Pre- and Postoperative Arrhythmias in Congenital Heart Disease
— From the results of surgery using surface-induced deep hypothermia —

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The incidence of arrhythmias in 3614 preoperative patients with congenital heart disease was 1.6 per cent. Pre- and postoperative serial ECGs in 186 patients with an isolated VSD and in 58 patients with TOF were reviewed.

The incidence of CRBBB and CRBBB-LAH following VSD repair was 33% and 6.6%, respectively, and for TOF was 55% and 5.2%, respectively. Complete A-V block (CAVB) was seen in 2 patients following VSD repair, and none of TOF patients developed CAVB. Late cardiac or dysrhythmic death was not recognized in our limited follow up period.

The indications for surgery in membranous VSD should be determined more carefully, because the risk of the surgical damage to the conduction system is still higher. Moreover, we must keep in mind that damage to the A-V condution system may occur naturally even in a patient with small VSD.

Finally, it can never be overemphasized that physicians must pay attention to the occurrence and course of postoperative arrhythmias.

Recently arrhythmias have been recognized as a major cause of late sudden death in patients undergoing intracardiac repair of congenital heart disease. Therefore, all physicians observing these patients should have a proper understanding of the degree of damage to the conduction system as well as the degree of the morphological and hemodynamic correction.

The purpose of this study was to evaluate the occurrence and incidence of arrhythmias in patients with ventricular septal defect (VSD) and/or tetralogy of Fallot (TOF) who had undergone intracardiac repair by using surface-induced deep hypothermia. Surface-induced deep hypothermia, which was developed mainly in Japan, has many advantages in open heart surgery. This is especially true for operations in infants and neonates with congenital heart disease. The details of the method and experiences with deep hypothermia have been given in a previous report!

PATIENTS AND METHODS
The occurrence and incidence of arrhythmias in preoperative patients must be clarified first, and it was studied in 3614 patients with congenital heart disease (mean age, 2.5 years old). Pre- and postoperative serial electrocardiograms in 186 patients with an isolated ventricular septal

Key Words:
Incidence of preoperative arrhythmias
Damage to the conduction system following intracardiac repair
Surface-induced deep hypothermia

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TABLE I  INCIDENCE OF VARIOUS ARRHYTHMIAS (per 10000)

<table>
<thead>
<tr>
<th></th>
<th>PVC</th>
<th>PAC</th>
<th>SVT</th>
<th>VT</th>
<th>JR</th>
<th>AVD</th>
<th>AVB</th>
<th>CAVB</th>
<th>CRBBB</th>
<th>CLBBB</th>
<th>WPW</th>
<th>SSS</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>CHD n=3614</td>
<td>27.1</td>
<td>13.5</td>
<td>32.5</td>
<td>0</td>
<td>10.8</td>
<td>2.3</td>
<td>10.8</td>
<td>13.5</td>
<td>18.9</td>
<td>5.4</td>
<td>16.2</td>
<td>13.5</td>
<td>1.65%</td>
</tr>
<tr>
<td>Preschool children n=266380</td>
<td>14.1</td>
<td>4.0</td>
<td>0.2</td>
<td>0.15</td>
<td>24.9</td>
<td>1.4</td>
<td>3.5</td>
<td>0.1</td>
<td>8.9</td>
<td>0.15</td>
<td>4.2</td>
<td>0</td>
<td>0.65%</td>
</tr>
</tbody>
</table>

Abbreviations: PVC = premature ventricular contracture; PAC = premature atrial contracture; SVT = supraventricular tachycardia; VT = ventricular tachycardia; JR = junctional rhythm; CAVB = complete A-V block; CRBBB = complete right bundle branch block; CLBBB = complete left bundle branch block; WPW = Wolff-Parkinson-White syndrome; SSS = sick sinus syndrome.

