

Fri. Jul 7, 2017

ROOM 3

AP Target Symposium

AP Target Symposium 1 (I-APT1)

Dealing with the borderline Left Ventricle - What are the requirements for biventricular circulation, and how to get there?

Chair: Kim Sung-Hae (Pediatric Cardiology, Shizuoka Children's Hospital, Japan)

Chair: Bing Jia (Department of Pediatric Cardiology, FuDan University, Shanghai, China)

2:35 PM - 4:05 PM ROOM 3 (Exhibition and Event Hall Room 3)

[I-APT1-01] The borderline left ventricle: circulation and morphology of fetus, neonate and infant

○ Kenichi Kurosaki¹, Akira Miyake¹, Masataka Kitano¹, Atsushi Hirota¹, Isao Shiraishi¹, Takaya Hoashi², Hajime Ichikawa² (1.Division of Pediatric Cardiology, National Cerebral and Cardiovascular Center, Osaka, Japan, 2.Division of Pediatric Cardiovascular Surgery, National Cerebral and Cardiovascular Center, Osaka, Japan)

2:35 PM - 4:05 PM

[I-APT1-02] Clinical decision-making process for biventricular circulation

○ Sung-Hae Kim (Department of Cardiology, Shizuoka Children's Hospital, Shizuoka, Japan)

2:35 PM - 4:05 PM

[I-APT1-03] Surgery for the mitral valve

○ Zsolt Prodán (Congenital Cardiac Surgery Budapest, Hungary)

2:35 PM - 4:05 PM

[I-APT1-04] The Borderline Left Ventricle: Surgery for the Aortic Valve and the Ventricular Outlet

○ James D St Louis¹ (Department of Surgery Children's Mercy Hospital, USA)

2:35 PM - 4:05 PM

[I-APT1-05] Role of Norwood procedure or bilateral pulmonary artery banding for biventricular repair in hypoplastic left heart complex

○ Shingo Kasahara, Yosuke Kuroko, Yasuhiro Kotani, Sadahiro Arai (Department of Cardiovascular surgery, Okayama University, Okayama, Japan)

2:35 PM - 4:05 PM

[I-APT1-06] What are the requirements for biventricular circulation, and how to get there ?

○ Shusheng Wen, Jimei Chen, Yong Zhang (Department of Pediatric Cardiac Surgery, Guangdong General Hospital, China)

2:35 PM - 4:05 PM

AP Target Symposium

AP Target Symposium 2 (I-APT2)

Dealing with the borderline Right Ventricle - Fontan vs One-and-a-Half Ventricle Repair vs Biventricular Repair: what are the criteria and how to get there -

Chair: Munetaka Masuda (Department of Cardiovascular Surgery, Yokohama City University Hospital, Japan)

Chair: Hiroyuki Yamagishi (Department of Pediatrics, Keio University School of Medicine, Japan)

4:15 PM - 5:45 PM ROOM 3 (Exhibition and Event Hall Room 3)

[I-APT2-01] Circulation and morphology of the borderline right ventricle

○ Hiroyuki Yamagishi (Department of Pediatrics, Keio University School of Medicine, Japan)

4:15 PM - 5:45 PM

[I-APT2-02] Fontan vs One-and-a-Half Ventricle Repair vs Biventricular Repair: what are the criteria and how to get there

○ Mark A. Fogel (Children's Hospital of Philadelphia, USA)

4:15 PM - 5:45 PM

[I-APT2-03] Decision-making process in theater: PAIVS

○ Shunji Sano¹, Yasuhiro Kotani², Takahiro Eitoku³, Kenji Baba³, Shingo Kasahara²

(1.Department of Pediatric Cardiothoracic Surgery, University California San Francisco, USA, 2.Department of Cardiovascular Surgery, Okayama University Hospital, Japan,

3.Department of Pediatrics, Okayama University Hospital, Japan)

4:15 PM - 5:45 PM

[I-APT2-04] Decision-making process in theatre:

4:15 PM - 5:45 PM

Decision-making process in theatre: Ebstein's anomaly (One and a half repair in Ebstein's anomaly)

○ Tae Gook Jun (Samsung Medical Center, Sungkyunkwan University School of Medicine,

Korea)

4:15 PM - 5:45 PM

[I-APT2-05] Decision-making process in theatre: AVSD

○Zsolt Prodán (Congenital Cardiac Surgery

Budapest, Hungary)

4:15 PM - 5:45 PM

[I-APT2-06] Late Cardiopulmonary Function after BVR,
1.5 VR, and Fontan Repair in Patients with
Borderline Right Ventricle

○Hideo Ohuchi (Departments of Pediatric
Cardiology and Adult Congenital Heart Disease

National Cerebral and Cardiovascular Center,

Osaka, Japan)

4:15 PM - 5:45 PM

Sat. Jul 8, 2017

ROOM 3

AP Target Symposium

AP Target Symposium 3 (II-APT3)

Dealing with congenitally corrected transposition of the great arteries - Efforts to minimize late development of systemic ventricular dysfunction
Chair:Yih-Sharng Chen(National Taiwan University Hospital, Taiwan)

Chair:Jun Yoshimoto(Pediatric Cardiology, Shizuoka Children's Hospital, Japan)

10:15 AM - 11:45 AM ROOM 3 (Exhibition and Event Hall Room 3)

[II-APT3-01] Morphological aspects

○Hideki Uemura (Congenital Heart Disease Center, Nara Medical University, Japan)

10:15 AM - 11:45 AM

[II-APT3-02] The late outcome of systemic right ventricle in congenitally corrected transposition of great arteries:
Functional repair or anatomical repair

○Hajime Ichikawa¹, Takaya Hoashi¹, Tomohiro Nakata¹, Masatoshi Shimada¹, Akihiko Higashida¹, Hideo Ohuchi², Kenichi Kurosaki², Isao Shiraishi² (1.Department of Pediatric Cardiovascular Surgery, National Cerebral and Cardiovascular Center, Japan, 2.Department of Pediatric Cardiology, National Cerebral and Cardiovascular Center, Japan)

10:15 AM - 11:45 AM

[II-APT3-03] Electrical issue of corrected TGA

○Jun Yoshimoto (Department of Cardiology, Shizuoka Children's Hospital, Shizuoka, Japan)

10:15 AM - 11:45 AM

[II-APT3-04] Congenitally Corrected Transposition:
Efforts to minimize late development of systemic RV Failure

○Sertaç M. Çiçek (Department of CV Surgery, Mayo Clinic, USA)

10:15 AM - 11:45 AM

[II-APT3-05] The Senning operation in anatomical repair of congenitally corrected transposition

○James S. Tweddell (Cardiothoracic Surgery, Cincinnati Children's Hospital Medical Center, USA)

10:15 AM - 11:45 AM

[II-APT3-06] Role of Fontan operation in cc-TGA

○Tae-Jin Yun (Asan Medical Center, University of Ulsan, Korea)

10:15 AM - 11:45 AM

Sun. Jul 9, 2017

ROOM 3

AP Target Symposium

AP Target Symposium 4 (III-APT4)

Optimizing results in staged surgical management of functionally univentricular hearts –

Preparation rather than Selection for Fontan –

Chair: Akio Ikai (The Cardiovascular Center, Mt. Fuji Shizuoka Children's Hospital, Japan)

Chair: Swee Chye Quek (Pediatrics, National University of Singapore, Singapore)

8:30 AM - 10:15 AM ROOM 3 (Exhibition and Event Hall Room 3)

[III-APT4-01] A Quantitative Analysis of the Systemic to Pulmonary Collateral Flow in Fontan Circulation by Cardiac Magnetic Resonance

