart rates tend to diminish with age and may not be of hemodynamic significance.[47] It is not a rarity to find patients in their 70's with resting rates < 60/minute. Agruss, *et al.*[48] studied asymptomatic bradycardia subjects (heart rates 41-51 beats/minute), ages 67-79, with no evidence of impaired cardiac performance and taking no drugs. These subjects were compared with 4 age-matched controls (heart rates 60-84 beats/minute). They found the following:



From Agruss[48]

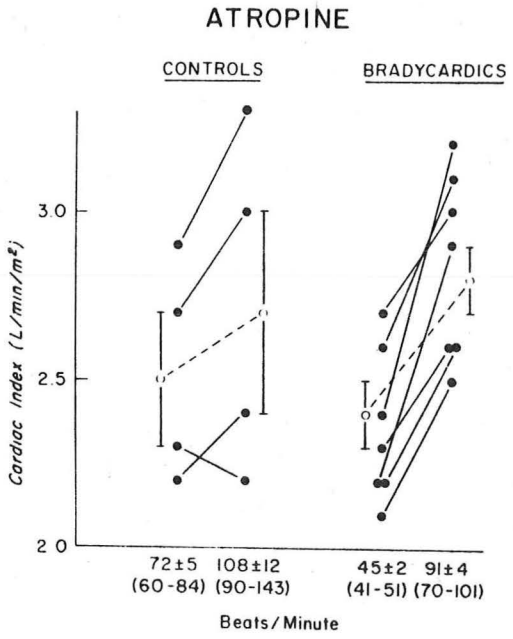
There was no significant difference in mean resting cardiac index between the groups. Both groups could be atrially paced. A significant increase in cardiac index occurred in the bradycardia group but not the controls. SNRT was normal in all patients.



From Agruss[48]

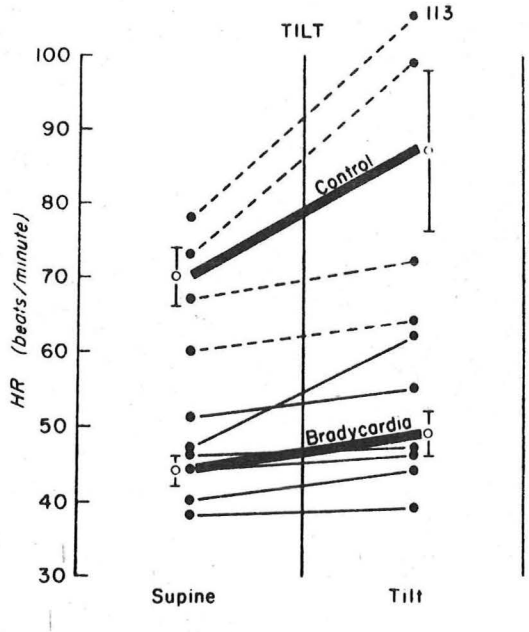
Changes in cardiac output during exercise are illustrated above. Heart rates and cardiac output response to exercise was normal for both groups with normal maximal exercise loads. The

bradycardia group increased their stroke volume more and heart rate less than the normal subjects with exercise to maintain the same cardiac output. This response is not present in patients with significant heart disease, *i.e.*, stroke volume does not increase with exercise



From Agruss[48]

The response of the cardiac index to Atropine (2 mg) was not different in both groups; however, the heart rate increase was significantly higher in the bradycardia group suggesting that increased vagal tone was an important factor in the slow heart rates in the bradycardia group.



From Agruss[48]

The response of the heart rate to head-up tilt revealed a greater increase in control subjects (24%) than in the bradycardia group (11%). Cardiac index increased similarly in both groups during tilt and mean brachial artery pressure did not change in either group.

Thus, significant degrees of sinus bradycardia may be associated with aging. Increased vagal tone may be a significant contributing factor to the bradycardia, and more importantly, normal cardiac performance can be present. Therefore, the implications and management of sinus bradycardia depend upon the setting in which it occurs.

This hemodynamically stable bradycardia may represent the earliest finding in sick sinus syndrome and slow progression may result or it may represent a totally unrelated (perhaps physiologic and benign) response to aging.

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COURSE, NATURAL HISTORY, AND PROGNOSIS OF SSS

Course

The sick sinus syndrome in its chronic form runs an erratic course with periods of normal node function alternating with abnormal behavior. This tends to cloud the homogeneity of the condition allowing separated single and vague episodes to slip by undiagnosed correctly. Some of the signposts of note along the way are inappropriate heart rates, dizzy spells, especially unexplained episodes of congestive failure, episodes tagged as convulsive disorders in the young, and finally syncopal or cardiac arrest attacks. Periodic or sustained sinus bradycardia can no longer go unchallenged even if asymptomatic and is a major evidence of this disorder in its early stage. Paroxysmal atrial fibrillation without evident heart disease may be another signal to explore the behavior of the sinus node. Since the provocative tests are not difficult to perform, such exploration must now be done in increasing numbers.

