Kartagener's Syndrome with Corrected Transposition

Conducting System Studies and Coronary Arterial Occlusion Complicating Valvular Replacement

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An 18-year-old man whose sister has classic Kartagener's syndrome was found to have sinusitis, bronchiectasis, and corrected transposition with normal visceral situs. Congenital complete heart block was secondary to absence of conducting-system pathways between a small posterior atrioventricular node and the transposed His bundle and bundle branches. No anterior atrioventricular node was present. Prosthetic valvular replacement of the left-sided (morphologic right) atrioventricular valve was complicated by coronary arterial occlusion by suture, with subsequent myocardial infarction. The case appears to represent an unusual variant of Kartagener’s syndrome with the abnormality of laterality being expressed as corrected transposition.

Kartagener’s syndrome, consisting of situs inversus, bronchiectasis, and chronic sinusitis, was first described in 1933.1 A review in 19622 collected a total of 334 cases from the world literature, and more cases have since been added.3 We have observed a unique variant of this syndrome, a patient with bronchiectasis, chronic sinusitis, and corrected transposition with situs solitus. In addition, we have described the anatomy of the conducting system and a complication of surgical replacement of the left-sided atrioventricular valve in this patient, and have reviewed the association of congenital heart disease with Kartagener’s syndrome.

CASE REPORT

A 4,340-gm (9 lb, 9 oz) boy was the product of a full-term pregnancy in a 30-year-old white woman. The fetal heart rate was less than 100 beats per minute, and congenital heart block was confirmed by an electrocardiogram after birth. An older sister had situs inversus, sinusitis, and bronchiectasis. The patient’s subsequent physical development was normal, although he had recurrent infections of the upper respiratory tract. At the age of 14 years, because of an enlarging heart, the patient underwent cardiac catheterization with His-bundle studies which were interpreted as showing absent conduction from the atrioventricular node to the ventricular myocardium. He did well until the age of 15 years, when progressive dyspnea, frequent respiratory infections, weight loss, and general malaise developed. At age 18,

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Figures 1. Left, Chest radiograph at age 11 years. Heart is enlarged. Splenic shadow is seen in left upper abdominal quadrant. Right, At age 18 years, patient’s heart shows marked enlargement. Scoliosis is also present.

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because of congestive heart failure, the patient was admitted to The Johns Hopkins Hospital, Baltimore. Physical examination revealed a thin, chronically ill-appearing adolescent. Tachypnea, a left ventricular heave, a grade-3/6 blowing holosystolic murmur at the left lower sternal border, a loud middiastolic apical murmur, and diffuse rales in both pulmonary fields were found. A chest radiograph (Fig 1) showed massive cardiomegaly and pulmonary congestion, and an ECG demonstrated complete heart block (Fig 2). Cardiac catheterization revealed ventricular inversion with transposition of the great vessels. The left-sided atrioventricular valve was grossly incompetent, and the left-sided atrium and ventricle were markedly dilated. The left atrioventricular valve was replaced with a 31 mm Björk-Shiley prosthesis, and an epicardial pacemaker was implanted. The patient initially did well; but on the seventh postoperative day, despite anticoagulation therapy, he developed a dense left hemiparesis. A brain scan demonstrated occlusion of the right middle cerebral artery. The patient subsequently did poorly, was troubled with multifocal premature ventricular contractions, and died with ventricular fibrillation on the 38th postoperative day.

At autopsy there was normal situs of the heart, lungs, and abdominal viscera. The heart weighed 680 gm and showed the features of corrected transposition. The aorta arose anteriorly from a left-sided (morphologic right) ventricle. A prominent crista supraventricularis was present between the aortic valve and the implanted Björk-Shiley valve. The pul-

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**Figure 2.** Electrocardiogram showing complete heart block with ventricular rate of 43.

