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CURRENT TOPICS REVIEW ARTICLE

Surgical treatment for adult congenital heart disease: consideration for indications and procedures

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Abstract The number of the adult patients with congenital heart diseases (ACHD) continues to grow owing to improvement of surgical results and medical management. Corrective surgery for complex CHD does not always mean complete cure. It is not rare that the patients will visit the cardiology institutes because of secondary lesions due to residua or sequela in adults. Some patients with CHD remain unrepairable with different degree of heart failure and pulmonary arterial hypertension. Association of arrhythmias is common in ACHD patients and sometimes critical. We experienced 265 surgical procedures for ACHD patients at our center between 1999 and 2015. Of these procedures, palliative surgery was performed in 3%, palliation to corrective surgery in 6%, primary repair in 57%, and redo surgery in 34%. Hospital mortality within 30 days in this period was 1.1%. Surgery for ACHD patients is safe, beneficial and low-risk treatment; however, tailored procedures for the individual patient are essential to obtain the optimal quality. A comprehensive multidisciplinary approach is required to fulfill this goal.

Keywords Adult congenital heart disease · Pulmonary arterial hypertension · Pulmonary valve replacement · Reoperation

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Introduction

The number of the adult patients with congenital heart diseases(ACHD) has been growing year by year due to improvement of surgical outcome and advances in medical management [1, 2]. In 1997, there were 304,000 children and adolescent patients whereas the number of adult patients was 318,000. Nowadays the adult patient population has increased to over 450,000 and it has surpassed that of young patients [3]. Corrective surgery for complex CHD does not always mean complete cure. Even among the patients who had lived asymptomatic lives after the prior corrective surgery, it is not rare that secondary lesions may emerge due to residua or sequela with time. Stenosis of the repaired left ventricular or right ventricular outflow tract, malfunction of the extracardiac conduit, development of pulmonary valve or atrioventricular valve regurgitation are frequently observed in the long-term period after the corrective surgeries. Because of the hereditary consequence of the CHD, surgical scars or chronic cardiac failure, atrial and ventricular tachyarrhythmia may be progressed. Since such arrhythmias often become responsible for cardiac dysfunction or sudden death [4], concomitant surgery for the secondary lesions and arrhythmia should be considered in ACHD patients. Sometimes the patients leave their cardiac disease untreated; however, they had pointed it out during childhood, or they remain palliative stage because of an inappropriate diagnosis or interruption of their treatment. Since there are few distinct evidences to determine the surgical indication for these ACHD patients with various morbidity at present, tailored treatment will be essential depends on individual conditions of the patients.

Although majority of the patients who underwent CHD care in the pediatric age are followed up in the children's centers, a few of them will visit to the adult cardiology

institutes. The adult cardiologists tend to be unprepared to treat CHD patients, it is the urgent task to build up the consulting system in this growing population of ACHD [5].

Primary repair for adults (Table 1)

Atrial septal defect (ASD) is the most common CHD remained unrepaired among the adult patients. Ventricular septal defect (VSD), patent ductus arteriosus (PDA), atrioventricular defect (AVSD), or Ebstein disease are also frequently presented unrepaired till adulthood. However congenitally corrected transposition of great arteries (ccTGA) is uncommon disease, 0.5% of all the CHD [6], adult cardiologists tend to encounter and are sometimes confused to diagnose correctly. Even nowadays, quite a few patients with cyanotic disease such as tetralogy of Fallot (TOF), or morphologically univentricular heart with well-balanced pulmonary obstruction are introduced for the first intervention.

ASD, VSD, pulmonary arterial hypertension (PAH)

In unrepaired ASD, left ventricular end diastolic volume (LVEDV) tends to dwindle with time because of an interaction between volume-overloaded RV and LV, and corresponding increase of left-to-right shunt. Since association of atrial tachyarrhythmia become frequent with years [7], ASD closure before 40 years of age is recommended [8]. Catheter closure using a septal occluder would be the first choice for the adult patients; however, surgical closure is considered to be more favorable if the patients have been associated with moderate to severe tricuspid regurgitation, or atrial tachyarrhythmia. These complications can be repaired concomitantly during ASD closure. It is well-described in the many reports that pulmonary vein isolation or maze procedure is effective to atrial fibrillation for adult ASD [8–10].