○Yoshihiko Kodama¹, Yuichi Ishikawa¹, Shiro Ishikawa¹, Ayako Kuraoka¹, Makoto Nakamura¹, Kouichi Sagawa¹, Toshihide Nakano², Hideaki Kado² (1. Department of Pediatric Cardiology, Fukuoka Children's Hospital, Japan, 2. Department of Cardiovascular Surgery, Fukuoka Children's Hospital, Japan)

8:30 AM - 10:15 AM

[III-APT4-02] Arrhythmia management as a preparation for a Fontan

○Heima Sakaguchi (Department of Pediatric Cardiology, National Cerebral and Cardiovascular Center, Osaka, Japan)

8:30 AM - 10:15 AM

[III-APT4-03] Atrioventricular valve anatomy and function in patients with single ventricle

○Masaki Nii (Shizuoka Children's Hospital, Shizuoka, Japan)

8:30 AM - 10:15 AM

[III-APT4-04] Surgical preparation for Fontan. Atrioventricular valve repair

○James S. Tweddell (Cardiothoracic Surgery, Cincinnati Children's Hospital Medical Center, USA)

8:30 AM - 10:15 AM

[III-APT4-05] Extending the indication of Damus anastomosis

○Makoto Ando, Yukihiro Takahashi (Department of Pediatric Cardiovascular

Surgery, Sakakibara Heart Institute, Tokyo, Japan)

8:30 AM - 10:15 AM

[III-APT4-06] Single ventricular repair strategy:

Controversy and some options and details in right heart bypass operation

○Hajime Ichikawa (Department of Pediatric Cardiovascular Surgery, National Cerebral and Cardiovascular Center, Japan)

8:30 AM - 10:15 AM

[III-APT4-07] Use of Mechanical Circulatory Assist

Devices and Cardiac Transplantation in the Failing Functional Single Ventricle Patients

○James, D St. Louis (Department of Surgery Children's Mercy)

8:30 AM - 10:15 AM

[III-APT4-08] TBA

○Sertaç M. Çiçek (Cardiovascular Surgery, Mayo Clinic, USA)

8:30 AM - 10:15 AM

AP Target Symposium

AP Target Symposium 1 (I-APT1)

Dealing with the borderline Left Ventricle - What are the requirements for biventricular circulation, and how to get there?

Chair: Kim Sung-Hae (Pediatric Cardiology, Shizuoka Children's Hospital, Japan)

Chair: Bing Jia (Department of Pediatric Cardiology, FuDan University, Shanghai, China)

Fri. Jul 7, 2017 2:35 PM - 4:05 PM ROOM 3 (Exhibition and Event Hall Room 3)

[I-APT1-01] The borderline left ventricle: circulation and morphology of fetus, neonate and infant

○ Kenichi Kurosaki¹, Akira Miyake¹, Masataka Kitano¹, Atsushi Hirota¹, Isao Shiraishi¹, Takaya Hoashi², Hajime Ichikawa² (1.Division of Pediatric Cardiology, National Cerebral and Cardiovascular Center, Osaka, Japan, 2.Division of Pediatric Cardiovascular Surgery, National Cerebral and Cardiovascular Center, Osaka, Japan)

2:35 PM - 4:05 PM

[I-APT1-02] Clinical decision-making process for biventricular circulation

○ Sung-Hae Kim (Department of Cardiology, Shizuoka Children's Hospital, Shizuoka, Japan)

2:35 PM - 4:05 PM

[I-APT1-03] Surgery for the mitral valve

○ Zsolt Prodán (Congenital Cardiac Surgery Budapest, Hungary)

2:35 PM - 4:05 PM

[I-APT1-04] The Borderline Left Ventricle: Surgery for the Aortic Valve and the Ventricular Outlet

○ James D St Louis¹ (Department of Surgery Children's Mercy Hospital, USA)

2:35 PM - 4:05 PM

[I-APT1-05] Role of Norwood procedure or bilateral pulmonary artery banding for biventricular repair in hypoplastic left heart complex

○ Shingo Kasahara, Yosuke Kuroko, Yasuhiro Kotani, Sadahiro Arai (Department of Cardiovascular surgery, Okayama University, Okayama, Japan)

2:35 PM - 4:05 PM

[I-APT1-06] What are the requirements for biventricular circulation, and how to get there ?

○ Shusheng Wen, Jimei Chen, Yong Zhang (Department of Pediatric Cardiac Surgery, Guangdong General Hospital, China)

2:35 PM - 4:05 PM

2:35 PM - 4:05 PM (Fri. Jul 7, 2017 2:35 PM - 4:05 PM ROOM 3)

[I-APT1-01] The borderline left ventricle: circulation and morphology of fetus, neonate and infant

○Kenichi Kurosaki¹, Akira Miyake¹, Masataka Kitano¹, Atsushi Hirota¹, Isao Shiraishi¹, Takaya Hoashi², Hajime Ichikawa² (1.Division of Pediatric Cardiology, National Cerebral and Cardiovascular Center, Osaka, Japan, 2.Division of Pediatric Cardiovascular Surgery, National Cerebral and Cardiovascular Center, Osaka, Japan)

Keywords: borderline left ventricle, hypoplastic left heart, biventricular repair

The “borderline left ventricle” (BLV) is considered as a left ventricle with an indeterminate position between normal heart and hypoplastic left heart syndrome. The BLV usually occurs in association with aortic valve stenosis, aortic coarctation, hypoplastic aortic arch, mitral valve stenosis and endocardial fibroelastosis. Fetal echocardiographic studies have demonstrated the progressive development of hypoplasia of left heart structures related to left-sided obstructive lesions.

In the presence BLV, we hesitate about decision of the treatment strategy, biventricular repair or univentricular palliation. We cannot ignore the mortality of the early postoperative period and subsequent reoperation after forcible biventricular repair, whereas serious complications such as protein losing enteropathy may occur after univentricular repair.

The predictor to determine the treatment strategy has been studied such as morphologic predictor (size of the mitral valve, end diastolic volume of left ventricle, endocardial fibroelastosis, size of the aortic valve, etc.) and functional predictor (ejection fraction, end diastolic pressure, pulmonary artery pressure, direction of the blood flow in the ascending aorta and the patent ductus arteriosus). However, the essential predictor is still unknown.

In late years, staged left ventricular recruitment to achieve biventricular repair after single ventricle palliation is suggested, while fetal intervention for aortic valve stenosis is proposed for catch-up growth of left heart in utero.

Patient with BLV is still in the in the grey zone.

2:35 PM - 4:05 PM (Fri. Jul 7, 2017 2:35 PM - 4:05 PM ROOM 3)

[I-APT1-02] Clinical decision-making process for biventricular circulation

○Sung-Hae Kim (Department of Cardiology, Shizuoka Children's Hospital, Shizuoka, Japan)

If the patient has a hypoplastic right ventricle and its associated lesions, you can wait and see whether the right heart components grow and gain better function during the initial palliative stage. On the other hand, in the cases with a marginally left ventricle, such challenge in monitoring can lead to persistent low cardiac output and respiratory failure. Even though he or she survived the life-threatening condition, the obstructive components beyond the left atrium possibly promote retrograde pulmonary hypertension and subsequent high pulmonary vascular resistance which prevent transformation to Fontan circulation. In this context, decision making process for the cases with a marginally left ventricle is completely different from that of the opposite side.