Natural History

The natural history of this syndrome of disordered sinus node function is only just now becoming apparent as the varied episodes are gathered together under one etiology. The knowledge is far

from complete, but at present it appears that the sinus node may die quite slowly, taking 5-10 years or more to cease functioning completely. Sinus bradycardia may progress to various forms of exit block or may simply become increasingly severe until no sinus beats are found. The escape rhythms, at first only periodic rescuers, eventually become the basic mechanism, particularly atrial fibrillation. When the latter exists with a slow ventricular rate in an undigitalized subject, it is probably an end stage of sinus node disease. Between these extremes of the sick sinus syndrome there may be many different physiologic adjustments. Because of this one would like to predict the crucial time for insertion of a pacemaker, since atrial fibrillation is probably the only stable long-term replacement rhythm and is not always the escape rhythm to arise. Other less stable escape rhythms often fail to fire after a while, especially if the disease process itself progresses to involve these other specialized tissues (junctional, atrial). Unfortunately such a prediction is not yet possible. The deterioration of sinus node dysfunction may proceed slowly at first then speed up and produce a disastrous asystole or some other marked change in the patient.

Prognosis

The long-term prognosis of the sick sinus syndrome cannot be stated with certainty in any one case since the end stage of sinus arrest cannot be predicted. However, since asystole can occur periodically and congestive failure can result also, the outcome is likely to be poor eventually. It does not appear likely that the failing sinus node can be cured, at least with present therapies. Hence replacement of its function by a demand pacemaker is inevitable in most patients.

49. Ferrer, MI: The sick sinus syndrome. Circulation 47:635, 1973.

TREATMENT

In drug-induced sick sinus syndrome the obvious method of treatment is withdrawal of the drug, *i.e.*, hypertensive medications, toxic doses of digitalis, Melloril, etc. However, the patient may require the specific drug for treatment of an underlying cardiovascular problem and therefore necessitate the use of a cardiac pacemaker.

In asymptomatic bradycardia, especially with no underlying serious heart disease, no treatment is necessary. An exercise test is indicated to evaluate the cardiac rhythm and function under stress to assure that no latent cardiac disease or stress is present.

In patients with thyrotoxicosis, sickle cell anemia, etc., the treatment should be directed toward the underlying cause.

In acute myocardial infarction, the sick sinus syndrome is usually transient and often vagally induced. This usually responds to Atropine; if not, then a temporary pacemaker should be used.

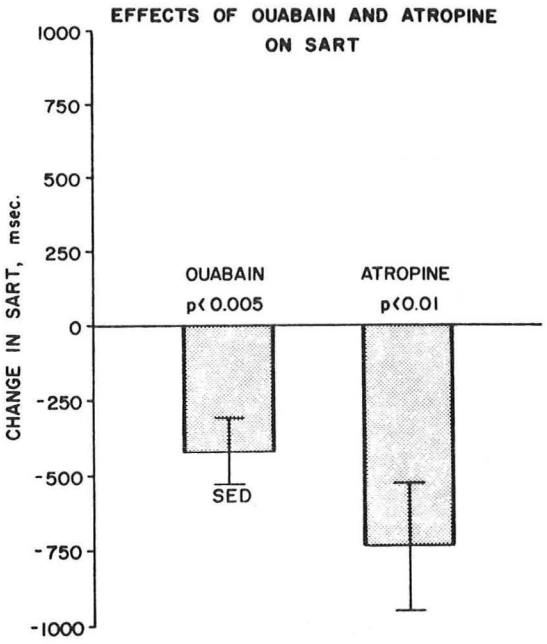
In patients with chronic sick sinus syndrome, drug therapy alone is unsatisfactory. The medications used to prevent or treat the atrial tachyarrhythmia - namely, Quinidine, procainamide, digitalis or propranolol - are all capable of promoting SA block or sinus arrest and depression of lower escape mechanisms. The dilemma is further complicated. Isoproterenol, useful for the treatment of sinus nodal disorders, may produce atrial fibrillation or atrial tachycardia. Furthermore, Atropine has usually been found to be ineffective for the treatment of SA block or sinus arrest. The dilemma can be resolved by the insertion of an artificial pacemaker. With a pacemaker in place, the bradycardia can be treated adequately and the concern of producing asystole by suppressing intrinsic impulse formation with drugs eliminated. At the same time, the rapid ventricular response to recurrent atrial tachyarrhythmias may be treated with digitalis or propranolol, or both, without fear of producing cardiac standstill; congestive heart failure, if present, may be handled safely with digitalis. In addition, ventricular pacing has been reported to diminish the rate of recurrence of supraventricular tachycardia. This beneficial effect appears to be due to the increased atrial rate as a result of 1:1 ventriculoatrial retrograde conduction.

Pathologic alterations of the sinus node, atria and AV junction are present and physiologic studies in patients with sinus nodal disorders would point toward simultaneous AV junctional abnormalities and the potential development of complete AV block. The tachycardia-bradycardia syndrome may represent multiple defects in the conduction system of the heart including the AV junction. Thus, transvenous fixed rate ventricular pacing might be preferable to demand pacing for patients with the tachycardia-bradycardia syndrome. Although demand pacing is safer than fixed-rate pacing, the latter maintains the capability of terminating recurrent reciprocating tachycardias, which may occasionally be present in the tachycardia-bradycardia syndrome. The advantages of both demand and fixed-rate pacing can be gained by the use of a demand pacemaker that can be converted to a fixed-rate pacemaker by the use of an external magnet or by radiofrequency transmission.

