

The paradox of choice in the surgical management of congenitally corrected transposition: what should we do with all of these options supported by little evidence?

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Despite the conceptually proper physiology consisting of independent in-series pulmonary and systemic circuits, the prognosis in patients with unoperated congenitally corrected transposition of the great arteries (ccTGA) is reserved, even in the absence of associated defects. This is attributable to several factors. Although reports have described unoperated patients with normal or near-normal systemic right ventricular (RV) function in their 60s and beyond (1,2), it is the exception rather than the rule. Faced with a systemic circulation, the morphologic RV fails in one third of patients with no associated lesions by the fifth decade of life, and in two thirds with prior surgery for concomitant defects by 45 years of age (3). When confronted with chronic pressure overload, the systemic RV invariably undergoes hypertrophic remodelling. Owing to its limited coronary reserve from the concordant predominantly single right coronary circulation, myocardial oxygen/supply mismatch can promote subendocardial ischemia, progressive fibrosis, and ultimately, overt heart failure (4-6). Second, the tricuspid valve, again part of the systemic circulation, frequently displays Ebsteinoid features with propensity for regurgitation. While the direction of the causal relationship between tricuspid insufficiency and RV dysfunction remains debated, RV failure often follows systemic atrioventricular (AV) regurgitation (7,8). Incompetence of the tricuspid valve correlates with excess

mortality, with poorer outcomes following AV valve replacement when the pre-operative RV ejection fraction is $\leq 40\%$ (9). In addition, the AV conduction system is displaced and susceptible to AV block (10,11). Long-term studies report a prevalence of complete AV block of 24-39% (3,12), with an annual incidence estimated to be approximately 2% per year (13). Pacemakers are, therefore, frequently indicated. In the absence of cardiac resynchronization therapy, pacing of the subpulmonary left ventricle can alter the ventricular depolarization pattern thereby precipitating systemic AV valve regurgitation and heart failure (14). This confluence of factors contributes to the risk for ventricular arrhythmias and sudden death (15).

In this context, it comes as no surprise that conventional repair techniques, which were commonly performed until the mid-1990s and only addressed associated lesions, led to mitigated results. In case series of older minimally symptomatic patients, so-called physiologic repairs yielded fair results. However, for the majority of children with early symptoms of heart failure at the time of intervention, such approaches did not appear to significantly improve long-term survival (16,17). Moreover, suboptimal midterm outcomes were achieved at the cost of a 2% to 5% perioperative mortality rate. In the hope of improving long-term outcomes in patients with septatable ccTGA hearts, specialized centers evolved towards technically challenging

biventricular repairs. Anatomic approaches address AV and ventriculoarterial discordance by means of a double switch operation, which typically consists of a Senning procedure combined with a Jatene, Bex-Nikaidoh or Rastelli repair. In recent years, promising long-term outcomes have been reported (18-21). However, given the paucity of evidence, ill-defined patient selection criteria, and publication bias favoring positive results, a great deal of controversy still surrounds claims of equivalent perioperative mortality, morbidity and reintervention rates. Children with excellent biventricular function and unobstructed pulmonary blood flow are generally considered well served by anatomic repairs performed in high-volume centers. In contrast, standardized preoperative evaluation and management of high-risk subgroups—such as those with an involuted left ventricle, borderline RV size and function, or grossly abnormal yet competent tricuspid valve—remain challenging, even in the most experienced hands.

There is, therefore, a trend towards simplifying anatomic repairs in an attempt to optimize their risk-benefit ratio, broaden their applicability, and incite lower-volume centers to adopt such approaches over higher-risk double switch surgery or avoidable Fontan palliation. The one-and-a-half ventricular repair technique, which substitutes the Senning component for a hemi-Mustard and bidirectional Glenn (BDG), is a good example of such a trend. It was proposed as early as 2004 as a means of addressing the small effective RV size that sometimes results from Rastelli variants of the double switch operation (22). The combined experience of 76 cases performed by three additional centers has suggested that it may be reasonable to perform a one-and-a-half ventricular repair in the context of relative surgical inexperience with the Senning procedure, borderline RV function and unfavorable tricuspid valve anatomy (23-26).

In a recent issue of the *European Journal of Cardiothoracic Surgery*, Zhang *et al.* contribute to the current evidence base a case-series of 31 consecutive patients who underwent a one-and-a-half ventricular repair for ccTGA at a single institution in Beijing, China, between 2011 and 2015 (27). To our knowledge, this manuscript is the first to report the systematic use of hemi-Mustard and BDG procedures in the setting of pulmonary stenosis coupled with cardiac malposition. The technique, performed in combination with a Rastelli procedure, was applied without individualized assessment of the need for RV decompression. In other words, its indication was predominantly driven by technical challenges anticipated as a result of malposition, as opposed to a diagnosis of preoperative RV dysfunction, tricuspid

insufficiency, or intraoperative recognition of severely reduced RV cavity size post intraventricular rerouting. In addition to introducing a new patient selection criterion that could potentially impact use of the one-and-a-half ventricular repair, Zhang *et al.* commented on their experience with staging of the superior cavopulmonary anastomosis in the context of ccTGA.

In their descriptive study of 31 patients, 11 underwent a staged procedure with a BDG performed prior to the hemi-Mustard and Rastelli (27). Consequently, most children were well over 1 year of age at the time of completion of ccTGA repair. Outcomes were reported at a mean follow-up of 3.3 years. Although there was 1 (3.2%) mid-term fatality, no perioperative death occurred. The incidence of atrial baffle obstruction, complete heart block and reoperation was 3.2%, 9.7% and 12.9%, respectively. Despite reintervention being rather common, the authors specified that all redo surgeries were in the perioperative phase, with no patient requiring a pulmonary conduit change. The absence of sinus node dysfunction is consistent with the relatively short follow-up period. While the authors are to be commended for these results that they considered favorable, the absence of controls precludes comparisons to other surgical techniques. Comparisons between one- and two-stage repair groups not surprisingly failed to yield significant findings given the limited sample size and lack of statistical power. It was noted, however, that a statistically greater incidence of postoperative pleural effusions occurred when the BDG, hemi-Mustard and Rastelli were performed concomitantly ($P=0.033$). This result is difficult to interpret if the incidence of pleural effusions in staged patients post-BDG is not taken into consideration.

In conclusion, Zhang *et al.*'s manuscript (27) describes a contemporary experience with the one-and-a-half ventricle repair for ccTGA in a large and highly specialized center. It raises important questions regarding the ability of lower-volume institutions to undertake complex ccTGA repairs in the context of malposition and the potential for BDG staging to improve outcomes in carefully selected patients. Furthermore, it comments on feasibility of the surgical technique (i.e., BDG, hemi-Mustard, and Rastelli) and reports encouraging results. Like most studies on the topic, the methodology does not allow direct comparisons to standard biventricular repair such that non-inferiority, let alone superiority, cannot be claimed. Lingering uncertainties remain regarding optimal patient selection criteria. While the experience shared on the management of a very complex disease is of great value, the manuscript

also serves as a reminder that congenital cardiac surgery is a field in which significant breakthroughs are becoming increasingly difficult to achieve without collaborative studies. In this perspective, we hope that the article by Zhang *et al.* will spark further interest in the evaluation of surgical options offered to patients with ccTGA and ultimately spur multicenter initiatives designed and powered to meaningfully impact current practice.

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Footnote

Conflicts of Interest: The authors have no conflicts of interest to declare.

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