See Article page 1080.

Commentary: Management of congenitally corrected transposition: Different strokes for different folks

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CENTRAL MESSAGE

Although anatomic repair is a favored approach for the management of congenitally corrected transposition, conclusively demonstrating the superiority of this pathway remains difficult.

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Congenitally corrected transposition of the great arteries (CCTGA) is a rare diagnosis consisting of combined atrioventricular and ventriculoarterial discordance with the morphologic right ventricle (mRV) and tricuspid valve within the systemic circulation. Historically, patients were managed with either observation or surgery to treat associated pathology, such as ventricular septal defect or pulmonary stenosis. This physiologic repair left the mRV and tricuspid valve to support the systemic workload. Recognizing the risks of progressive mRV dysfunction and tricuspid regurgitation, operations were pioneered to restore the morphologic left ventricle (mLV) and mitral valve to the systemic circulation.[1] Many centers have subsequently adopted this anatomic repair strategy as the preferred approach.[2-7]

In this issue of the *Journal*, Barrios and colleagues[7] present one of the largest, most diverse series of patients with CCTGA reported to date. The review includes 5 cohorts in distinctively different age groups: young children managed with anatomic repair or Fontan palliation and adults managed with physiologic repair, observation, or primary transplantation.

For adults who presented with preserved function and without associated lesions, simple observation was a successful option; the prototypical noninterventional patient presented at a median age of 35 years and demonstrated a further 20-year survival of 88%. As the authors note, this apparent excellent long-term survival may have benefited from both an unknown denominator and transition into the physiologic repair or transplant cohorts. For adults presenting with residual lesions, operative intervention usually occurred within a few years; this prototypical patient underwent physiologic repair at age 25 years and had a 15-year survival of 71%. Finally, for patients presenting with end-stage heart failure, primary heart transplantation represented the last resort and occurred at a median age of 46 years. Thus, most patients who presented in adulthood were managed primarily with observation or physiologic repair and survived well into middle age.

Among the children, a minority presented early with anatomy not amenable to biventricular repair and were directed toward Fontan palliation with excellent outcomes, similar to other studies.[8] For the large majority of children, anatomic repair was the strategy of choice, with preparatory mLV training as needed. The anatomic repair group showed several positive outcomes. Most anatomic repair patients demonstrated preserved systemic ventricular function, in contrast to the physiologic repair patients. Patients who required mLV training or underwent repair at an older age did not show worse outcomes, unlike in other studies.[4,5] Finally, these patients demonstrated very good survival (80% at 15 years), consistent with other encouraging reports.[2-6]

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For young children with favorable anatomy, the authors, like others, advocate for anatomic repair whenever feasible. However, they are reluctant to offer a universal recommendation for anatomic repair. Instead, they advise this strategy for patients with good biventricular and atrioventricular valve function. Although anatomic repair has become a favored choice, and the supporting rationale is sensible, intuitive, and backed by accumulating experience, there remains some room for equipoise. As this study shows, there was no clear winner among the multiple treatment pathways for this complex and heterogeneous disease.

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See Article page 1080.

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Commentary: Incomplete data and inertia: Neither silences the tolling bell of corrected transposition

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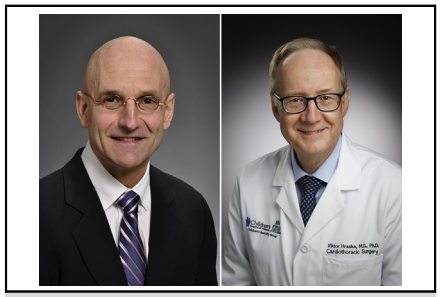

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CENTRAL MESSAGE

The Cleveland Clinic's report on corrected transposition offers very interesting reading but lacks convincing data to provide new insight into optimal management strategies.

Barrios and colleagues have taken on a truly massive effort with the ultimate goal of improving our understanding of the long-term outcomes associated with various treatment strategies for corrected transposition of the great arteries (cTGA). They have capitalized on both statistical expertise and a large cohort spanning the interval from 1995 to 2000. We applaud them for their effort and hard work and also express our respect and gratitude to Dr Roger Mee.

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