



Recognising sick sinus syndrome

KAREN PHILLIPS MB BS(Hons), FRACP

Sick sinus syndrome is not a life-threatening condition but it can cause significant morbidity, especially in the elderly. Permanent pacemakers remain the treatment of choice.

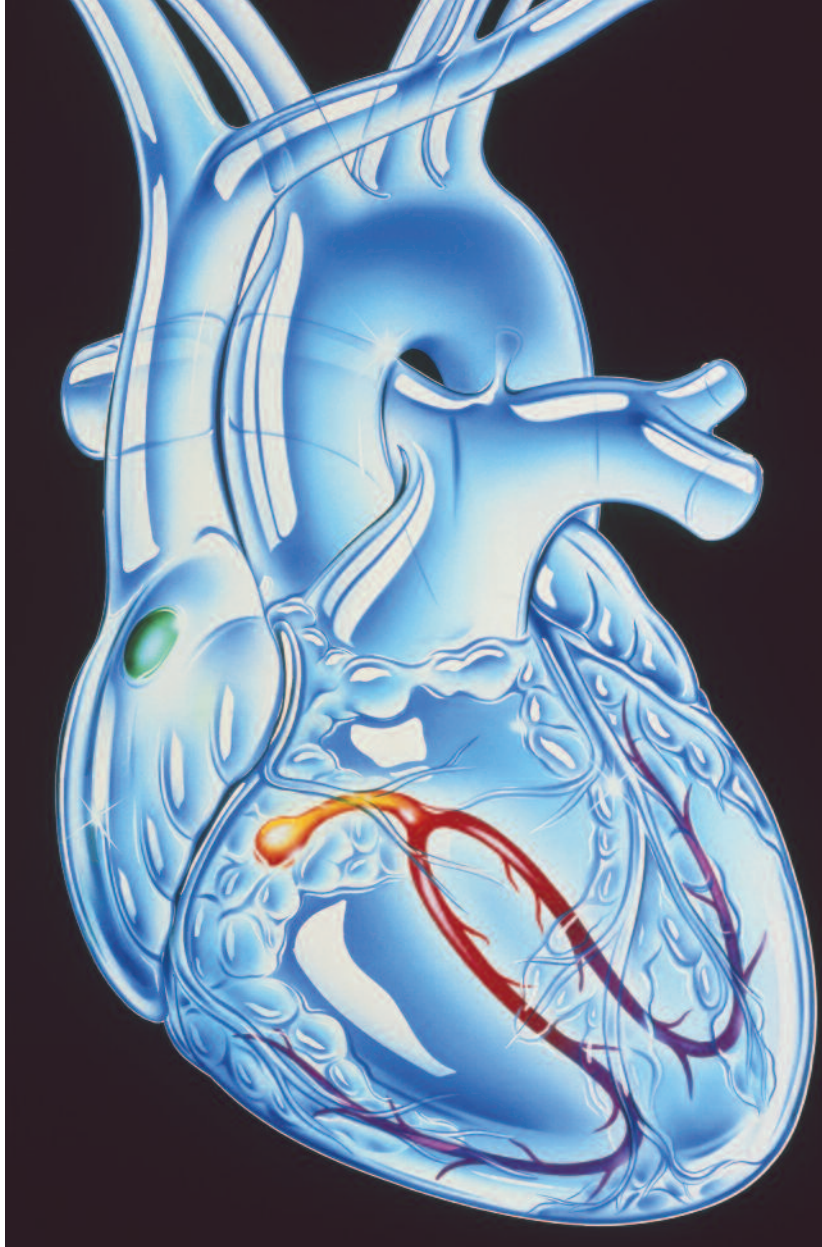
Sick sinus syndrome is a condition caused by dysfunction of the sinus node that results in physiologically inappropriate sinus bradycardia. Although not life-threatening, the symptoms of sick sinus syndrome can lead to significant morbidity and poor quality of life, especially in the ageing population.

