The Importance of Ventriculoatrial Malalignment in Anomalies of the Atrioventricular Valves, Illustrated by "Mitral Atresia" and Congenital Mitral Stenosis with Large Left Ventricle

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The concept of segmental malalignment is fundamental to the understanding of complex congenital heart disease. For example, it is now widely understood that the infundibular (conal) septum can be importantly malaligned relative to the ventricular septum.

However, ventriculoatrial (VA) malalignment, which also is very important, is much less well understood. It is hardly surprising that the ventricles often are malaligned relative to the atria because, from the developmental standpoint, the ventricular tube is a "professional contortionist." Normally, the human straight heart tube starts to loop and twist to the right during Streeter’s horizon 10, (i.e., 20–22 days of age), and D-loop formation is completed during horizon 11 (i.e., 22–24 days of age). The left ventricle (LV) develops faster than the right ventricle (RV), and the ventricular apex swings from right to left—levocardia normally being achieved by horizon 18 (i.e., 36–38 days of age).

Although the bulboventricular part of the heart is highly mobile, as described above, the sinoatrial segment is comparatively fixed; it is anchored in position by the venae cavae, pulmonary veins, diaphragm, and lungs. Consequently, VA malalignments almost always are due to malpositions of the mobile ventricles—not of the fixed atria. The atrioventricular (AV) valves are "caught" in between the looping, twisting, and turning ventricles on

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the one hand, and the relatively static atria on the other. Hence, it is no surprise that ventricular maldevelopment, leading to ventricular malposition and hence to VA malalignment, can have catastrophic effects upon those “middlemen,” the AV valves, that develop from the AV canal or junction between the ventricles and the atria and from the ventricular myocardium.

*When the RV fails to develop*, the result is single LV with infundibular outlet chamber (IOC) [1, 2]. In this situation, the ventricular septal remnant lies to the right of the tricuspid valve (TV) with a D-loop or to the left of the TV with an L-loop, resulting in double-inlet LV.

*When the LV fails to develop*, the result is single RV [1, 2]. The ventricular septal remnant lies to the left of the mitral valve (MV) with a D-loop or to the right of the MV with an L-loop, resulting in double-inlet RV.

*When the RV is underdeveloped*, the ventricular septum (VS) underlies the TV, resulting in straddling TV, double-outlet right atrium (DORA), or tricuspid atresia (TAt). In DORA, the straddling TV becomes adherent to the VS, with part of the TV opening into the small RV and the rest opening into the large LV. In TAt, the expected site of the tricuspid orifice sits right over the posterior portion of the VS.

*When the VS underlies the MV*, the result can be straddling MV. In this situation, the LV often is somewhat underdeveloped, but the malalignment between the mitral orifice and the VS for whatever reason (not necessarily because the LV is small) is crucial. In typical mitral atresia (MAt), the LV is diminutive.

But what about MAt or severe congenital mitral stenosis (MS) with a large LV? Is it really possible to have MAt with a large LV, as was first reported by Quero [3]?

Briefly, the entity reported by Quero [3] certainly does exist. In 2,504 autopsied cases of congenital heart disease in the cardiac registry of the Boston Children’s Hospital, we have found 12 cases of MAt with large LV (0.5%) and seven cases of congenital MS with large LV (0.3%). Limitation of space makes it impossible to report these 19 cases in detail. Suffice it to say that in MAt with large LV, the VA malalignment was very marked. The angle between the VS and the atrial septum (i.e., the VA septal angle) was measured using needles and a protractor, as viewed from the front of the heart, projected on the frontal plane. The VA septal angle ranged from 20–100°, with the mean being 60° (normal mean, 5°). The VA septal angle was thus much greater than normal. Typically, although the atrial septum was vertical (normal), the VS was semihorizontal (abnormal).

Even more important was the lateral displacement of the ventricular part of the heart: 1) *rightward displacement* in eight of nine cases with D-loop (89%), so that the expected site of the orifice of the MV was above the free wall of the large LV; or 2) *leftward displacement* of the ventricular part of the heart in all three cases with an L-loop, so that the expected site of the orifice of the MV was located above the right-sided LV free wall.

Of these 12 cases of “MAt” with a large LV, five had single LV with