CASE REPORT

Left Anterior and Left Posterior Hemiblock in Tricuspid Atresia and Transposition of the Great Vessels

Observations and Electrocardiographic Nomenclature and Electrophysiologic Mechanisms

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SUMMARY A patient with tricuspid atresia and characteristic electrocardiographic features of counterclockwise and superiorly oriented frontal plane QRS loop (left anterior hemiblock) is presented. Operative intervention resulted in a clockwise and inferior rotation of the frontal QRS loop (left posterior hemiblock) without the development of complete left bundle branch block. This observation suggests that the electrocardiographic pattern of left anterior hemiblock may result from other mechanisms in addition to block of left bundle branch fibers oriented toward the anterior part of the left ventricle. The case further suggests that electrocardiographic patterns of apparent A-V conduction defects may not be at all associated with true block in the A-V conduction system. Further, it emphasizes the fact that various electrophysiologic mechanisms may account for identical electrocardiographic patterns.

THE ELECTROCARDIOGRAM of patients with tricuspid atresia typically shows a superior and counterclockwise rotation of the mean frontal QRS loop or "left axis deviation." This electrocardiographic finding is considered to represent a left anterior hemiblock, originating from destructive lesions of the anterior portion of the left bundle branch, and delayed activation of the anterolateral part of the left ventricle.

It has been suggested previously that a counterclockwise and superiorly oriented frontal plane QRS loop may not necessarily represent block in the anterior part of the left bundle branch but may result from asynchronous activation of the left ventricle. Such may be the case for different forms of congenital heart defects in which there are different arrangements of the left ventricular conduction system such that the posterior left ventricular wall is activated prior to the anterior left ventricular wall. A similar pattern of left ventricular activation sequence also may result from premature activation of the posteroinferior left ventricular wall as has been shown to occur with premature ventricular depolarizations.

In this report we present a case of tricuspid atresia with serial electrocardiograms showing a pattern of both a left anterior and a left posterior hemiblock but no complete left bundle branch block at any time. Our observations on this case lend further support to the hypothesis that a counterclockwise and superiorly oriented QRS loop in the frontal plane need not necessarily result from block in the anterior part of the left bundle branch and that almost identical electrocardiographic patterns may be caused by different electrophysiologic mechanisms.

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Case Report

MM is a seven-year-old girl for whom the diagnosis of tricuspid atresia, transposition of the great arteries, ventricular septal defect, infundibular pulmonic stenosis and juxtaposition of the atrial appendages was established immediately after birth by cardiac catheterization.

The electrocardiogram (fig. 1) demonstrated notched P waves with a mean frontal P axis of 0°, a counterclockwise and superiorly oriented frontal plane QRS complex (−40°), prominent anterior forces in V1 and V2 and ST-segment depression with T wave inversion in V6, which were consistent with a pattern of left ventricular strain.

Cardiac catheterization and angiocardiography confirmed the original findings and, in addition, showed a marked pressure gradient between the left and right ventricles by a restrictive ventricular septal defect.

The patient was referred for open heart surgery because of severe congestive heart failure, markedly limited physical activity and recurrent chest pain.

At surgery a right ventriculotomy was performed. The right ventricle was small with heavy trabeculations obstructing the aortic outflow tract. These obstructing trabeculations were removed. A muscular ventricular septal defect was identified and the interventricular septum was carefully electrophysiologically delineated from the right side.11 Electrograms of the specialized ventricular conduction system could not be identified in the operative field and the ventricular septal defect was moderately enlarged to increase systemic flow. In addition, a second muscular ventricular septal defect was created in the subaortic position. The heart remained in normal sinus rhythm during these procedures without axis shift. However, subsequent palpation in the subaortic area through the newly created ventricular septal defect produced a brief period of heart block. When sinus rhythm resumed, the frontal axis of the QRS complex had shifted markedly to the right. The remainder of the operation was well tolerated.