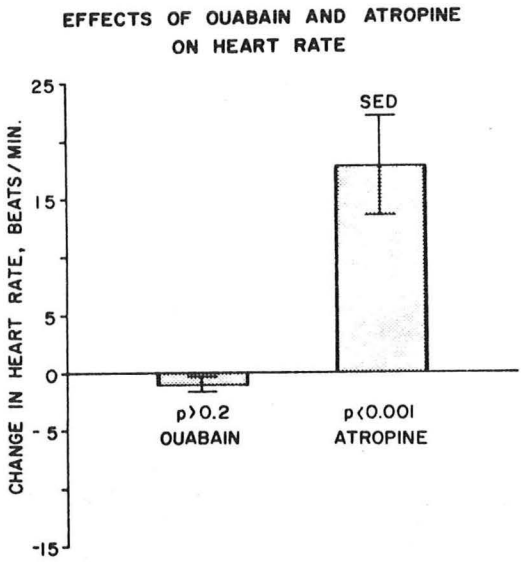
All patients with the tachycardia-bradycardia syndrome should be given maintenance doses of digitalis after pacemaker implantation. In addition, propranolol may also be necessary to slow the rapid ventricular response of recurrent atrial tachyarrhythmias. In some instances, Quinidine, procainamide or propranolol, singly or in combination, may be needed to suppress the ectopic atrial rhythm disturbances.

The Use of Digitalis in Sick Sinus Syndrome

Engel and Schaal studied the effect of digitalis in patients with sick sinus syndrome.[50] They compared the sinus rate change and SNRT (SART) before and after digitalis and Atropine.



Changes are expressed as the mean differences between control SART and those obtained following ouabain administration to 14 patients and those obtained after subsequent administration of atropine to 12 patients. SART = *sino-atrial recovery time;* SED = *standard error of the difference.*



From Engel and Schaal[50]

Digitalis (Quabain) produced no change in heart rate. The shortening of the SNRT by digitalis is indicative of an increase in automaticity of the SA node. Digitalis has a vagotonic effect on the SA node which results in a negative chronotropic effect (decreased heart rate), which can be eliminated by combined vagal and sympathetic blockade. There is no evidence that digitalis decreases sinus node automaticity directly in therapeutic ranges.[51,52] The lack of a decrease in heart rate with digitalis in patients with sick sinus syndrome may indicate an imbalance in sympathetic and parasympathetic tone in these patients resulting in no response to the digitalis-induced vagotonia.

When clinically indicated, digitalis may be used for congestive heart failure or tachyarrhythmias in the sick sinus syndrome in the absence of significant AV conduction disease.

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51. Sherlag, BJ, Abelleira, JL, Narula, OS, and Samet, P: The differential effects of ouabain on sinus, A-V nodal, His bundle and idioventricular rhythms. Amer. Heart J. 81:227, 1971.
52. Ten Eick, RE, and Hoffman, BF: Chronotropic effect of cardiac glycosides in cats, dogs, and rabbits. Circulation Res. 25:365, 1969.

The Use of Anticoagulants

Rubenstein, *et al.*[53] reported systemic embolization in 8 of 33 patients with the brady-tachy syndrome. Presumably, stasis in the atria associated with the tachyarrhythmias promotes mural thrombus formation, and they recommend anticoagulants in these patients if their general condition warrants it and if there is no contraindication to such therapy.

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CASE REPORT #1

Brady-Tachycardia Syndrome With SA and AV Node Disease

[redacted] [redacted] [redacted]/73

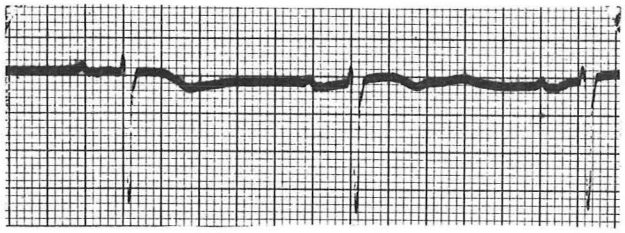
Fifty-two-year-old [redacted] female admitted for evaluation of sick sinus syndrome. She has noted palpitations since the age of 16 and has been followed in medicine clinics for mild to moderate hypertensive cardiovascular disease. Her treatment has been phenobarbitol and hydrochorothiazide. Several of her electrocardiograms demonstrated first degree heart block with a shifting atrial pacemaker. During the past 5 years she has noted progressive symptoms of palpitations. These are often associated with substernal crushing chest pain. The palpitations appear quite suddenly and disappear suddenly as well. A 24-hour Avionics recording on [redacted]/73 demonstrated shifting atrial pacemaker from the SA node with first degree heart block to the AV junctional area. There were numerous junctional and atrial premature beats. There were runs of slow atrial and junctional tachycardia. After shifting from the atrial to junctional rhythm, there was an increase in rate. In several areas of junctional tachycardia there was progressive prolongation of the R-R interval suggesting Wenckebach exit block out of the junctional focus. Some areas of the Avionics tracing revealed AV dissociation. Her resting heart rate during the day ranged between 50 to 60 and at night would drop as low as 45/minute. It was also noted that during a rather prolonged episode of junctional tachycardia, which persisted for over an hour, that she had inverted T waves and S-T depression suggesting ischemia. Physical examination revealed a pleasant, well-developed lady whose blood pressure was 140/90 and her heart rate was 55 and regular.