Since 1991 until recently, a number of scoring systems for the marginally hypoplastic left ventricle to discriminate the decision have been reported. These are calculated by combination of structural indices

measured on echocardiography such as mitral valve annulus diameter or area, long axis length of the ventricle, aortic root diameter, grade of tricuspid regurgitation, and so on. Some of which involve the degree of endocardial fibroelastosis (EFE) as well. Coexistence of EFE does compromise the diastolic function even though the ventricle has adequate size and shortening. However, it is difficult to qualify the actual impact. All the scoring systems are designed to predict the temporary accomplishment of biventricular repair, but they have limitations in estimating the long-term outcome.

Hypoplastic left ventricles are frequently associated with obstructive lesions in its inflow and outflow tracts. Therefore, to predict the outcome, you need to extrapolate the effect of surgical and/or interventional recruitment including EFE resection. The current management and thought process differ among countries and institutions. In this presentation, realistic data will be reviewed on neonates with a marginally left ventricle over our past 15 years' experience, and we will discuss about this evolving subject.

2:35 PM - 4:05 PM (Fri. Jul 7, 2017 2:35 PM - 4:05 PM ROOM 3)

[I-APT1-03] Surgery for the mitral valve

○Zsolt Prodán (Congenital Cardiac Surgery Budapest, Hungary)

TBA

2:35 PM - 4:05 PM (Fri. Jul 7, 2017 2:35 PM - 4:05 PM ROOM 3)

[I-APT1-04] The Borderline Left Ventricle: Surgery for the Aortic Valve and the Ventricular Outlet

○James D St Louis¹ (Department of Surgery Children's Mercy Hospital, USA)

Keywords: Cardiac Surgery, Left Ventricle, Aortic Valve

Hypoplasia of left heart structures, including the left ventricular mass, the left ventricular outflow tract, aortic valve, and the aorta is associated with both an increase mortality following surgical intervention as well as the need for single ventricle palliation strategies. Studies have indicated that hypoplasia of left heart structures are related to left sided obstructive lesions. In patients with borderline left ventricular mass and obstruction at various levels of the left ventricular outflow tract, correction of these lesions may allow for successful biventricular repair. This presentation will explore both palliative and definitive surgical procedures that address these left sided obstructive lesions in the setting of a small left ventricle.

2:35 PM - 4:05 PM (Fri. Jul 7, 2017 2:35 PM - 4:05 PM ROOM 3)

[I-APT1-05] Role of Norwood procedure or bilateral pulmonary artery banding for biventricular repair in hypoplastic left heart complex

○Shingo Kasahara, Yosuke Kuroko, Yasuhiro Kotani, Sadahiro Arai (Department of Cardiovascular surgery, Okayama University, Okayama, Japan)

Background: Hypoplastic left heart syndrome(HLHS) is spectrum of structural cardiac malformations characterized by variable underdevelopment of the left heart syndrome.

Hypoplastic left heart complex (HLHC) is widely understanding the structural milder cardiac malformations than HLHS. These patients may be candidates for biventricular repair. Objective of this study was to assess the outcome of the biventricular approach in HLHC.

Method: Retrospective study of 9 HLHC patients who underwent biventricular repair. The cardiac dimensions (mitral and aortic valve annulus, left ventricular internal diastolic dimension) were measured before and after biventricular repair.

Results: There was no early and late mortality. Six cases were underwent Norwood procedure as a first palliation and three cases were underwent bilateral pulmonary artery banding as a first palliation.

Conclusions: Biventricular repair is successful in HLHC patients, even with preoperative mitral and LVEDd of 80 % of normal respectively. Inflow augmentation strategy (regulate ASD size, TAPVC repair etc.) makes LV grow. Norwood procedure or bilateral pulmonary artery banding as first palliation for HLHC attributes biventricular repair in selected cases.

2:35 PM - 4:05 PM (Fri. Jul 7, 2017 2:35 PM - 4:05 PM ROOM 3)

[I-APT1-06] What are the requirements for biventricular circulation, and how to get there ?

○Shusheng Wen, Jimei Chen, Yong Zhang (Department of Pediatric Cardiac Surgery, Guangdong General Hospital, China)

“ Hypoplastic left heart syndrome” is an unsatisfactory term describing lethal underdevelopment of the left ventricle (LV). It represents the more severe end of a spectrum of LV hypoplasia, mandating single-ventricle palliation or cardiac transplantation. Less severe “ borderline” ventricular hypoplasia may instead allow various biventricular therapeutic strategies and better long-term outcomes. These “ borderline ventricles” of moderate hypoplasia represent a clinical decision-management problem, because the decision to pursue biventricular repair or univentricular repair must frequently be made in the first few days of life. This decision is difficult to reverse and may prove fatal if incorrect. The LV rehabilitation procedure was associated with low operative mortality, immediate improvement in left atrial and right ventricular pressures, and maintenance of biventricular circulation at mid-term follow-up. Risk factors that have been associated with poor outcome after biventricular repair include the size and multiplicity of the left-sided obstructive lesions and the presence of EFE. Higher grade of EFE has been shown to be a strong predictor of mortality after biventricular repair. The poor prognosis in patients with circumferential EFE may be due to impairment of both systolic and diastolic ventricular performance. Primary LV rehabilitation procedure, when applied to patients with borderline left heart structures and severe EFE, allows maintenance of biventricular circulation with low operative mortality.

AP Target Symposium

AP Target Symposium 2 (I-APT2)

Dealing with the borderline Right Ventricle - Fontan vs One-and-a-Half Ventricle Repair vs Biventricular Repair: what are the criteria and how to get there –

Chair: Munetaka Masuda (Department of Cardiovascular Surgery, Yokohama City University Hospital, Japan)

Chair: Hiroyuki Yamagishi (Department of Pediatrics, Keio University School of Medicine, Japan)

Fri. Jul 7, 2017 4:15 PM - 5:45 PM ROOM 3 (Exhibition and Event Hall Room 3)

[I-APT2-01] Circulation and morphology of the borderline right ventricle

○ Hiroyuki Yamagishi (Department of Pediatrics, Keio University School of Medicine, Japan)

4:15 PM - 5:45 PM

[I-APT2-02] Fontan vs One-and-a-Half Ventricle Repair vs Biventricular Repair: what are the criteria and how to get there

○ Mark A. Fogel (Children's Hospital of Philadelphia, USA)

4:15 PM - 5:45 PM

[I-APT2-03] Decision-making process in theater: PAIVS

○ Shunji Sano¹, Yasuhiro Kotani², Takahiro Eitoku³, Kenji Baba³, Shingo Kasahara²
(1. Department of Pediatric Cardiothoracic Surgery, University California San Francisco, USA, 2. Department of Cardiovascular Surgery, Okayama University Hospital, Japan, 3. Department of Pediatrics, Okayama University Hospital, Japan)

4:15 PM - 5:45 PM

[I-APT2-04] Decision-making process in theatre: Ebstein's anomaly (One and a half repair in Ebstein's anomaly)

○ Tae Gook Jun (Samsung Medical Center, Sungkyunkwan University School of Medicine, Korea)

4:15 PM - 5:45 PM

[I-APT2-05] Decision-making process in theatre: AVSD

○ Zsolt Prodán (Congenital Cardiac Surgery Budapest, Hungary)

4:15 PM - 5:45 PM

[I-APT2-06] Late Cardiopulmonary Function after BVR, 1.5 VR, and Fontan Repair in Patients with Borderline Right Ventricle

○ Hideo Ohuchi (Departments of Pediatric Cardiology and Adult Congenital Heart Disease National Cerebral and Cardiovascular Center, Osaka, Japan)

4:15 PM - 5:45 PM

4:15 PM - 5:45 PM (Fri. Jul 7, 2017 4:15 PM - 5:45 PM ROOM 3)

[I-APT2-01] Circulation and morphology of the borderline right ventricle

○Hiroyuki Yamagishi (Department of Pediatrics, Keio University School of Medicine, Japan)

As the normal heart has two separate ventricles, namely right and left ventricle, “two-ventricle repair” is an ideal surgical strategy for complex congenital heart diseases. There are, however, defects that cannot be easily septated surgically commonly undergo the Fontan operation, or “one-ventricle repair.” And occasionally, there are hearts with two ventricular cavities and two atrioventricular valves, but the morphologic and physiologic characteristics of the right-sided ventricle are insufficient and subject to the “one and a half ventricle repair.” The strategy how to deal with the borderline right ventricle is extensively discussed in the session.

