Acquired Right Bundle-Branch Block and Left Anterior Hemiblock in Ostium Primum Atrial Septal Defect

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SUMMARY
Over a 3-year period of observation, the electrocardiogram in an adult patient with an ostium primum atrial septal defect changed from first-degree atrioventricular block alone to right bundle-branch block and left-axis deviation (left anterior hemiblock) with variable atrioventricular block. Coronary atherosclerosis was excluded by selective coronary arteriography. Although right bundle-branch block and left anterior hemiblock are commonly present at birth in patients with endocardial cushion defect, this is believed to represent the first reported instance in which these electrocardiographic abnormalities have developed under observation in later life. It is postulated that chronic hemodynamic stress contributed to the progression of disease in the conduction system.

Additional Indexing Words:
First-degree atrioventricular block
Complete atrioventricular block
Endocardial cushion defect

Second-degree atrioventricular block
Coronary arteriography
Left-axis deviation

LEFT-AXIS DEVIATION with right bundle-branch block is of value in distinguishing patients with ostium primum atrial septal defect (incomplete endocardial cushion defect) from those with ostium secundum atrial septal defect.1-3 Blount and co-workers2 ascribed the left-axis deviation to left ventricular hypertrophy resulting from associated mitral regurgitation. Toscano-Barbosa and colleagues,8 on the other hand, held that the conduction disturbances in this condition were due to a congenital abnormality of the conduction system, unrelated to mitral regurgitation or other pathophysiologic disturbances.

The present report describes a 44-year-old woman with an ostium primum atrial septal defect in whom the electrocardiogram changed from first-degree atrioventricular block to variable atrioventricular block associated with complete right bundle-branch block and left anterior hemiblock, over a 3-year period of observation. This is believed to be the first reported case of this congenital defect in which the typical conduction disturbances developed in later life.

Case Report
A 44-year-old female was first noted to have a heart murmur at 12 years of age. She bore four children without difficulty, but at 35 years of age she developed easy fatigability and dyspnea on exertion, which was slowly progressive over the 3 years prior to admission. She also noted chest discomfort, not...
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typical of angina pectoris, but continued to do most of her housework.

The blood pressure was 120/70. The V wave in the jugular venous pulse was increased. There was a left ventricular lift 1 cm lateral to the mideclavicular line in the fifth intercostal space. The second heart sound was widely split, and the splitting was fixed during the respiratory cycle. A grade III/VI high-pitched, blowing holosystolic murmur at the cardiac apex, which radiated to the left axillae, and a grade I/VI short ejection murmur at the left second intercostal space were audible. Chest roentgenograms demonstrated left ventricular enlargement, slight left atrial enlargement, and mildly increased pulmonary vascular markings.

Cardiac catheterization studies at 42 and 43 years of age demonstrated a low-lying atrial septal defect with a 1.6 to 1 left-to-right shunt at the atrial level. The mean left and right atrial pressures were equal, but there was an elevated V wave in the left atrial tracing (20 mm Hg). Pulmonary artery pressure was normal. Left ventricular cineangiograms demonstrated mild mitral regurgitation and a typical "gooseneck" deformity of partial endocardial cushion defect with cleft mitral valve. Coronary arteriography revealed no demonstrable atherosclerotic disease.

Representative electrocardiograms are shown in figure 1. The first electrocardiogram, dated 10/30/59, was considered to show only minimal prolongation of the P-R interval, minor right ventricular conduction delay, and counterclockwise rotation. The second tracing shown, dated 10/3/61, was comparable to the first, except for a slight increase in the QRS amplitude in leads II and aV1, suggestive of left ventricular hypertrophy; there was no evidence of an rSr' complex in V1. The third tracing, on 8/15/66, demonstrated first-degree atrioventricular block, right bundle-branch block, and left-axis deviation. Other electrocardiograms taken in 1969 (not shown) demonstrated second-degree block and on one occasion transient complete atrophicventricular block. The most recent electrocardiogram, taken 2/2/70, showed slight further deviation of the mean frontal QRS axis superiorly and to the left.

