Sick sinus syndrome
Symptomatic cases in children

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Radford, D. J., and Izukawa, T. (1975). Archives of Disease in Childhood, 50, 879. Sick sinus syndrome: symptomatic cases in children. In 20 children needing treatment for symptomatic sick sinus syndrome, the average age at presentation was 7-1 years and ranged from 9 months to 18 years. Symptoms were never precise but, in retrospect, 5 children had syncope, 7 had a rapid heart action, 6 had dyspnoea or tachypnoea, 2 had nonspecific chest pains, 2 had pale spells, and 1 had a sudden hemiplegia. Symptoms followed cardiac surgery in 15 cases and were related to unoperated congenital heart disease in 2 and to myocarditis in 2. The aetiology was unknown in 1 case. The type of cardiac surgery resulting in the development of the sick sinus syndrome was predominantly related to atrial suturing.

Both tachy- and bradydysrhythmias were found, including wandering atrial pacemaker (9 cases), junctional rhythm (19 cases), supraventricular tachycardia (9 cases), atrial flutter (11 cases), and atrial fibrillation (2 cases). Both atrial (8 cases) and ventricular (7 cases) premature beats were seen.

All patients were given trials of drug therapy but difficulties were encountered. Cardioversion was used for tachyarrhythmias in 11 cases without serious problems. Six children had permanent cardiac pacemakers inserted with good results. Recognition of the sick sinus syndrome in childhood is important and treatment must be regulated by the severity of symptoms.

More interest in the sick sinus syndrome has been shown recently and it has been recognized more often (Ferrer, 1973; Kaplan et al., 1973; Lloyd-Mostyn, Kidner, and Oram, 1973; Moss and Davis, 1974; Rubenstein et al., 1972; Radford and Julian, 1974). The condition presents a wide spectrum of clinical manifestations and of problems with management. Most reports have been of adult patients, mostly elderly, and recordings of cases in children are few (Nugent et al., 1974; Scott and Macartney, 1974; Greenwood, et al., 1974). However, the condition occurs in children and its recognition and treatment are important.

Lown (1967) defined the sick sinus syndrome as a defect in elaboration or conduction of sinus impulses characterized by chaotic atrial activity, changing P-wave contour, and bradycardia interspersed with multiple and recurrent ectopic beats and runs of atrial and nodal tachycardia. The diagnosis is essentially an electrocardiographic one. Using Easley and Goldstein's (1971) subclassification, type I sick sinus syndrome is diagnosed in patients exhibiting inappropriate sinus bradycardia or sinus arrest, and type II in those showing a bradycardia-tachycardia combination. We present here 20 cases of the sick sinus syndrome in children who had symptoms associated with their dysrhythmias and who have required treatment.

Patients and methods

A computer write-out of coded arrhythmias in children seen in the Cardiology Department of the Hospital for Sick Children, Toronto, was obtained and the electrocardiograms (ECG) and case records of the patients surveyed. Sequential ECGs of all patients at the hospital who had had the Mustard operation (Mustard et al., 1964) of an intra-atrial baffle for transposition of the great vessels were also reviewed and the dysrhythmias documented (Mulholland et al., 1975). Those with atrial arrhythmias occurring only in the immediate postoperative period were excluded.

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as also were children who had isolated atrial dysrhythmias without symptoms. The remaining 20 patients who had ECG evidence of the sick sinus syndrome and symptoms needing therapy were reviewed.

**Results**

Clinical details of the 20 patients (13 boys, 7 girls) are shown in Table I. Their mean age at onset of symptoms was 7.1 years with a range of 9 months to 18 years. Three had bradyarrhythmias alone (type I sick sinus syndrome) and the other 17 had symptomatic brady-tachycardia (type II).

