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EDITORIAL COMMENT

Electrophysiology Follows Anatomy?*

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In 1936, Toscano-Barbosa et al. (1) at the Mayo Clinic called attention to 3 electrocardiographic features of what came to be known as atrioventricular septal defects (AVSDs): 1) the PR interval; 2) the frontal plane QRS axis; and 3) the sequence of ventricular activation. Toscano-Barbosa et al. (1) recognized certain distinctive features of the QRS in standard 12-lead electrocardiograms, stating, ". . .the similarity that may exist in the precordial leads may lead one astray from the differences that almost universally exist in the extremity leads which are of real discriminatory value." These features are more clearly illustrated by vector analysis (2) (see below).

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Left-axis deviation in AVSDs ranges from moderate to extreme, with the QRS axis directed superior and to the left or superior and to the right. The mean axis can reach −180°—an extreme left-axis deviation (3,4) (Fig. 1). Counterclockwise depolarization results in Q waves in leads I and aVL. An abrupt change in the sequence of terminal force ventricular activation causes notching of the S waves in leads II, III, and aVF, also best illustrated in the vector loop (2) (Fig. 2). The mechanisms responsible for the characteristic patterns of the QRS were believed to stem from congenital alterations in the excitation pathways into the ventricles, as originally proposed by Toscano-Barbosa et al. (1,3).

The left anterior division of the left bundle branch in AVSDs is increased in length and has fewer fibers than normal. The left posterior division is shorter than normal and provides small branches to the posterobasal wall of the left ventricle. These features of the left bundle branch result in early activation of the posterobasal left ventricular wall and in delayed activation of the anterior superior wall (3), anatomic and electrophysiologic characteristics that have long been regarded as explanations for the left-axis deviation and depolarization patterns of AVSDs.

Why then did Hakacova et al. (5), in this issue of *iJACC*, propose a new explanation, namely that leftward deviation of the QRS axis in AVSDs is the result of (correlates with) an imbalance in the positions of left ventricular papillary muscles? Papillary muscle locations relative to the interventricular septum and left ventricular free wall are examples of recent clinical interest in the positions of these structures.

The mitral apparatus consists of 5 components: the left atrium, the annulus, the leaflets, the papillary muscles, and the contiguous left ventricular wall (6). Ostium primum atrial septal defects (primum AVSDs), and indeed all forms of AVSDs, are congenital malformations of the atrioventricular apparatus and atrioventricular junction, i.e., the mitral apparatus, and have recently been studied in normal individuals and related to the QRS axis and to the location of the conduction system, specifically the left fascicles as determined by the positions of left ventricular papillary muscles relative to borders of the fascicular fen. The left bundle branch and its divisions fan out just beyond their origins, and constitute a transition zone, or fen, between the papillary muscles and the ventricular septum and free wall. Leftward deviation of the QRS axis associated with primum AVSDs has now been related to anatomic displacement of the papillary muscles that is believed to alter the activation sequence of the left ventricle.

The study by Hakacova et al. (5) was based on 2 assumptions: 1) the positions of the left ventricular papillary muscles between the interventricular septum and the left ventricular free wall localize the

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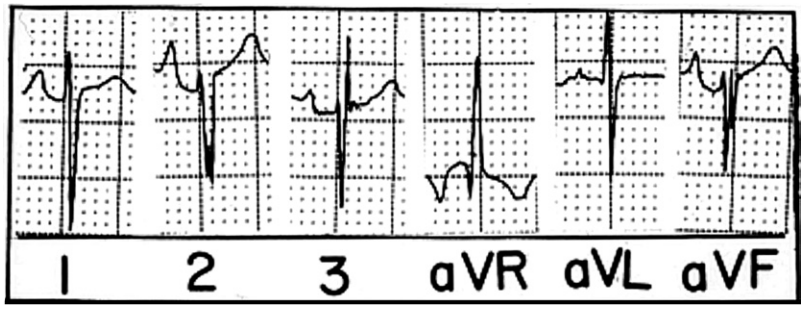

Figure 1. Limb Leads in a Patient With Down Syndrome and an Atrioventricular Septal Defect

Left-axis deviation is extreme, with the axis directed upward and to the right. A small Q-wave in lead aVL suggests counterclockwise depolarization. The S waves are notched in leads II and aVF.

