

Sick sinus syndrome in childhood

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SUMMARY The clinical and electrocardiographic findings in five children with the sick sinus syndrome and an otherwise normal heart are described. There were three boys and two girls. Their age at onset of either bradycardia or symptoms ranged from 1 day to 7 years. In one patient, the youngest ever reported with this syndrome, bradycardia was noted before birth. Four children presented with neurological symptoms—attacks of dizziness, fainting spells, or syncope. One boy, treated for epilepsy before the underlying arrhythmia was diagnosed, died suddenly while playing. One child had near-fatal syncope caused by ventricular tachycardia. Continuous 24-hour electrocardiographic monitoring is the best method of assessing the severity of the condition. Sinus bradycardia, sinuatrial block, and periods of sinus arrest up to 4.8 seconds were recorded. Two patients had associated atrioventricular block and were therefore presumed to have binodal disease. Atrial fibrillation or flutter occurred in three patients. Isolated sick sinus syndrome may be a life-threatening condition in childhood for which, in selected cases, the insertion of a permanent pacemaker is indicated.

Most reports on the sick sinus syndrome deal exclusively with adults or the elderly¹⁻⁴; in childhood the condition is rare. It has been reported to occur after intra-atrial operations with injury to the sinuatrial node or its blood supply, particularly after the Mustard operation⁵⁻⁷ and also in children with various congenital cardiac malformations,⁸⁻¹⁰ but occasionally it occurs in children and adolescents with no other evidence of heart disease.^{10 11} The latter condition, which is probably of congenital origin, is not sufficiently recognised for a number of reasons. The presenting symptoms, produced by intermittent cerebral ischaemia, usually mimic central nervous disorders. With the exception of bradycardia, which is often intermittent, there is frequently no abnormality on physical examination and routine electrocardiograms. Finally, the syndrome is generally considered to occur only in adults. Its diagnosis in children, however, is important, for the arrhythmia may threaten life and need specific treatment.

In this study, five children with the sick sinus syndrome are presented. The wide range and variability of symptoms and the value of continuous electrocardiographic monitoring in diagnosis are emphasised.

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Patients and methods

Between January 1967 and January 1979, five children with severe bradycardia were seen. None had clinical evidence of structural heart disease. Only one, being treated for epilepsy, was taking drugs. All had routine electrocardiograms taken and, four, continuous 24-hour electrocardiographic tape recordings. Each patient met at least two of the following three criteria: (1) long periods of bradycardia, defined as a heart rate of 40 beats/min or less lasting one minute or more, (2) episodes of sinus arrest of three seconds or longer, (3) episodes of 2:1 or 3:1 sinuatrial block. There were three boys and two girls. Their ages, at the onset of bradycardia or symptoms, ranged from 1 day to 7 years (Table 1).

For 24-hour monitoring a two-channel Medilog tape recorder with build-in reference time signal (Oxford Medical Instruments) was used. Recordings were played back on a semiautomatic high-speed analyser, giving a visual display of the heart rhythm on a 40 seconds memory oscilloscope. Arrhythmias were analysed on direct write-out traces. The children over 6 years of age had an exercise test on a bicycle ergometer. The younger children were asked to run a distance of 150 metres. Electrophysiological studies were performed in three

Table 1 Clinical data in five children with isolated sick sinus syndrome

Case No.	Age at onset of symptoms (y)	Age at which bradycardia first noted (y)	Symptoms	Treatment
1	5	8	Epileptic fits, died suddenly aged 10	None
2	7	7	Syncope, dizziness	Isoprenaline, anticoagulants
3	5	4	First syncope near-fatal	Permanent pacemaker
4	—	1 day	None	Permanent pacemaker
5	6	12	Syncope	Isoprenaline

children. His bundle recordings were obtained by the method of Scherlag *et al.*¹² Sinus recovery times were recorded after high atrial pacing for three minutes. For programmed electrical stimulation the JSI-stimulator (Janssen Scientific Instruments) was used. In two patients, studied after the implantation of a permanent cardiac pacemaker, the underlying escape rhythm was assessed by inhibition of the pacemaker using chest wall stimuli produced by a Grass stimulator with variable impulse duration and voltage.

