What do we really know about the management of patients with congenitally corrected transposition of the great arteries?

James S. Tweddell, MD

From the Division of Cardiothoracic Surgery, Department of Surgery, Cincinnati Children's Hospital and Medical Center, Cincinnati, Ohio; and the Division of Cardiothoracic Surgery, Department of Surgery, University of Cincinnati, Cincinnati, Ohio.

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Address for reprints: James S. Tweddell, MD, Cincinnati Children's Hospital Medical Center, 3333 Burnet Ave, MLC 2013, Cincinnati, OH 45229-3026 (E-mail: James.Tweddell@cchmc.org).

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Decision making concerning the treatment of congenitally corrected transposition of the great arteries (ccTGA) is challenging because of the rarity of the lesion, combined with wide anatomic variability and the fact that many of the outcomes occur in the distant future. Initially, it was thought that the morphologically right ventricle (RV) could sustain systemic work; indeed, the finding of apparently healthy older individuals with ccTGA without associated lesions was reassuring justification that atrial-level correction of dextrotransposed great arteries would yield an acceptable long-term outcome. Ultimately, however, more systematic analysis of the outcomes of individuals with ccTGA, even those without associated lesions, demonstrated high incidences of heart failure and premature death.¹ Results of several large series have demonstrated a 10-year survival between 68% and 88% after physiologic repair (PR) of ccTGA.²⁻⁶ Reoperation-free survival has been between 50% and 80%. After PR, patients remain at risk for RV dysfunction with associated tricuspid regurgitation. By 20 years after PR, most patients will have both tricuspid regurgitation and RV dysfunction. Furthermore, survivors of PR have markedly reduced exercise capacity.⁷

In 1990, Ilbawi and colleagues⁸ first reported anatomic correction of ccTGA with a combination of atrial-level switch operation and the Rastelli procedure to restore the left ventricle (LV) and mitral valve into the systemic circulation. Shortly after the report by Ilbawi and colleagues,⁸ the combination of the atrial-level switch and the arterial switch operations to achieve anatomic repair (AR) of ccTGA was reported by groups in Japan, the United Kingdom, and Australia.⁹⁻¹¹ Several single-center experiences demonstrated 10-year survivals of 78% to 100% for patients undergoing the atrial plus arterial switch and 60% to 96% for the atrial switch plus the Rastelli procedure.¹²⁻¹⁸ Ten-year freedom from reoperation after the double switch is 75% to 86%, and that for the atrial switch

James S. Tweddell, MD

Central Message

Despite innovative surgical procedures, decision making for congenitally corrected transposition of the great arteries remains a challenge.

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plus the Rastelli procedure is 57% to 91%. At least 4% of patients will require a pacemaker within 5 years of AR, and pacemaker implantation is strongly associated with LV dysfunction. In addition, sinus node dysfunction is common after the atrial switch operation, occurring in as many as 40% of patients after 20 years, with tachyarrhythmias occurring in 20% by 10 years.^{19,20} Baffle obstruction also remains a challenge, and the risk of baffle complications is 12% at 25 years.²¹⁻²³ Reoperation and need for a pacemaker affect the quality of life of survivors of AR.²⁴

Retraining of the morphologic LV has been proposed for patients with ccTGA to make them candidates for AR. This strategy has been applied not only to individuals who have failing PR but also prophylactically to individuals with ccTGA without associated lesions. LV retraining has achieved a "proof of concept" status, but as yet we do not know the best strategies or who is a candidate. Despite a variety of strategies, LV retraining remains a risk factor for long-term outcome.²⁵

Certainly, the published single-center series represent some of the best efforts at AR; however, there appear to be some real challenges with this strategy. In this issue of *The Journal of Thoracic and Cardiovascular Surgery*, Alomair and colleagues²⁶ from Toronto summarize a 34-year experience with this challenging patient population. They report on the outcomes of 89 patients undergoing PR and 38 patients undergoing AR. The survival trend for AR

was not encouraging, with a 20-year survival of 53.6% compared with 65.7% in the PR group. Indeed, for the entire cohort, AR was the only predictor of death or transplant, although AR has not been a predictor of mortality since 2000. Furthermore, there appears to be some advantage to AR with regard to sustained systemic ventricular dysfunction and atrioventricular valve regurgitation. The largest experience comparing the outcomes of PR and AR comes from Japan and includes 189 patients.⁴ Similar to the Toronto experience, the series from Tokyo Women's Medical University did not find a survival advantage to the AR strategy except among patients with tricuspid regurgitation.

In this issue of the *Journal*, Hraška and colleagues,²⁷ my friends and colleagues from the Medical College of Wisconsin, have provided a comment on the article by Alomair and colleagues²⁶ from the University of Toronto. Certainly, Hraška and colleagues²⁷ are experts in the management of this challenging group of patients and have advocated for AR. They have offered some well-thought out suggestions that they believe will improve the outcome of AR. Among these are improvements in the atrial switch operation aimed at limiting superior vena cava (SVC) and pulmonary venous baffle obstruction. SVC baffle obstruction remains a challenge because of the unique anatomy of ccTGA. Some have advocated for a bidirectional superior caval connection as part of the correction of ccTGA, but I agree with Hraška and colleagues that this is less than optimal because it limits SVC access for transvenous pacemaker placement and it subjects the patients to a chronically elevated SVC pressure, potentially with venovenous collaterals and potentially with a volume-loading shunt to the RV. Although patch augmentation of the SVC-RV junction might limit the potential for obstruction, other groups have reported very good results without this addition. Hraška and colleagues²⁷ have suggested an improved method for LV retraining. Although this well-reasoned strategy, which combines both pressure and volume loading seems to make sense, there are as yet inadequate data to recommend any specific LV-retraining strategy. Although heart block among patients with normal ventricular looping is rarely associated with LV dysfunction, the association between heart block and LV dysfunction among patients with ccTGA is a troubling reality. Hraška and colleagues²⁷ are correct that every effort should be made to avoid surgical heart block. While the role of biventricular pacing for congenital heart disease in general and for ccTGA in particular, has not been completely defined the addition of biventricular pacing for patients with ccTGA and heart block would seem to add little additional risk and should be considered. The addition of a Damus-Kaye-Stansel repair as part of the atrial switch Rastelli procedure is an innovative strategy to limit residual or recurrent LV outflow tract obstruction and should be

considered as a routine part of the procedure. The combination of the Damus-Kaye-Stansel repair with the Rastelli procedure may limit the need for more radical and higher risk procedures, such as aortic root translocation, that carry a high risk of surgical heart block. Despite all these well-reasoned and thoughtful modifications to the AR, however, it is unclear whether such change can improve results or extend the use of AR among patients with ccTGA.

To summarize, treatment of ccTGA remains a challenge because it is rare, there is wide anatomic variability, and we are most concerned with late outcomes. What we know comes from single-institution retrospective studies that span a long period and in which the surgical strategies were often used in different eras. Interinstitutional outcomes are likely to vary widely because of differences in patient selection and surgical technique. Finally, the LV and perhaps the remaining myocardium of patients with ccTGA may have additional intrinsic abnormalities that render them more prone to development of dysfunction, accelerated by development of conduction abnormalities and complete heart block.²⁸ Can we make evidence-based recommendations on the management of patients with ccTGA on the basis of the reported worldwide experience? I think that we can say that for the patient with ccTGA with a nonrestrictive VSD, good RV function and without tricuspid regurgitation, PR or a Fontan procedure is not a wrong choice. For those with tricuspid valve regurgitation or RV dysfunction with good LV function at systemic pressure, an AR is the best choice. The role of LV retraining remains to be defined.

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