Coronary Sinus Rhythm in Anomalous Systemic Venous Connection

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SUMMARY
(1) In 1,244 patients with various congenital heart disease, 12 cases of coronary sinus rhythm were recognized.
(2) Four cases were found in 33 patients with persistent left superior vena cava, and 6 cases were encountered in 9 patients with absence of the inferior vena cava.
(3) The intimate correlation between coronary sinus rhythm and these systemic venous anomalies was emphasized.

Additional Indexing Words:
Persistent left superior vena cava Absence of inferior vena cava Frontal P wave axis Left atrial rhythm

SYSTEMIC venous anomalies, such as persistent left superior vena cava and absent inferior vena cava, are not so rare in various congenital heart diseases. Preoperative identification of these anomalous systemic venous connections is clinically important, because their presence necessitates modified technique for venous cannulation of cardiopulmonary bypass.

In cases of persistent left superior vena cava, usually entering the right atrium via the coronary sinus, coronary sinus rhythm was occasionally shown in routine electrocardiograms, resulting from persistent pacemaker of the left horn of the sinus venosus. In addition, typical coronary sinus rhythm was frequently found in patients with absent inferior vena cava in our experience.

The detailed description of these observations is the purpose of this report.

MATERIALS AND METHODS

Scaler electrocardiograms of 1,244 cases with various congenital heart diseases in our Department were reviewed. This series consisted of atrial septal defect (323 cases), tetralogy of Fallot (319), interventricular septal defect (274), patent ductus arteriosus (226), pulmonic stenosis (53), endocardial cushion defect (33), and double outlet right ventricle (16).
Systemic venous anomalies associated with these cardiac malformations were persistent left superior vena cava (33 cases), absence of the inferior vena cava (9), anomalous left superior vena cava draining into the left atrium (2), and anomalous inferior vena cava draining into the left atrium (1). The detailed anatomic classification is shown in Table I.

The preoperative 12-leads scalar electrocardiograms were reviewed in all cases. The P waves were examined in height, contour, duration and frontal axis. In some cases P loops of vectorcardiograms were examined at the same time.

Criteria for recognition of coronary sinus rhythm accepted in the present series are as follows:3)

1) Frontal P wave axis ranges from \(-30^\circ\) to \(-90^\circ\).
2) PR interval is over 0.12 sec.
3) P wave in lead V\(_6\) is not negative.

## RESULTS

Twelve patients showed coronary sinus rhythm as summarized in Table II. Cardiac anomalies associated with this rhythm were tetralogy of Fallot (4 cases), interventricular septal defect (3), atrial septal defect (2, one of which was sinus venosus defect), endocardial cushion defect (2), and double outlet right ventricle (1). Moreover, 10 of 12 cases were confirmed to have such venous anomalies as persistent left superior vena cava (4 cases), absence of the inferior vena cava (4), and both (2) (Fig. 1 and 2).

Conversely, coronary sinus rhythm was seen in 4 out of 33 patients with persistent left superior vena cava (12%) and in 6 out of 9 with absent inferior vena cava (67%). Nodal rhythm was found in one of persistent left superior vena cava and one of absent inferior vena cava respectively. Left atrial rhythm

<table>
<thead>
<tr>
<th>Table I. Anomalous Systemic Venous Connection</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>No. of Cases</strong></td>
</tr>
<tr>
<td>Persistent LSVC</td>
</tr>
<tr>
<td>33</td>
</tr>
<tr>
<td>Absent IVC</td>
</tr>
<tr>
<td>Anomalous IVC to LA</td>
</tr>
<tr>
<td>Anomalous LSVC to LA</td>
</tr>
<tr>
<td>Total</td>
</tr>
</tbody>
</table>

Abbreviations: LSVC=Left superior vena cava, IVC=Inferior vena cava, LA=Left atrium, ASD=Atrial septal defect, TF=Tetralogy of Fallot, VSD=Ventricular septal defect, PDA=Patent ductus arteriosus, PS=Pulmonic stenosis, ECD=Endocardial cushion defect, DORV=Double outlet right ventricle
Fig. 1. Electrocardiogram in a case (M.T.). Frontal axis of P wave was $-60^\circ$ and PR interval was 0.15 sec.