A tri-electrode catheter was inserted into the femoral vein percutaneously and advanced up into the right ventricle. This was positioned across the tricuspid valve where His bundle potentials were recorded. After obtaining rest recordings the patient was paced at 71, 83 and 97 beats/minute. The resting recordings demonstrated prolongation of the A-H interval ranging from 165 to 200 milliseconds. The H-V interval remained constant at 45. At the two higher pace rates there was progressive prolongation of the A-H interval with drop beats characteristic of Wenckebach Type I heart block. The sinus node recovery time was 2.0 seconds (Normal < 1.5 sec).

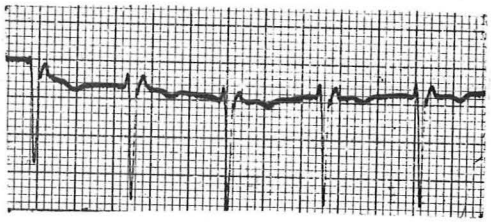
The study confirmed that the patient had AV junctional disease. Her prolonged sinus node recovery time was compatible with her clinical findings of sick sinus syndrome.

A permanent demand transvenous pacemaker with right ventricle electrode placement was inserted on [redacted]/73 with a pacing rate of 70/minute. She has subsequently had no further symptoms and her electrocardiograms show continuous pacing.

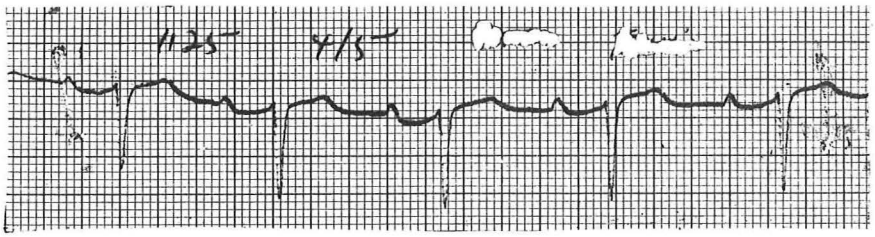
1963 Sinus Rate 50



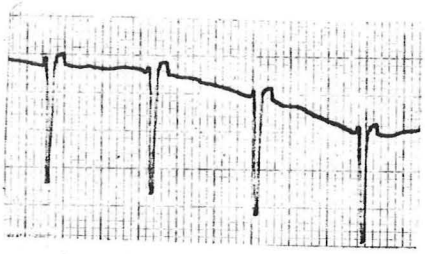
1963 Junctional Tachycardia Rate 118



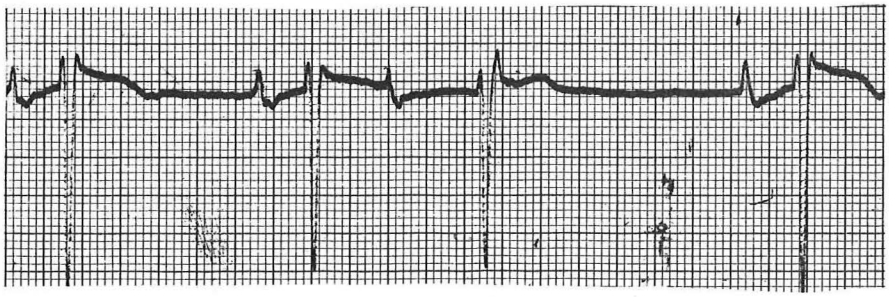
[redacted]/73 First Degree Block
Rate 70