It is uncommon that VSD with significant left-to-right shunt might be carried over into adulthood; however, some patients live, on the other hand diseased lives with different degree of congestive heart failure and pulmonary arterial hypertension (PAH). Recently medicines for PAH treatment have been dramatically developed. Several endothelin receptor antagonists, PDE-5 inhibitors and prostacyclin analogues are now available for clinical use. Both specific drug therapy and combination therapy with these medicines have been successfully applied in the treatment of severe PAH [11–14]. Therefore, advanced and dynamic criteria have been indicated for ASD and VSD with PAH in the recent guidelines concerning the management of ACHD, that is, closure of the defect is suggested when pulmonary arterial pressure and pulmonary vascular resistance significantly decrease to targeted figure with inhalation of oxygen, inhalation of nitrogen monoxide, or treatment with PAH medicines [15]. Based on these guidelines, the concept of "treat and repair" has been practiced for the patients with severe PAH when they respond to the treatment for several months with PAH medicines [16, 17] (Fig. 1).

Table 1 Distribution of surgical cases in the adult CHD (1999–2015 at CCVC)

Primary correction in adult age (including patients with palliation) *N* = *166*	
Atrial septal defect (with or without PAPR)	44.0%
Ventricular septal defect	18.1
Ebstein's disease	4.2
Tetralogy of Fallot	2.4
Congenitally corrected TGA	1.8
Univentricular morphology with PS	3.0
Reoperation in the long-term after correction *N* = 90	
PVR in repaired TOF	24.4%
Conduit exchange in Rastelli type operation	12.2
AVV replacement for repaired AVSD	8.9
Conversion to extracardiac TCPC for classical Fontan	14.4

PAPVR partial anomalous pulmonary vein return, *TGA* transposition of great arteries, *ccTGA* congenitally corrected TGA, *PS* pulmonary stenosis, *PVR* pulmonary valve replacement, *AVV* atrioventricular valve, *AVSD* atrioventricular septal defect, *PVR* pulmonary valve replacement

Ebstein's disease in adults

Ebstein's disease has a wide spectrum of severity depend on tricuspid valve dysfunction, degree of atrialization of the RV, association of other cardiac defects, arrhythmia and so on. Majority of the patients with a mild form can live asymptomatic lives till adulthood. However, not a few adult patients will present with cardiomegaly, exercise intolerance or palpitation due to progression of TR over time. There will appear LV dysfunction which is caused by compression from the dilated RV [18]. Hemodynamic condition may abruptly worsen; occasionally corrupt seriously when tachyarrhythmia, especially rapid atrial fibrillation associates. Most of the adult patients are expected to have adequate RV contractility and a "sail-like" large anterior leaflet which are categorized in type A or B of Carpentier's classification [19], so that they can accept tricuspid valve plasty (TVP) applying monocuspidization [20] such as Carpentier's method [19] or Hetzer's method [21]. In 2007 da Silva et al. introduced a modification of the Carpentier's method [22]. Detachment of the anterior and posterior leaflet and longitudinal RV plication are performed in the same manner with Carpentier's method. The displaced septal leaflet is also partially mobilized and sutured to the free edge of the posterior leaflet forming a "cone". The newly created cone-shaped valve is attached to the diminished true annulus (Fig. 2). If there are severe perforations or deformities in the anterior leaflet,