Congenital heart defects involving hypoplasia of the right or left ventricle account for 25% of all mortality from congenital heart disease in children and may be the result of defects in expansion of a precursor pool of ventricular cardiomyocytes. Cell lineage analyses have demonstrated that two progenitor cell populations, the first heart field (FHF) and second heart field (SHF), are derived from the lateral plate and splanchnic mesoderm, respectively. The FHF forms the crescent shaped heart primordium that gives rise to the linear heart tube and later contribute to most of the left ventricle. The SHF cells, initially medial and caudal to the FHF, migrate through the pharyngeal mesoderm into the heart tube and contribute to the outflow tract, right ventricle and atria.

As for the ventricular development, a “ballooning” model has been proposed in which growth of the ventral aspect of the linear heart tube gives rise to the outer curvature of the looped heart and results in ventricular expansion. Our observation using model mice represents some of the more convincing functional evidence supporting this model. Dissection of the complex molecular pathways involved in ventricular specification, differentiation and growth would provide the basis for understanding the pathogenesis of hypoplastic ventricle syndromes.

4:15 PM - 5:45 PM (Fri. Jul 7, 2017 4:15 PM - 5:45 PM ROOM 3)

[I-APT2-02] Fontan vs One-and-a-Half Ventricle Repair vs Biventricular Repair: what are the criteria and how to get there

○Mark A. Fogel (Children's Hospital of Philadelphia, USA)

Children born with varying degrees of hypoplasia of one or both ventricles can occur with many different anatomic configurations. In many instances, the choice is clear whether to reconstruct the heart with a single ventricle (SV) or a biventricular repair. There remains, however, a group of patients where that decision is not as clear cut (such as the borderline left ventricle) and reconstruction of cardiovascular system can proceed as either. In addition, if the SV route is chosen, supplemental pulmonary blood flow from the ventricle (the “one and a half ventricle” repair) may be an option. Congenital heart diseases such as double outlet right ventricle, critical aortic stenosis and malaligned atrioventricular canal are 3 such cases where these decisions come in to play. Clinical outcomes are not straightforward in these cases and the clinician needs to rely on a limited dataset in the literature along with their own

experience as well as newer ideas and imaging modalities. This lecture will discuss focus on these 3 types of congenital heart lesions when the operative decision is complex and review the literature on the topic. In addition, newer data and ideas about how to make this decision will be discussed. Ultimately, this will stimulate the discussion of which is better - a "good" Fontan or "poor" biventricular repair?

4:15 PM - 5:45 PM (Fri. Jul 7, 2017 4:15 PM - 5:45 PM ROOM 3)

[I-APT2-03] Decision-making process in theater: PAIVS

○Shunji Sano¹, Yasuhiro Kotani², Takahiro Eitoku³, Kenji Baba³, Shingo Kasahara² (1.Department of Pediatric Cardiothoracic Surgery, University California San Francisco, USA, 2.Department of Cardiovascular Surgery, Okayama University Hospital, Japan, 3.Department of Pediatrics, Okayama University Hospital, Japan)

Pulmonary atresia with an intact ventricular septum (PAIVS) is an uncommon congenital heart disease with a variable degree of right ventricular (RV) hypoplasia and coronary artery anomalies. Patients with RV-dependent coronary circulation should be better managed with single ventricular circulation, however achieving biventricular repair with good functional status and low systemic venous pressure is the ideal goal even to the patients with PAIVS. For biventricular circulation to be achieved, the tricuspid valve and RV must have adequate size to support pulmonary blood flow. Previous studies of PAIVS showed that RV and tricuspid valve diameters had poorer growth when RV– pulmonary artery continuity was not achieved. To aim a growth of right-sided heart structures, our repair strategy of the first palliation for patients with PAIVS includes modified BTS with pulmonary valvotomy.

We reviewed the data at our institution and analyzed the impact of the first palliation on the growth of right-sided heart and factors associated with a choice of definitive surgical procedure.

Methods and Results: Fifty patients with PAIVS underwent a staged surgical approach in Okayama university hospital since 1991. Six (12%) patients died after 1st palliation or inter-stage. Thirty patients could achieve a biventricular repair (BVR group), 6 patients had a 1+1/2 ventricular repair (1+1/2V group), and 5 patients had Fontan completion (Fontan group). After modified BTS with pulmonary valvotomy, normalized tricuspid valve(TV) diameter did not increase in any of group (BVR: pre 80% vs. post 83%, 1+1/2V: pre 63% vs. post 51%, Fontan: pre 57% vs. post 49%). Normalized RVEDV increased in only BVR group (BVR: pre 32% vs. post 64%, 1+1/2V: pre 43% vs. post 42%, Fontan: pre 29% vs. post 32%). Major coronary artery fistula was a strong factor with proceeding single-ventricle palliation (BVR 4/30 (13%) patients, 1+1/2V 1/6 (17%), and Fontan 4/5 (80%)).

Conclusions: TV growth was not obtained by modified BTS with pulmonary valvotomy, therefore TV size at birth appeared to be a predictor for achieving BVR. Proportionate RV growth was seen only in patients achieved BVR. However, RV growth was not seen in patients having 1+1/2 ventricular repair. Therefore, indication of biventricular repair is TV size of more than 80% of normal and Fontan is TV size of less than 50% of normal. Major coronary artery fistula was a strong predictor for proceeding single-ventricle palliation.

4:15 PM - 5:45 PM (Fri. Jul 7, 2017 4:15 PM - 5:45 PM ROOM 3)

[I-APT2-04] Decision-making process in theatre: Ebstein' s anomaly

(One and a half repair in Ebstein's anomaly)

○Tae gook Jun (Samsung Medical Center, Sungkyunkwan University School of Medicine, Korea)

Since the original contribution of Billingsley et al[1] about the one and a half repair in hypoplastic right ventricle, the indications of this strategy have been extended to a variety of congenital heart disease. The clinical situation for the one and a half repair include right ventricular outflow obstruction with a hypoplastic right ventricle(RV), pulmonary atresia with intact ventricular septum, Ebstein's anomaly, complex atrioventricular septal defect, inlet VSD with straddling of tricuspid valve, and the complex anomaly with difficulty factors during intracardiac repair[2].

In patients with Ebstein's anomaly, additional bidirectional Glenn (one and a half repair) with tricuspid valve repair or replacement achieves: it decrease the volume loading on the enlarged and dysfunctional RV, provides adequate preload to LV, reduces the hemodynamic stress on a complex TV repair, permits a more aggressive valve repair while avoiding valve stenosis. tolerates longer intervals between repeat TV operations for progressive TR and failing TV prostheses, and it provides less mortality[3-5].