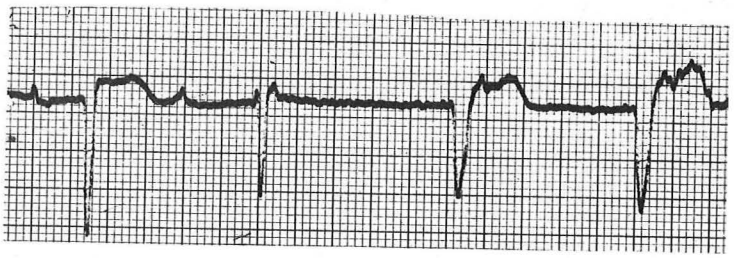


[redacted]/73 Junctional Tachycardia
Rate 110

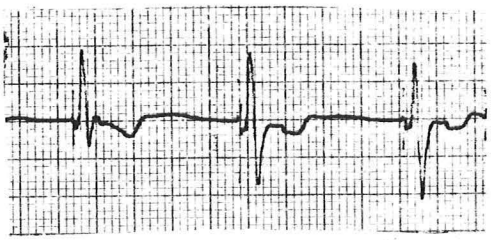


[redacted]/73 SA Block and AV Node Wenckebach Block With Ventricular Escape





[redacted]/73 Permanent Pacer Rate 70



CASE REPORT #2

Ischemia of the SA Node Secondary to Coronary Artery Disease

[redacted] [redacted] [redacted]/73

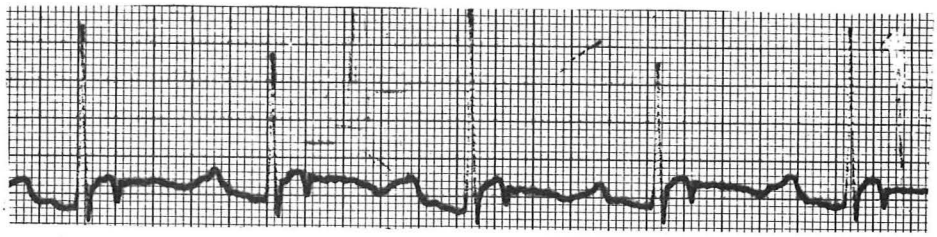
Fifty-three-year-old lady admitted with a 2-month history of increasing angina pectoris (precordial chest pain radiating to her left arm) occurring several times daily with a recent onset of angina decubitus. The night of admission the chest pain lasted for 2 hours, unrelieved by nitroglycerin.

On admission her electrocardiogram showed inverted T waves V_1-V_6 with a sinus rate of 60.

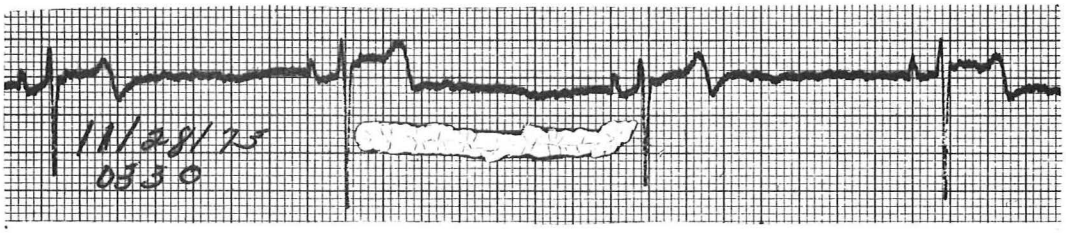
She developed episodes of sinus bradycardia to rates of 33/minute during her hospital course with no evidence of escape rhythms or other conduction blocks. She had several episodes of chest pain responding to vasodilators not necessarily associated with her bradyarrhythmias. The ECG pattern fluctuated during the first week showing anterior ischemia with T wave inversions and S-T elevations that resolved within a week. The serum enzymes were nondiagnostic for an acute MI.

On [redacted]/73 she was atrially paced to a rate of 110 beats/minute resulting in S-T segment depression (3 mm), prolonged P-R interval (.24 sec) and angina. Sinus node recovery time was 1.6 seconds (normal < 1.4 sec) after a pacing rate of 110 beats/minute. She was placed on large doses of isosorbide dinitrate sublingually and orally after which she had no more episodes of angina or bradycardia.

