The study by Miyoshi et al. published in this issue of the Journal adds a new piece to the puzzle of fetal bradycardia associated with congenital heart defects (CHD). They reviewed the outcome of 29 fetuses with CHD associated with fetal bradycardia over a 6-year period and investigated potential outcome predictors in those populations. Prognosis is still poor and that there are different clinical manifestations, such as outcome and escape rhythm rate, among the CHDs: congenitally corrected transposition of the great arteries (CCTGA) and heterotaxy (polysplenia) in fetal congenital heart block (CHB). Are the differences in outcomes attributable to different histopathologies of fetal bradycardia between the populations?

From an electrophysiological viewpoint, the site of the block theoretically may be different in these CHDs. Generally, ECG data on the QRS width and the configuration of conducted beats and/or escape beats are of value in localizing the site of the block. Although a narrow QRS complex is most compatible with an atrioventricular (AV) nodal or intra-His problem, a wide QRS complex is most compatible with an infra-His problem. In CHB, the rate of the escape rhythm provides some information about the site of the block. Precise identification of the site of the block based on the ECG (escape rhythm rate and QRS width) may not be possible because considerable overlap is observed. However, presumed localization of a conduction disturbance may be possible and aid the clinician in planning the therapeutic approach.

CHB can occur with CHDs or in otherwise normal hearts. When occurring in a structurally normal heart, the pattern of the cardiac conduction system can take 1 of 3 anatomic forms: atrial-axis discontinuity, nodal-ventricular discontinuity, or intraventricular discontinuity (Figure). The association between CHB and maternal connective tissue disease is well documented and a previous study has shown the lack of an AV node or nodal-ventricular discontinuity.

CHB associated with CHDs is most frequently observed in the anomaly of CCTGA, isomerism of the atrial appendages (heterotaxy), and with some AV septal defects. In other words, the derangements in AV conduction are related to malalignment of the ventricular septum.

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CCTGA

The histopathology of CHB is summarized in Table. The anomalous AV node and His bundle are generally anterior, and the long penetrating bundle is somewhat tenuous or vulnerable to fibrosis with advancing age.\(^6,7\)

Histologic investigation has disclosed that fibrosis and disruption in an elongated non-branching His bundle, together with fibrosis, occurs at the junction with the anterior node in situs solitus with L-loop CCTGA (without pulmonary atresia).\(^8-10\)

Electrophysiologic observations are consistent with the histological studies of CCTGA.\(^11\)

Heterotaxy (Polysplenia)

Heterotaxy patients with CHB may have a discontinuity between the AV node and His-Purkinje system.\(^12\) either because of an initial lack of fusion between the AV nodal tissue and the His bundle or a secondary interruption of the AV conduction axis.\(^13\) However, we currently do not have sufficient histological evidence of the infra-Hisian conduction system in heterotaxy syndrome.

CHD patients have a conduction axis (His-Purkinje system) abnormality that includes sling-like tissues. In particular, patients with heterotaxy syndrome may have a tendency to such abnormalities. Notably, an association of sick sinus syndrome (SSS) with CHDs is high in the present study. The incidence of SSS was 14/20 (70%) in polysplenia, 1/2 (50%) in asplenia, 1/4 (25%) in CCTGA, and 0/3 (0%) in critical pulmonary stenosis. Hypoplasia or absence of the sinus node results in sinus bradycardia in polysplenia syndrome. Namely, AV block in polysplenia syndrome is associated with the absence of the sinus node. On the other hand, sinus node(s) are generally present in asplenia. The pathology of AV block in heterotaxy may be different even between polysplenia and asplenia. However, we do not have sufficient evidence to confirm this either histologically or electrophysiologically thus far.

The present information may be helpful in the challenging management of fetal bradycardia in association with CHDs.\(^14\)

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