EDITORIAL COMMENTARY

Doubly unusual in double discordance: Appendage-based accessory pathway in congenitally corrected transposition

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In this issue of *Heart Rhythm Case Reports*, Radhakrishnan and colleagues[1] present a case of a very unusual accessory pathway in the setting of congenitally corrected transposition of the great arteries (ccTGA, also termed L-transposition of the great arteries, L-TGA, or double discordance, referring to combined atrioventricular [AV] and ventriculoarterial discordance).[1,2]

ccTGA is a complex congenital heart disease—well known to electrophysiologists owing to high rates of heart block—where the ventricles are inverted, with the left ventricle on the right side of the heart (taking deoxygenated blood from the right atrium through a mitral valve, and ejecting to the pulmonary artery), and the right ventricle on the left side of the heart (taking oxygenated blood from the left atrium through a tricuspid valve, and ejecting to the aorta). In ccTGA, the left-sided tricuspid valve often has features of Ebstein anomaly, and accessory pathways tend to be located on its annulus or in the septal/paraseptal region (Table 1).[3–5]

In the present report, a young adult with ccTGA and Wolff-Parkinson-White syndrome presented for a fifth electrophysiology study owing to intractable supraventricular tachycardia, unusually attributed to a right free wall accessory pathway. The authors ultimately identified and successfully ablated a pathway connecting the right atrial appendage and the free wall of the right-sided left ventricle.

Such appendage-based pathways are rare but do occur in patients with structurally normal hearts, as reviewed in a recent issue of the journal.[6] They are more commonly associated with right than with left atrial appendages, possibly owing to the larger surface area of the right atrial appendage in contact with the ventricular epicardium. These pathways are challenging to identify and ablate owing to their rarity, location within a relatively low-flow structure, and seemingly broad nature.[7] Some may require epicardial or surgical approaches, with extensive dissection sometimes needed to completely divide a band-like connection between the appendage and ventricle.[7,8]

Although right-sided accessory pathways are rare in ccTGA, generally this refers to traditional accessory pathways, located on the fibrous valve annulus and believed to derive embryologically from incomplete development of the valve annulus. However, appendage-based pathways have different origin and anatomy, which may explain why they seem to require more intense ablation than typical pathways. The pathway in the present case finally blocked when the power was increased to 40 watts with an irrigated tip catheter.

This case serves as an excellent reminder to consider alternative substrates when a case is not proceeding as expected. Clues about the presence of an appendage-based pathway include a relatively broad area of "early" electrograms on the valve annulus in the appendage region, less early electrogram timing than would be expected, a far-field appearance to "early" electrograms, progressively earlier activation if mapping away from the annulus, and of course difficulty achieving successful ablation.

As in most congenital heart diseases, individual anatomy must be considered, as the conduction system in ccTGA varies based on atrial sidedness, associated defects, and other factors. The authors note that prior operators had hesitated to attempt ablation owing to uncertainty about the location of the conduction system. Indeed, in patients with ccTGA and usual atrial arrangement (situs solitus), the AV node is usually shifted superiorly to the "anterior" aspect of the right-sided mitral valve, near the region where this patient's accessory pathway was initially mapped. As described by the authors, ablation at the anterior mitral valve annulus or right atrial appendage base may put the AV node at risk in patients with ccTGA. Cryoablation can be a valuable tool to reduce risk of AV block in patients with congenital heart disease, especially when the conduction system may be displaced owing to ccTGA, ventricular septal defect, or especially atrioventricular septal defect. Cryoablation may also be useful if ablating near coronary arteries,[9] as could be the case in an atrial appendage, especially a left atrial appendage.

While not a focus of the case report, the use of Stereotaxis (St. Louis, MO) for this case deserves mention. Robotic magnetic navigation has been found to be as safe and effective as manual ablation of accessory pathways, and has some

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Table 1 Published series of accessory pathways in congenitally corrected transposition of the great arteries

Reference	Patients	Pathways	Left-sided pathways	Right-sided pathways
Takeuchi et al[3]	11	13	13	0
Tseng et al[4]	5	10	9	1, posteroseptal
Iturralde et al[5]	3	4	2	2, posteroseptal and midseptal
Total	19	27	24 (89%)	3 (11%)

potential advantages.[10–13] Catheters used for robotic magnetic navigation are more flexible and atraumatic than manual catheters and may thus reduce procedural risks when ablating in an appendage.

In conclusion, Radhakrishnan and colleagues' very interesting and thought-provoking report nicely emphasizes the importance of maintaining an appropriate index of suspicion for an atypical pathway when a case does not proceed as expected, of considering an individual patient's anatomy and conduction system, and of thoughtfully using our growing number of tools to optimize benefit and risk. This case is one the authors, and perhaps the readers, will not soon forget!

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