TABLE II  THE INCIDENCE OF ARRHYTHMIAS IN POSTOPERATIVE TOF PATIENTS

<table>
<thead>
<tr>
<th>Type</th>
<th>VSD</th>
<th>NSR</th>
<th>IRBBB</th>
<th>CRBBB</th>
<th>CRBBB + LAH</th>
<th>LAH</th>
<th>CAVB</th>
<th>total</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td></td>
<td>32</td>
<td>7</td>
<td>8</td>
<td>(17.0%)</td>
<td></td>
<td></td>
<td>47</td>
</tr>
<tr>
<td>II</td>
<td></td>
<td>34</td>
<td>35</td>
<td>41</td>
<td>8</td>
<td>1</td>
<td>2</td>
<td>121</td>
</tr>
<tr>
<td></td>
<td></td>
<td>3</td>
<td>5</td>
<td>2</td>
<td>(20.0%)</td>
<td></td>
<td></td>
<td>10</td>
</tr>
<tr>
<td>III</td>
<td></td>
<td>2</td>
<td>2</td>
<td>3</td>
<td>(37.5%)</td>
<td>1</td>
<td>2</td>
<td>8</td>
</tr>
<tr>
<td>total</td>
<td></td>
<td>71</td>
<td>49</td>
<td>54</td>
<td>(29.0%)</td>
<td>9</td>
<td>1</td>
<td>2</td>
</tr>
</tbody>
</table>

TABLE III  THE INCIDENCE OF CRBBB AND CAVB IN POSTOPERATIVE VSD PATIENTS

<table>
<thead>
<tr>
<th>TOF</th>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
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<th></th>
<th></th>
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<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>CRBBB</td>
<td>CRBBB+LAH</td>
<td>NSR &amp; IRBBB</td>
<td>A-V block 1°</td>
<td>Junctional rhythm</td>
<td>PVC</td>
<td>A-V dissociation</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
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<td></td>
<td></td>
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<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>TOF</td>
<td>58 cases</td>
<td>32 (55.2%)</td>
<td>3 (5.2%)</td>
<td>23 (39.7%)</td>
<td>2</td>
<td>3</td>
<td>2</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>8 cases (13.8%)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Defect repaired via a right ventriculotomy and in 58 patients with tetralogy of Fallot were reviewed. The follow up period ranged up to 14 years (mean 6 years). Intracardiac electrophysiologic studies were performed to evaluate the electrophysiologic properties of the sinus node, atria, A-V node and His-Purkinje system in 13 patients with tetralogy of Fallot 1 month to 9 years after operation.

RESULTS

The incidence of arrhythmias in 3614 preoperative patients with congenital heart disease was 1.6 per cent. Compared with the incidence of arrhythmias in healthy preschool children, that in congenital heart disease was about three times higher. Supraventricular tachycardia, W-P-W syndrome, premature atrial beats, complete left bundle branch block and sick sinus syndrome were seen more frequently in the latter (Table I). Of the congenital heart diseases examined, vis-

ceroatrial situs discordant syndrome (asplenia and polysplenia syndromes), corrected transposition of the great arteries, secundum atrial septal defect, endocardial fibroelastosis and mitral valve prolapse syndrome were more likely to have arrhythmias.

ARRHYTHMIAS FOLLOWING INTRACARDIAC REPAIR

186 patients with isolated ventricular septal defect underwent intracardiac repair. Their ages ranged from 1 month to 6 years (mean 11

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Fig. 2. HBE from Case 10.
Top, one month postoperatively. First and second degree A-V block were seen together. HBE revealed a long AH interval of 140 msec and Wenckebach type AH block, suggestive of a damage to the A-V nodal region. Bottom, four years postoperatively. As compared to the top, HBE showed a normal AH interval (72 msec) and a long HV interval (60 msec), indicative of infra-Hisian block. However, his serial ECGs have maintained CRBBB with right axis deviation.

(months). Ventricular septal defects were divided into three types according to Kirklin’s classification. No fatalities occurred in the postoperative and the follow up period. Of 139 patients undergoing intracardiac repair of membranous ventricular septal defect (type II and III), 46 (33.1%) developed complete right bundle branch block, 8 (6.6%) had combined complete right bundle branch block and left anterior hemiblock, one had isolated left anterior hemiblock, and 2 developed complete A-V block. One complete A-V block occurred immediately after the oper-
Fig. 3. Refractory period measurement from Case 10. Refractory periods of the right atrium and A-V node were normal, while those of His-Purkinje system were remarkably prolonged.

Fig. 4. Surgery for membranous VSD. Surgery for the membranous VSD is decreasing with the times. However, the surgery under age one year, which requires patch closure, has had an absolute majority in the membranous VSD repair.