Fig. 2. Angiocardiogram of the same case (M.T.), demonstrating absence of the inferior vena cava. Right heart catheterization disclosed interventricular septal defect with severe pulmonary hypertension. At operation, a large cannula was inserted into the superior vena cava and two small cannulas into two hepatic veins.

was seen in one of absent inferior vena cava.

Frontal P wave axis in coronary sinus rhythm in the present series, was
Table II. Coronary Sinus Rhythm

<table>
<thead>
<tr>
<th>Name</th>
<th>Age</th>
<th>Sex</th>
<th>Cardiac Anomalies</th>
<th>Venous Anomalies</th>
<th>Frontal P-Axis</th>
<th>PR Interval</th>
</tr>
</thead>
<tbody>
<tr>
<td>I.T.</td>
<td>5</td>
<td>M</td>
<td>TF</td>
<td>LSVC</td>
<td>-90°</td>
<td>0.13 sec.</td>
</tr>
<tr>
<td>A.M.</td>
<td>24</td>
<td>F</td>
<td>Sinus Venosus Defect</td>
<td>LSVC</td>
<td>-45</td>
<td>0.17</td>
</tr>
<tr>
<td>T.Y.*</td>
<td>17</td>
<td>M</td>
<td>ECD+PS</td>
<td>LSVC</td>
<td>-45</td>
<td>0.22</td>
</tr>
<tr>
<td>H.T.</td>
<td>21</td>
<td>M</td>
<td>VSD</td>
<td>LSVC</td>
<td>-40</td>
<td>0.20</td>
</tr>
<tr>
<td>M.A.</td>
<td>22</td>
<td>F</td>
<td>ECD+PS+DORV**</td>
<td>AIVC+LSVC</td>
<td>-150</td>
<td>0.17</td>
</tr>
<tr>
<td>H.Y.</td>
<td>14</td>
<td>M</td>
<td>DORV</td>
<td>AIVC</td>
<td>-60</td>
<td>0.15</td>
</tr>
<tr>
<td>H.S.*</td>
<td>2</td>
<td>F</td>
<td>TF</td>
<td>AIVC+LSVC</td>
<td>-60</td>
<td>0.19</td>
</tr>
<tr>
<td>K.T.</td>
<td>10</td>
<td>M</td>
<td>TF</td>
<td>AIVC</td>
<td>-90</td>
<td>0.16</td>
</tr>
<tr>
<td>I.Y.</td>
<td>13</td>
<td>M</td>
<td>ASD</td>
<td>AIVC</td>
<td>-50</td>
<td>0.17</td>
</tr>
<tr>
<td>M.T.</td>
<td>10</td>
<td>M</td>
<td>VSD</td>
<td>AIVC</td>
<td>-60</td>
<td>0.15</td>
</tr>
<tr>
<td>Y.T.</td>
<td>7</td>
<td>M</td>
<td>TF</td>
<td>None</td>
<td>-35</td>
<td>0.15</td>
</tr>
<tr>
<td>S.A.</td>
<td>4</td>
<td>F</td>
<td>VSD</td>
<td>None</td>
<td>-35</td>
<td>0.14</td>
</tr>
</tbody>
</table>

* Intermittent coronary sinus rhythm
** Dextrocardia

Located at $-35^\circ$ or less. PR interval ranged from 0.13 to 0.22 sec.

In the absence of systemic venous anomalies, mean frontal axes of P wave in atrial septal defect, interventricular septal defect, and tetralogy of Fallot were found at $+40.8^\circ$, $+40.0^\circ$, and $+46.5^\circ$ respectively, whereas the same axes in cardiac anomalies with persistent left superior vena cava, absent inferior vena cava, and atrial septal defect of sinus venosus type were directed towards left, locating at $+22.0^\circ$, $-42.1^\circ$, and $+13.0^\circ$ respectively (Fig. 3).

