Coronary Sinus Rhythm in the Polysplenia Syndrome*

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A leftward and superior P wave axis, the so-called coronary sinus rhythm, was found in nine of 12 patients with the typical polysplenia syndrome. These nine patients had the usual developmental abnormalities of the sinus venosus, including absence of the renal to hepatic portion of the inferior vena cava, other associated cardiovascular lesions, visceral heterotaxia, and multiple spleens. The absence of the superior portion of the inferior vena cava complicated cardiac catheterization in all patients, making it impossible to perform from the inguinal approach in four. The finding of coronary sinus rhythm in a patient with congenital heart disease and visceral heterotaxia should alert the clinician to consider the diagnosis of polysplenia and the attendant constellation of anomalies. In contrast, the asplenia syndrome, which has many similar developmental defects, except that absence of the superior portion of the inferior vena cava is very rare, has a very low incidence of this atrial arrhythmia. In a critically ill infant who has coronary sinus rhythm, with congenital heart disease and heterotaxia, it might be expedient to approach cardiac catheterization from the upper extremity.

A superior and leftward P axis, the so-called coronary sinus rhythm, has been reported in various congenital cardiac lesions. It has most commonly been reported in association with sinus venosus atrial septal defects1 and persistent left superior vena cava,1 but may also occur with single atrium2 and absence of the renal to hepatic portion of the inferior vena cava withazygous extension.3 More recently, Momma and Linde4 have documented coronary sinus rhythm in four patients with the polysplenia syndrome.5 This syndrome is characterized by polysplenia or multiple spleens, congenital heart disease with absence of the hepatic segment of the inferior vena cava, bronchopulmonary isomerism, and visceral heterotaxia. Momma and Linde4 suggested that this ectopic atrial rhythm might serve to distinguish between the asplenia and polysplenia syndromes.

It is the purpose of this communication to further document the association of coronary sinus rhythm with the polysplenia syndrome and to comment on possible clinical implications.

Materials and Methods

Twelve cases of the polysplenia syndrome were confirmed at The Children's Hospital Medical Center, Boston, between 1954 and 1972. Nine cases were documented by complete postmortem examination. Polysplenia was demonstrated by splenic scintigraphy in two living patients with complex congenital heart disease, visceral heterotaxia, and normal splenic function. The remaining case was found at exploratory laparotomy in a patient with congenital heart disease and heterotaxia, who presented with clinical signs of peritonitis.

The diagnosis of coronary sinus rhythm was based on an electrocardiographic record showing a leftward and superior P wave axis, between -30° and -90°, with inversion of P waves in leads 2, 3, and aVF; a P-R interval of normal duration for age and heart rate; and a P wave preceding each QRS complex, compatible with requirements of an A-V rhythm.6

Nine of the 12 patients with polysplenia had electrocardiographic evidence of coronary sinus rhythm prior to digitalis administration. Seven of the nine patients have died, and two are alive. All nine have undergone diagnostic cardiac catheterization, and the two living patients have, in addition, undergone peripheral radionuclide venography. These nine patients comprise the study group, and their age, sex, and associated cardiovascular, bronchopulmonary, and visceral

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anomalies are presented in Table 1. The three patients without coronary sinus rhythm had similar cardiac anomalies, and two of the three had electrocardiographic evidence of nodal rhythm.

Standard 12-lead electrocardiograms were recorded in all patients prior to the administration of digitalis, but all available tracings were studied. The P waves were examined for morphology, height, contour, duration, and frontal plane axis. The P-R interval was examined with regard to heart rate and age. The triaxial reference frame was used to determine frontal P wave and QRS axis. The electrocardiographic features are summarized in Table 2.

**RESULTS**

**Anatomic Findings**

As shown in Table 1, all patients had complex cardiovascular disease, polysplenia and visceral heterotaxia. The visceral heterotaxia was manifested most typically as an abnormally symmetric liver, common gastrointestinal mesentery with malrotation and a nonretroperitoneal pancreas. Five patients had anomalies of bronchopulmonary lobation, including four with bilateral hyparterial bronchi, and one with inverse normal bronchopulmonary lobation.