The prevalence of sick sinus syndrome increases with age, with a mean age of 68 years at time of diagnosis.¹ It is the most common indication for the implantation of a permanent pacemaker in Australia, accounting for about 40% of new implants. It can be a common cause of syncope leading to falls in the elderly. Although it may be associated with or result from other cardiac conditions, sick sinus syndrome, by itself, is not associated with an increased risk of mortality. Diagnosis and treatment, therefore, has a strong emphasis on recognition and management of symptoms.

Causes

The incidence of sinus node dysfunction increases exponentially with age. Sick sinus syndrome may be caused by a number of conditions, all of which result in either intrinsic or extrinsic impairment of sinus node impulse creation or propagation. Degenerative or fibrotic age-related changes in the heart account for most cases of sick sinus syndrome. Other potential causes of sick sinus syndrome include infiltrative diseases, metabolic conditions, medication use and neuromuscular syndromes (see the box on page 30).

There is increasing interest in the role of genetic abnormalities to explain apparently inheritable forms of sinus node dysfunction or sporadic forms that affect young adults or even children. Abnormalities in cardiac ion channels, which are responsible for normal pacemaker depolarisation currents and cell excitability, have been



Key points

- The prevalence of sick sinus syndrome increases with age and it is the most common indication for implantation of a permanent pacemaker.
- Symptoms of sick sinus syndrome may be intermittent and nonspecific – for example, fatigue, confusion and exercise intolerance.
- ECG monitoring and a high level of suspicion remain the cornerstones of diagnosis.
- Exercise testing can be helpful to diagnose chronotropic incompetence causing exertional symptoms.

CARDIOLOGY TODAY 2013; 3(1): 29-31

Dr Phillips is a Cardiologist and Cardiac Electrophysiologist in private practice at Heart Care Partners, Brisbane; and based predominantly at Greenslopes Private Hospital, Brisbane, Qld.



Symptoms of sick sinus syndrome

- Fatigue
- Lightheadedness and 'dizziness'
- Syncope
- Exertional symptoms such as dyspnoea and exhaustion
- Confusion or irritability
- Exacerbation of cognitive dysfunction (e.g. 'worsening' dementia and memory loss)

Causes of sick sinus syndrome

- Idiopathic/degenerative fibrosis
- Cardiomyopathy/heart failure
- Atrial tachyarrhythmias
 - 'tachycardia-bradycardia syndrome'
- Infiltrative
 - sarcoidosis
 - amyloidosis
 - hereditary haemochromatosis
- Inflammatory
 - myocarditis
 - collagen vascular disease
 - hyperkalaemia
- Medications
 - β -blockers
 - calcium channel blockers
 - digoxin
 - antiarrhythmic drugs
 - clonidine
 - methyl dopa
 - lithium
- Neuromuscular
 - Friedreich's ataxia
 - muscular dystrophy
- Genetic
 - SCN5a sodium channel mutation
 - HCN4 pacemaker ion channel mutation
 - ankyrin-B mutation (membrane protein for sodium pump)

identified in some cases of familial sick sinus syndrome. Potentially reversible detrimental changes in sinus node function are also known to occur in patients with congestive cardiac failure or atrial fibrillation and are mediated by changes in cardiac ion channel function in the presence of cardiac pathology. In patients with these conditions, sinus node dysfunction is frequently exacerbated by use of medications with 'rate-lowering' effects, such as β -blockers, diltiazem and verapamil, which are required to treat these patients.

Clinical manifestations

Sinus node dysfunction may manifest as either inappropriate resting sinus bradycardia, abrupt sinus node pauses or chronotropic incompetence during activity. In some patients, there may be atrial, junctional or ventricular 'escape rhythms' that compensate for bradycardia. The dysfunction, and therefore the symptoms, may be intermittent and unpredictable especially in the earlier stages of sick sinus syndrome (see the box on this page). Frequently, symptoms may also be vague and nonspecific, especially in the elderly, leading to difficulty or a delay in establishing the diagnosis.