Case reports

The salient clinical and electrocardiographic features are summarised in Tables 1, 2, and 3. In view of the scarcity of similar published reports and the wide spectrum of symptoms, the case histories will be presented in detail.

Table 2 Electrocardiographic findings on routine and 24-hour electrocardiograms

Case No.	Age (y)	Slowest heart rate (beats/min)	Sinuatrial block	Longest asystole (s)
1	8	40	Sinus arrest	1.5
2	12	24	2:1 and 3:1	4.2
3	4	22	3:1	3.5
4	4	30	2:1	4.8
5	12	32	2:1	2

CASE 1

A 5-year-old boy began to have short attacks of syncope which were thought to be epilepsy and were treated with diphenylhydantoin. In July 1967 at the age of 8 routine examination disclosed irregularity of the pulse. Apart from the apparent epileptic fits he was free of symptoms. There was no history of syncope or sudden death in other members of the family. On examination the only abnormality was a slow and irregular pulse between 40 and 50 beats/minute. The electrocardiogram showed slow atrioventricular junctional rhythm

with long periods of bigeminy (Fig. 1). Although long traces were recorded, at no time could normal sinus rhythm be recorded. In this boy, the possibility of a link between the arrhythmia and the syncopal attacks was not entertained. At the age of 10 he died suddenly while playing.

CASE 2

A 7-year-old white girl was first examined at the University Hospital of Kinshasa, Zaire, in 1968, because of attacks of dizziness and a slow heart rate. The family history was unremarkable. The electrocardiogram showed slow sinus rhythm, often below 40/min, atrial premature beats, and atrioventricular junctional escape beats. The bradycardia and fainting attacks were attributed to an excess of vagal tone. Isoprenaline, six times daily, was given and seemed to decrease the dizziness. In 1968 and 1969, however, two short episodes of loss of consciousness unrelated to effort were witnessed by her parents. When she was first seen by us in July 1969, the heart rate was 40 to 50/minute. A grade 2/6 ejection murmur and a short diastolic rumble at the apex were heard. As there was no other clinical evidence of heart disease, these murmurs were attributed to the slow heart rate and increased stroke volume. The electrocardiogram showed slow sinus rhythm, a PQ interval of 0.18–0.20 second and long atrial pauses of 1.8 to 2 seconds. On exercise the heart rate increased to 120/minute but, within 15 seconds, it slowed down and became irregular, a result of second and third degree sinuatrial block. From 1969 to 1974 she took no drugs. In 1974 she was admitted for further in-

Table 3 Arrhythmias in five children with congenital sick sinus syndrome

Type of arrhythmia	No. of patients
Sinus bradycardia 40/min or below	5
Sinuatrial block (2nd or 3rd degree)	5
Predominant atrioventricular junctional rhythm	1
Atrial flutter or fibrillation	3
Ventricular tachycardia and fibrillation	1
Atrioventricular block (first or second degree)	2

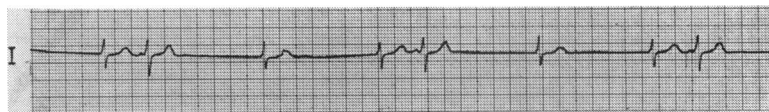


Fig. 1 Electrocardiogram lead I from 8-year-old boy (case 1) showing slow junctional rhythm. The intermittent P waves and occurrence of bigeminy may be explained by slow retrograde conduction to the atria and reciprocal beats. Though long traces were obtained, at no time could normal sinus rhythm be recorded. The small irregularity of the T wave in complex 3 is artefactual.

vestigation. Continuous monitoring showed extremely long pauses, up to four seconds, particularly at night (Fig. 2). In view of the extreme bradycardia, the insertion of a permanent pacemaker was recommended but this was refused by the parents as being unnecessary in an apparently healthy girl. In view of the risk of thromboembolic complications anticoagulants were started in 1974 and continued until the present. Cardiac rhythm remained unstable. In 1975 atrial flutter with 3:1 and varying atrioventricular conduction was recorded and a few weeks later slow sinus bradycardia with pauses up to three seconds. In 1976 and when last seen in 1978, once again she had atrial flutter, but now with varying atrioventricular conduction (Fig. 3). In the last 9 years under our care, she has had no further attacks of syncope. Her only symptom is an occasional episode of dizziness.