Indication of one and a half repair in Ebstein's anomaly include severe cyanosis at rest, severe RV dilatation or dysfunction (cardiothoracic ratio >65%, RVEDV >250 mL/m², RVEF <0.25), small, squashed, D-shaped left ventricle(leftward shift of IVS), postrepair RAP:LAP >1.5:1, postrepair low cardiac output (persistent metabolic acidosis, low urine output, increasing creatinine, poor peripheral perfusion, and low mean arterial blood pressure <50 mm Hg), stenotic TV orifice after repair(mean gradient >8mmHg)[4-8]. The patient should have good LV function and good pulmonary conditions: LVEDP less than 15mmHg, good pulmonary artery pressure (less than 18 – 20 mmHg), and transpulmonary gradients less than 10 mmHg[7]. For the good hemodynamic after one and half repair, the right ventricular outflow tract obstruction must be avoided and the pulmonary valve is needed[9].

Contraindications [6] for one and a half repair are profound RV dilatation or dysfunction (RV ejection fraction [EF] <10%), left ventricular dilatation with dysfunction (LV EF <25%), , mean PA pressure >20 mm Hg, pulmonary arteriolar resistance >4 Woods units, LVEDP or LA pressure >12 mm Hg, and significant pulmonary artery hypoplasia.

Concerns after one and a half repair include possible development of venovenous collaterals due to high CPV, facial suffusion, pulmonary arteriovenous fistulae, central venous thrombosis, and difficulty access for right ventricle for electrophysiology study, ablation, and pacing lead insertion.

The one and a half ventricle repair seems to be a valid alternative to Fontan and biventricular repairs in selected patients with Ebstein's anomaly.

References

1. Billingsley, A.M., et al., *Definitive repair in patients with pulmonary atresia and intact ventricular septum*. J Thorac Cardiovasc Surg, 1989. **97**(5): p. 746-54.
2. Chowdhury, U.K., et al., *One and a half ventricle repair with pulsatile bidirectional Glenn: results and guidelines for patient selection*. Ann Thorac Surg, 2001. **71**(6): p. 1995-2002.
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4:15 PM - 5:45 PM (Fri. Jul 7, 2017 4:15 PM - 5:45 PM ROOM 3)

[I-APT2-05] Decision-making process in theatre: AVSD

○Zsolt Prodán (Congenital Cardiac Surgery Budapest, Hungary)

TBA

4:15 PM - 5:45 PM (Fri. Jul 7, 2017 4:15 PM - 5:45 PM ROOM 3)

[I-APT2-06] Late Cardiopulmonary Function after BVR, 1.5 VR, and Fontan Repair in Patients with Borderline Right Ventricle

○Hideo Ohuchi (Departments of Pediatric Cardiology and Adult Congenital Heart Disease National Cerebral and Cardiovascular Center, Osaka, Japan)

Long-term outcome of patients with the borderline right ventricle (RV) depends on the severity of abnormalities (size and function) of the tricuspid valve (TV) and RV. The final options of these patients mainly include Fontan operation, one-and a-half repair (1.5VR), and biventricular repair (BVR) and each of procedure may have significant effect on the long-term morbidity and mortality. Since 2006, we have evaluated cardiopulmonary function of patients with critical RV outflow tract obstruction with intact ventricular septum, consisting of 16 those after Fontan (24 ± 6 years), 4 after 1.5VR (24 ± 6 years), and 15 after BVR (24 ± 6 years). According to our data, peak oxygen uptake (VO₂) was highest in the BVR patients, while the lowest in 1.5VR patients ($p < 0.01$), and same trend of plasma levels of brain natriuretic peptide were observed ($p < 0.01$), indicating superior long-term outcome of BVR patients over the other two groups. In BVR patients, however, small RV volume was associated with low peak VO₂ ($p < 0.05$), implying some of BVR patients with borderline RV volume might have benefited from Fontan operation.

AP Target Symposium

AP Target Symposium 3 (II-APT3)

Dealing with congenitally corrected transposition of the great arteries - Efforts to minimize late development of systemic ventricular dysfunction

Chair:Yih-Sharng Chen(National Taiwan University Hospital, Taiwan)

Chair:Jun Yoshimoto(Pediatric Cardiology, Shizuoka Children's Hospital, Japan)

Sat. Jul 8, 2017 10:15 AM - 11:45 AM ROOM 3 (Exhibition and Event Hall Room 3)

[II-APT3-01] Morphological aspects

○Hideki Uemura (Congenital Heart Disease Center, Nara Medical University, Japan)
10:15 AM - 11:45 AM

[II-APT3-02] The late outcome of systemic right ventricle in congenitally corrected transposition of great arteries: Functional repair or anatomical repair

○Hajime Ichikawa¹, Takaya Hoashi¹, Tomohiro Nakata¹, Masatoshi Shimada¹, Akihiko Higashida¹, Hideo Ohuchi², Kenichi Kurosaki², Isao Shiraishi² (1.Department of Pediatric Cardiovascular Surgery, National Cerebral and Cardiovascular Center, Japan, 2.Department of Pediatric Cardiology, National Cerebral and Cardiovascular Center, Japan)
10:15 AM - 11:45 AM

[II-APT3-03] Electrical issue of corrected TGA

○Jun Yoshimoto (Department of Cardiology, Shizuoka Children's Hospital, Shizuoka, Japan)
10:15 AM - 11:45 AM

[II-APT3-04] Congenitally Corrected Transposition: Efforts to minimize late development of systemic RV Failure

○Sertaç M. Çiçek (Department of CV Surgery, Mayo Clinic, USA)
10:15 AM - 11:45 AM

[II-APT3-05] The Senning operation in anatomical repair of congenitally corrected transposition

○James S. Tweddell (Cardiothoracic Surgery, Cincinnati Children's Hospital Medical Center, USA)
10:15 AM - 11:45 AM

[II-APT3-06] Role of Fontan operation in cc-TGA

○Tae-Jin Yun (Asan Medical Center, University of Ulsan, Korea)
10:15 AM - 11:45 AM

10:15 AM - 11:45 AM (Sat. Jul 8, 2017 10:15 AM - 11:45 AM ROOM 3)

[II-APT3-01] Morphological aspects

○Hideki Uemura (Congenital Heart Disease Center, Nara Medical University, Japan)

It is essential to clarify the concept behind the terms 'congenitally corrected transposition of the great arteries', 'discordant atrioventricular connections', 'l-transposition', and so on. Overall understanding then will require recognition of morphological diversity and architectural spectra within this entity; not only for atrioventricular/ventriculo-arterial connections, but also atrial arrangement, nature of a ventricular septal defect (if any), obstruction across the ventricular outflow tracts (particularly to the pulmonary arteries), and anatomy of the coronary circulation. The feature of the conduction system has been well documented in this setting.

Such background knowledge should contribute to better comprehension of functional results of the systemic right ventricle or those subsequent to the so-called anatomic biventricular repair in the longer term. The investigators may not necessarily discuss the outcome through a uniform language. Thus, confusion or misleading analysis could go further in this difficult clinical area.

10:15 AM - 11:45 AM (Sat. Jul 8, 2017 10:15 AM - 11:45 AM ROOM 3)

[II-APT3-02] The late outcome of systemic right ventricle in congenitally corrected transposition of great arteries: Functional repair or anatomical repair

○Hajime Ichikawa¹, Takaya Hoashi¹, Tomohiro Nakata¹, Masatoshi Shimada¹, Akihiko Higashida¹, Hideo Ohuchi², Kenichi Kurosaki², Isao Shiraishi² (1.Department of Pediatric Cardiovascular Surgery, National Cerebral and Cardiovascular Center, Japan, 2.Department of Pediatric Cardiology, National Cerebral and Cardiovascular Center, Japan)

The poor outcome of systemic right ventricle (RV) in congenitally corrected transposition of great arteries (ccTGA) is a well-known fact. Our strategy for the treatment of ccTGA is to place the left ventricle in a systemic position since 1990. The outcome is compared with the functional repair.