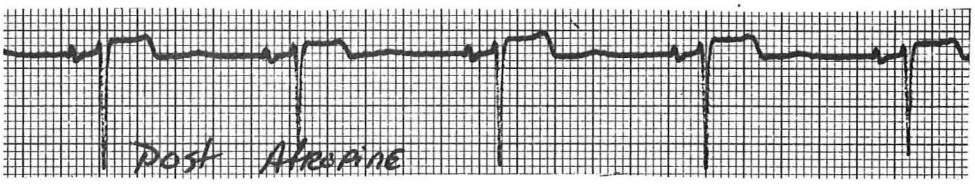
[redacted]/73 Admission Rate 58



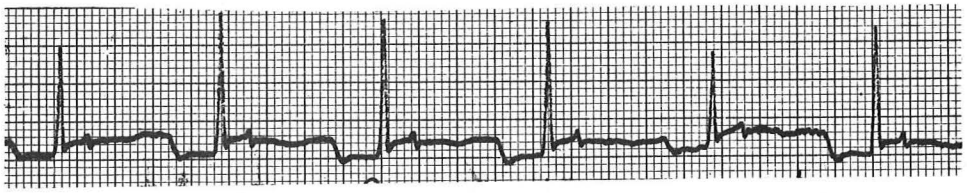
BP 170/80 Asymptomatic, Asleep Rate 38



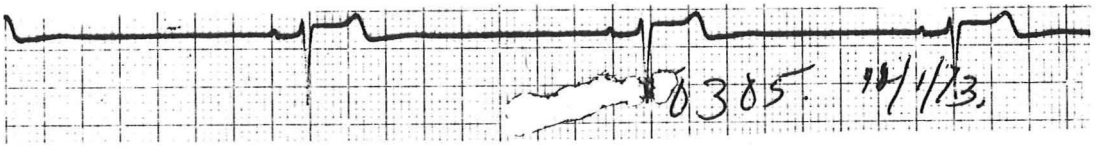
0.5 mg Atropine I.V. Rate 57

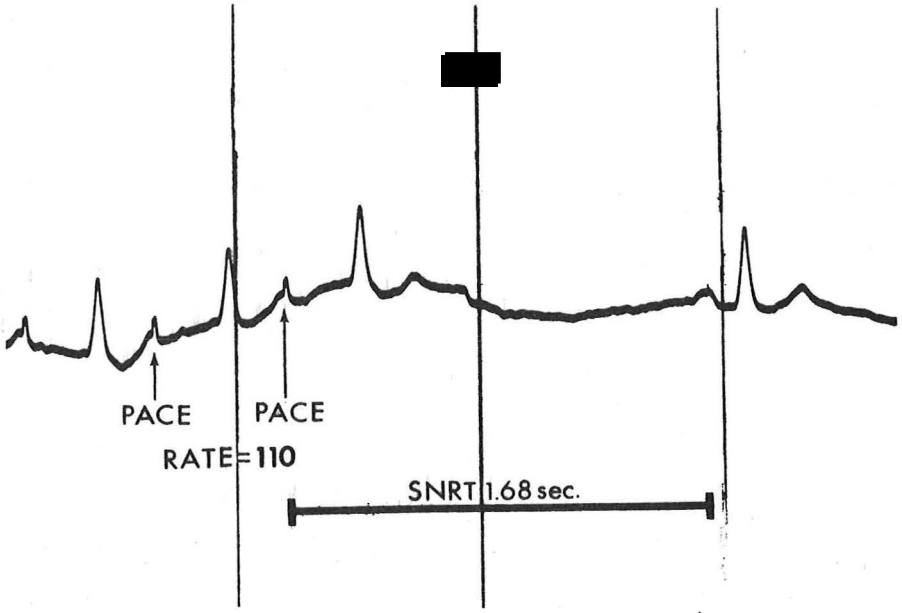


[redacted]/73 During Chest Pain Rate 69



[redacted]/73 Asleep BP 160/80 Rate 32





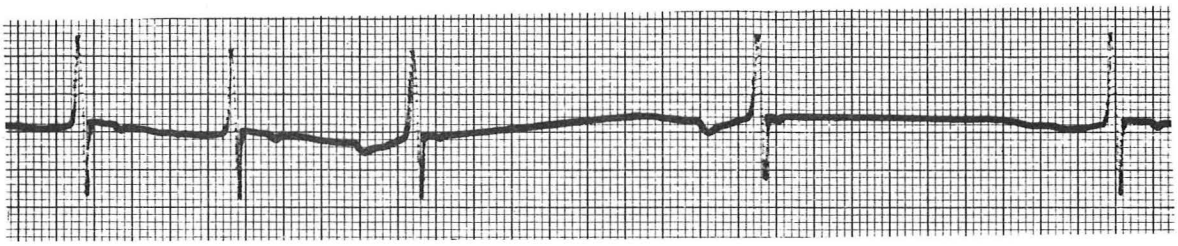
CASE REPORT # 3

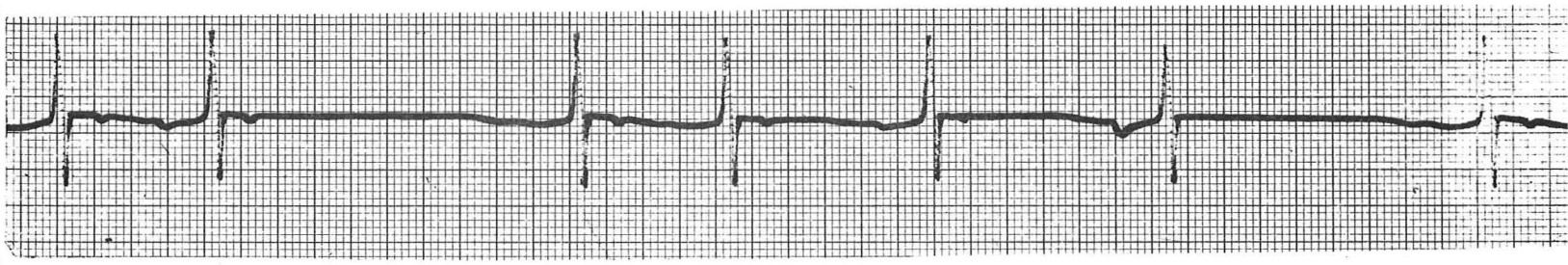
Sick Sinus Syndrome Secondary to Sickle Cell Disease

[redacted] [redacted] [redacted] [redacted]/73

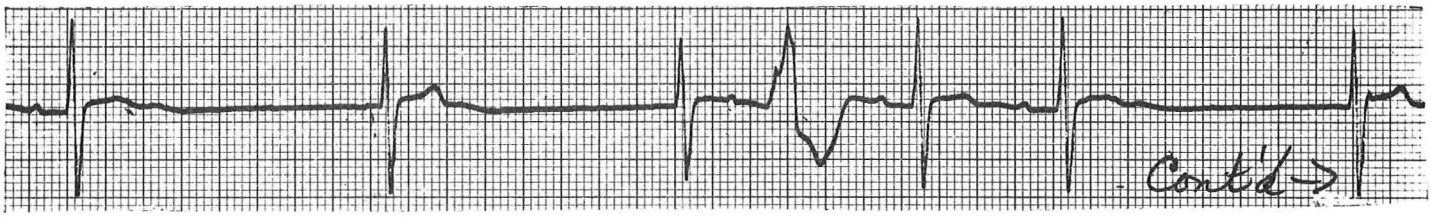
Twenty-four-year-old [redacted] male presented with acute onset of chills, fever, malaise and chest pain. He was known to have sickle cell disease (SS) but had been asymptomatic for 4 years. He was thought to be in sickle crisis and was admitted to the intensive care unit because of a cardiac arrhythmia which was asymptomatic.

