Arrhythmias in children with pulmonary stenosis

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Arrhythmias have been recorded in association with congenital pulmonary stenosis. Of six patients who had arrhythmias before operation, anatomical conduction defects were present in two and the arrhythmias persisted after operation. In four, the arrhythmic episodes have not been recorded after operation. No specific causative factors were evident in the pre-operative group.

Acute and often fatal arrhythmias have occurred in the early post-operative period. High risk patients were those with cyanosis, high right ventricular pressure (in excess of 175 mm. Hg), and gross right ventricular hypertrophy. There is evidence to suggest that these arrhythmias develop in association with myocardial ischaemia.

Arrhythmias are a well-known feature in certain types of congenital heart disease, such as Ebstein's anomaly (Taussig, 1947; Engle et al., 1950; Mayer, Nadas, and Ongley, 1957), corrected transposition (Schiebler et al., 1961), and endocardial cushion defects (Somerville, 1965). Arrhythmias in the post-operative period have occurred after corrective procedures for most congenital cardiac lesions.

This paper studies the incidence and significance of arrhythmias occurring in pulmonary stenosis with an intact ventricular septum from a group of 195 children between the ages of 2 weeks and 16 years.

Subjects and methods

Operations were carried out on 237 children for pulmonary stenosis between 1953 and March 1960. Because of inadequate pre-operative clinical information, 42 patients are excluded from this review.

Arrhythmias were suspected from the clinical history or physical examination but only electrocardiographic evidence was accepted as proof. Cardiac catheterization was performed before operation in all patients with arrhythmias. Patients had electrocardiograms recorded by a standard direct-writing machine before and after operation. Six standard leads and chest leads V4R, V1, V3, V5, and V7 were recorded.

In addition, patients had continuous monitoring of the electrocardiogram on an oscilloscope after operation.

Results

Pre-operative arrhythmias A total of 195 children between the ages of 2 weeks and 16 years had operations for pulmonary stenosis. All the children had an intact ventricular septum.

Arrhythmias were recorded before operation in six children (3.5%).

Three patients had episodes of supraventricular tachycardia (Fig. 1, 2, and 3). One of these patients had Wolff-Parkinson-White syndrome (Fig. 4). Three other patients had arrhythmias. One patient had a wandering atrial pacemaker (Fig. 5). One patient had nodal rhythm (Fig. 6) and one patient had complete heart block (Fig. 7).

All these six children survived the operation, and have done well. There were no episodes of arrhythmia in the immediate post-operative period. During post-operative observation over a mean period of 4.1 years, further episodes of arrhythmia have occurred in the child with Wolff-Parkinson-White syndrome (Wolff, Parkinson, and White, 1930); complete heart block has persisted in this child. The remaining four children are free from symptoms and electrocardiograms show sinus rhythm. To date no suspected or recorded arrhythmias have occurred in these four children.

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The clinical details of the six children are summarized in Table I. All three children with defects of the atrial septum were cyanosed. None of the six children had other associated cardiac lesions.

Cases 5 and 6 had no symptoms and the abnormality in rhythm was found on a routine pre-operative electrocardiogram.

No correlation has been shown between the development of arrhythmias and right ventricular pressure, the presence or absence of an atrial septal defect, or the presence of central cyanosis.

Post-operative arrhythmias Nine of the 195 children developed arrhythmias after operation. The onset of arrhythmias was between 3 and 48 hours after operation in all patients. In two children ventricular fibrillation developed as a terminal event following low cardiac output. One of these children had multivalvular myxomatous disease and one had tricuspid valve stenosis.

The onset of arrhythmia in the other seven children was sudden. Three of these seven children died.

Data of the nine children are recorded in Table 2. The right ventricular pressure was high in most patients, and at operation gross right ventricular hypertrophy was present. In two children the degree of hypertrophy had reduced the right ventricular cavity to a slit. All except Case 7 were cyanosed.

Discussion

Pre-operative arrhythmias Standard paediatric cardiological texts do not describe the association of arrhythmias with isolated pulmonary stenosis (Nadas, 1963; Keith, Rowe, and Vlad, 1967; Moss and Adams, 1968; Watson, 1968). The incidence of arrhythmias in this series is low (3.5%) but is of special interest in that four of six patients have had relief from episodes of arrhythmia after operation.

FIG. 2 Supraventricular tachycardia (Case 2).