Levocardia was present in seven and dextrocardia in two patients. All patients had a normal ventricular relationship (D-ventricular loop), and the great arteries were normally related in all patients, but one (No. 8, Table 1) in whom D-transposition of the great arteries was present. Atrial situs solitus was felt to be present in all patients, despite absence of

<table>
<thead>
<tr>
<th>Case, No.</th>
<th>Age at Death</th>
<th>Sex</th>
<th>Cardiovascular Abnormalities</th>
<th>Heterotaxia</th>
<th>Bronchopulmonary Abnormalities</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>2 days</td>
<td>F</td>
<td>levo cardia; absent IVC; ASD I*; severe Ao. stenosis; subvalvar Ao. stenosis with accessory endocardial cushion tissue; mitral stenosis; normal coronary sinus</td>
<td>symm. liver rt. sided stomach common G-I mesent.</td>
<td>inversus</td>
</tr>
<tr>
<td>2</td>
<td>14 days</td>
<td>M</td>
<td>levo cardia; absent IVC; ASD II*; VSD, small; hypoplasia of ascending Ao. &amp; severe coarct. of Ao.; PDA; normal coronary sinus</td>
<td>symm. liver rt. sided stomach common G-I mesent.</td>
<td>hypart.</td>
</tr>
<tr>
<td>3</td>
<td>5-1/2 yrs.</td>
<td>M</td>
<td>levo cardia; absent IVC; common atrium; CAVC; TOF; aberrant rt. subclavian artery; normal coronary sinus</td>
<td>symm. liver rt. sided stomach common G-I mesent.</td>
<td>hypart.</td>
</tr>
<tr>
<td>4</td>
<td>15 yrs.*</td>
<td>M</td>
<td>levo cardia; absent IVC; common atrium; CAVC; PAPVR</td>
<td>symm. liver rt. sided stomach</td>
<td>prob. normal</td>
</tr>
<tr>
<td>5</td>
<td>1 day</td>
<td>F</td>
<td>dextrocardia; absent IVC; common atrium; CAVC; mild pulm. stenosis; normal coronary sinus; PAPVR</td>
<td>symm. liver rt. sided stomach common G-I mesent.</td>
<td>hypart.</td>
</tr>
<tr>
<td>6</td>
<td>12-8/12 yrs.</td>
<td>F</td>
<td>levo cardia; absent IVC; Bil. SVC; LSVC to left atrium; common atrium; CAVC; absent coronary sinus</td>
<td>symm. liver midline stomach common G-I mesent.</td>
<td>none</td>
</tr>
<tr>
<td>7</td>
<td>3 mos.</td>
<td>F</td>
<td>levo cardia; absent IVC; Bil. SVC; LSVC to left atrium; CAVC; PDA; absent coronary sinus; PAPVR</td>
<td>symm. liver rt. sided stomach common G-I mesent.</td>
<td>hypart.</td>
</tr>
<tr>
<td>8</td>
<td>7-8/12 yrs.</td>
<td>M</td>
<td>levo cardia; absent IVC; Bil. SVC; LSVC to coronary sinus; Transposition of great arteries; CAVC; pulmonic stenosis; PDA</td>
<td>normal liver rt. sided stomach common G-I mesent.</td>
<td>none</td>
</tr>
<tr>
<td>9</td>
<td>30 yrs.*</td>
<td>F</td>
<td>dextrocardia; absent IVC with hemiazygous to LSVC to coronary sinus; common atrium; CAVC; pulmonic stenosis</td>
<td>symm. liver rt. sided stomach</td>
<td>prob. normal</td>
</tr>
</tbody>
</table>

*Living

Abbreviations: Absent IVC = absence of renal to hepatic portion of the inferior vena cava with azygous extension to right superior vena cava; Ao. = Aortic; ASD I* = ostium primum atrial septal defect; ASD II* = secundum atrial septal defect; Bil. = bilateral; CAVC = complete common atrioventricular canal; Coarct. = coarctation; Hypart. = bilateral hyparterial bronchial anatomy; LSVC = left superior vena cava; G-I mesent. = gastro-intestinal mesentery; PDA = patent ductus arteriosus; PAPVR = partial anomalous pulmonary venous return; Symm. = abnormally symmetric; SVC = superior vena cava.