Discussion

Between the ages of 38 and 41 years, this patient's scalar electrocardiograms changed from mild first-degree atrioventricular block, to variable, more-advanced atrophicventricular block associated with complete right bundle-branch block and left-axis deviation. The patient's most recent electrocardiogram (fig. 1) satisfied the criteria for the diagnosis of bifascicular disease involving the right bundle branch and the anterior superior division of the left bundle branch. These features included: (1) typical, complete right bundle-branch block; (2) left-axis deviation, and a mean vector for the first half of the QRS interval at about -60° with a small rS in lead II; and (3) small, normal Q waves in leads I and aVl. Although the presence of first-degree atrioventricular block is compatible with concomitant disease in the posterior division of the left bundle branch (trifascicular block), this diagnosis cannot be made with confidence from scalar electrocardiograms. It is of interest, however, that complete atrioventricular block occurred transiently in this patient.

There are several possible explanations for progression of disease of the conduction tissue in patients with ostium primum atrial septal defect, as documented in the present report. The right bundle branch and the anterior superior division of the left bundle are thinner than the posterior division of the left bundle and therefore vulnerable to injury by passive stretch resulting from atrial or ventricular enlargement. Several studies have demonstrated that the right bundle branch and the anterior division of the left bundle lie near the rim of the endocardial cushion defect. Progressive, chronic endocardial fibrosis, caused by turbulence in the region of the defect could directly damage these contiguous conduction fascicles. Of course, bifascicular disease also could develop as a coincidental feature in an older patient. However, in the present patient coronary atherosclerosis was excluded by selective coronary arteriography. Other
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causes of disease in the conduction fascicles, such as sclerosis of the cardiac skeleton\(^{10}\) and scleral degenerative changes\(^{11}\) of the cardiac conduction tissue, would be unusual in a young patient.

Lev,\(^7\) Visioli,\(^8\) and Feldt\(^6\) and their associates have shown that anatomical displacement of the normal conduction tissue is a relatively constant finding in endocardial cushion defect. Rossi\(^{12}\) demonstrated disruption, interruption, and absence of fascicles in endocardial cushion defect, and Visioli and co-workers\(^8\) described interruption of conduction fascicles with anomalous insertion of chordae tendinea directly into the free wall of the left ventricle.

The fact that left-axis deviation is present at birth in many infants with incomplete endocardial cushion defect was used by Burchell and associates\(^{13}\) as circumstantial evidence that left-axis deviation results from a congenital abnormality of conduction tissue. An explanation for left-axis deviation was also presented by Durrer and associates\(^{14}\) who found abnormally early activation of the posterior-inferior surface of the left ventricle, which they ascribed to early takeoff and foreshortening of the posterior division of the left bundle branch from the bundle of His. Burchell and co-workers,\(^{13}\) using similar techniques, were unable to corroborate these findings, however. This mechanism seems unlikely in our patient since the earliest electrocardiographic tracings showed normal left ventricular activation.

The findings in the present patient indicate that in rare instances right bundle-branch block and left anterior hemiblock will not be present at birth in patients with ostium primum atrial septal defect but will develop later, presumably as a result of chronic abnormal hemodynamic stresses. Although this is believed to be the first reported case in which these conduction disturbances have developed after birth, partial regression of conduction abnormalities has been reported following surgical repair. Thus, Levy and associates\(^{15}\) reported that in 28 of 32 patients left-axis deviation was less marked following surgical repair of the lesion, and three patients with left-axis deviation preoperatively had normal mean frontal QRS axes postoperatively. These observations support the view that the conduction disturbances in some patients are at least in part dependent upon longstanding physiologic stresses. Finally, the present findings indicate that acquired trifascicular block may occur, and should be watched for, during the natural course of patients with ostium primum atrial septal defect.

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Figure 1

Serial electrocardiograms demonstrating progression of conduction disturbances in a patient with ostium primum atrial septal defect. See text for details.

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