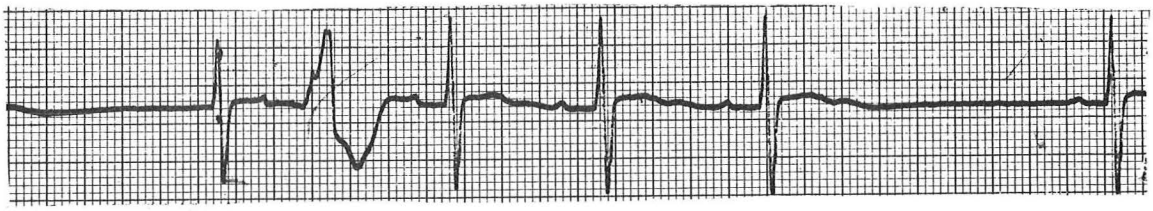
This rhythm strip was obtained on admission.



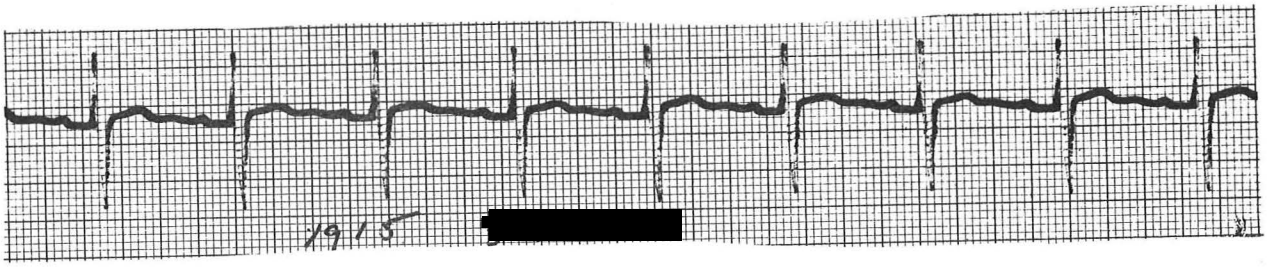


Thorazine and fluids were administered

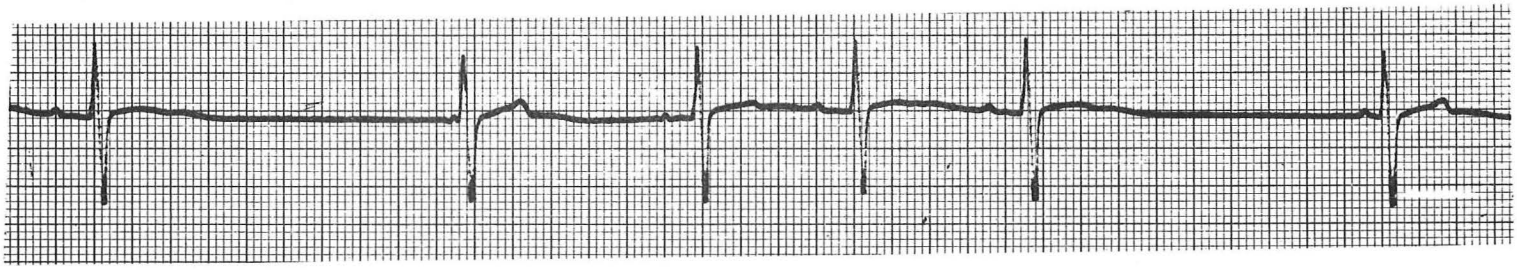




He was given Atropine 1 mg I.V. and briefly developed a sinus tachycardia.

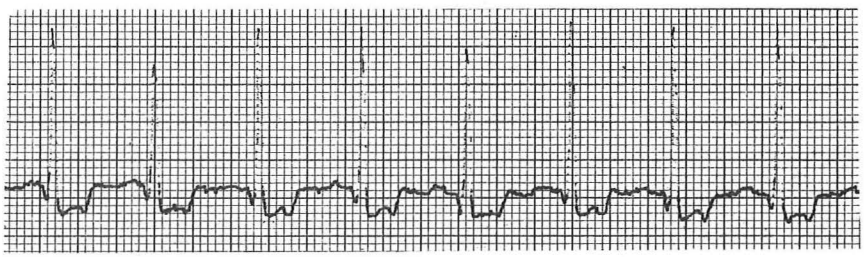


He returned to his atrial arrhythmia.



Since he maintained a normal blood pressure and urine output, was not orthostatic and did not develop dizziness or syncope, it was elected not to use a transvenous pacemaker. It was assumed that he had developed ischemia of his SA node secondary to sickle cell sludging in his sinus nodal artery.

Within 2 days he had returned to a normal sinus rhythm and was asymptomatic.