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inferior vena caval-right atrial direct continuity.

An endocardial cushion defect, usually a complete common atrioventricular canal, was present in all but one patient (No. 2, Table 1); this patient had both a secundum atrial defect and a membranous ventricular septal defect.

Abnormality of pulmonary venous return was present in three patients, and was manifested as right pulmonary veins to right atrium and left pulmonary veins to left atrium. In the other cases, pulmonary venous return was normal.

Developmental anomalies of the sinus venous were present in all of the patients. All showed absence of the renal to hepatic portion of the inferior vena cava, with either an aygous or hemiazygous extension. The coronary sinus was felt to be normal.

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**Table 2—Electrocardiographic Findings in Polysplenia**

<table>
<thead>
<tr>
<th>Case, No.</th>
<th>Rhythm</th>
<th>P-R</th>
<th>Heart Rate</th>
<th>Frontal Plane Axis</th>
<th>QRS Inscription</th>
<th>Other</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>cor. sinus</td>
<td>0.14</td>
<td>125</td>
<td>-60°</td>
<td>+90° CW</td>
<td>RAE</td>
</tr>
<tr>
<td>2</td>
<td>cor. sinus</td>
<td>0.12</td>
<td>140</td>
<td>-50°</td>
<td>-120° CCW &amp; sup.</td>
<td>intermittent nodal rhythm; RVH</td>
</tr>
<tr>
<td>3</td>
<td>cor. sinus</td>
<td>0.13</td>
<td>96</td>
<td>-70°</td>
<td>-120° CCW &amp; sup.</td>
<td>RVH</td>
</tr>
<tr>
<td>4</td>
<td>cor. sinus</td>
<td>0.14</td>
<td>88</td>
<td>-60°</td>
<td>-90° CCW &amp; sup.</td>
<td>RVH</td>
</tr>
<tr>
<td>5</td>
<td>cor. sinus</td>
<td>0.12</td>
<td>120</td>
<td>-80°</td>
<td>+45° CW</td>
<td>RVH, RAE</td>
</tr>
<tr>
<td>6</td>
<td>cor. sinus</td>
<td>0.14</td>
<td>70</td>
<td>-80°</td>
<td>+270° CW</td>
<td>intermittent nodal rhythm; RVH, severe</td>
</tr>
<tr>
<td>7</td>
<td>cor. sinus</td>
<td>0.14</td>
<td>135</td>
<td>-75°</td>
<td>-125° CCW</td>
<td>RAE</td>
</tr>
<tr>
<td>8</td>
<td>cor. sinus</td>
<td>0.14</td>
<td>140</td>
<td>-60°</td>
<td>+150° CW</td>
<td>RAE, RVH</td>
</tr>
<tr>
<td>9</td>
<td>cor. sinus</td>
<td>0.14</td>
<td>90</td>
<td>-60°</td>
<td>-90° CCW &amp; sup.</td>
<td>RVH</td>
</tr>
</tbody>
</table>

Abbreviations: Cor. Sinus = coronary sinus rhythm; CW = clockwise; CCW = counterclockwise; RAE = right atrial enlargement; RVH = right ventricular hypertrophy; Sup. = superior.
in four of the seven patients on whom autopsies were performed. In two other patients on whom autopsies were performed, the coronary sinus was absent and the left superior vena cava terminated in the left atrium. These two patients also had coronary sinus-septal defects. In two additional patients, one on whom autopsy was performed and one living, a persistent left superior vena cava terminated in a normal coronary sinus. The status of the coronary sinus in the other living patient (No. 4, Table 1) is not known.