CASE 3

In December 1976 a 4-year-old boy was referred by the family doctor who had noted a slow and

irregular heart rate during a fever. He had never fainted or had a fit. On examination the only abnormality was a bradycardia between 30 and 40/minute. The electrocardiogram showed sinus bradycardia and high-grade sinoatrial block (Fig. 4). In February 1977 regular sinus rhythm at 72/minute was noted. Four months later long periods of bradycardia were again recorded. Exercise increased the heart rate to 80 to 84/minute but this was followed within seconds by bradycardia, with long pauses caused by 3:1 sinoatrial block.

In September 1977 he collapsed while playing at school. According to his teacher he fell suddenly, and was found to be unconscious, and not breathing. He was given mouth-to-mouth respiration and within 10 minutes rushed to the emergency department of the hospital. On arrival he was still unconscious. The electrocardiogram showed periods of ventricular tachycardia (Fig. 5). He was intubated and cardioversion was carried out. Stable sinus rhythm, however, could not be restored. A few hours later a temporary transvenous pacemaker

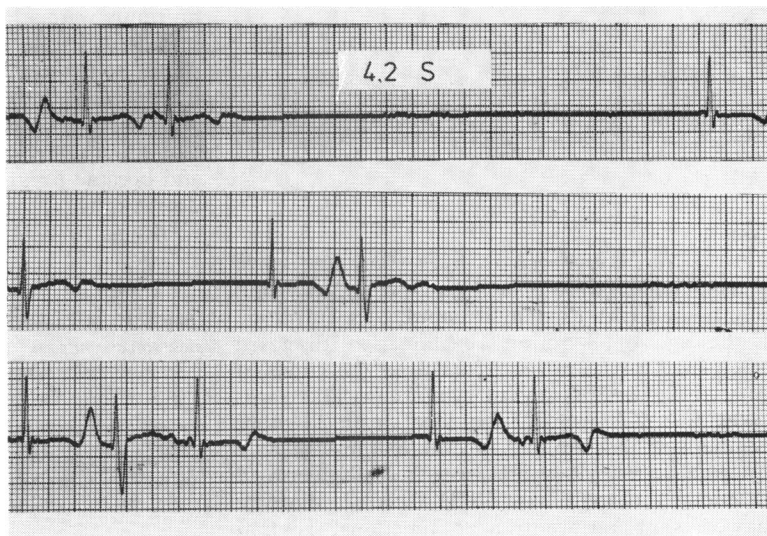


Fig. 2 Continuous strip of a monitoring lead from a 12-year-old girl (case 2). Nocturnal heart rate dropped below 30 beats/min. The longest asystolic period (upper panel) was 4.2 seconds.

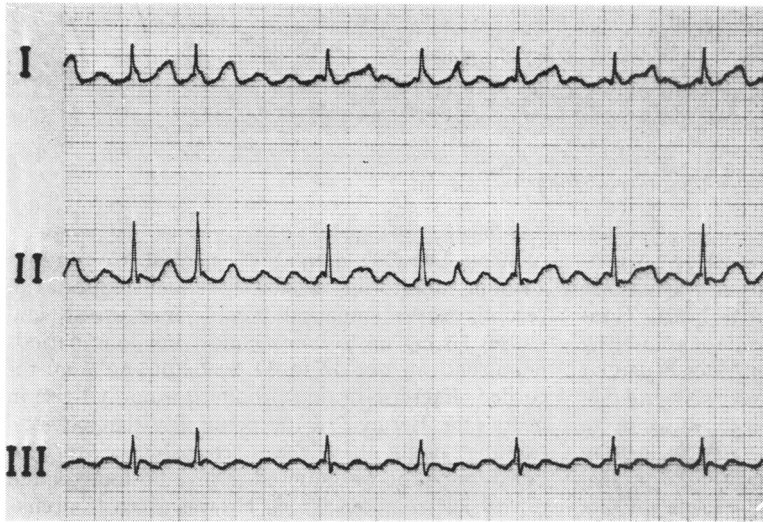


Fig. 3 Leads I, II, and III in the same patient (case 2) two years later, showing atrial flutter with varying atrioventricular conduction.

had to be introduced because of repeated episodes of ventricular tachycardia alternating with periods of atrial and ventricular standstill. The next day a permanent lithium demand pacemaker was inserted and extubation was possible. For three days he remained semiconscious and his gait was uncertain for several weeks. He eventually recovered completely. Subsequent electrocardiograms showed that the pacemaker was intermittently inactive, because of atrial flutter or fibrillation with a rapid ventricular response. This was treated with digoxin and quinidine.

