Commentary: Time for a multi-institutional study for congenitally corrected transposition of great arteries?

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Chavez and colleagues¹ present their outstanding outcomes of the double switch (DS) operation in 103 patients with congenitally corrected transposition of great arteries (cc-TGA). It provides strong evidence for the benefit of anatomical repair and that excellent midterm survival can be achieved with a consistent and aggressive approach to left ventricle retraining and correction at a young age (where possible).

Their key findings were a median age at DS of 2.1 years. A ventricular septal defect was present in 70% of cases and initial pulmonary artery banding (PAB) was required in 61% (one-third of these were to prevent overcirculation and two-thirds were for left ventricle retraining, n = 40) for a median of 1.1 years. Transplant-free survival was 94.6% at 5 years and 93% were free from significant left ventricle dysfunction. Once again, older age at DS was found to be a risk factor for death/transplant and the outcomes fuel the debate over the optimum age and indication for this formidable procedure. The authors note that patients younger than age 2 years had the best outcomes, yet many older patients also did well without any absolute age ceiling. Other important and positive findings were that the need to retrain the left ventricle was not found to be a categorical risk factor for late dysfunction, as had been suggested by previous studies,¹ including earlier reports on this cohort.³ This may reflect the fact that the criteria for adequate left ventricle preparedness have become stricter (including a morphological left ventricle pressure >90%), yet it is important to note that older patients still took longer to retrain.

On the negative side, the study highlights that reinterventions on the neoaortic valve are an important early and late complication—especially if there is intervention on the left ventricular outflow tract at the time of DS. The burden of pacing interventions is high, and although many of these are an intrinsic component of cc-TGA, the preference for cardiac resynchronization therapy is an important observation, especially in the setting of any left ventricle dysfunction. Overall, the positives still greatly outweigh the negatives, but achieving such outstanding outcomes for DS reflects the strengths of a high-volume center and the fact that surgeon and institutional experience play an important role.

The report gives powerful support for the role of DS, but in an institution where the authors acknowledge their bias toward anatomical repair (an understandable philosophy with such good outcomes). Furthermore, they do not provide any comparative outcome data for either conservative management or conventional repair—which could have an equally important role for certain groups of patients undergoing cc-TGA.

The eternal dilemma with cc-TGA is that we know the natural history is generally very poor, yet the behavior of the systemic right ventricle and tricuspid valve is unpredictable and there are subgroups of uncorrected patients who are able to sustain good systemic right ventricle function. This is further clouded within the group of patients with a
ventricular septal defect: Once these patients have undergone PAB then the destination is usually DS, even if right ventricle function remains good. There are not enough data to know which of these patients are best served by DS and which are best served by conventional repair. Chavez and colleagues\(^1\) correctly refer to many of the publicized disappointing outcomes with conventional repair, but we should also be aware that small series of good outcomes with conventional repair continue to be published in selected groups,\(^4,5\) and that not every center has been able to reproduce such good outcomes with DS. Recent data from the Boston group looked specifically at cc-TGA with intact septum and again favored anatomical repair but still recognized that some self-selected older patients sustained good right ventricle function with no intervention.\(^6\)

The most controversial area is whether prophylactic PAB should be offered to younger patients with intact septum, as has been suggested by Raisky and colleagues\(^7\) and also acknowledged as a management option by the Boston team.\(^6\)

Despite these impressive outcomes for DS, many questions around best management of cc-TGA remain unanswered, especially around the ideal age of intervention, the best way (and age) to retrain the left ventricle, and the management of cc-TGA/intact septum. The questions may be too convoluted to readily design a randomized trial, but we desperately need a multicenter study to gather large numbers of contemporary outcomes across all these controversial areas to define best practice and match the right procedure to the right patient.

References

7. Raisky O. Early pulmonary artery banding in congenitally corrected transposition of the great arteries with intact ventricular septum: a winning strategy? Accessed April 2021. [https://www.aats.org/aatsims/AATSWeb/Association/Meetings/Annual_Meeting/100th_Annual_Meeting_Virtual/Program_Highlights.aspx](https://www.aats.org/aatsims/AATSWeb/Association/Meetings/Annual_Meeting/100th_Annual_Meeting_Virtual/Program_Highlights.aspx)