Editorial

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Anatomic Repair Including Hemi-Mustard in Congenitally Corrected Transposition of the Great Arteries: What We know and Still Need to Know

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Congenitally corrected transposition of the great arteries (ccTGA) is a complex congenital heart defect characterized by both atrio-ventricular and ventriculo-arterial discordance, which results in a physiologically "normal" circulation. There are various anatomical and clinical combinations in this disease entity. The natural course of ccTGA depends largely on the presence of associated defects such as ventricular septal defects (VSDs), pulmonary stenosis or atresia, more appropriately referred to as left ventricular outflow tract (LVOT) obstruction, and tricuspid valve (TV) anomalies.

Surgical option for the management of ccTGA

The optimal surgical option for ccTGA remains controversial because, fundamentally, the options are dependent on the cardiac anatomy as well as the operating surgeon and the surgical team. The known surgical treatment strategies are as follows; 1) conventional repair, 2) anatomic repair, 3) single ventricle palliation (Fontan operation).

The conventional repair is focused on addressing the associated lesions without correcting the discordant connections, which finally leaves the morphological right ventricle (RV) and TV to serve as the systemic circulation. These patients usually suffer from progressive RV failure and tricuspid regurgitation after repair. Biliciler-Denktas et al.[1] from the Mayo Clinic reported that overall survival after conventional repair was 77% at 5 years and 67% at 10 years, and late survival was adversely affected by prior operations, more severe preoperative functional class, and cardiac rhythm other than sinus. Hraska et al.[2] from the Boston Children's Hospital reported that the 1- and 15-year survivals after conventional repair were 84% and 61%, respectively, and those requiring TV replacement at any time during follow-up had a significantly worse outcome. These two reports drew a similar conclusion that long-term outcome of conventional repair is unsatisfactory and alternative surgical approaches should be considered to achieve better outcomes. Even with no associated lesions, the natural history of ccTGA is poor, with only 50% survival at 40 years. Therefore, ccTGA itself should be considered to be far from being 'corrected', and true correction, in terms of restoring the discordant connections has been achieved by anatomic repair.[3]

The anatomic repair places the morphological left ventricle (LV) and mitral valve in the systemic circulation. Since Ilbawi et al.[4] first introduced the concept of anatomic repair, numerous reports have been published, showing its encouraging early- and intermediate-term results. Anatomic repair represents a group of procedures in which the atrio-ventricular discordance is 'corrected' by an atrial switch (Senning or Mustard), or hemi-Mustard, and ventriculo-arterial discordance is 'corrected' by an arterial switch operation, by the Rastelli procedure, or by aortic root translocation, depending on the underlying anatomy of the LVOT and/or morphology of the VSD.[5]

Lastly, the single ventricle palliation might be the preferable option if there is an unfavorable anatomy for anatomic repair, or if there is dysfunction of the LV or complex systemic and/or pulmonary venous anatomy. Hsu et al.[6] from Taiwan reported 17-year surgical experience of ccTGA, suggesting that anatomic repair is the choice of operation, only for patients with favorable anatomy, but single

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ventricle palliation should be considered in those with complex and unfavorable anatomy (e.g. remote type VSD, small RV, chordae straddling, and unusual coronary artery pattern). This palliation provides favorable early and mid-term surgical outcomes and avoids any complications related to intracardiac repair procedure. However, the Fontan circulation affects organs outside the heart and may lead to liver cirrhosis, protein-losing enteropathy, or plastic bronchitis. Overall treatment options for this circulatory Fontan failure are disappointing; avoidance of problems is most important, because once the Fontan circuit is created, it "runs on auto-pilot," and allows little modulation.[7]

Anatomic repair: true biventricular repair versus one and a half repair using a hemi-Mustard modification

Traditionally anatomic repair involves Senning/Mustard procedure combined with Rastelli operation or arterial switch. Recently, however, a modified technique called hemi-Musatard and bidirectional Glenn procedure, which only involves re-routing the inferior vena cava to TV and connecting the superior vena cava (SVC) directly to the pulmonary artery was introduced as a type of one and a half repair. This hemi-Mustard strategy has been reported to have multiple following theoretical benefits compared to a full atrial switch;[8] 1) it avoids suture lines in the sinus node area. Data from Senning and Mustard series demonstrate a late incidence of sinus node dysfunction in excess of 60%, 2) the technique shortens cross-clamp time, 3) it eliminates the possibility of superior vena cava baffle obstruction or leaks, 4) it prolongs the longevity of the right ventricle to pulmonary artery conduits used in the Rastelli repair, 5) it simplifies the procedure in patients with dextroposition or other positional abnormalities.

Shim et al.[9] reveals favorable early and intermediate-term outcomes of anatomic repair using this modification in selected patients with ccTGA. Of total 16 patients, 11 patients underwent atrial switch combined with a Rastelli-type procedure and 5 with arterial switch and 11 patients had the hemi-Mustard procedure. What the authors tried to emphasize in their study is the modification of the atrial switch technique from a Senning or Mustard procedure to a hemi-Mustard procedure to overcome high early mortality and morbidity. Also, another more important suggestion noted in the discussion states that the application of the hemi-Mustard procedure should be limited to patients who need Rastelli-type operation. The hemi-Mustard modification could be applied to those with more complex ccTGA to decrease the surgical challenge, a real advantage for surgeons and patients. However, it still has several drawbacks such as the limited transvenous access for pacing, long-term moderate elevation of the SVC pressure, need for a competent pulmonary valve, and impaired functional capacity.[10] With their experience accumulated, Senning procedure becomes a preferred method in patients without LVOT obstruction.

Other issues of anatomic repair to be solved in the future

The indications for and the optimal timing of anatomic repair should be reconsidered in wo subgroups of patients with ccTGA. The first group is about patient with ccTGA, VSD, and LVOT obstruction. The indication for anatomic repair for those with a "balanced" systemic and pulmonary blood flow without clinically significant cyanosis is still unclear. Further technical improvement and longer follow-up of this group might change the treatment strategy in the future because the conduit replacement can be performed more safely.[11] Another group is asymptomatic small babies with isolated ccTGA. Current indications for double switch operation include patients who have developed symptoms attributable to tricuspid regurgitation or RV dysfunction or both. However, Mainwaring and Hanley.[8] reported their experience of the preventive approach using a double-switch procedure in 5 patients, and also, Metton et al.[12] reported an article about early prophylactic pulmonary artery banding in asymptomatic young infants with isolated ccTGA. These challenging practices will provide the possibility on whether we can expand the indication for anatomic repair in those patients who suffer from potential late LV failure related to LV retraining.

There is still no consensus on the surgical method for ccTGA associated with VSD and significant LVOT obstruction. When anatomic repair is considered, a Restelli-type procedure is inevitably performed instead of a double switch, but its long-term results have been reported to be less than optimal.[13] When a Rastelli procedure is likely to be complicated by increased morbidity and mortality due to the unfavorable anatomy, alternative procedures including Fontan operation should be considered.[6][13] However, aortic translocation in ccTGA with a complex LVOT obstruction should be considered if the anatomy is inadequate for an intraventricular baffle as part of a Rastelli operation, including the presence of an inlet and/or restrictive VSD, the association with straddling of the atrioventricular valves, and borderline RV volume, which are considered contraindications for a Rastelli operation.[14][15] As many fields of surgical techniques, perioperative managements, and cardiopulmonary bypass evolves, this complicated procedure is expected to be performed more safely and more competently.

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