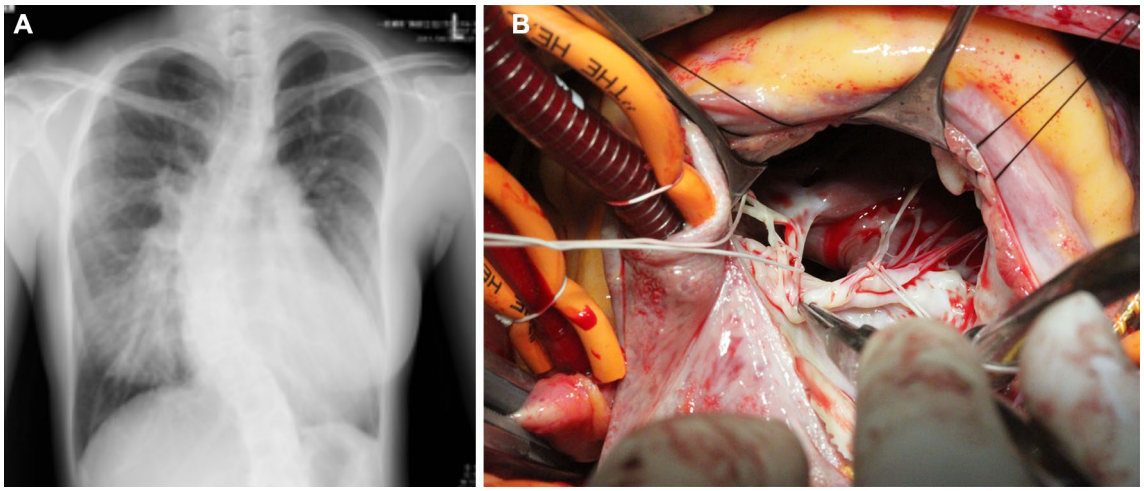

Fig. 1 **a** Chest X-ray of a 35-year-old women. CTR was 65%. VSD with severe PAH. Preoperative PAp was 112/23 (mean 59) mmHg and PVR was 11 WU. After O_2 inhalation, PVR decreased to 6.7 WU. **b** Intraoperative photo of the patient. Large perimembranous VSD, of which diameter was 15 mm, was closed with a patch. Oral ET receptor antagonist and PDE5-inhibitor were given after the surgery. Peak PAp decreased to 40 mmHg. *PAH* pulmonary arterial hypertension, *PAp* pulmonary arterial pressure, *PVR* pulmonary vascular resistance, *WU* wood unit, *ET* endothelin, *PDE* phosphodiesterase

tricuspid valve replacement (TVR) may be mandatory. For a better quality of life without anticoagulation and provability of valve-in-valve treatment in the future, TVR with a tissue valve would be more preferable than with a mechanical valve [23, 24]. The authors from Mayo clinic notified that preoperative LV dysfunction is an independent predictor of late mortality, so that prompt restore of TV function should be considered when LV function is decreasing [25].

Congenitally corrected transposition of the great arteries (ccTGA); morphological RV in systemic circulation

CcTGA is an uncommon complicated cardiac anomaly accounting for 1% or less of all CHD. Morphologically, ccTGA comprises atrioventricular and ventriculoarterial "double discordance" maintains the appropriate blood flow. There are two major groups in ccTGA according to surgical point of view, one is with only minor or no significant lesions, the other with VSD plus pulmonary stenosis (PS) or atresia. Regardless of associated lesions, the morphological RV and the tricuspid valve support systemic circulation. Patients only with minor lesions are usually asymptomatic and survive well into adulthood. However, more than 80% of the adult patient tend to have TR and over 30% of them experiences RV dysfunction or congestive heart failure in their middle age and the rate increases with age [26].

Surgical outcome of ccTGA has been reported not satisfactory in the previous studies. Hraska et al. [27] examined 123 cases of surgical treatment and divided them in three groups, that is, Fontan pathway group, VSD group (VSD closure or VSD closure with LVOT repair) and TV group (TVR or TVP). Survival rate of all the groups of 5, 10 and 15 years were unsatisfactory as 75, 68 and 61%, respectively. Among them, the patients who underwent TV surgery had a significantly worse result. Mongeon et al. [28] reviewed 46 case of TVR performed at a single center. They found that 63% of the patients who had preoperative RV ejection fraction (RVEF) $\geq$ 40% maintained over postoperative RVEF $\geq$ 40%, whereas, only 10.5% of the patient with RVEF $<$ 40% kept preoperative value. The authors concluded that earlier stage operation should be considered before RVEF deteriorates below 40%. It is essential to follow up asymptomatic ccTGA continuously and to refer surgical consultation at earlier timing after diagnosis of new onset TR is made. Scherptong et al. [29] examined 16 patients who underwent TV surgery in systemic RV (8 patients with TVP, 8 with TVR). 36% of the patients with TVP needed reoperation due to recurrence of TR, and survival rate at 2 years after TVP was significantly worse than the patients with TVR. Since residual TR would deteriorate RV function, TVR would be the first choice for the adult patients with ccTGA and TR [26, 28, 29].