lateral borders of the fascicular fen, and therefore localize 2 of the starting points of left ventricular activation; and 2) an anterior papillary muscle that is closer to the septum than the posterior papillary muscle results in delayed activation of the anterior left ventricular free wall and in leftward deviation of the QRS axis. The objective of the study by Hakacova et al. (5) was to test the hypothesis that in primum AVSDs, an anatomic imbalance in the positions of left ventricular papillary muscles correlates with leftward deviation of the QRS axis. Imbalance between papillary muscle positions was defined as an anterior papillary muscle closer to the ventricular septum than the posterior papillary muscle. The anterior muscle was considered closer to the septum when its distance from the midportion of the septum was less than its distance from the midpoint of the free wall. The posterior papillary muscle was considered abnormal when its distance from the midseptum was smaller than its distance from the midpoint of the free wall. Echocardiograms were used to determine the positions of the anterior and posterior papillary muscles relative to the ventricular septum and left ventricular free wall. The accuracy of these measurements hinged on the accuracy of identifying the origin of the papillary muscles. However, echocardiography has its limitations in this regard because adjacent structures may have similar echogenicity. It was reassuring that echocardiographic determinations of left ventricular papillary muscle positions relative to the ventricular septum and the left ventricular free wall were validated by precise magnetic resonance imaging of papillary muscle positions. These positions served as markers of the borders of the endocardial area supplied by the left fascicles that is activated earlier than the remainder of the left ventricular endocardium.

The degree of shift of the electrical axis was numbered in a clockwise direction (positive axis) and in a counterclockwise direction (negative axis). Vectocardiograms would have better served this purpose.

Primum AVSDs were divided into 3 groups according to the direction of the QRS: 1) typical left-axis deviation; 2) horizontal axes; and 3) inferior axes. Forty-six percent of the patients in the study by Hakacova et al. (5) had typical left deviation of the QRS, 37% had a horizontal QRS axis, and 17% had an inferior axis. When the anterior and posterior papillary muscle positions in these 3 groups were compared, significant differences emerged. When the position of each papillary muscle was compared separately with deviation of the QRS axis, displacement

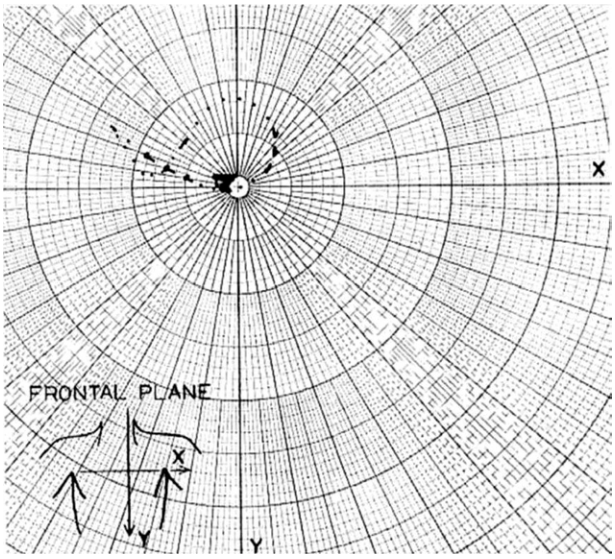

Figure 2. Vectorcardiogram in a Patient With an Ostium Primum AVSD

Depolarization is counterclockwise, the frontal plane axis is upward and to the right, and there is an abrupt change in terminal force direction.

of the posterior papillary muscle further from the septum correlated significantly with the axis ($r = 0.4$, $p = 0.03$), but correlation between the position of the anterior papillary muscle and the QRS axis was not significant ($r = 0.1$, $p = 0.4$).

A relationship between papillary muscle positions and QRS characteristics was recently described in normal subjects. Interestingly, 17% of primum AVSDs in the study by Hakacova et al. (5) had an inferior QRS axis with papillary muscle positions similar to normal. Electroanatomic abnormalities in primum AVSDs may therefore represent a continuum, with primum AVSDs, normal QRS axes, and normal papillary muscle positions a distinct category. Accordingly, the most significant finding in the study was a connection between papillary muscles and electrophysiological characteristics that implied a developmental relationship between the conduction system and the trabecular structures.

The relationship between papillary muscle positions and the degree of leftward deviation of the QRS axis was convincingly shown. The authors might now consider extending their study to patients with Down syndrome and extreme left-axis deviation. They might also comment, even if the comments are speculative, on the distinctive change in sequence of ventricular activation reflected in the abrupt alteration in terminal force direction of the QRS that notches the S waves in leads II, III, and aVF, and is best represented in the vectorcardiogram. It could also be argued that use of vectorcardiograms throughout the study coupled with the scalar electrocardiogram would have served the authors' purpose better.

Hakacova et al. (5) focused on primum AVSDs defined as malformations of the atrioventricular junction above the level of the atrioventricular valve, with no detectable defect below the valve. This focus was not a limitation, however, because the specialized conduction pathways in all forms of AVSDs are believed to have the same distribution (7).

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Key Words: atrioventricular septal defect ■ electrophysiology ■ left axis deviation.