In general, only small numbers of patients present with classic dizzy spells or syncope to suggest a bradyarrhythmia. Common symptoms include fatigue, complaints of low energy level and exercise intolerance, including exertional breathlessness or lightheadedness and exhaustion. Sinus node dysfunction can give rise to a range of neurological symptoms due to cerebral hypoperfusion, especially in the elderly. These include episodic confusion and irritability, exacerbation of cognitive dysfunction, such as apparent worsening dementia, and memory loss. Sinus node dysfunction does not infrequently occur in patients with paroxysmal atrial fibrillation and may manifest most conspicuously as symptomatic prolonged sinus node pauses following termination of an episode of atrial fibrillation (Figure 1). Sick sinus syndrome in this setting may be referred to as 'tachycardia-bradycardia syndrome.'

Investigations and diagnosis

The cornerstone of diagnosis of sick sinus syndrome remains ECG monitoring and correlation of symptoms with inappropriate sinus node function. Sick sinus syndrome may be suspected from marked resting bradycardia on examination (heart rate less than 50 beats per minute). A 12-lead ECG and 24-hour Holter or multiday cardiac rhythm monitoring should be considered as basic investigations. Exercise testing may be considered for patients with exertional symptoms. In some instances, invasive cardiac electrophysiological testing of sinus node function and autonomic testing can be considered. Echocardiography is usually an important ancillary investigation to assess for other structural heart diseases.

The diagnosis of sick sinus syndrome requires correlation of symptoms with documented evidence of abnormal sinus node function. Frequent or significant sinus node pauses (greater than three seconds) in the awake patient or the presence of junctional escape rhythms (i.e. the atrioventricular junction generates a spontaneous rhythm to compensate for the lack of atrial rhythm;

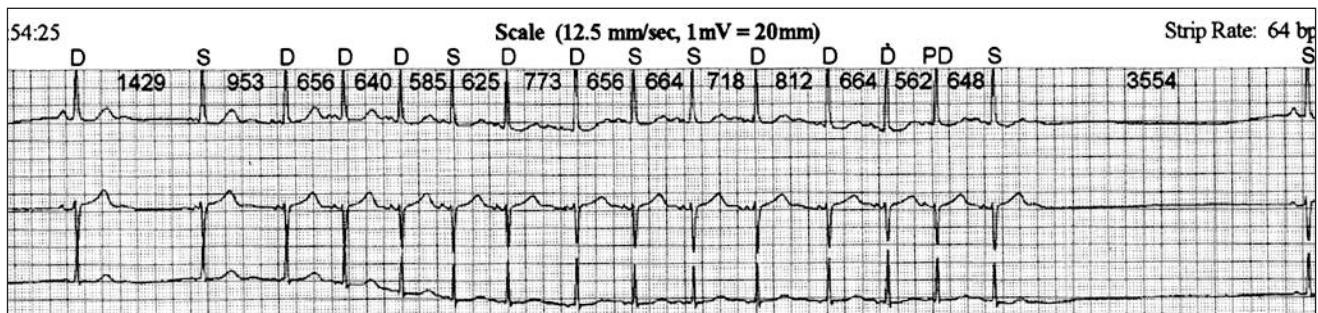


Figure 1. ECG showing an episode of tachycardia bradycardia. A prolonged sinus pause follows a burst of atrial tachycardia. Timing intervals are in msec.