Double switch operation [30] is an alternative approach to avoid late systemic RV dysfunction. In this procedure for ccTGA with minor legions, discordant connections will be anatomically corrected by atrial switch and arterial switch. To obtain optimum outcome, training of the morphological LV by pulmonary arterial banding is required to tolerate systemic pressure load. However, LV training should start before 15 years old because myocardial fibrosis would progress rather than increase of LV mass after the age of

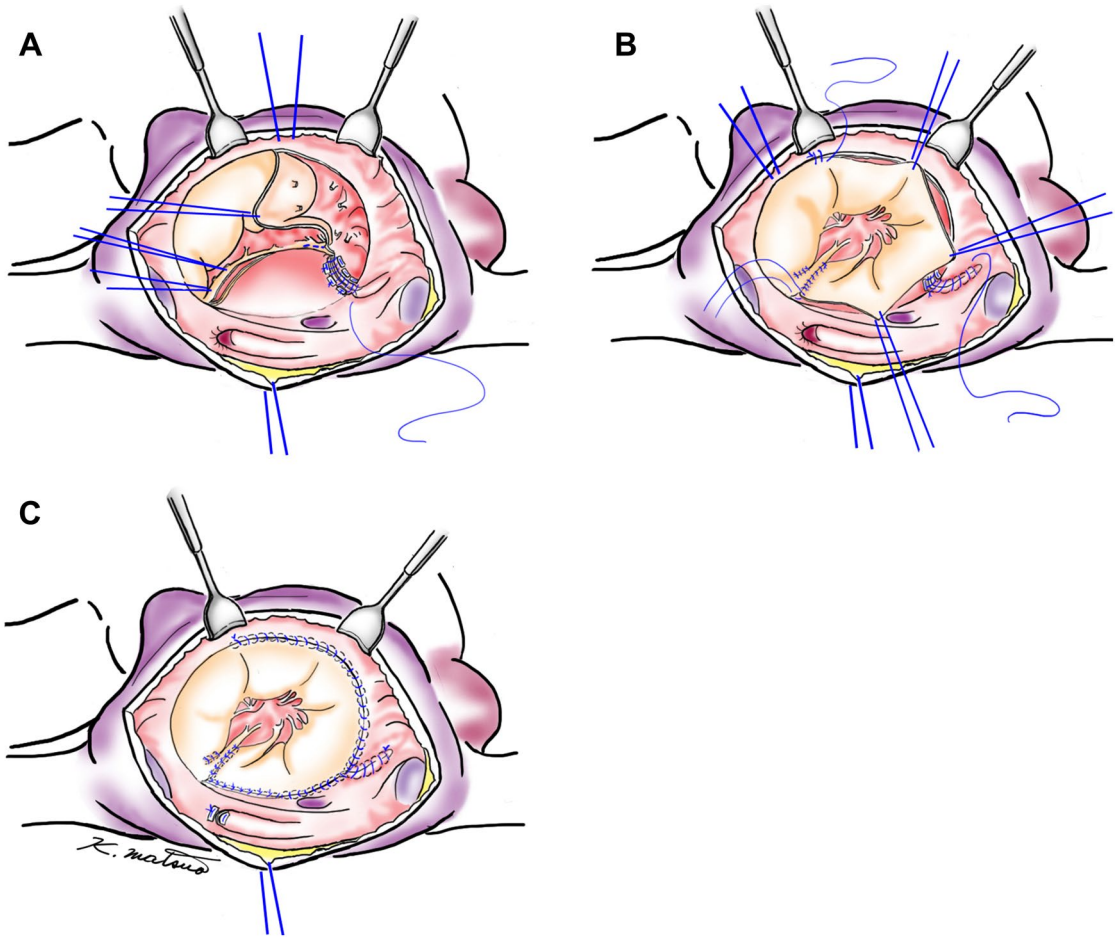

Fig. 2 Operative illustration of Cone operation. **a** 1/3 ~ 1/2 of the anterior leaflet is detached. The posterior and septal leaflet is detached till the apex. Plication of atrialized RV and the true annulus. **b** The posterior leaflet edge is sutured to the septal edge forming a "cone". **c** Clockwise rotation of the "cone" and suture to the true annulus

adolescence [31]. In VSD/PS group, the anatomical repair is completed with atrial switch, interventricular re-routing and conduit repair between the morphological RV and the PA.