Fallot, 7 died in the operative period, for an operative mortality of 11 per cent. Of the 58 survivors, 32 patients (55.2%) developed complete right bundle branch block, and 3 (5.2%) developed bifascicular block (CRBBB-LAH). The other 23 patients (39.7%) had normal sinus rhythm or incomplete right bundle branch block (QRS width ≤ 0.08 sec). Late cardiac or dysrhythmic death was not recognized in our limited follow-up period ranged from 1 to 13 years. Other postoperative arrhythmias were as follows: first degree A-V block was seen in 2 patients, junctional rhythm in 3, premature ventricular beat in 2, and A-V dissociation in one (Table III).

Intracardiac electrophysiologic studies were performed on 13 patients who underwent intracardiac repair of tetralogy of Fallot. Normal sinus node function was recognized in all patients. Sinus node recovery time (SNRT) and sinoatrial conduction time (SACT) were ranged from 580 to 1286 msec and from 46 to 119 msec, respectively. Prolongation of AH interval was detected in 2 patients and prolongation of HV interval was detected in 2 other patients (Table IV). One patients (Case 10) demonstrated a mild prolongation of HV interval, and furthermore showed a markedly prolonged His-Purkinje effective refractory period of 578 msec. These data suggested a
# TABLE IV  ELECTROPHYSIOLOGIC FINDINGS IN POSTOPERATIVE TOF PATIENTS

<table>
<thead>
<tr>
<th>No.</th>
<th>pat. sex</th>
<th>age at examination (yr)</th>
<th>age at operation (yr)</th>
<th>postop. QRS pattern</th>
<th>resting HBE (msec)</th>
<th>SN function</th>
<th>refractory period</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>JS M</td>
<td>5</td>
<td>5</td>
<td>IRBBB</td>
<td>645  66  63  50</td>
<td>825 128</td>
<td>180 192 295 352</td>
</tr>
<tr>
<td>2</td>
<td>KK M</td>
<td>10</td>
<td>3</td>
<td>IRBBB</td>
<td>735 39 101 20</td>
<td>112 1032 140</td>
<td>297 226 F460</td>
</tr>
<tr>
<td>3</td>
<td>KK F</td>
<td>10</td>
<td>5</td>
<td>CRBBB</td>
<td>664 24 83 50</td>
<td>119 1000 151</td>
<td>336 242 &lt;242 432</td>
</tr>
<tr>
<td>4</td>
<td>KK M</td>
<td>6</td>
<td>4</td>
<td>CRBBB</td>
<td>507 19 96 33</td>
<td>77 643 127</td>
<td>136 153 153 350</td>
</tr>
<tr>
<td>5</td>
<td>TK F</td>
<td>7</td>
<td>4</td>
<td>IRBBB</td>
<td>757 31 78 47</td>
<td>75 1049 139</td>
<td>292 223 F311</td>
</tr>
<tr>
<td>6</td>
<td>AK M</td>
<td>13</td>
<td>4</td>
<td>CRBBB</td>
<td>625 36 68 36</td>
<td>82 820 131</td>
<td>195 194 330 381</td>
</tr>
<tr>
<td>7</td>
<td>RK F</td>
<td>7</td>
<td>7</td>
<td>IRBBB</td>
<td>662 16 81 36</td>
<td>59 823 124</td>
<td>161 220 &lt;220 401</td>
</tr>
<tr>
<td>8</td>
<td>YS M</td>
<td>8</td>
<td>2</td>
<td>CRBBB PR†</td>
<td>540 21 111 61</td>
<td>87 755 140</td>
<td>215 200 246 374</td>
</tr>
<tr>
<td>9</td>
<td>KS M</td>
<td>11</td>
<td>4</td>
<td>CRBBB</td>
<td>828 21 94 42</td>
<td>99 1286 155</td>
<td>458 203 F346</td>
</tr>
<tr>
<td>10</td>
<td>KI M</td>
<td>6</td>
<td>2</td>
<td>IRBBB</td>
<td>808 26 72 60</td>
<td>105 1057 131</td>
<td>249 204 232 264</td>
</tr>
<tr>
<td>11</td>
<td>FW F</td>
<td>3</td>
<td>4</td>
<td>IRBBB</td>
<td>540 23 70 35</td>
<td>46 580 107</td>
<td>40 160 245 295</td>
</tr>
<tr>
<td>12</td>
<td>HK M</td>
<td>5</td>
<td>3</td>
<td>N</td>
<td>510 20 90 40</td>
<td>56 645 126</td>
<td>135 215 300 375</td>
</tr>
<tr>
<td>13</td>
<td>TY F</td>
<td>5</td>
<td>5</td>
<td>N</td>
<td>730 15 80 45</td>
<td>62 1040 143</td>
<td>310 285 &lt;320 420</td>
</tr>
</tbody>
</table>