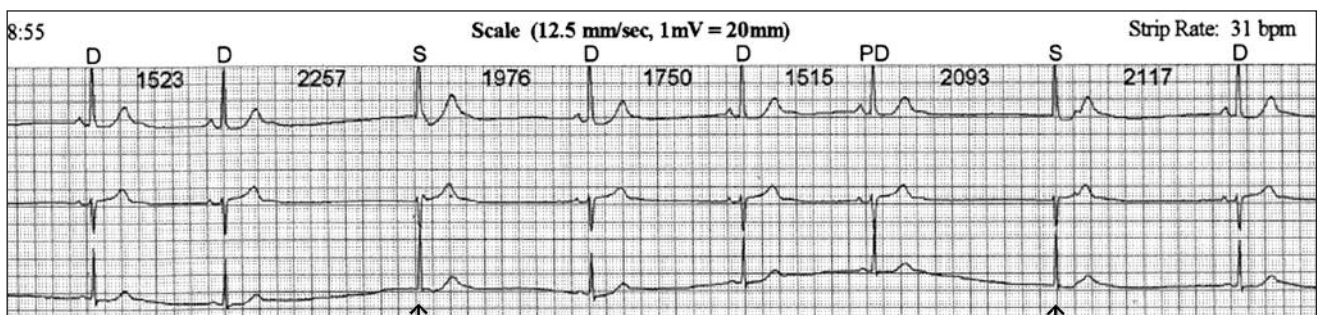


Figure 2. ECG showing the onset of junctional escape rhythm following progressive slowing in the sinus rate. Timing intervals are in msec.

Figure 2) strongly suggest sinus node dysfunction, and may be detected on resting 12-lead ECG or Holter monitors. Abnormal chronotropic response to exercise testing (in the absence of rate-lowering medications) is characterised by the inability to achieve 80% of the maximal age-predicted heart rate (calculated using the formula $220 \text{ beats per minute} - \text{patient age}$). Alternatively, chronotropic incompetence may be suggested by a 'blunted' daily heart rate range on Holter monitoring – a rule of thumb is a maximum heart rate of under 100 beats per minute during normal activity in the absence of rate-lowering medications.

Management

When managing patients with sick sinus syndrome, withdrawal of all medications contributing to sinus bradycardia, where appropriate, should be considered in the first instance. Recognition and management of any underlying conditions is also important (see the box on page 30). For most patients, however, the condition will be idiopathic and the single most effective therapy to relieve symptoms and improve quality of life is implantation of a permanent pacemaker. The most 'physiological' mode of permanent pacing is atrial based (either a single-chamber atrial pacemaker or dual-chamber atrioventricular pacemaker) with 'rate-responsive' capabilities to best mimic the usual sinus node function. Contemporary pacemakers are equipped with 'sensors' that can detect when patients are exercising and increase the pacing rate appropriately.

Although uncommon, complications from permanent pacemaker

therapy include pneumothorax, cardiac perforation, venous thrombosis, infection and malfunction of pacemaker leads. The risk to benefit ratio for this therapy is greatly enhanced with careful diagnosis and recognition of symptoms caused by sick sinus syndrome.²

Future challenges

As we improve our understanding of the structure and function of the sinus node, there is the future prospect that specific treatments may become available for sick sinus syndrome that will avoid the requirement for an artificial permanent pacemaker. Research is currently investigating whether manipulation or replacement of defective cardiac ion channel function with pharmacological agents or gene therapy might provide future therapy options for patients with sick sinus syndrome.³

CT

References

- Rodriguez RD, Schocken DD. Update on sick sinus syndrome, a cardiac disorder of aging. *Geriatrics* 1990; 45: 26-30.
- Epstein AE, DiMarco JP, Ellenbogen KA, et al. ACC/AHA/HRS 2008 guidelines for device-based therapy of cardiac rhythm abnormalities: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Revise the ACC/AHA/NASPE 2002 Guideline Update for Implantation of Cardiac Pacemakers and Antiarrhythmia Devices) developed in collaboration with the American Association for Thoracic Surgery and Society of Thoracic Surgeons. *J Am Coll Cardiol* 2008; 51: e1-62.
- Dobrzynski H, Boyett MR, Anderson RH. New insights into pacemaker activity: promoting understanding of sick sinus syndrome. *Circulation* 2007; 115: 1921-1932.

COMPETING INTERESTS: None.