Unrepaired and palliative stage cyanotic diseases in ACHD

Except for Eisenmenger syndrome, majority of adult patients with cyanotic CHD have well-regulated pulmonary blood flow owing to a large VSD, and proper size of PS or a systemic-pulmonary shunt. Tetralogy of Fallot (TOF), TGA with VSD and PS, or univentricular morphology with PS may present unrepaired in adults. Some patients are introduced at the palliative stage such as systemic aorto-pulmonary shunts, pulmonary arterial banding, or bidirectional cavopulmonary shunt (BCPS) [32–34].

Because of chronic exposure to hypoxemia, those patients are prone to have hyperviscosity symptoms like headache, dizziness or muscle pain due to erythrocytosis, bleeding tendency and thrombosis caused due to coagulation abnormalities [35]. They also may have innumerable collateral vessels from the descending aorta or bronchial artery to the pulmonary artery. Since these collateral vessels might make surgical procedures hazardous, they should be examined in detail and be closed as possible before surgical intervention.

Despite these secondary impairments adding primary CHD, there remains possibility of surgical treatment for these patients. It should be advocated to re-examine the patients who have previously been diagnosed inoperable by multiple modalities and the latest consideration for surgical indications [33, 34]. Even for the patients with cyanotic CHD in the middle age or later, corrective surgery might be safely applicable [36, 37].

Reoperation (Table 1)

Corrective surgery for CHD does not always mean curative. Repaired left or right ventricular outflow tract tends to be stenotic. Abnormal valves may shrink or calcify and, as a result, valve regurgitation or stenosis will develop with age. Pulmonary valve regurgitation (PR) in repaired TOF and malfunction of the conduits are the most frequent reasons that need reoperation in the long follow-up period [38]. AV valve regurgitation after AVSD repair, aortic valve regurgitation after atrial switch procedure, failing classical Fontan pathway and so on also will be presented for redo surgery in the adult age.

Pulmonary valve replacement (PVR) for repaired TOF

Pulmonary valve regurgitation is inevitable when RVOT in TOF has been repaired with a transannular patch. Even in the patients whose pulmonary valve was preserved, regurgitation may appear due to cusp prolapse, cusp shrinkage or commissural widening with time (Fig. 3). Consequent RV dilatation will trigger RV dysfunction, wide QRS and sometimes ventricular tachyarrhythmia (VT). Gatzoulis et al. described VT would increase 25 to 30 years after TOF repair and strong relevant of PR to VT and sudden death [39]. Indications of PVR for repaired TOF are now considered as moderate to severe PR, RVEDVI over 160–180 ml/m^2, QRS duration exceeding 180 ms and so on [40–42]. Our colleague previously reported 15 cases of PVR for the adult patients with repaired TOF [43]. Their preoperative mean RVEDVI and ESVI was 165 and 91.3 ml/m^2, respectively. These figures were significantly decreased to 106 and 63 ml/m^2 after PVR. Mean QRS duration was also shortened from 176 to 165 ms (Fig. 4). Despite our patients presented at relatively late stage, maximum RVEDVI was 278 ml/m^2, impact of PVR was satisfactory regarding the RV function. However, we thought earlier timing of reoperation was recommendable since recovery of LVEF was slightly impaired in our series [43]. PVR is considered more preferable than PV plasty for such patients with dysfunctional RV. Usually a tissue valve is selected for PVR [44, 45] because long durability can be expected at the low-pressured pulmonary position and unnecessity of anticoagulant. Relatively large size tissue valve of 23 or 25 mm are of choice to reduce stress to the cusps. If the annular diameter is inadequate for the prosthesis, RVOT can be augmented with a patch.

Exchange of extracardiac conduit

Regardless valved or non-valved, the conduit used for RV-PA connection become malfunction due to somatic growth, or degeneration of the conduit valve with time. When the patients are associated with symptoms like breathlessness on excretion, fatigue, abdominal distension, and objective symptoms as QRS widening, moderate to severe regurgitation or pressure gradient excessing 50 mmHg across the conduit, conduit exchange would be recommended.