**Symptoms.** Although the symptoms were often imprecise at the time, in retrospect they can be summarized as follows: 5 children had

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**TABLE I**

Clinical data of 20 children with sick sinus syndrome (SSS)

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Sex</th>
<th>Age at onset (years)</th>
<th>Symptoms</th>
<th>Associated conditions</th>
<th>Cardiac surgery</th>
<th>Time relation of symptoms to surgery</th>
<th>Type of SSS</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>M</td>
<td>7</td>
<td>Syncope, dizziness, Palpitations, dyspnoea (congestive heart failure)</td>
<td>Normal heart (catheter study)</td>
<td>-</td>
<td>I</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>2</td>
<td>M</td>
<td>13</td>
<td>Syncope, dizziness, Palpitations, dyspnoea (congestive heart failure)</td>
<td>Normal heart (clinically)</td>
<td>-</td>
<td>II</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>3</td>
<td>F</td>
<td>4</td>
<td>Syncope, dizziness, Palpitations, dyspnoea (congestive heart failure)</td>
<td>Coxackie viral myocarditis</td>
<td>-</td>
<td>II</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>4</td>
<td>F</td>
<td>5</td>
<td>Mother noted rapid heart failure</td>
<td>Secundum ASD</td>
<td>+</td>
<td>Pre-op</td>
<td>II</td>
<td>+</td>
</tr>
<tr>
<td>5</td>
<td>M</td>
<td>14</td>
<td>Syncope, chest pain</td>
<td>Primum ASD</td>
<td>+</td>
<td>7 years</td>
<td>I</td>
<td>+ +</td>
</tr>
<tr>
<td>6</td>
<td>M</td>
<td>12</td>
<td>Syncope, dizziness, palpitations, Palpitations, dyspnoea</td>
<td>Sinus venous ASD with PAPVD</td>
<td>+</td>
<td>8 years</td>
<td>II</td>
<td>+</td>
</tr>
<tr>
<td>7</td>
<td>M</td>
<td>9</td>
<td>Brief chest pains</td>
<td>Sinus venous ASD with PAPVD</td>
<td>+</td>
<td>2 years</td>
<td>II</td>
<td>+ +</td>
</tr>
<tr>
<td>8</td>
<td>F</td>
<td>12</td>
<td>Palpitations, dyspnoea, Palpitations, dyspnoea</td>
<td>Common atrium with PAPVD</td>
<td>+</td>
<td>Immediate postop</td>
<td>II</td>
<td>+ +</td>
</tr>
<tr>
<td>9</td>
<td>M</td>
<td>5</td>
<td>Palpitations, dyspnoea, Palpitations, dyspnoea</td>
<td>TAPVD</td>
<td>+</td>
<td>2 years 1 year</td>
<td>II</td>
<td>+ +</td>
</tr>
<tr>
<td>10</td>
<td>M</td>
<td>9</td>
<td>Palpitations, dyspnoea, Palpitations, dyspnoea</td>
<td>T of F with left SVC draining to coronary sinus</td>
<td>+</td>
<td>2 years</td>
<td>II</td>
<td>+ +</td>
</tr>
<tr>
<td>11</td>
<td>M</td>
<td>18</td>
<td>Syncope</td>
<td>T of F with pulmonary cusp agenesis</td>
<td>+</td>
<td>11 years</td>
<td>I</td>
<td>+ -</td>
</tr>
<tr>
<td>12</td>
<td>F</td>
<td>11</td>
<td>Syncope</td>
<td>Single ventricle pulmonary atresia, PDA</td>
<td>-</td>
<td>-</td>
<td>II</td>
<td>- +</td>
</tr>
<tr>
<td>13</td>
<td>F</td>
<td>3</td>
<td>Tachypnoea</td>
<td>Single ventricle with TGA double orifice tricuspid</td>
<td>+</td>
<td>2 years</td>
<td>II</td>
<td>+ +</td>
</tr>
<tr>
<td>14</td>
<td>F</td>
<td>5</td>
<td>Dyspnoea (cardiac failure)</td>
<td>Double outlet RV with TGA</td>
<td>+</td>
<td>4 years</td>
<td>II</td>
<td>+ -</td>
</tr>
<tr>
<td>15</td>
<td>F</td>
<td>9/12</td>
<td>Hemiplegia</td>
<td>TGA</td>
<td>+</td>
<td>8/12 I</td>
<td>II</td>
<td>+ -</td>
</tr>
<tr>
<td>16</td>
<td>M</td>
<td>17/12</td>
<td>Pale spells, partial anomalous pulmonary venous drainage</td>
<td>TGA</td>
<td>+</td>
<td>3/12 II</td>
<td>II</td>
<td>+ +</td>
</tr>
<tr>
<td>17</td>
<td>M</td>
<td>3</td>
<td>Partial anomalous pulmonary venous drainage</td>
<td>TGA</td>
<td>+</td>
<td>2 years</td>
<td>II</td>
<td>+ +</td>
</tr>
<tr>
<td>18</td>
<td>M</td>
<td>3</td>
<td>Partial anomalous pulmonary venous drainage</td>
<td>TGA</td>
<td>+</td>
<td>1/12 II</td>
<td>II</td>
<td>+ +</td>
</tr>
<tr>
<td>19</td>
<td>M</td>
<td>4</td>
<td>Partial anomalous pulmonary venous drainage</td>
<td>TGA</td>
<td>+</td>
<td>Immediate postop</td>
<td>II</td>
<td>+ +</td>
</tr>
<tr>
<td>20</td>
<td>M</td>
<td>4</td>
<td>Partial anomalous pulmonary venous drainage</td>
<td>TGA</td>
<td>+</td>
<td>5/12 II</td>
<td>II</td>
<td>+ +</td>
</tr>
</tbody>
</table>