Patients and method: One hundred sixty one ccTGA patients with balanced ventricle were included in this retrospective non-randomized analysis. There were 81 patients with conventional repair including simple VSD closure or complex LV-PA conduit with interventricular rerouting. Seventy patients underwent double switch operation of either Senning/Mustard plus Jatene/Rastelli type operation. Ten patients underwent only pacemaker implantation or palliative surgery. **Results:** Survival rates in the conventional group were poor with 10, 20 and 30 year freedom from death after surgery of 75, 71 and 65%, respectively. The age at initial surgical intervention inversely correlated with the survival (expired 9 ± 16 vs survived 19 ± 20). The survival rate of simple tricuspid valve replacement in 22 patients (average 29 years old) were 91, 91 and 91% in 10, 20 and 30 years after the operation, respectively. Since 1997, the survival rates after double switch operation were 97 and 91.3% at 10 and 20 years, respectively. The median age at the initial surgical intervention was 1.8 year old. **Conclusion:** When earlier surgical intervention is needed, conventional repair only provide poor outcome. Earlier decision of treatment strategy may improve the clinical outcome of patients with ccTGA.

10:15 AM - 11:45 AM (Sat. Jul 8, 2017 10:15 AM - 11:45 AM ROOM 3)

[II-APT3-03] Electrical issue of corrected TGA

○Jun Yoshimoto (Department of Cardiology, Shizuoka Children's Hospital, Shizuoka, Japan)

In congenitally corrected TGA (ccTGA), there are several electro-anatomical issues. No.1 is an anatomical issue. SA node lies in normal position, but AV node (AVN) is in abnormal position. The actual position and course of AVN and His bundle differs case by case, especially because of straddling and DORV. Also connective tissue lies superficially and fragile. This seems to be the reason of occurrence of congenital and postsurgical complete AV block. No.2 is electrical and mechanical dyssynchrony. Bundle branch block of systemic ventricle causes electrical and mechanical dyssynchrony and they can be the cause of heart failure. In the case of univentricular physiology, however, interventricular dyssynchrony can cause severe heart failure. No.3 is tachy arrhythmia post double switch operation. Both Senning and Mustard operation can cause intra atrial reentry tachycardia(IART). Ablation of IART of ccTGA has difficult problem because of anatomical abnormality of AVN, therefore brockenbrough puncture of intraatrial septum or baffle. Ablation of AVnodal Reentry tachycardia is quite difficult because of anatomical abnormality.

10:15 AM - 11:45 AM (Sat. Jul 8, 2017 10:15 AM - 11:45 AM ROOM 3)

[II-APT3-04] Congenitally Corrected Transposition: Efforts to minimize late development of systemic RV Failure

○Sertaç M. Çiçek (Department of CV Surgery, Mayo Clinic, USA)

Congenitally corrected transposition of the great arteries is a rare defect representing approximately 0.5% of all congenital heart disease. ccTGA is characterised by the combination of atrioventricular discordance and ventriculo-arterial discordance. This combination, atrioventricular discordance and ventriculoarterial discordance, produces a unique set of management challenges.

Patients with congenitally corrected transposition of the great arteries have a morphological right ventricle that sustains the systemic circulation. In these patients, regurgitation of the systemic atrioventricular valve, the tricuspid valve, is a common finding. When TV regurgitation becomes severe, it is associated with RV failure and decreased survival. Associated anomalies such as ventricular septal defect, morphologic left ventricular outflow tract obstruction and tricuspid valve abnormalities occur in the majority of these patients. Although the presence of these associated conditions influences the clinical presentation of this condition and the type and timing of surgical intervention, ***the ability of the morphologic right ventricle and the tricuspid valve to withstand a lifetime of exposure to systemic pressure largely determines the ultimate outcome of these patients.***

Progressive dysfunction of the tricuspid valve and right ventricle occurs after prolonged exposure to systemic pressure in a substantial percentage of patients who have undergone an atrial switch operation for transposition of the great arteries with increased risk for patients who have accompanying lesions. If sensitive measures of ventricular function are used, an even larger percentage of cc-TGA patients can be shown to have subclinical evidence of limited cardiovascular reserve. A significant percentage of cc-TGA patients had perfusion defects during exercise stress testing with radionuclide imaging.

Increasing morphologic LV pressure either naturally by PS or banding may maintain RV geometry & prevent progression of TR. Shifting the ventricular septum leftward, reduces left ventricular (LV) end-diastolic volume and causes LV to fall within the Frank-Starling curve and reduces LV filling dynamics

and end-diastolic pressure.

Earlier intervention with tricuspid regurgitation even with minimal severity could reverse the future RV failure. Recent data supports the use of tricuspid valve replacement as an alternative to repair, especially in adult patients. The use of the double switch operation is predicated on the assumption that long-term outcomes will be better with the morphologic left ventricle supporting the systemic circulation.

Surgery for congenitally corrected transposition of the great arteries has evolved overtime. Anatomical repair of cc-TGA is now performed with quite satisfactory outcomes in children. The double switch operation has become the preferred surgical procedure in selected cases. However, Systemic A-V valve regurgitation strongly associated with RV dysfunction and CHF whether this is causative or secondary remains speculative.

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10:15 AM - 11:45 AM (Sat. Jul 8, 2017 10:15 AM - 11:45 AM ROOM 3)

[II-APT3-05] The Senning operation in anatomical repair of congenitally corrected transposition

○James S. Tweddell (Cardiothoracic Surgery, Cincinnati Children's Hospital Medical Center, USA)

Current controversies in the management of the patient with congenitally corrected transposition of the great arteries (ccTGA) revolve around the best choice of procedure among the choices of physiologic repair, anatomic repair and single ventricle palliation.(1-3) While anatomic correction of ccTGA aims to improve long term outcome by placing the morphologic left ventricle (LV) as the systemic ventricle and avoiding systemic right ventricular failure the long-term outlook is complicated by a high rate of re-intervention, conduction abnormalities combined with systemic left ventricular dysfunction. Options for the atrial level switch component of the anatomic correction include; the Senning operation, the Mustard procedure or the so called hemi-Mustard combined with a bidirectional Glenn shunt.(4) To maximize the benefit of anatomic repair and limit reoperations, an atrial level switch that minimizes sinus node dysfunction, baffle obstruction and supraventricular arrhythmias is essential.(5-7) While, potentially more complicated to perform the Senning procedure uses native tissue and results in a low rate of late baffle obstruction. Technical strategies to avoid sinus node dysfunction can be successfully applied.(8) The Senning, as opposed the hemi-Mustard, maintains the superior caval vein connection to the atrium which may be important for access for pacemaker and arrhythmia management. The steps of the Senning procedure will be demonstrated with a video and the late outcomes reviewed.