**Electrocardiographic Findings**

The electrocardiographic findings of the polysplenia patients with coronary sinus rhythm are presented in Table 2.
All patients had a leftward and superiorly oriented P wave axis on initial tracings with negative P waves in leads 2, 3, and aVF, upright P waves in leads 1 and aVL, and in all initial tracings the P wave was positive in V6. The frontal P wave axis varied from $-50^\circ$ to $-80^\circ$ in all patients prior to digoxin administration. The P-R interval corrected for age and rate was normal in all instances. Figure 1a is representative of typical coronary sinus rhythm in a patient (Pt. 2, Table 2) with polysplenia. Subsequent to digitalis (digoxin) administration, a tracing taken one day after the initial recording (Fig 1b) revealed a further leftward and superior shift in the P wave axis, and the P wave in V6 had become negative, suggesting left atrial rhythm.9

In all patients for whom there were serial electrocardiographic records, the tracings showed a transition from one atrial ectopic focus to another. An occasional patient even showed variation from coronary sinus rhythm to nodal rhythm (Fig 2) in the same tracing. In the patients on digoxin therapy, toxicity was suspected, but even when digitalis was withheld for several days, the nodal rhythm would persist.

The mean frontal QRS axis appeared leftward and superior in five patients, and the QRS inscription was counterclockwise and superior. In the remainder, the mean frontal QRS axis was either in the normal range or showed abnormal right axis deviation.

**DISCUSSION**

The electrocardiographic features characteristic of coronary sinus rhythm include P waves preceding each QRS complex, compatible with requirements of an AV rhythm; inversion of the P waves in leads 2, 3, and aVF; and a P-R interval usually of normal duration.8 Although the incidence of coronary sinus rhythm in adults has been estimated to range from 1.3 per thousand to 3.4 per thousand,8 its incidence in patients with congenital heart disease is not known.

The differentiation of coronary sinus rhythm from left atrial rhythm is often difficult, and the electrocardiographic P wave changes often overlap in these two rhythms,9 as illustrated by one of our patients. Although a diagnosis of coronary sinus rhythm is made from a leftward and superior P wave axis, it has been shown that the origin of this rhythm may be in the inferior left atrium,6,10 bundle of His,11 or lower right atrium, including the orifice of the coronary sinus.9,12 Lau and co-workers9 have paced the inferior left atrium in close proximity to the coronary sinus and have produced P wave and P loop changes which were indistinguishable from those of coronary sinus rhythm. In addition, inversion of the P wave in V6, thought to be almost diagnostic of left atrial rhythm,9 was consistently noted by these authors both in coronary sinus and left atrial pacing. Based on these observations, Lau and colleagues9 suggest that a common intra-atrial conduction pathway is utilized in the genesis of both coronary sinus and left atrial rhythms.

A recent review of all electrocardiographic records from 30 patients with the congenital asplenia syndrome on whom autopsies were performed at The Children’s Hospital Medical Center yielded only one with coronary sinus rhythm.13 Momma and Linde4 did not document coronary sinus rhythm in their six asplenia patients, but rather found a vertical and inferior P wave axis. Ruttenberg and his colleagues14 make no mention of abnormalities of atrial rhythm in their clinicopathologic review of 17 patients with the asplenia syndrome.

Why should coronary sinus rhythm be seen more frequently in patients with polysplenia than asplenia? Ongley and his associates15 noted inverted P waves in leads 2, 3, and aVF in three of five patients with a developmental complex characterized by anomalous connection of pulmonary veins to right atrium, anomalous inferior vena cava, and multiple spleens (polysplenia syndrome) and suggested the P wave abnormalities could be attributed to the low position of the sinoatrial node near the coronary sinus.

Momma and Linde4 have suggested that as polysplenia is a syndrome of bilateral left-sidedness with suppression of normal right-sided structures,8 the sinoatrial node, a right-sided structure, is poorly developed. Hence, the coronary sinus node (derived from conduction tissue of the left sinus horn16) becomes dominant, resulting in coronary sinus rhythm. This hypothesis seems unlikely for two reasons: (1) Despite the absence of the coronary sinus in two of our patients, and in two of the patients reported by Momma and Linde,4 coronary sinus rhythm was still the dominant atrial rhythm in these patients. (2) In patients with incomplete development of the left atriovenous fold, the left side of the sinus venosus maintains continuity with the left atrium and is represented grossly as termination of the left superior vena cava in the left atrium, absence of the coronary sinus, and the characteristic posteroinferior coronary sinus-septal defect.17 There may be sinoatrial conduction tissue adjacent to the left superior vena cava–left atrial junction.16 If this left-sided conduction tissue becomes the dominant atrial pacemaker, as it did in one of Van Mierop's18 asplenia patients, the frontal P wave vector should...
be directed from left to right and inferiorly, suggesting atrial inversion, not leftward and superiorly, as in coronary sinus rhythm.

