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Surgical Intervention for Adult Congenital Heart Disease: Emerging Population

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Abstract

Improvement of surgical results and medical management for Congenital Heart Diseases (CHD) led emerging population of the adult patients. However, corrective surgery for complex CHD does not always mean complete cure. Increasing number of adult patients with CHD (ACHD) will visit the cardiology facilities because of secondary or residual lesions. Conduit malfunction, valvular insufficiency, association of arrhythmias and so forth are 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 and beneficial treatment, however tailored procedures for the individual patient are essential to obtain the optimal quality.

Keywords: Adult congenital heart disease; Congenitally corrected transposition of great artery; Pulmonary valve replacement; Reoperation

Introduction

Due to improvement of surgical outcome and medical management, majority of the patients with congenital heart disease (CHD) can reach their adulthood [1,2]. In our country, the population of adult patients with CHD has increased to over 450,000 and it has surpassed that of young patients (Figure 1a) and a percentage of complex CHD has also been increasing among ACHD patients (Figure 1b) [2]. This situation is the same current in the most of advanced countries [3,4]. However, corrective surgery for complex CHD does not always mean curative. Quite a few patients leave their cardiac defects untreated or remain palliative stage. These patients are prone to visit the tertiary referral facilities associated with severe symptoms. Since there are few distinct evidences to determine the surgical intervention for the ACHD patients with various morbidity at present, tailored treatment will be essential depends on individual conditions of the patients.

Primary Repair

Atrial Septal Defect (ASD) is the most common CHD remained unrepaired among the adult patients. Ventricular Septal Defect (VSD), Patent Ductus Arteriosus (PDA), or Ebstein diseases are also frequently presented unrepaired till adulthood. However congenitally corrected transposition of great arteries (ccTGA) is uncommon disease, 0.5% of all the CHD [5], adult cardiologists tend to encounter and are sometimes confused to diagnose correctly. Even nowadays, it is not seldom that the patients with cyanotic disease such as tetralogy of Fallot (TOF) (Figure 2), or morphologically univentricular heart with well-balanced pulmonary obstruction are introduced for the first intervention.

Recently medicines for pulmonary arterial hypertension (PAH) treatment have been dramatically developed. Several endothelin receptor antagonists, Phospodiesterase-5 inhibitors and prostacyclin analogues are now available for clinical use. 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 in halation of oxygen, inhalation of nitrogen monoxide, or treatment with PAH medicines [6]. Based on these guidelines, the concept of

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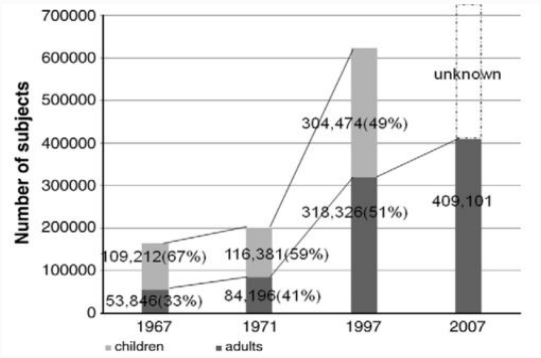

Figure 1a: Change of total number of CHD. The ACHD patient population has increased to over 450,000 and it has surpassed that of young patients in 2007 [2].

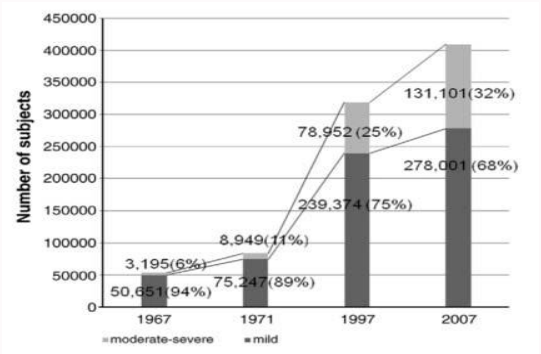

Figure 1b: Severity of CHD in adult patients. A percentage of complex CHD has been increasing since 1997 [2].

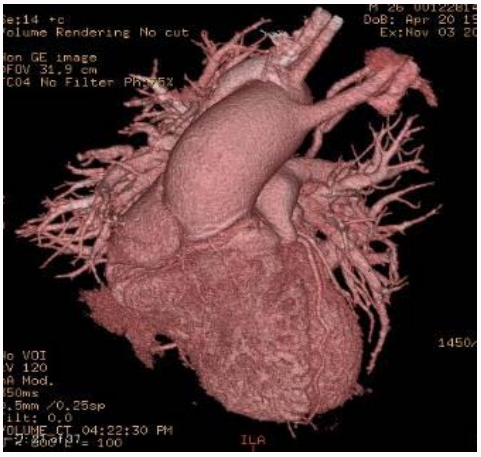

Figure 2: Enhanced CT image of 26 year-old man with TOF, pulmonary atresia and major aorto-pulmonary collateral arteries (MAPCAs). Pulmonary blood flow was supplied mainly through three MAPCAs (blue, green and red arrows). Since the ascending aorta was enlarged to 57 mm in diameter associated with grade 2 of aortic valve regurgitation, unifocalization of the pulmonary artery, Rastelli procedure and ascending aortic replacement were performed concomitantly.