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age at operation (yr.)</th>
<th>Length of history of arrhythmia (yr.)</th>
<th>Cardiac catheterization</th>
<th>Arrhythmia</th>
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<tbody>
<tr>
<td>1</td>
<td>9</td>
<td>8</td>
<td>—</td>
<td>130</td>
</tr>
<tr>
<td>2</td>
<td>1½</td>
<td>6mth.</td>
<td>—</td>
<td>135</td>
</tr>
<tr>
<td>3</td>
<td>11</td>
<td>7</td>
<td>R→L 230</td>
<td>Supraventricular tachycardia; intermittent right bundle-branch block Supraventricular tachycardia (Wolff-Parkinson-White) syndrome) Complete heart block Nodal rhythm</td>
</tr>
<tr>
<td>4</td>
<td>5</td>
<td>5</td>
<td>R→L 100</td>
<td>Complete heart block Nodal rhythm</td>
</tr>
<tr>
<td>5</td>
<td>6</td>
<td>—</td>
<td>—</td>
<td>100</td>
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<tr>
<td>6</td>
<td>8</td>
<td>—</td>
<td>R→L 100</td>
<td>Wandering atrial pacemaker Nodal rhythm</td>
</tr>
</tbody>
</table>

RVP—Peak systolic right ventricular pressure in mm. Hg. R→L—Right to left.
TABLE 2  Data in 9 children who developed arrhythmias after operation

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age at operation (yr.)</th>
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<th>Alive or dead</th>
<th>Clinical details</th>
</tr>
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<td></td>
<td></td>
<td></td>
<td></td>
<td>RVP</td>
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<tr>
<td>1</td>
<td>10</td>
<td>R→L</td>
<td>225</td>
<td>Dead</td>
</tr>
<tr>
<td>2</td>
<td>2 10/12</td>
<td>R→L</td>
<td>110</td>
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</tr>
<tr>
<td>3</td>
<td>13</td>
<td>R→L</td>
<td>250</td>
<td>Dead</td>
</tr>
<tr>
<td>4</td>
<td>1 1/12</td>
<td>R→L</td>
<td>*</td>
<td>Alive</td>
</tr>
<tr>
<td>5</td>
<td>10</td>
<td>R→L</td>
<td>100</td>
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</tr>
<tr>
<td>6</td>
<td>8</td>
<td>R→L</td>
<td>180</td>
<td>Dead</td>
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<tr>
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<td>5</td>
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<td>220</td>
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<td>8</td>
<td>1 10/12</td>
<td>R→L</td>
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<tr>
<td>9</td>
<td>3</td>
<td>R→L</td>
<td>230</td>
<td>Dead</td>
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</tbody>
</table>

RVP—Peak systolic right ventricular pressure in mm. Hg.
R→L—Right to left.
RVH—Right ventricular hypertrophy.
* Not measured.

Stern and Borman (1967) described atrial flutter as a complication of pulmonary stenosis in adulthood, and felt that this arrhythmia was related to the presence of tricuspid insufficiency and right atrial enlargement. Atrial flutter was recorded in one patient in this series after operation, and in this patient aneurysmal dilatation of the right atrium was noted at operation.

However, in the pre-operative group right atrial dilatation was not noted. Two of the patients had presumed anatomical conduction defects—Wolff-Parkinson-White syndrome, and complete heart block, respectively. Episodes of arrhythmia have continued to occur in the first of these patients while the heart block has persisted in the second.

In the four patients without conduction defects, no correlation has been noted between the arrhythmia and right ventricular pressure, right ventricular hypertrophy, right atrial dilatation, patency of the atrial septum, or arterial desaturation.

Though episodes of supraventricular tachycardia may occur in normal children, it is more common to find them in association with congenital cardiac lesions.

In otherwise normal children who have an episode of supraventricular tachycardia, recurrences are common. In this series frequent episodes of supraventricular tachycardia occurred before operation in two children but have not been recorded in the post-operative period.
Post-operative arrhythmias  Acute post-operative arrhythmias have mainly occurred in patients with cyanosis, high right ventricular pressure, and severe right ventricular hypertrophy. Twenty-eight of the 195 patients in the series had a pre-operative peak systolic right ventricular pressure greater than 175 mm. Hg, and five of these patients (18%) developed acute arrhythmias after operation. Right ventricular pressure after operation in these patients was still raised, usually being close to systemic level.

Macroscopical ischaemic lesions of the myocardium were noted at necropsy in two children. Myocardial ischaemic lesions in association with congenital heart disease and ventricular hypertrophy are now well documented (Esterly and Oppenheimer, 1967; Berry, 1967; Tawes et al., 1969).

Arrhythmias in the early post-operative period may be related to myocardial ischaemia. This ischaemia develops because the grossly hypertrophied right ventricular myocardium is inadequately perfused.

Intensive supervision is essential after operation in these patients, because arrhythmias usually develop suddenly.

Arrhythmic episodes in those patients with a very high right ventricular pressure have not been recorded later than 48 hours after operation.

Arrhythmias in the late post-operative period have occurred in three patients in this series. Two of these had pre-operative arrhythmias associated with conduction defects. One patient developed atrial flutter after operation but successful cardioversion was performed three years later.

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References