ASD, atrial septal defect; PAPVD, partial anomalous pulmonary venous drainage; TAPVD, total anomalous pulmonary venous drainage; T of F, tetralogy of Fallot; TGA, transposition of great arteries; RV, right ventricle.
Sick sinus syndrome

referred to as ‘syncope of infancy’ or ‘syncope fugax’. It is important to distin-
guish this condition from vasovagal syncope, which is often seen in children
and adults and is not associated with structural heart disease. Since the 1960s,
the term ‘sick sinus syndrome’ has been used to describe patients with
symptoms of syncope, palpitations, or cardiac arrest who have slow heart
rates or arrhythmias. However, the underlying causes and mechanisms of
this condition are not fully understood.

Associated conditions. Associated cardiac defects were recognized in
7 of the 18 cases. Of the 7 children, 2 had had surgical implantation of
atrial septal defect and 5 had had previous pacemaker insertions. These
children had atrial septal defects and had undergone surgery for cardiac
defects. However, not all children with sick sinus syndrome have a
history of cardiac surgery or pacemaker implantation.

Surgery. The type of cardiac surgery resulting in the development of the
sick sinus syndrome was almost entirely related to manipulation and suturing
of the atrial chambers. The three patients with atrial septal defects and the
one with a common atrium had had patch reconstructions of the atrial
septum. The boy with total anomalous pulmonary venous drainage into the
left superior vena cava had had surgical implantation of the anomalous common
venous chamber into the left atrium. The patient with a single ventricle and
transposed relationship of the great vessels had had a surgical creation of an
atrial septal defect (Blalock and Hanlon, 1950). Seven children with transposition
of the great arteries had had redirection of the venous return by the
insertion of an intra-atrial baffle (Mustard et al., 1964). In only one patient was the Mustard
operation the primary surgery and the other five had had previous palliative operation. Two of the
six had had preceding Blalock-Hanlon atrial

surgical implantation and four had had the operation described by Edwards and Barger (1965),
of creating partial anomalous venous return into the right atrium by moving the atrial septum. The
patient who developed a hemiplegia did so 8 months after her Sterling Edwards operation and before the
Mustard repair.

The two patients with tetralogy of Fallot had had total correction operations with patch closure of the
ventricular septal defect and infundibulotomy. The associated abnormality of a left superior vena
cava draining into the coronary sinus had been left untreated in Case 10. One patient (Case 11) had
had more complicated surgery four years after the initial repair, as he had required a further infundi-
bulotomy and replacement of the pulmonary valve (which had cusp agenesis) with a homograft valve.
A third operation was necessary to replace the homograft valve (which calcified) with a prosthetic valve.

Electrocardiograms. The ECG features of the 20 children are shown in Table II: and the
fastest and slowest heart rates recorded are given. Atrial flutter was more common than atrial fibrilla-
tion, which occurred in only two cases. Ventricular ectopic beats were recorded in 7 patients, two of
whom had documented ventricular tachycardia. Illustrative examples of the various rhythms in one
patient (Case 8) are shown in the Fig.