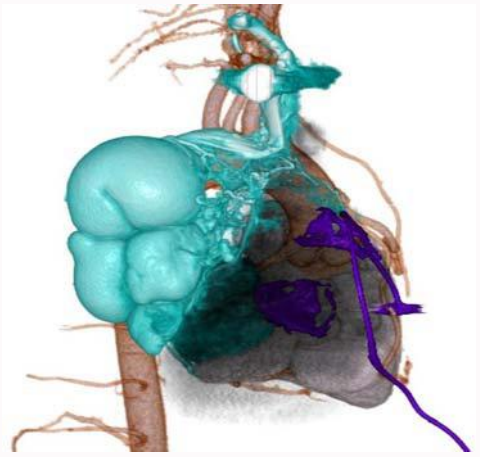

Figure 3: CT image of 35 year-old women who underwent atrio-pulmonary connecting Fontan operation at 5 years old. She suffered from easy fatigue and hypoxemia around 80% of SpO_2. She developed extraordinarily dilated veno-venous collateral from SVC to right lower pulmonary vein. Conversion to extracardiac TCPC and ligation of collateral vessels were performed.

"treat and repair" has been practiced for the patients with CHD and severe PAH [7,8].

Double discordance between atrio-ventricular and ventriculo-arterial connection is conspicuous characteristic of ccTGA. The morphological RV and the tricuspid valve accept systemic circulation. Therefore 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 [9]. Surgical outcome of ccTGA has been reported not satisfactory in the previous studies [10]. Mongeon et al. [11] reviewed 46 case of tricuspid valve replacement (TVR) performed at a single center. They found that 63% of the patients who had pre-operative RV ejection fraction (RVEF) ≥40% maintained over post-operative RVEF ≥40%, whereas only 10.5% of the patient with RVEF <40% kept pre-operative value. Asymptomatic ccTGA should be continuously followed-up and be referred to surgical consultation at earlier timing after diagnosis of new onset TR is made [12]. TVR would be the first choice for the adult patients with ccTGA and TR [9,11,13].

Reoperation

Corrective surgery for CHD does not always mean curative. Pulmonary valve Regurgitation (PR) in repaired TOF and malfunction of the conduits are the most frequent reasons that need re-operation in the long follow-up period [14]. PR is inevitable when RVOT in TOF has been repaired with a transannular patch. Consequent RV dilatation will trigger RV dysfunction, wide QRS and sometimes ventricular tachyarrhythmia (VT). Gatzoulis et al. [15] described VT would increase 25 to 30 years after TOF repair and strong relevant of PR to VT and sudden death. Indications of Pulmonary Valve Replacement (PVR) for repaired TOF are now considered as moderate to severe PR, RVEDVI over 160~180 ml/m2, QRS duration exceeding 180 ms and so on [16-19]. Usually at issue valve is selected for PVR because long durability can be expected at the low-pressured pulmonary position and unnecessity of anticoagulant [20,21]. 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.

In classical atrio-pulmonary connecting Fontan (APC-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. Reentrant

tachyarrhythmia around the surgical scar, atrial flutter or atrial fibrillation will appear in significant population of the patients [22]. Some patients develop veno-venous collateral vessels owing to high CVP and these vessels cause significant desaturation when they return to the pulmonary vein (Figure 3). Mavroudis et al. [23] advocated that conversion to extra cardiac total cavo-pulmonary connection (TCPC) concomitant arrhythmia surgery rescued such “failing” classical Fontan. The authors excised the sinus node together with damaged RA wall because it is nonfunctional in vast majority of the patients with failing Fontan [23,24]. However, some authors preserved the sinus node [25,26] and implanted DDD type pacemaker to prevent bradycardia. The route between inferior vena cava and the PA is reconstructed with an ePTFE tube of 18 mm to 22 mm diameter.

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. It is indispensable to examine retrosternal space and anatomical characteristics preoperatively by CT or MRI. 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 [27].

Stellin et al. [28] 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. We experienced 265 surgical procedures for ACHD patients at our center between 1999 and 2015. Our data shows 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%. Despite progressive changes in complexity and severity of ACHD, hospital mortality within 30 days in this period was 1.1%. We conclude that surgery for ACHD patients is a safe and beneficial treatment.

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 are required to achieve this goal [27,29].

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