Anatomic repair for congenitally corrected transposition: the promise and the reality

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Physiological repair of congenitally corrected transposition (ccTGA, atrioventricular and ventriculoarterial discordance, discordant transposition) has been available for well over 50 years. ‘Anatomic’ repairs, which address both discordant connections, were first performed in the late 1980s by Imai, Yaghara, Mee, Ilbawi and others. The Melbourne concept of pulmonary artery banding for left ventricular (LV) retraining in patients with low LV pressure was applied to ccTGA early in the switch conversion experience. These pioneering operations were adopted almost immediately by many teams, based on a conceptual advantage not yet supported by mid- or long-term data. Anatomic corrections have proved to have important surgeon-specific and institutional learning curves, magnified by the rarity of the lesion. For example, the EACTS Congenital Database reports a 30-day mortality of 16.4% for the atrial switch + Rastelli procedure (73 cases), despite the fact that 5 centres (contributing 34 cases) had no mortality. Limitations of the anatomical strategy are now well recognized.

Hsu et al. (National Taiwan University Hospital, Taipei) have reported their experience with a variety of procedures for ccTGA. The patients were a mixed group from the anatomical viewpoint, and the indications for each procedure evolved during the study period [1]. Results with repairs leaving the right ventricle as the sole systemic pump, were disappointing (as in nearly every previous report). The ‘anatomic’ repairs (atrial switch and Rastelli, atrial and arterial switch) had an even worse outcome over the 13-year follow-up interval, whether or not operative mortality was included in the calculation.

The reporting team is competent and experienced, working in one of the top Asian centres. The outcome data reflect the experience in many other units worldwide, despite optimistic reports from a limited number of centres whose results seem difficult to consistently reproduce. Clearly, the general anatomical repair strategy has been suboptimal for some cases of ccTGA, despite the theoretical advantages.

This brings us to the use of the Fontan operation for ccTGA, the central discussion point of the present paper by Hsu et al. Historically, we have a strong evidence base favouring a Fontan approach for many types of complex biventricular hearts in which efficient septation is impeded by remote ventricular septal defect (VSD), abnormalities of ventricular balance and/or valve-tensor complex, coronary anomalies etc. (availability of the Lecompte procedure, REV, Nikaidoh, truncal rotation etc notwithstanding). The authors have used these established criteria to assign some of their patients to a univentricular approach. Short- and long-term outcome (whether viewed in context or independently) have been excellent. Most of their cases had natural restriction of pulmonary blood flow. We have followed a similar strategy for the past 15 years with superimposable results, and have more recently extended the concept to some cases which were also suitable for septation [2]. Event-free survival and possibly quality of life do seem to be superior with the Fontan approach, at least out to the second decade.

As in other anatomical situations, ‘biventricular repair at any cost’ is probably not the answer for ccTGA. Despite the familiar long-term Fontan issues, patients are extremely likely to survive the definitive operation, and those who maintain sinus rhythm will usually have an acceptable haemodynamic status and enjoy their lives, with a minimal risk of reoperation. Can we really say the same for all complex ccTGA patients undergoing biventricular repair? Perhaps not, although the usual caveats relating to a non-randomized series without standardized surgical indications apply. The main issue is that historically, most ccTGA patients treated with a Fontan strategy have been considered unsuitable for a biventricular repair. Paradoxically, then, the best results have been realized in the worst subset of patients.

Some important questions remain regarding ccTGA:

(i) Which anatomical arrangements are sufficiently complex to justify a univentricular approach to ccTGA?
(ii) Is the Fontan approach ever advisable in patients with ccTGA who start life with high pulmonary blood flow?
(iii) When should we consider the Fontan strategy for cases of ccTGA with LV outflow tract obstruction and VSD who have no contraindication to septation?
(iv) Can we expect the long-term results of the Fontan operation in ccTGA to be as good (or as bad) as in other anatomic subgroups?
Will the survival curves for the two approaches eventually converge, and if so when?

As Hsu et al. suggest, the limitations of our understanding must be taken into account when recommending the best operative strategy for ccTGA, as for other inherently complex lesions.

REFERENCES