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10:15 AM - 11:45 AM (Sat. Jul 8, 2017 10:15 AM - 11:45 AM ROOM 3)

[II-APT3-06] Role of Fontan operation in cc-TGA

○Tae-Jin Yun (Asan Medical Center, University of Ulsan, Korea)

Patients with congenitally corrected transposition of the great arteries (cc-TGA) can be categorized into discrete three subgroups with respect to the surgical strategies: 1) cc-TGA with ventricular septal defect (VSD) and no pulmonary stenosis (PS) 2) cc-TGA with intact ventricular septum (IVS), and 3) cc-TGA, VSD and PS or pulmonary atresia (PA). The first subset is thought to be the best candidates for double switch operation (DSO), because patients rapidly develop severe congestive heart failure which necessitates urgent surgical interventions (i.e. pulmonary artery banding for staged DSO or early primary DSO). In the second subgroup, pulmonary artery banding for the training of the morphologic left ventricle is commonly indicated when regurgitation of the atrioventricular valve of the systemic right ventricle (i.e. tricuspid regurgitation) develops. As long as the tricuspid valve is competent, however, it is very difficult to determine the exact timing of any surgical intervention. The best surgical option for the third group is still under debate. Anatomical repair (i.e. Intraventricular rerouting from the morphologic LV to the aortic valve in association with atrial switch operation and right ventricle to pulmonary artery extracardiac conduit interposition) has been attempted for this subset, but it has been pointed out that the potential risks of morbidities associated with atrial switch operation, RV-PA conduit stenosis, and left ventricular outflow tract obstruction, particularly in case of restrictive VSD,

may outweigh the potential benefits of recruiting the morphological left ventricle to the systemic circulation. Furthermore, superiority of anatomical repair over the Fontan procedure does not seem to be fully substantiated by the clinical reports pertaining to the long-term outcome of anatomical repair for cc-TGA with VSD and PS.

AP Target Symposium

AP Target Symposium 4 (III-APT4)

Optimizing results in staged surgical management of functionally univentricular hearts – Preparation rather than Selection for Fontan

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Chair: Akio Ikai (The Cardiovascular Center, Mt. Fuji Shizuoka Children's Hospital, Japan)

Chair: Swee Chye Quek (Pediatrics, National University of Singapore, Singapore)

Sun. Jul 9, 2017 8:30 AM - 10:15 AM ROOM 3 (Exhibition and Event Hall Room 3)

[III-APT4-01] A Quantitative Analysis of the Systemic to Pulmonary Collateral Flow in Fontan Circulation by Cardiac Magnetic Resonance

○Yoshihiko Kodama¹, Yuichi Ishikawa¹, Shiro Ishikawa¹, Ayako Kuraoka¹, Makoto Nakamura¹, Kouichi Sagawa¹, Toshihide Nakano², Hideaki Kado² (1. Department of Pediatric Cardiology, Fukuoka Children's Hospital, Japan, 2. Department of Cardiovascular Surgery, Fukuoka Children's Hospital, Japan)

8:30 AM - 10:15 AM

[III-APT4-02] Arrhythmia management as a preparation for a Fontan

○Heima Sakaguchi (Department of Pediatric Cardiology, National Cerebral and Cardiovascular Center, Osaka, Japan)

8:30 AM - 10:15 AM

[III-APT4-03] Atrioventricular valve anatomy and function in patients with single ventricle

○Masaki Nii (Shizuoka Children's Hospital, Shizuoka, Japan)

8:30 AM - 10:15 AM

[III-APT4-04] Surgical preparation for Fontan. Atrioventricular valve repair

○James S. Tweddell (Cardiothoracic Surgery, Cincinnati Children's Hospital Medical Center, USA)

8:30 AM - 10:15 AM

[III-APT4-05] Extending the indication of Damus anastomosis

○Makoto Ando, Yukihiro Takahashi (Department of Pediatric Cardiovascular Surgery, Sakakibara Heart Institute, Tokyo, Japan)

8:30 AM - 10:15 AM

[III-APT4-06] Single ventricular repair strategy: Controversy and some options and details in right heart bypass operation

○Hajime Ichikawa (Department of Pediatric Cardiovascular Surgery, National Cerebral and Cardiovascular Center, Japan)

8:30 AM - 10:15 AM

[III-APT4-07] Use of Mechanical Circulatory Assist Devices and Cardiac Transplantation in the Failing Functional Single Ventricle Patients

○James, D St. Louis (Department of Surgery Children's Mercy)

8:30 AM - 10:15 AM

[III-APT4-08] TBA

○Sertaç M. Çiçek (Cardiovascular Surgery, Mayo Clinic, USA)

8:30 AM - 10:15 AM

8:30 AM - 10:15 AM (Sun. Jul 9, 2017 8:30 AM - 10:15 AM ROOM 3)

[III-APT4-01] A Quantitative Analysis of the Systemic to Pulmonary Collateral Flow in Fontan Circulation by Cardiac Magnetic Resonance

○Yoshihiko Kodama¹, Yuichi Ishikawa¹, Shiro Ishikawa¹, Ayako Kuraoka¹, Makoto Nakamura¹, Kouichi Sagawa¹, Toshihide Nakano², Hideaki Kado² (1.Department of Pediatric Cardiology, Fukuoka Children's Hospital, Japan, 2.Department of Cardiovascular Surgery, Fukuoka Children's Hospital, Japan)

Keywords: Cardiac Magnetic Resonance, Fontan, Systemic to Pulmonary Collateral

With the prevalence of cardiac magnetic resonance (CMR), a considerable amount of systemic to pulmonary collateral flow (SPC) in Fontan patients has been widely recognized. An aim of this study is to clarify causal relationships between SPC and other various parameters of Fontan circulation. Out of 655 consecutive patients in the Fukuoka Fontan Study database, 288 pts whose Qp/Qs by CMR was 1.0 or over were recruited. The median age at CMR was 13.5 (2.2-40.4) years old. The %SPC was calculated as $(Q_p - Q_s) \times 100 / Q_s$, and its median was 13.8% (0-95.7%). %SPC was significantly correlated to the smaller pulmonary artery index (Nakata index) before Fontan ($p=0.02$) and comorbidity of total anomalous pulmonary vein drainage ($p=0.04$). There was positive correlations between %SPC and the end-diastolic volume of the ventricle ($p=0.006$), and plasma BNP levels ($p<0.001$). Furthermore, an increase in %SPC was related to a deterioration in NYHA functional classification ($p=0.02$). It is concluded that a noninvasive measurement of SPC by CMR is highly effective in an evaluation of Fontan circulation.

8:30 AM - 10:15 AM (Sun. Jul 9, 2017 8:30 AM - 10:15 AM ROOM 3)

[III-APT4-02] Arrhythmia management as a preparation for a Fontan

○Heima Sakaguchi (Department of Pediatric Cardiology, National Cerebral and Cardiovascular Center, Osaka, Japan)

In patients with a single ventricular physiology, arrhythmias can be a risk factor for surgical treatment including a bidirectional Glenn (BDG)/total cavopulmonary connection (TCPC) completion. Cases with hypoplastic right or left heart syndrome especially are often complicated with atrial arrhythmias due to a unilateral atrial load continuing from the fetal period. Herein we reviewed 8 cases (12 sessions) who underwent radiofrequency catheter ablation (RFCA) prior to a BDG/TCPC completion in our hospital over the last decade. Two patients were excluded from this study because they did not have an indication for a TCPC completion at that time. Of the 6 patients, underlying congenital heart defects consisted of a pulmonary atresia (PA) with an intact ventricular septum s/p Blalock Taussig (BT) shunt in 2, hypoplastic left heart syndrome (HLHS) s/p Norwood in 2, tricuspid atresia PA s/p BT shunt in 1, and atrio-ventricular discordance and mitral atresia s/p BDG in 1. Among 10 sessions, the median age and body weight at the time of the RFCA were 7 months and 5.3 kg, respectively. The target arrhythmias that were ablated were focal atrial tachycardia (AT) in 4 cases (8 sessions), intra-atrial re-entrant tachycardia in 1, and AVRT in 1. Although 3 out of 6 patients required multiple sessions because of difficulty inducing AT, their arrhythmia substrates were successfully eliminated. After the RFCA, all underwent a BDG/TCPC completion. It is very useful option as a preparation for the Fontan to tenaciously attempt to ablate any atrial arrhythmias prior to the BDG/TCPC.