CASE 4

A healthy newborn girl had had a slow fetal heart rate of 60/minute during the last days of pregnancy and during labour. At birth the rate remained at

60/minute but it increased to 90/minute over the next few days. When she was first examined at our department at 2 months of age, she was in regular sinus rhythm at 80/minute. Her rate increased to only 100/minute when she cried. At 14 months of age slow sinus rhythm at 46/minute was recorded. The PQ interval was 0.16 second. A few blocked P waves were noted. At the age of 3 fine atrial fibrillation with a ventricular response of 42/minute was observed, and this increased to only 52/minute when she ran 150 metres. On auscultation a grade 3/6 ejection murmur at the pulmonary area and a diastolic rumble at the apex were heard. These murmurs and cardiomegaly on the chest x-ray were attributed to the extreme bradycardia. Cardiac catheterisation showed no abnormality except a slightly increased systolic pressure in the pul-

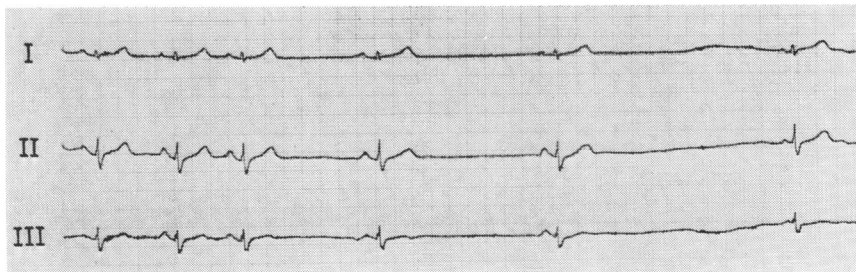


Fig. 4 Leads I, II, and III from a 4-year-old boy (case 3). Progressive shortening of the sinus cycle (complexes 1 to 3) is followed by overt sinoatrial block. The third PP interval is exactly twice the second PP interval. The fourth PP interval equals three times the second PP interval minus 220 ms. This suggests coexisting first degree and 3:1 sinoatrial block. The morphology of the sixth P wave is different. It is probably an atrial escape beat.



Fig. 5 Lead II recorded in a 4-year-old boy (same case as in Fig. 4) during an episode of coma, after sudden collapse. Accelerated idioventricular rhythm leads to ventricular tachycardia.

monary artery (36/12, mean 18 mmHg). Angiocardiology showed enlargement of the cardiac chambers but an otherwise normal heart. Continuous 24-hour electrocardiographic monitoring showed fine atrial fibrillation with an irregular ventricular response varying between 30 and 96/minute by day, with occasional longer pauses up to four seconds. During the night the ventricular rate varied between 15 and 45/minute with exceptional peaks at 70/minute. One pause of 4.8, one of 4.6, and numerous pauses of more than 3 seconds were recorded. In spite of the extreme bradycardia the child was active and completely free of symptoms. This made management difficult as the parents were reluctant to accept that the child's condition was serious and potentially lethal, but in view of our experience with the previous patients insertion of a permanent pacemaker was advised. This has been carried out recently.

CASE 5

A 12-year-old boy gave a history of 10 to 12 brief syncopal attacks which had started at the age of 6 years. There had been no prodromal symptoms nor was there any relation to effort. He was a keen footballer and took part in various sports without symptoms. Physical examination was normal. The electrocardiogram showed regular sinus rhythm at 76 beats/minute and a prolonged PR interval

(0.30 s). The QT interval was normal. Continuous 24-hour monitoring disclosed frequent episodes of second degree sinuatrial block, sinus arrest, and pauses lasting up to two seconds. For long periods nocturnal rates of 32 to 34 beats/minute were recorded. Exercise testing produced a normal rise in heart rate. Because of the possibility that physical training might have been contributing to this bradycardia, he was advised to give up football and to restrict strenuous effort. He was given isoprenaline 15 mg 12-hourly and has had no further syncopal attacks in the last two years.