CASE REPORT #4

Glossopharyngeal Neuralgia with Cardiac Syncope

[redacted]. [redacted]/69

A 46-year-old woman was admitted to [redacted] for the first time in [redacted], 1969. She had been seen as an outpatient at another hospital 7 years previously when she complained of paroxysms of severe pain below the right ear radiating down to the neck and up into the face, lasting no more than a minute. A diagnosis was not made and the symptoms disappeared spontaneously within a few days.

She remained symptom free for a year. The pain then began to recur at irregular intervals, lasting for one to two weeks with remissions varying from four to six months. Three years after the onset of her pain symptoms, her tonsils and upper right teeth were removed because of chronic infection which was thought to be the cause of the pain but no relief of her paroxysms of pain was noted.

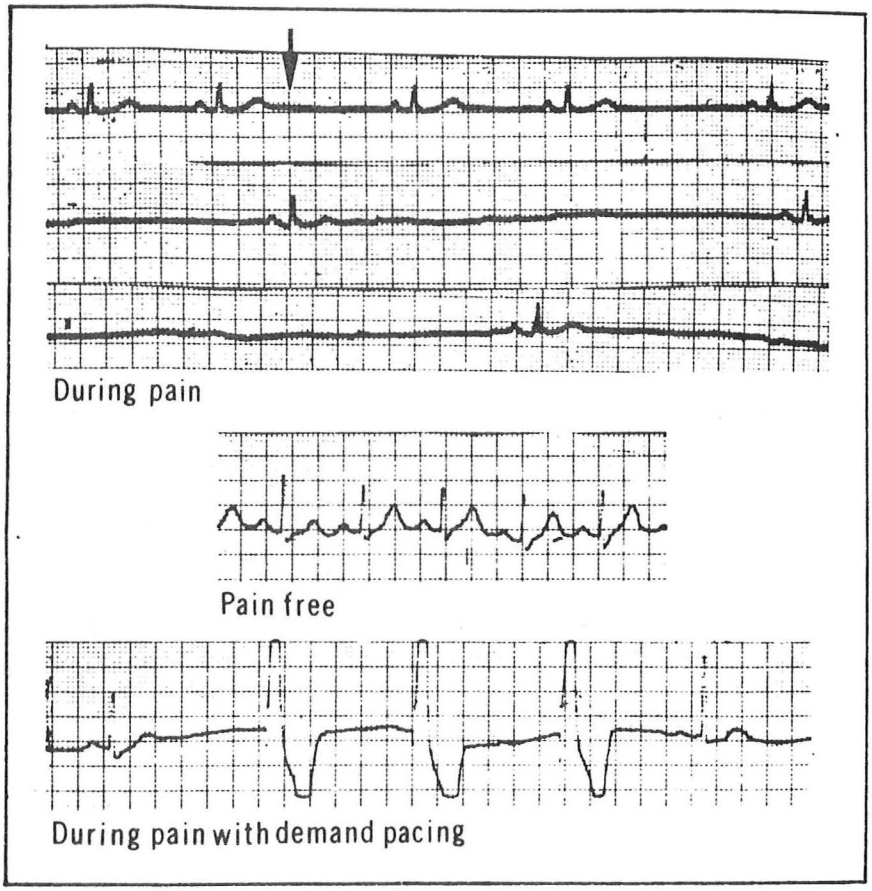
In [redacted], 1969, the patient developed severe pain which was precipitated by swallowing of either solid or liquid foods, especially cold liquids. The symptoms became progressively more severe and in the 15 days prior to admission, paroxysms of pain appeared six to eight times a day, lasting one to two minutes. For the first time, the paroxysms of pain were associated with an occasional loss of consciousness, but no convulsions occurred. The syncopal attacks occurred several times, always following severe episodes of pain, and were unassociated with other neurological manifestations.

While in the emergency room waiting to be examined, the patient had an attack of pain which resulted in loss of consciousness. An ECG recorded sinus arrest and sinus bradycardia. She was given 0.4 mg Atropine intravenously and responded with a sinus rate of 100 beats/minute. Massage of the carotid sinus did not cause bradycardia, but when pain was produced by touching the right tonsillar fossa, sinus bradycardia developed. A diagnosis of glossopharyngeal neuralgia was made.

Other findings on physical examination were within normal limits. The white blood cell count, and levels of fasting blood glucose, blood urea nitrogen, and serum electrolytes, were normal; findings of serologic study and urinalysis were normal.

A temporary transvenous demand pacemaker was inserted via the subclavian vein percutaneously to the right ventricular apex for use as a standby pacemaker in the event of paroxysms of pain and bradycardia. She was placed on a continuous cardiac monitor and started on diphenylhydantoin 100 mg three times daily. During

this period, the patient continued to have spontaneous attacks of neuralgia associated with bradycardia, but syncopal attacks were prevented by demand pacing. After two weeks of ineffective medical therapy with dosage of diphenylhydantoin up to 500 mg daily, right occipital craniotomy and resection of the right glossopharyngeal nerve were performed without complications. She has had no further attacks following the resection and has remained symptom free for 12 months without medication.



Patient's electrocardiogram. Top, *Taken during an attack of pain showing sinus bradycardia and sinus arrest;* center, *when free of pain;* bottom, *during pain after insertion of a pacemaker. Arrow* (top) *indicates time of onset of pain.*