Recent reports have documented that both asplenia and polysplenia share in common a high incidence of bilateral superior vena cavae, absent coronary sinus with left superior vena cava-left atrial communication and coronary sinus-septal defects, and abnormalities of the atrioventricular canal. Conotruncal abnormalities including transposition of the great arteries and pulmonary stenosis or atresia are seen in both, but are more common in asplenia than polysplenia. The most consistent and striking anatomic difference, however, is that absence of the superior portion of the inferior vena cava is very common in polysplenia and is quite rare in asplenia, although asplenia and an absent inferior vena cava may rarely coexist. Perhaps this abnormality of the sinus venosus either directly or indirectly affects the development of the right-sided sinoatrial node, predisposing to ectopic and possibly a more primitive atrial rhythm. This is only speculative, of course, and does not explain those instances of coronary sinus rhythm when there is no abnormality of the sinus venosus. In this regard it should be mentioned that while absence of the superior portion of the inferior vena cava is the usual finding in polysplenia, a normal inferior vena cava may occasionally be present. Momma and Linde and Jue and Edwards have described patients with typical polysplenia syndrome, but with a normal inferior vena cava. Their patients showed coronary sinus rhythm, although Jue and Edwards referred to the P wave abnormality as “an ectopic right atrial pacemaker.” A histopathologic study of the sinoatrial conduction system in cardiac patients with asplenia and polysplenia is in preparation.

Absence of the renal to hepatic portion of the inferior vena cava with either an aszygous or hemi-aszygous extension not infrequently makes cardiac catheterization from the leg difficult, if not impossible. Although difficulty in cardiac catheterization was encountered in all of our patients, in four the inguinal approach had to be abandoned, and the axillary approach used. In this regard, it is of interest that McLaughlin, Krovetz, and Schiebler describe difficulty in cardiac catheterization of a patient with the Laurence-Moon-Biedl-Bardet syndrome. At postmortem examination the patient (their Case 3) had polysplenia and typical cardiovascular abnormalities, including aszygous continuation of the inferior vena cava. Although the authors did not comment on the P waves of this patient’s electrocardiogram (their Fig 9), the P waves are positive in leads 1 and aVL, negative in 3 and aVF, and isoelectric in aVR, thus typical of coronary sinus rhythm.

The recognition of coronary sinus rhythm in a patient with congenital heart disease and visceral heterotaxia should alert the clinician to the consideration of polysplenia and absence of the inferior vena cava. In a critically ill infant with cardiac disease, excessive time spent in cardiac catheterization may be deleterious. In such a situation, it may be expedient to proceed from a right axillary approach. We are currently performing radionuclide peripheral venography in the not critically ill and the older patient to visualize the inferior vena cava prior to cardiac catheterization. This has proved a rapid, safe, and valuable diagnostic aid in these patients and will be subject of a forthcoming communication.

REFERENCES

Freaks in Nomenclature

The evolution of citrus probably began in the Malay Archipelago at least twenty million years ago. The ancestral plant apparently made its way to the Asian mainland, and from it developed the modern fruit. Among the first citrus varieties to go on into the Mediterranean basin was the citron which acquired its name because of an early confusion with another tree. The large rough-skinned citron resembled the greenish-yellow cone of the cedars of Lebanon, and since citron trees and orange trees are nearly identical in shape and foliage, the confusion inevitably expanded. The Greeks called the citron a kedromelon, or cedar apple. The Romans turned this into malum citreum, and applied the term, often shortened merely citreum, to all of the various fruits of citrus trees. In the eighteenth century, the Swedish botanist, Linnaeus (1707–1778), made the name “citrus” official for the genus. So lemons, limes, citrons, oranges, grapefruit, and tangerines are now grouped under a name which means cedar. The word “orange” evolved from Sanskrit. The Hindus called an orange naranga. This became the Persian narans, Neo-Latin aurantium or aurantium, arancia in Italy, and orange in France.