Postoperatively the patient had low cardiac output and severe heart failure which required vigorous treatment with
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Dopamine and isoproterenol for several days. Her condition improved gradually and she was maintained on digoxin and diuretics. Throughout the postoperative period her electrocardiogram (fig. 2) demonstrated a mean QRS axis in the frontal plane of $+120^\circ$, and superior and leftward initial forces which were followed by maximal rightward and inferior forces and were consistent with a left posterior hemiblock. This electrocardiographic pattern was seen at heart rates of 111 to 160 beats/min. Pre-existing anterior forces in $V_1$ and $V_2$ had diminished with further depression of ST segments and disappearance of the Q waves in $V_4$ and $V_6$.

Thirteen days postoperatively, the electrocardiogram reverted to the preoperative pattern with a mean frontal QRS axis of $-60^\circ$ with small Q waves in $V_4$ and $V_6$ consistent with a left anterior hemiblock pattern (fig. 3). Heart rate was 115 beats/min.

Discussion

The counterclockwise superiorly oriented frontal plane QRS loop in the preoperative electrocardiogram for this patient has been ascribed to impaired conduction in the left anterosuperior fibers of the left bundle branch system. Although the electrocardiogram is considered by many to indicate block to left bundle branch fibers oriented toward the anterior part of the left ventricle, it is important to realize that the electrocardiographic pattern of left anterior hemiblock does not distinguish between cases with early activation of the posterior left ventricular wall, slowed conduction without complete block to anterior left bundle branch fibers or a different distribution of the left bundle branch with an elongated path for left bundle branch fibers oriented toward the anterior part of the left ventricle. Were the electrocardiogram due to a true block to the anterior fibers of the left bundle, it would be reasonable to assume that any additional injury to the remaining posterior fibers should produce an electrocardiographic pattern of a complete left bundle branch block. Numerous electrocardiograms recorded during the postoperative period repeatedly demonstrated only a left posterior hemiblock pattern. A left bundle branch block pattern was not observed throughout the entire period.
period of hospitalization. In addition the posterior hemiblock pattern occurred at rates of 111 to 160 beats/min. As conduction improved in the posterior portion of the left bundle system, the anterior hemiblock pattern was again seen at a rate of 115/min. We therefore suggest that some other electrophysiologic mechanism, not associated with block to the anterior part of the left bundle branch, accounted for the altered modes of left ventricular activation in this patient.

The argument can be made that functional impairment with decreased conduction velocity in the anterior portion of the left bundle may account for delayed activation of the anterior part of the left ventricle and result in a superiorly oriented frontal plane QRS axis. However, in our patient, serial electrocardiograms obtained following surgery showed a posterior hemiblock pattern at heart rates of 111 to 160 beats per minute. We therefore feel that the presence of normal A-V conduction at these high rates along the anterior fibers of the left bundle branch strongly suggests the lack of functional impairment of this structure.

Studies on patients with other forms of congenital heart defects and with a characteristic superiorly oriented counterclockwise QRS loop in the frontal plane suggest other possible mechanisms for such an electrocardiographic pattern. It has been shown with the use of epicardial mapping techniques that in patients with counterclockwise superiorly oriented frontal plane QRS loops, posteroinferior left ventricular wall activation occurs early in relation to activation of the anterosetal left ventricular wall. Spach et al. postulated that this represented premature stimulation of the posterior left ventricular wall rather than late activation of the anterior left ventricular wall due to congenital anatomic abnormality in the distribution of the left bundle branch.

Boineau and co-workers studied a dog with congenital ostium primum defect. From correlations of electrophysiologic and anatomic data, they demonstrated premature activation of the posterior left ventricular wall. Thus, an unusual anatomic distribution of the specialized conduction system had been invoked to explain a markedly counterclockwise and superiorly oriented frontal plane QRS complex. This mechanism is distinctly different from pathologic interruption of anterior fascicles of the left bundle.