Fontan conversion (conversion to extracardiac total cavopulmonary connection; TCPC)

In classical atrio-pulmonary connecting Fontan (APC-Fontan) and Bjork Fontan, the RA is disposed to be excessively dilated with time because of chronic high CVP. Resultant stagnant blood flow in the RA may cause decreased cardiac output and thrombus formation. Substrate of atrial arrhythmia will be made in the stretched wall. Reentrant tachyarrhythmia around the surgical scar, atrial flutter or atrial fibrillation will appear in significant population of the patients [46]. Mavroudis et al. reported that conversion to

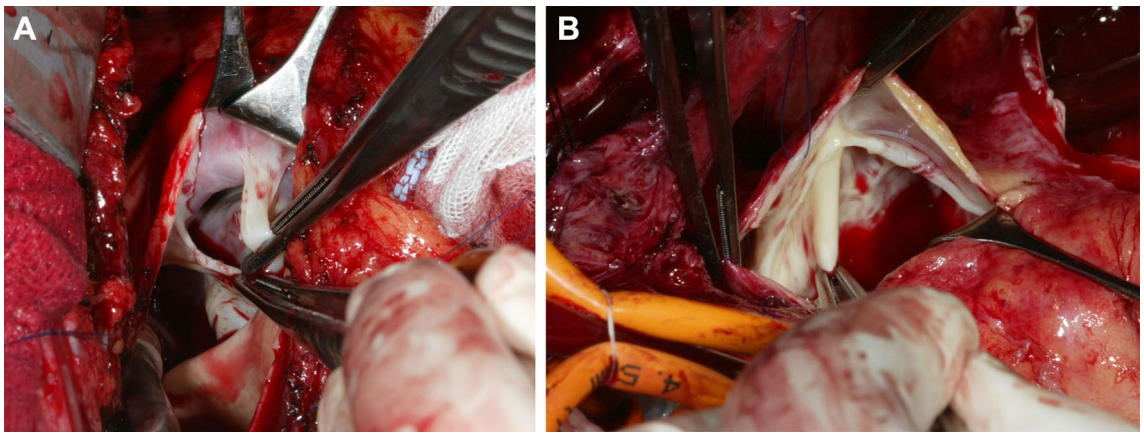

Fig. 3 Intraoperative photo of the pulmonary valve in the repaired TOF patients. They underwent only commissurotomy on the pulmonary valve. **a** A 36-year-old man repaired at 2 years old. The commissure is markedly widened. **b** A 41-year-old man repaired at 2 years old. The cusps shrank and completely lost coaptation

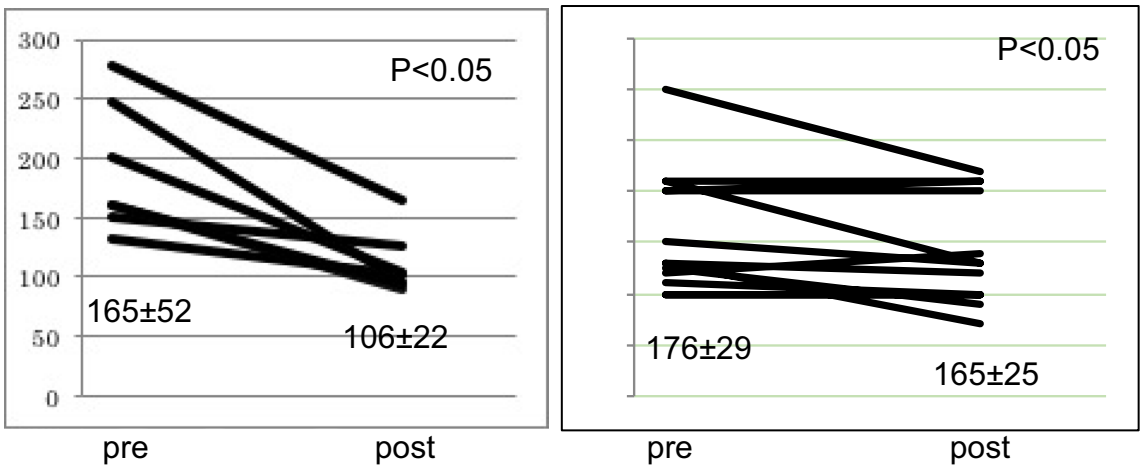

Fig. 4 Changes in RVEDVI and QRS duration pre- and post-PVR for repaired TOF. RVEDVI was decreased from 165 to 106 ml/m^2. Mean QRS duration was also shortened from 176 to 165 ms. *PVR* pulmonary valve replacement