**Figure 3.** Top left, Apical portion of transversely sectioned heart shows massive myocardial infarction involving posterior wall of both ventricles and interventricular septum. Top right, base of transversely sectioned heart viewed from atrial aspect shows Björk-Shiley prosthesis in place in left-sided atrioventricular valvular ring. Center right, Postmortem radiograph of base of heart oriented as above. Arrow 1 indicates point of suture occlusion of left-sided circumflex coronary artery. Arrows 2 and 3 indicate positions of serial histologic sections of conducting system shown below. Note soft tissue shadow of crista supraventricularis between anterior aortic valve and left-sided atrioventricular valve containing prosthesis. Center left, Section from level of arrow 1 showing markedly narrowed left-sided circumflex coronary artery at site of suture transfixation (arrows) of wall. Part of lumen is still present at left and filled with black-staining injection mass. Remainder of lumen at right is compressed and filled with organized thrombus (hematoxylin-eosin, original magnification × 25). Lower left, Section from level of arrow 2. Small posterior atrioventricular node is at right of artery filled with black-staining injection mass in interatrial septum. Prosthesis was sutured into atrioventricular ring at left (hematoxylin-eosin, original magnification × 10). Lower right, Section from level of arrow 3. His bundle gives rise to left (morphologic right) and right (morphologic left) bundles but has no connection with atrial muscle. No anterior atrioventricular node was found (hematoxylin-eosin, original magnification × 25).
monary artery arose from the right-sided (morphologic left) ventricle. The right atrioventricular valve resembled a mitral valve and had fibrous continuity between its anterior leaflet and the pulmonic valve.

Postmortem coronary arteriographic studies revealed subtotal occlusion of the left-sided coronary artery (Fig 3). A healing myocardial infarct in the distribution of this artery involved the posteroseptal wall of the left-sided ventricle and was histologically consistent with an age of five to six weeks. Serial histologic sections of the occluded segment of the left circumflex coronary artery revealed suture material transfixing the walls of the artery and an associated organizing luminal thrombus.

Serial histologic sections of the entire atrioventricular septal area showed a small atrioventricular node subendocardially on the right side of the interatrial septum just anterior to the coronary sinus in the usual position. No anterior node, as previously described in corrected transposition,6-7 was present. The posterior node was separated by fibrous tissue from the His bundle, which was entirely below the central fibrous body. The His bundle gave rise to a single large bundle supplying the left-sided ventricle and several smaller bundles fanning out to the right-sided ventricle. No connections between atrial and ventricular muscle were found.

The lungs were arranged normally with three right lobes and two left. Bronchiectasis, with peribronchial fibrosis and chronic inflammation, was most marked in the lower lobes. The sphenoidal sinus contained mucoid material and showed sinusitis. Thromboemboli, probably arising from the mural thrombus overlying the myocardial infarct, accounted for a cystic infarct in the distribution of the right middle cerebral artery and for a small organizing splenic infarct.

**DISCUSSION**

Kartagener's syndrome is a triad which has been suggested to have recessive inheritance and incomplete penetrance.2-4 It is commonly seen in siblings, but vertical transmission from an affected individual to a child has not been described. There have been numerous reports5,6-15 of siblings who manifest all possible combinations of the three phenotypic features of the syndrome. Variability of organ-system involvement may be explained by a single gene with a pleiotropic effect.16 An alternative hypothesis that several linked genes are responsible seems statistically less likely; however, at present the mode of inheritance has not been established.17 Although the combination described in this report has not been previously recorded, we believe that it does represent a variant of Kartagener's syndrome, particularly since the patient's only sibling demonstrated the classic triad.

Congenital heart disease is uncommon in Kartagener's syndrome. Of the cases reported,5,16-20 no single lesion seems to be characteristic, as far as can be judged from the incomplete descriptions (Table 1). Cases 1, 3, and 4 appear to be, at least in part, malformations of laterality of the cardiovascular system. Our case is of interest in that corrected transposition represents the sole manifestation of a laterality malformation. Corrected transposition is thought to result from looping of the primitive heart tube to the right side, instead of to the left, as occurs normally.21 The mechanism of heart looping is poorly understood. Butler22 demonstrated that looping occurs in the absence of fixation of the proximal and distal ends of the heart tube, and Manasek and Monroe23 have shown that looping will occur in the absence of blood flow in a noncontracting tube. Mechanical and hemodynamic factors are not primary determinants in the mechanism of looping; rather, an intrinsic property of the myocardial cell determines the side to which the heart loops. The corrected transposition in this patient, whose sister has complete situs inversus, may represent a partial expression of genetically determined situs inversus, rather than a fortuitous finding.