ELECTROPHYSIOLOGICAL STUDIES

Electrophysiological data were available in three patients. In case 2 the atrioventricular conduction and the sinus recovery time were normal. In case 3 only data obtained by chest wall stimulation were available. Inhibition of the permanent pacemaker produced atrial and ventricular standstill for 3.7 seconds. This observation shows the unreliability of subsidiary escape mechanisms when the sinus node is failing. In case 4 intracardiac electrocardiography disclosed fine atrial fibrillation. After three minutes of right ventricular stimulation, ventricular standstill of 5.4 seconds occurred when pacing was stopped (Fig. 6). Similarly, when the permanent pacemaker was inhibited by chest wall stimulation, a pause of 3.5 seconds was recorded.

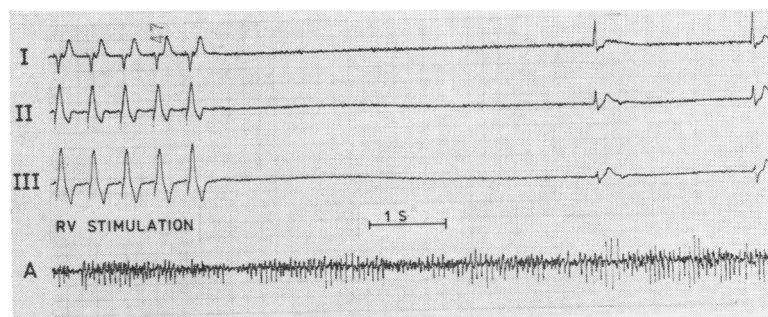


Fig. 6 Leads I, II, III, and an intracardiac right atrial lead (A) in a 3-year-old girl (case 4) known since birth to have sick sinus syndrome and atrioventricular node dysfunction. The basic rhythm is fine atrial fibrillation. When intracardiac right ventricular stimulation (the first five beats) is stopped, it takes 5.4 seconds before a junctional escape beat appears.

Discussion

The sick sinus syndrome in childhood is a well-known complication of cardiac surgery.^{5,6} Its association with cardiac malformations⁸⁻¹⁰ and with myocarditis^{7,10} is equally well documented. In the present series, however, no underlying or associated heart disease could be detected. The term isolated sick sinus syndrome seems therefore appropriate. Approximately 20 similar cases have been published, a few well-documented case reports¹³⁻¹⁷ and one extensive study.¹¹ In two other papers, dealing mainly with postoperative sinus node dysfunction, the occurrence of this syndrome in a few otherwise healthy children is mentioned.^{7,10}

For inclusion in our small series deliberately strict criteria were applied, as little is known about the physiological variations of cardiac rhythm in normal infants and children. According to a recent study¹⁸ the slowest heart rate in 134 randomly selected neonates was 93 ± 12 beats per minute. In only one was bradycardia below 50 beats per minute and a systolic pause of 1.8 seconds recorded. In sleeping young adults rates may be as low as 33 beats per minute, but in only two out of 50 individuals were pauses of two seconds recorded.¹⁹ It is therefore unlikely that any of our patients had physiological bradycardia.

The age at onset of symptoms in our patients (Table 1) is younger than in most of those previously reported. In one (case 4) bradycardia was noted before birth by the obstetrician, who suspected congenital atrioventricular block. The question therefore arises whether our patients and other young children previously reported^{13,15,16} differ aetiologically from the adolescent group. All the patients reported by Scott *et al.*,¹¹ for instance, were athletic boys between 10 and 15 years. In our series, however, the hypothesis that excessive training might have produced bradycardia can be excluded, as most patients were below the age of 10 and, with the exception of one boy (case 5), did not take part in sports.