![Fig. 3 Frontal axis of P wave in various congenital heart diseases. Left axis deviation was frequently seen in patients with persistent left superior vena cava, absence of the inferior vena cava, and atrial septal defect of sinus venosus type.](image)

Intermittent coronary sinus rhythm was found in 2 cases. In a patient (H.S.) coronary sinus rhythm was easily converted into regular sinus rhythm. Direction of the frontal axis of P wave was $0^\circ$ in sinus rhythm and $-60^\circ$ in
Fig. 4. Electrocardiogram of Case T.Y. showing an abrupt change from coronary sinus rhythm to left atrial rhythm. Deep inspiration or carotid sinus pressure appeared to be associated with the abrupt change of rhythms. Angiocardiography demonstrated persistent A-V canal and left superior vena cava. At operation, the third cannula was inserted from the right atrium to the left superior vena cava via the coronary sinus, as no left innominate vein was found in this case.

coronary sinus rhythm. PR interval was normal and almost equal in both rhythms. In another case (T.Y.) both coronary sinus rhythm and left atrial rhythm, instead of sinus rhythm, were observed. An abrupt change in P wave axis could be recorded during a single continuous electrocardiographic record (Fig. 4 and 5).

DISCUSSION

As far as the definition of coronary sinus rhythm, upper nodal rhythm, and left atrial rhythm is concerned, there are many controversies. But the frequent association of such abnormal cardiac rhythm with the anomalous systemic venous return observed in the present study is worthy of special note.

Scherf noted that coronary sinus rhythm showed electrocardiogram exhibiting deeply inverted P wave in lead II and III, accompanied by normal or slightly shortened PR interval, and that the focus of origin of such stimulus was situated in the upper part of the A-V node.

Hancock and associates more extended the criteria for recognition of this ectopic rhythm, saying that the P wave with left axis deviation (inverted P wave in lead III; mean frontal P axis less than +15°) was thought to arise from an accessory pacemaker in the coronary sinus region of the A-V node, which was persisting from embryonic life when the left superior vena cava persisted, or taking over pacemaking function when the S-A node was anatomically absent, as observed in atrial septal defect of sinus venosus type.

In the present experience, the left axis deviation of P wave less than +15° was recognized in 67 per cent of patients with absent inferior vena cava, and
Fig. 5. Vectorcardiogram of the same case (T.Y.). Mean P-loop vector oriented to left superior in coronary sinus rhythm (upper record) and to right inferior anterior in left atrial rhythm (lower record).
in 33 per cent of cases with persistent left superior vena cava. However, the same finding was observed in 15 per cent of patients with atrial septal defect and in 14 per cent with ventricular septal defect even in the absence of systemic venous anomalies. On the basis of these results, it seems better to use the term "coronary sinus rhythm" in such a limited sense as that described by Scherf.

Differentiation of upper nodal rhythm from coronary sinus rhythm is primarily based on the PR interval, and is not of prime importance. However, since the intimate correlation was found between coronary sinus rhythm and systemic venous anomalies, the recognition of this rhythm is very helpful in preoperative diagnosis of anomalous systemic venous return, which can be confirmed by angiocardiography.

Mirowski emphasized the importance of differentiating coronary sinus rhythm from left atrial rhythm, and proposed that the electrocardiogram, showing short PR interval and inverted P wave in leads II, III, and V6, should be defined as left atrial rhythm. In the record of Spordick and associates, however, 5 out of 9 patients with coronary sinus rhythm revealed negative P wave in lead V6. In our opinion such rhythms were classified as left atrial rhythm. In 2 cases of left atrial rhythm in our experience, however, retrograde conduction as described by Mirowski was not recognized, as P waves were upright in leads II and III, whereas inverted in leads I and V6 in them.

Patten noted the presence in embryonic heart of symmetrically located pacemakers at the junctional area of the common cardinal veins with the right and left horns of the sinus venosus. He assumed that the S-A node developed from the right cardinal vein pacemaker and the A-V node developed from the left cardinal vein pacemaker.

Since the left superior vena cava represents the postnatal persistence of embryonic left cardinal vein, left-sided pacemaker may be able to persist and function as an abnormally active pacemaker in case of persistent left superior vena cava. In the absence of the inferior vena cava, persisting bilateral superior venae cavae are common findings in case of hemiazygos continuation, but the same venous anomaly is not infrequent also with azygos continuation. The function of left cardinal vein pacemaker can remain in the presence of combined left superior vena cava. Presumably the absent inferior vena cava can also be accompanied by persistence of left cardinal vein pacemaker alone even when left superior vena caval communication is not retained.

References