extracardiac TCPC concomitant arrhythmia surgery rescued such "failing" classical Fontan [47]. The anterior wall of dilated RA is widely excised after take-down of atriopulmonary connection. When the patient is associated with atrial fibrillation, Cox III maze procedure is performed in the LA. The authors excised the sinus node together with damaged RA wall because it is nonfunctional in vast majority of the patients with failing Fontan [47, 48]. However, some authors described that they preserved the sinus node [49, 50] and implanted DDD type pacemaker to prevent bradycardia. After suture-closure of the reduced RA, the route between inferior vena cava and the PA is reconstructed with an ePTFE tube of 18–22 mm diameter.

General consideration in the reoperation

Reentry of sternotomy is always challenging in ACHD patients. Dilated RV, anteriorly positioned ascending aorta or calcified RV-PA conduit may lie immediately beneath, or sometimes adhered to the sternum. There always exists concern of arterial or heart injury during re-sternotomy, that consequently causes catastrophic hemorrhage. Therefore, it is indispensable to examine retrosternal space and anatomical characteristics preoperatively by CT or MRI. Previous operation records also provide the important information to realize the anatomical structure. If the risk of heart injury deemed high, femoro-femoral partial bypass should be started to reduce cardiac volume and to perform sternal reentry safely. Any intracardiac shunt (residual ASD, or VSD) must be carefully inspected to avoid hazardous air embolism in case that heart injury might take place [51]. Since it is not seldom that critical tachyarrhythmia occurs during sternal reentry or dissection of the heart, putting defibrillation patches is recommendable.

Summary

Srinathan et al. [52] examined 149 of surgical procedures for ACHD patients from 1998 to 2002 and reported its mortality was 3.4%. Stellin et al. [53] analyzed 1,247 of ACHD patients enrolled in the multicenter study during 5 years from 1997. The patients were divided into 3 groups; 4.4% of palliative procedures, 79.3% of repair (corrective surgery) and 16.3% of reoperation. There was 2.4% of hospital mortality within 30 days. The authors concluded that surgery for ACHD patients is a safe, beneficial and low-risk treatment. We experienced 265 surgical procedures for ACHD patients at our center between 1999 and 2015. Our data show almost the same tendency as the Stellin's report; palliation in 3%, palliation to correction in 6%, repair in 57%, and redo surgery in 34%. Hospital mortality within 30 days in this period was 1.1%. There were 3 post-surgery deaths. A 33-year-old man who had ccTGA, VSD and PS could not walk even a short distance because of deep cyanosis. His morphological RV was as small as 70% of normal, therefore conventional Rastelli procedure was thought unsuitable. Double switch procedure was successfully performed; however, 3 days after the operation, he needed conversion to a hemi-anatomical repair by Glenn anastomosis and pulmonary septation to reduce RV volume load. Lung edema ensued and consequently he died of hypoxemia and multiple organ failure (MOF) on postoperative day 20. The second case was a 56-year-old man with polysplenia syndrome associated with incomplete AVSD, Cor triatriatum and severe PH. He well tolerated a palliative atrial septation using a fenestrated patch. However, he was diagnosed with constrictive pericarditis 2 months after the operation. He died from acute LV failure and lung edema next day after pericardiectomy procedures. The third patient was a 26-year-old man with

DORV, complete AVSD, AV valve regurgitation and left peripheral PS. He had undergone bidirectional Glenn procedure at the age of 13 following bilateral systemic-pulmonary shunt. He had been hospitalized due to severe cyanosis, congestive heart failure and Protein-losing enteropathy (PLE). Biventricular repair was thought indispensable to resolve the present condition. We performed AVSD repair with two-patch method, left pulmonary artery plasty, RV-PA conduit reconstruction and septation of the right pulmonary artery. MVR was added for residual mitral regurgitation on post-operative day 7. The patient eventually succumbed to heart failure and MOF 21 days after the first operation.

Standardized and evidence-driven treatment and care should be perused; however, tailored treatment is essential to obtain the optimal quality for the individual patient. A comprehensive multidisciplinary approach by congenitally trained surgeons, pediatric and adult cardiologist, anesthesiologist and other medical subspecialists are required to achieve this goal [51, 54].

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