Treatment. All patients were given trials of drug therapy. The most widely used drug was
digoxin, which was given to 16 patients, often for its inotropic action as well as for its chronotropic
action. The other often used antiarrhythmic drug was propanolol. Eight patients who had sympto-
matic tachycardia were treated with combined digoxin and propanolol, but when bradycardia
occurred this had to be stopped. Case 2 had a total of 8 different antiarrhythmic drugs prescribed
in varying combinations (over several years) in an attempt to control his episodic tachycardia. The
symptom of dizziness due to bradycardia in Case 1 improved with ephedrine, but the patient
developed syncope and a permanent pacemaker was inserted. Two other patients with syncope
were given sustained-action tablets of isoprenaline hydrochloride (Saventrine) while under close
observation as inpatients. However, the drug caused supraventricular tachycardia in one and a
short run of self-terminating ventricular fibrillation in the other and was withdrawn. Both patients had
a permanent pacemaker inserted.
Cardioversion was performed in 11 of the patients on 1 to 7 different occasions. No serious problems such as embolism or collapse were recorded, though sinus rhythm was not always restored.

**Pacemakers.** Permanent cardiac pacemakers were inserted in 6 patients. Five of these had syncope and the sixth had severe cardiac failure worsened by episodic tachyarrhythmia. Of those with syncope three had type I sick sinus syndrome with bradycardia only. The other two had both bradyarrhythmia and tachycardia, but after the pacemaker was inserted they had no further palpitations and required no drugs. The sixth patient had only recently had a further cardiac operation to relieve a left ventricular outflow obstruction and to repair the mitral valve, and the permanent pacemaker was inserted to help to control her longstanding dysrhythmias.

One patient was given an atrial epicardial fixed rate (asynchronous type) pacemaker. The others were given ventricular synchronous (demand) systems, of which two have transvenous electrodes and three have electrodes sutured to the epicardium.

Hist-bundle studies were attempted in four of the pacemaker patients. Adequate recordings could not to be obtained in one and in another the conduction intervals accorded with the normal values of Abella et al. (1972). In the other two intra-atrial conduction was prolonged by 50% and 100% respectively and the H-V interval (His-bundle to ventricular depolarization time) was also prolonged by 5% and 15%. In a fifth patient atrial pacing showed a 2:1 atrioventricular block at rates of 130 to 150/minute.

### Discussion

The many previous reports of the sick sinus syndrome have been of the condition in adults mostly elderly and with ischaemic heart disease as the commonest aetiologic factor (Radford and Julian, 1974; Chokshi et al., 1973). Recognition of the syndrome in children is recent and important (Nugent et al., 1974; Scott and Macartney, 1974; Greenwood et al., 1974).

Most of our symptomatic cases followed cardiac surgery. This is of particular interest because of the increase in cardiac surgery in childhood and the reported development later of arrhythmias. Varying atrial dysrhythmias of the sick sinus syndrome after surgical closure of atrial septal defects (Young,
Fig.—Case 8. Illustrative examples of the various sick sinus syndrome rhythms (lead II) over a period of 8 years.
emboli in children with congenital heart disease, to which the disease is more susceptible, has been documented in several studies. The occurrence of emboli in children with congenital heart disease is well documented, and the incidence of emboli in children with congenital heart disease is unknown. The risks of cardioversion in the sick sinus syndrome are known (Lown, 1967). We used it with caution in 11 cases and there were no significant problems.

Almost one-third of our patients were given permanent pacemakers. That reflects the severity of the symptoms and the difficulty of control with other therapy. Pacemakers eliminate bradycardia and have also been observed to diminish or even eliminate the tachycardias (Radford, and Julian, 1974; Cheng, 1968; Sigurd et al., 1973). Nevertheless, they have their own complications and the decision to commit a child to long-term cardiac pacing is a difficult one. Syncpe is life-threatening and therefore an absolute indication for pacing.

The prognosis and natural history of the syndrome have not yet been determined. Symptoms have resolved with the development of permanent atrial fibrillation (Cohen, 1973; Stock, 1970; Radford and Julian, 1974). Interest has been shown recently in developing provocative tests to define the condition better (Mandel et al., 1972; Strauss et al., 1972; Gupta et al., 1974). Hopefully, these may help to predict which cases may prove the most severe. This should be particularly useful in cases resulting from cardiac surgery. With better understanding of the atrial conduction mechanisms of the sick sinus syndrome dysrhythmias after cardiac surgery may be avoided by changes in surgical technique. Meanwhile, the severity of symptoms and the potential risks must be weighed against the difficulties of the types of therapy available.

The purpose of this report is to provide a retrospective survey of the clinical picture presented by children with ECG features of the sick sinus syndrome, with a view to understanding the mechanisms of the sick sinus system, to determine the best treatment, and if possible, to discover how it may be prevented.

References
Sick sinus syndrome


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