Four out of the five children presented with neurological symptoms—dizziness, fainting spells, or syncope (Table 1). One boy (case 1), who later died suddenly while playing, was treated for epilepsy before the underlying arrhythmia was diagnosed. Another (case 3), aged 5, had no symptoms until he suddenly collapsed with ventricular tachycardia, again while playing. Syncopal attacks on exertion were also noted by Scott *et al.*¹¹ and sudden death reported in two further cases.^{15,20} Cerebrovascular accidents, common in adults with the bradycardia-tachycardia syndrome,² were not encountered in our patients but Onat *et al.*¹³ described right-sided

hemiparesis and facial weakness, presumably secondary to cerebral embolism, in a 3-year-old girl.

Three of the four children exercised were unable to increase their heart rate more than 30 per cent. An inappropriate response to exercise is generally considered to be a simple and reliable clue to the diagnosis.^{9,11,15} Similarly, the response to the intravenous administration of atropine, which was not studied in our patients, is reported to be abnormal.^{3,9,11} Continuous electrocardiographic monitoring, however, provides, in our experience, the best tool for evaluating patients suspected of having sinus node dysfunction. The information gained by this method is important from two points of view. In the first place, bradycardias and long pauses (Table 2), often unsuspected on the routine electrocardiograms, may be documented—in three of the children nocturnal rates dropped below 30 beats per minute and asystoles as long as 4.2, 3.5, and 4.8 seconds were recorded. Secondly, the full spectrum of arrhythmias is detected (Table 3)—atrial flutter or fibrillation was observed in three patients.

Supraventricular arrhythmias are so common in adults with the sick sinus syndrome²⁻⁴ that it led to the use of the alternative term the bradycardia-tachycardia syndrome. They are also prominent in children with the postoperative sick sinus syndrome.^{5,6}

By contrast, ventricular tachycardia or fibrillation have not been documented in large series of adults,^{2,4} but seem to occur in children. In one patient (case 3, Fig. 5), the former caused a near-fatal syncope. Chaotic ventricular rhythm has also been reported in six previous cases.^{7,14,15} Obviously, the association is too common to be fortuitous. It has been suggested that bradycardia favours desynchronisation of cardiac activation and thus the occurrence of ventricular tachycardia.^{21,22} The finding that two patients had delayed atrioventricular conduction and therefore, presumably, binodal disease is of particular interest. This has also been found in three other children with this syndrome.^{7,13,16}

Electrophysiological studies were of limited value. In one patient with severe bradycardia in whom a complete study was performed, atrioventricular conduction and sinus node recovery times were normal.

A previous study found a considerably prolonged sinus node recovery time in one child, but a normal one in another.²³ The occurrence of "false negative" electrophysiological testing and its interpretation have recently been reviewed in adults.²⁴ From the practical viewpoint, we accepted the diagnosis of the sick sinus syndrome on the basis of abnormal 24-hour electrocardiographic monitoring and exercise testing, even in the presence of a normal

sinus node recovery time. In children with a permanent pacemaker, the spontaneous activity of the sinus node or subsidiary pacemakers can be assessed when the artificial pacemaker is temporarily inhibited by chest wall stimuli.

The histopathology of isolated sick sinus syndrome in children and adolescents has not been defined. The only pathological study, by James *et al.*,²⁵ found intimal proliferation and medial hypertrophy in the sinus node artery of two young athletes who died suddenly during exertion. The pre-existing rhythm, unfortunately, was not well documented. It appears reasonable, for a number of reasons, to postulate a congenital defect of the sinus node as the basic lesion, namely: (1) the syndrome may present in early childhood and, as documented for the first time in our study, even at birth, (2) it is occasionally associated with other cardiac malformations,⁷⁻⁹ (3) more than one case may occur in a single family.^{14 26 27} Moreover, a few families have been described in which both sinuatrial and atrioventricular node dysfunction were inherited.^{26 28}

Because of the small number of children with isolated sick sinus syndrome so far described, and the wide spectrum of symptoms, a therapeutic policy is difficult to establish. Based on our experience in which one child died suddenly and another suffered a life-threatening ventricular arrhythmia we consider that permanent cardiac pacing is the only reliable treatment if symptoms are present or if they are not but the heart rate is very slow. Williams *et al.*²⁰ concur. In patients with tachycardia antiarrhythmic drugs and, possibly, anticoagulants should be used.

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