Tricuspid atresia characteristically demonstrates a markedly counterclockwise and superiorly oriented frontal plane QRS complex. Electrocardiographic and vectorcardiographic analyses have confirmed this finding in from 53 to 92% of cases. Several mechanisms have been suggested to explain the electrocardiographic pattern in these patients: lack of development of right ventricle, increased left ventricular mass, fibrosis of left bundle branch system, destructive lesions of the anterior-superior fibers of the left bundle branch system and unusual distribution of the conduction system resulting in asynchrony of left ventricular activation. Increased left ventricular mass and fibrosis of the left bundle branch system may result in frontal leftward deviation, but do not explain a marked shift in the frontal QRS complex such as seen in association with disruptive lesions of the anterior-superior fibers of the left bundle branch. Indeed, this has been amply documented in experimental studies. In addition, significant fibrosis is not observed in histologic examination of the conduction system in tricuspid atresia. Although fibrosis may be observed in various forms of congenital heart disease, this mechanism is more likely to progress with time rather than to be a cause for a counterclockwise and superiorly oriented frontal plane QRS loop which is, in many, present from birth.

The findings of Guller et al. demonstrating early origin of the left bundle branches near the atrioventricular node-His bundle junction, suggest that the unusual distribution of the conduction system in patients with tricuspid atresia is the most likely explanation for the observed counterclockwise superiorly oriented frontal plane QRS loop in these patients. This abnormal anatomic distribution of the left bundle branch may lead to either early activation of the posterior left ventricular wall as noted by Boineau, Spach and Durrer et al. in patients with ostium primum defects, or delayed activation of the left anterior ventricular wall due to a prolonged path for the anterior fibers of the left bundle branch. As yet, similar electrophysiologic observations have
not been made in patients with tricuspid atresia. The counterclockwise and superiorly oriented frontal plane QRS loop initially found for our patient may therefore be explained on the basis of asynchronous activation of the left ventricle with delayed activation of the anterior left ventricular wall in relation to the posterior left ventricular wall rather than conduction impairment of the anterior fibers of the left bundle branch. Such a mechanism would also explain our patient's serial electrocardiograms and the development of a posterior hemiblock pattern rather than a complete left bundle branch block pattern.

The case presented in this study emphasizes the fact that various electrophysiologic mechanisms may account for identical electrocardiographic patterns. For example, proximal right bundle branch lesions currently cannot be differentiated from peripheral lesions of the right bundle branch.17 A right bundle branch block pattern in association with a counterclockwise and superiorly oriented QRS loop may result from His bundle lesions, a lesion of the right bundle branch and the anterior fibers of the left bundle branch, a congenital difference in distribution of the left bundle branch or a combination of these.18

Further, electrocardiographic patterns of apparent A-V conduction defects may not be associated at all with block in the A-V conduction system but rather with electrocardiographically unrecorded (concealed) premature beats.19,20

The term pseudoblock previously has been referred to electrocardiographic patterns suggesting the presence of a conduction disturbance but resulting from mechanisms other than impaired conduction. We propose that pseudoblock due to concealed depolarizations19,20 should be referred to as "functional pseudoblock." In addition, apparent A-V conduction impairment may be caused by a congenitally different arrangement of the specialized A-V conduction system as seen in some patients with congenital heart disease and electrocardiograms with a counterclockwise superiorly oriented frontal QRS loop or in patients with P-R prolongation — first degree A-V block — in whom it has been shown to occur without impairment to the functional capacity of their specialized A-V conduction system.21 Such pseudoblocks are anatomic in origin.

Various electrophysiologic mechanisms leading to the genesis of identical electrocardiograms have been predicted with remarkable accuracy for many years from scalar electrocardiograms.18,20,22 However, attempts to indicate a precise electrophysiologic mechanism from the scalar electrocardiogram may lead to oversimplification and misinterpretation of the true underlying cardiac abnormality. The use of clinical electrophysiologic techniques such as His bundle recordings and localized atrial and ventricular electrograms for determination of the precise electrophysiologic mechanism may be warranted in cases in which danger to life, mode of treatment or prognosis are affected by the electrocardiographic interpretation.

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