HBE = His bundle electrogram;  SN = sinus node;  CL = cycle length;  SACT = undivided sinoatrial conduction time;  SNRT = sinus node recovery time;  %SNRT = SNRT/CL (%);  CSNRT = corrected sinus node recovery time;  RAERP = effective refractory period of right atrium;  AVNFRP = functional refractory period of A-V node;  F = fast A-V nodal pathway;  S = slow A-V nodal pathway.
latent conduction disturbance of His-Purkinje system (Fig. 2, 3). Discontinuous $H_1 - H_2$ curve, indicative of the presence of dual A-V nodal pathways, was detected in 3 patients (Case 2, 5 and 9), but none of them had developed supraventricular tachycardia.

DISCUSSION

The incidence of arrhythmias in preoperative patients with congenital heart disease was 1.6 per cent. This incidence was about three times higher than that in healthy preschool children. Such arrhythmias as supraventricular tachycardia, W-P-W syndrome, premature atrial beat, complete right bundle branch block and sick sinus syndrome were seen more frequently in patients with congenital heart disease.

Surface-induced deep hypothermia has the following excellent advantages. Firstly, the risk of damage to the A-V node, His bundle and bundle branches can be reduced under a quiet heart and a bloodless field. Secondly, the injury to the sinus node, internodal tracts and atrial muscle can be avoided, owing to the fact that this technique does not require both atriotomy and cannulation for cardiac bypass. However, recognition of the location of His bundle based on intracardiac mapping technique is impossible under surface cooling. Consequently, surgeons who would adopt the surface cooling technique must have a thorough knowledge of the anatomy of the conduction system in each congenital heart disease.

Transatrial repair of membranous ventricular septal defect and transpulmonary repair of supravalvular aortic stenosis are now widely recommended. The reported incidence of complete right bundle branch block was 0.4 to 4.4 per cent when a right atrial approach was used. The mechanism of production of postoperative complete right bundle branch block has been studied extensively. Many investigators have attributed it to damage to the proximal bundle branch at the time of operation, and others have suggested that the complete right bundle branch block results from the right ventriculotomy, not from the damage to the proximal right bundle branch. The overall occurrence of postoperative complete right bundle branch block probably represents a combination of both proximal and peripheral damage. The reported incidence of complete right bundle branch block and CRBBB-LAH following repair of membranous ventricular septal defect is from 10 to 60% and from 0.5 to 8%, respectively, and for tetralogy of Fallot is from 40 to 100% and from 5 to 23%, respectively. The reported incidence of complete A-V block following intracardiac repair of ventricular septal defect and tetralogy of Fallot is from 0.2 to 1.7% and from 0.6 to 2.0%, respectively.

The late cardiac death following repair of tetralogy of Fallot has been reported at 0.7 to 2.1%. The combination of CRBBB and LAH described as bifascicular block has been attributed by many to be the result of surgical damage to both the proximal right bundle branch and the anterior division of the left bundle branch. The clinical significance and prognosis of CRBBB-LAH is by no means clear because of different genesis of complete right bundle branch block. Some authors have reported a high incidence of complete A-V block and sudden cardiac death, but many reports have documented a generally good prognosis, and the critical factor in a poor late prognosis in patients with bifascicular block may be the history of transient postoperative complete A-V block. A long H-V interval in patients with bifascicular block is indicative of trifascicular block and the risk of future complete A-V block.

In conclusion, the indications for operation of membranous ventricular septal defect should be determined more carefully. We recommend medical treatment instead of immediate surgery even in patients with a pulmonary to systemic flow ratio of above 2.0, if their pulmonary artery pressure is normal or only mildly elevated. Most of these patients will have decreasing left to right shunts and normal pulmonary artery pressure after the first year of life. They will never require surgical intervention. If these patients continue to have large left to right shunts by 4 to 5 years of age, then surgery may be indicated (Fig. 4).

We must keep in mind that damage to the A-V conduction system may occur naturally even in a patient with ventricular septal defect of small left to right shunts (Fig. 5).

Finally, it can never be overemphasized that physicians must always pay attention to the occurrence and course of postoperative arrhythmias. Holter monitoring, exercise electrocardiogram and intracavitary electrophysiologic studies, if necessary, should be performed to evaluate the clinical significance of postoperative arrhythmias.

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