Occlusion of the left coronary artery at surgery resulted from suture material transfixing the artery in the annulus of the systemic atrioventricular valve. This artery courses in the left atrioventricular groove but has the distribution of a right coronary artery.4 Occlusion resulted in a posteroseptal myocardial infarct, a complication described in another patient with corrected transposition undergoing anuloplasty for a regurgitant left-sided atrioventricular valve.4 These aberrant locations of the right coronary artery, therefore, represents a definite hazard during surgery involving the systemic atrioventricular valve in corrected transposition.

Reports of the anatomy of the conducting system in corrected transposition are few and at variance.5-7 Anderson et al6,6 describe a small posterior atrioventricular node in the normal position and a second node anteriorly which gives rise to the His bundle and subsequently the bundle branches. The morphology of the bundle branches is inverted; a large single bundle (normal right) is found in the left-sided ventricle, and the fascicles forming the morphologic left bundle are in the right-sided ventricle. This case is of particular interest in that the patient had congenital heart block and serial histologic studies revealed no communication between the small posterior atrioventricular node and the His bundle and no atrioventricular connection or evidence of an anterior atrioventricular node. Anderson et al6 attribute the development of heart block, seen commonly.

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**Table 1—Congenital Heart Disease in Kartagener's Syndrome**

<table>
<thead>
<tr>
<th>Case</th>
<th>Reference</th>
<th>Malformation</th>
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<tbody>
<tr>
<td>1</td>
<td>Dickey18</td>
<td>Transposition of great vessels and ventricular septal defect</td>
</tr>
<tr>
<td>2</td>
<td>Divekar19</td>
<td>&quot;Cyanotic congenital heart disease&quot;</td>
</tr>
<tr>
<td>3</td>
<td>Holmes et al10</td>
<td>Transposition of great vessels and biventricular or tricuspid heart</td>
</tr>
<tr>
<td>4</td>
<td>Holmes et al10</td>
<td>Asplenia syndrome with single ventricle, atrial septal defect, pulmonary stenosis, and anomalous pulmonary venous drainage to portal vein</td>
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<tr>
<td>5</td>
<td>Pomerleau et al20</td>
<td>Tetralogy of Fallot</td>
</tr>
<tr>
<td>6</td>
<td>Solomon et al</td>
<td>Corrected transposition</td>
</tr>
</tbody>
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in adult patients with corrected transposition, to fibrous replacement of the conducting tissues connecting the anterior node to the His bundle. In our patient with congenital heart block, the fibrous tissue between the atrioventricular node and His bundle, which may represent a distorted central fibrous body, was presumably present prior to birth and provides an anatomic explanation for heart block.

In conclusion, this patient with Kartagener’s syndrome had corrected transposition as the manifestation of abnormal laterality. The cardiac conducting system was unusual in that congenital heart block was present and no anterior atrioventricular node was found. Prosthetic valvular surgery was complicated by myocardial infarction from suture injury and thrombosis of the left circumflex coronary artery.

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Pericarditis with Effusion Caused by Actinomyces israelii*

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Actinomyces israelii is a rare cause of pericarditis. Infection spreads to pericardium from an intrathoracic focus. Branching gram-positive filaments are seen on microscopic examination of clinical specimens. Cultures should be performed under anaerobic conditions. Both surgical drainage and antibiotics (penicillin) are required for treatment. Nocardiosis may resemble actinomycosis in many respects.

Actinomyces israelii infrequently causes disease in man. Pericarditis caused by this organism is even rarer.1 We recently cared for a patient who had signs and symptoms of pericarditis with effusion and a pulmonary infiltrate resulting from infection with A israelii.

CASE REPORT

A 47-year-old man was admitted to the Atlanta Veterans Administration Hospital because of chest pain and cough.

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