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EDITORIAL COMMENT

Congenitally Corrected Transposition of the Great Arteries

Long-Term Benefits of Tricuspid Valve Replacement*

Rafael Alonso-Gonzalez, MD, MSc, Danielle Massarella, MD

Congenitally corrected transposition of the great arteries (cc-TGA) is characterized in its simplest form as atrioventricular and ventriculoarterial discordance. The anomaly is rare and in simple cases may be asymptomatic. In those presenting with symptoms or associated lesions, the optimal surgical approach has yet to be defined. In the case of physiologic repair or in simple cc-TGA where no surgical intervention is necessary, the tricuspid valve remains in the systemic position, as does the right ventricle (RV). Over time, patients will develop heart failure symptoms due to severe tricuspid valve regurgitation and/or systemic right ventricular dysfunction; this is the most common presentation in adults diagnosed with cc-TGA.[1,2] The mechanism of tricuspid regurgitation in this patient population is commonly associated with an Ebstein-like tricuspid valve, but dilatation of the systemic RV also plays a significant role. At present, recommendations for surgical intervention on the tricuspid valve are limited to symptomatic patients or asymptomatic patients with severe tricuspid regurgitation and progressive right ventricular dilation. In both indications, patients must have preserved or mildly impaired systemic RV systolic function (ejection fraction $> 40\%$).[3,4]

In this issue of *JACC: Advances,* Egbe et al[5] present a retrospective study of patients with cc-TGA having undergone tricuspid valve replacement (sTVR). The effect of surgical intervention was defined by presurgical vs postsurgical functional and echocardiographic parameters. Forty-eight patients having undergone sTVR were propensity-matched to controls with cc-TGA and moderate tricuspid regurgitation and similar age, sex, body mass index, and presence of cardiac implantable electronic devices. Not surprisingly, those patients who did not undergo intervention experienced declining of peak oxygen consumption (VO_2), worsening of the systemic RV systolic function, and increase in neurohormonal activation. Following sTVR, however, patients were noted to exhibit improvement in peak VO_2 and echocardiographic indices of right ventricular function such as global longitudinal strain and dP/dtic.

Symptom status is an important factor in surgical decision-making in the cc-TGA population. However, the congenital population is unique in that individuals born with heart disease are known to underestimate their functional limitations and may report no symptoms despite significant objective functional limitation. The current study reinforces this fact, noting a poor correlation between New York Heart Association (NYHA) functional class and peak VO_2 in both the sTVR and control groups. Furthermore, despite clear decline of peak VO_2 in the control group, the NYHA functional class did not change overtime as one might have expected. These findings suggest that subjective symptom status is not a reliable marker of significant valvopathy and stress the value of serial cardiopulmonary exercise testing in this population.

In patients with cc-TGA given the intrinsic limitations of the RV in supporting the systemic circulation, some degree of right ventricular systolic dysfunction is expected and tolerated. Although cardiac magnetic

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From the Toronto ACHD Program, Peter Munk Cardiac Centre, University Health Network, Toronto, Ontario, Canada.

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resonance is the technique of choice to assess right ventricular volumes and right ventricular systolic function in patients with systemic RV, echocardiography is the most common technique used in this setting. With echocardiography, right ventricular systolic function can be evaluated using multiple parameters, including tricuspid annular plane systolic motion, Doppler tissue imaging-derived tricuspid annular systolic velocity wave (S'), RV index of myocardial performance, fractional area change, three-dimensional echocardiography ejection fraction, longitudinal strain, and strain rate.[6] Egbe et al[5] showed that global longitudinal strain improves after sTVR in patients with cc-TGA and this correlates with improvement in subjective, NYHA functional class, and objective, peak VO_2, exercise capacity. On the contrary, a classic parameter like fractional area change declined after sTVR in this population. In addition, a longitudinal strain $<-16\%$ was associated with improvement in objective exercise capacity after sTVR. This raises the question whether global longitudinal strain should be included as part of the standard echocardiographic assessment in this population. As the tricuspid valve supports the systemic circulation in cc-TGA, perhaps it would be prudent to maintain a lower threshold for valve replacement in order to preserve ventricular function and aerobic capacity in the intermediate and long term. Using global longitudinal strain to assess the systolic function of the systemic RV might help identify asymptomatic patients with significant tricuspid regurgitation who might benefit from elective tricuspid valve replacement.

While this study importantly suggests that sTVR may benefit even asymptomatic cc-TGA patients, there were several limitations. Ascertainment and selection bias are related to the retrospective, single-center study design. Though improvement in right ventricular function is suggested by echocardiographic indices, the results would have been strengthened had gold standard methods (cardiac magnetic resonance) of ventricular functional assessment in widespread clinical use been used in addition. A recent study suggested that right ventricular volumetric thresholds predict outcomes of morbidity and mortality in those with systemic RV physiology,[7] and the combination of volumetric and functional assessments to study the benefits of sTVR as a future direction would be highly informative.

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ADDRESS FOR CORRESPONDENCE: Dr Rafael Alonso-Gonzalez, The Bitove Family Professor of Adult Congenital Heart Disease, Toronto ACHD Program, Division of Cardiology, Peter Munk Cardiac Centre-Toronto General Hospital, 5N-525, 585 University Avenue, Toronto, Ontario M5G 2N2, Canada. E-mail: Rafa.Alonso@uhn.ca.

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