Anatomic repair for congenitally corrected transposition of the great arteries: easier is better?

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Sojak et al. [1] reported their experience with bidirectional cavopulmonary anastomosis (BCPA) and the semi-mustard procedure for an anatomic correction of a congenitally corrected transposition of the great arteries (ccTGA), a strategy that somewhat simplifies the classic atrial switch procedure as already reported by several groups [2].

In this recent small series (eight patients), the authors made the choice to use this approach in selected patients with atrio-apical discordance to technically simplify the atrial switch component of the surgery. For the functionally borderline right ventricle, and/or tricuspid valve dysfunction, a one-and-a-half-type repair was chosen to unload the RV. Of the patients who benefited from this modified atrial switch, six had an associated Rastelli procedure and two had an arterial switch procedure. The early mortality was 12.5% (one patient); two patients were treated for supraventricular tachycardia and three patients developed pleural effusion but no superior vena cava syndrome was notably observed. Only one patient needed re-operation for a conduit replacement at 6 years, from a mean follow-up of 4.5 years. All except one patient were in NYHA class I at the last follow-up. Systemic or pulmonary venous obstruction, sinus node dysfunction or atrial baffle leak were not observed.

Successful early outcomes with anatomic repair in ccTGA were first reported by several centres in the mid-1990s. Nevertheless, 20 years later, it remains a challenging surgical procedure and the anatomic variability observed poses technical difficulties. The excellent results published by most of the experienced centres in this field [3, 4] probably do not reflect the real morbidity and mortality observed worldwide for the anatomic repair of ccTGA. Therefore, a strategy that reduces the surgical complexity of the repair, whilst maintaining good early and long-term outcomes, has to be considered as an important alternative, particularly for patients presenting for surgery at lower volume centres.

This one-and-a-half repair approach definitely reduces the technical challenge of the operation, especially when atrio-apical discordance is present (difficult surgical access to the atrial chambers and narrowed free wall of the right atrium) even though flipping the heart over into the left chest by opening the left pleura usually allows one to perform a Senning. Less complex surgery is associated with a shorter cross-clamp time, something that can be helpful in preserving the ventricular function in this subset of patients. This one-and-a-half strategy, perhaps offers the possibility that anatomic repair, even in the most complex forms of ccTGA, could be achieved by less-experienced surgeons. Another theoretical advantage of the procedure is the avoidance of a superior vena cava stenosis, though this complication has become rare, with almost no reported instances of re-operation in the most recent series [3, 4]. For the pulmonary venous pathway, one might expect a decrease in the rate of obstruction with this technique, although it appeared that, for all except one case in this latter series, the atrial suture lines (easier surgical procedure, sinus node dysfunction or atrial baffle leak) were not observed.

Nevertheless, staged approach, with initial BCPA alone, is an option for complex ccTGA with LVOTO that avoids iterative systemic-pulmonary shunts. This strategy allows a complete repair to be delayed at a time when construction of the intraventricular LV to aorta baffle would otherwise be hazardous and shortens the surgery at the time of a subsequent full repair. BCPA might also allow re-operation to be delayed in cases where the RV to pulmonary artery conduit becomes stenotic. However, the benefits (easier surgical procedure, sinus node rhythm, unloading of the RV) are counterbalanced by the numerous adverse effects of this technique.

BCPA precludes any superior venous access for endovascular electrophysiological procedures, including pacing. Complete heart block is a frequent complication after the closure of the ventricular septal defect in ccTGA or during the natural history of the condition, and epicardial leads are therefore mandatory in this setting.

The one-and-a-half repair is a haemodynamically non-physiological state, whereby the antegrade pulsatile pulmonary blood flow can compete with the cavopulmonary Anastomosis. We have learned from our experience in single-ventricle patients that an additional source of pulmonary blood flow in addition to the BCPA can lead to disabling headaches, superior
vena cava syndrome and pleural effusion [5]. We have dealt with this problem by a slight banding of the proximal right pulmonary artery to flatten the systolic wave. Another strategy in this situation would be to keep patent the azygos vein to avoid high venous pressure in the superior vena cava and to increase the antegrade flow by superior-to-inferior caval anastomosis. Patients with high pulmonary pressure are not eligible for BCPA, but there is also concern for patients who have initially benefited from this strategy and who might later develop moderate elevation of pulmonary artery pressure (diastolic and systolic LV dysfunction, mild pulmonary venous obstruction and mild mitral regurgitation). Another question emerges as to the mid-term haemodynamics of patients who will be repaired with a REV-type procedure (instead of a Rastelli) without a pulmonary valve and an associated BCPA. Last but not the least, there is evidence that functional capacity with a one-and-a-half repair is significantly less than a true biventricular repair [6], a fact that has to be considered before offering this alternative strategy.

The hemi-Mustard/bidirectional cavopulmonary anastomosis is a widely applicable approach for the anatomic repair of ccTGA, particularly for atrio-apical discordance and the functionally borderline RV and TV. The main point of this strategy is that it decreases the surgical challenge, a real advantage for the surgeon and the patient. Drawbacks are mainly represented by the limited transvenous access for pacing, long-term moderate elevation of the superior vena cava pressure, need for a competent pulmonary valve and impaired functional capacity.

In conclusion, we think that this strategy should be taken into consideration for selected patients only and not for all cases of atrio-apical discordance or RV and TV dysfunction.

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REFERENCES