8:30 AM - 10:15 AM (Sun. Jul 9, 2017 8:30 AM - 10:15 AM ROOM 3)

[III-APT4-03] Atrioventricular valve anatomy and function in patients with single ventricle

○Masaki Nii (Shizuoka Children's Hospital, Shizuoka, Japan)

Atrioventricular valve (AVV) function is one of the most important determinants of prognosis of patients with single ventricular physiology. Therefore, how to treat AVV regurgitation is a key for the preparation to have Fontan operation. Echocardiography is the 1st line imaging modality to assess AVV function before operation. The mechanisms of AVV regurgitation in patients with single ventricle and how to assess AVV function by two and three-dimensional echocardiography is discussed.

8:30 AM - 10:15 AM (Sun. Jul 9, 2017 8:30 AM - 10:15 AM ROOM 3)

[III-APT4-04] Surgical preparation for Fontan. Atrioventricular valve repair

○James S. Tweddell (Cardiothoracic Surgery, Cincinnati Children's Hospital Medical Center, USA)

A competent non-stenotic atrioventricular valve is essential for optimal long term outlook. Risk of atrioventricular valve regurgitation is greatest for individuals with a single tricuspid valve or a common atrioventricular valve, followed by patients with two atrioventricular valves and least for those with a single mitral valve.(1) Patients with a common atrioventricular valve are a challenging group to repair due to wide variation of valve morphology which includes patients with heterotaxy syndrome and unbalanced atrioventricular canal defect. Successful repair nearly always includes approximation of the central zone of apposition or "cleft" between the bridging leaflets.(2, 3) Additional strategies include suture annuloplasty and partial commissural closure.(4, 5) The approximation of the central zone of apposition may be combined with the addition of a structural support using a polytetrafluoroethylene strip to decrease the anterior-posterior dimension and take stress off the central leaflet suture line and may be useful in smaller, younger patients.(6, 7) Especially among individuals with heterotaxy syndrome, abnormal cardiac position such as mesocardia and dextrocardia may complicate exposure of the valve and intraoperative assessment of the source of regurgitation may not reflect the mechanism when the heart is in normal position. Therefore, careful study of the preoperative studies are necessary to identify the areas of regurgitation and surgery should be planned according to this evaluation rather than solely on the intraoperative assessment. For the patient with hypoplastic left heart syndrome, tricuspid regurgitation may be due to both intrinsic abnormalities of the tricuspid valve as well as acquired abnormalities including annular dilatation as well as elongation of subvalvar apparatus due to ischemia resulting in leaflet prolapse. Partial or complete suture annuloplasty will reduce annular dilatation and treatment of leaflet prolapse can be managed with partial or complete commissural closure.(8, 9) As repair strategies become more radical such as requiring leaflet augmentation or artificial cords the durability of repair decreases. Mitral valve repairs are the least common. Repairs may be directed at approximation of a persistent zone of apposition or "cleft". Additional procedures include partial or complete annuloplasty either suture or ring, partial commissural closure and edge-to-edge repair. Most

patients with single ventricle anatomy can achieve a reasonable outcome with valve repair and although reoperation is common this can often be combined with staged palliation.

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8:30 AM - 10:15 AM (Sun. Jul 9, 2017 8:30 AM - 10:15 AM ROOM 3)

[III-APT4-05] Extending the indication of Damus anastomosis

○Makoto Ando, Yukihiko Takahashi (Department of Pediatric Cardiovascular Surgery, Sakakibara Heart Institute, Tokyo, Japan)

Keywords: Single ventricle, Fontan, Damus

Background

Damus-Kaye-Stansel procedure (DKS), generally performed in the presence of subaortic stenosis, may be beneficial in case of Fontan candidate with a two well-developed ventricles.

Patients

Patients undergoing either Glenn anastomosis (BDG) or modified Fontan operation (F) (1978-2016, N=567) was the object of study. The staged Fontan operation was primarily performed (N=418). The remaining 95 patients are presently awaiting the Fontan operation after the Glenn anastomosis.

Results

Patients undergoing DKS anastomosis had a better mortality (5.1% vs. 11.2%) and medication free rate (73.9% vs. 64.9%) compared with the others. The best actuarial survival and Fontan achievement rate

was seen in the DKS group compared with the Norwood and the other groups. The timing of the DKS (BDG or F) did not affect cardiac catheterization data after the Fontan operation. Especially, in the recent years (2006-) more patients have undergone DKS procedure after the preceding pulmonary banding (57.7%); the primary reason includes the increase in patients having two-well developed ventricles. Our data also indicated that transaction of the pulmonary arterial associated a higher incidence of late ventricular dyssynchrony.

Conclusions

DKS can be performed safely at any stage after the BDG procedure. Even without a concern for subaortic stenosis, DKS may be offer better results for patients with two well-developed ventricles, providing systemic outflow from both ventricles.

8:30 AM - 10:15 AM (Sun. Jul 9, 2017 8:30 AM - 10:15 AM ROOM 3)

[III-APT4-06] Single ventricular repair strategy: Controversy and some options and details in right heart bypass operation

○Hajime Ichikawa (Department of Pediatric Cardiovascular Surgery, National Cerebral and Cardiovascular Center, Japan)

The following 4 subjects are discussed by request.

Antegrade flow

Additional antegrade pulmonary flow (AAPF) after bidirectional Glenn is applied for hepatic factor and pulsatility. In the articles, in which AAPF is recommended, the age of Fontan completion is high. After 2010, the number of articles describing the benefit decreased, possibly because Fontan completion is getting earlier, the role of AAPF may no more exists.

Lateral tunnel (LT) vs. Extracardiac (EC): Systematic review by Dr. Li. shows the data of 3499 patients with either LT or EC, there was almost no difference except the increased incidence of late arrhythmia in lateral tunnel group.

Fenestration In 1990, fenestration was considered to be an savior and employed in many centers as an routine practice. After 2010, many big centers tend to publish their excellent outcome after non-fenestrated Fontan operation. The application of EC tended to avoid fenestration. However, the effect of non-fenestration has influence on a long term outcome including liver dysfunction.

Age at Fontan As we reported more than 20 years ago, better ventricular function could be achieved with a younger age at Fontan completion. This was confirmed in many other centers. However, recent north America's multicenter study shows that the age at Fontan does not predict the long term outcome.

Conclusion: The role of AAPF might be ended in the era of earlier Fontan. EC Fontan is the mainstream after 2010. Fenestration might revive. We need multicenter prospective study to elucidate the optimal age at Fontan.

8:30 AM - 10:15 AM (Sun. Jul 9, 2017 8:30 AM - 10:15 AM ROOM 3)

[III-APT4-07] Use of Mechanical Circulatory Assist Devices and Cardiac Transplantation in the Failing Functional Single Ventricle Patients

○James, D St. Louis (Department of Surgery Children's Mercy)

Palliative interventions are commonly utilized for patients with a functionally single ventricular congenital cardiac anatomy, where definitive procedures are not possible. Failure to progress through these series of palliative procedures occur for a number of reasons. When this failure occurs, utilization of mechanical circulatory assist devices and eventual listing for cardiac transplantation may be required. This presentation will address the complications which may occur in patients undergoing stages palliative procedures for functionally single ventricular anatomy, use of mechanical circulatory assist devices, cardiac transplantation as well as addressing the outcomes of these procedures.

8:30 AM - 10:15 AM (Sun. Jul 9, 2017 8:30 AM - 10:15 AM ROOM 3)

[III-APT4-08] TBA

○Sertaç M. Çiçek (Cardiovascular Surgery, Mayo Clinic, USA)

TBA