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 Kurosaki[1], Akira Miyake[1], Masataka Kitano[1], Atsushi Hirota[1], Isao Shiraishi[1], Takaya Hoashi[2], Hajime Ichikawa[2] (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 Louis[1] (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 Sano[1], Yasuhiro Kotani[2], Takahiro Eitoku[3], Kenji Baba[3], Shingo Kasahara[2] (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

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 Kurosaki[1], Akira Miyake[1], Masataka Kitano[1], Atsushi Hirota[1], Isao Shiraishi[1], Takaya Hoashi[2], Hajime Ichikawa[2] (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 Louis[1] (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 Kurosaki[1], Akira Miyake[1], Masataka Kitano[1], Atsushi Hirota[1], Isao Shiraishi[1], Takaya Hoashi[2], Hajime Ichikawa[2] (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 Louis[1] （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 all 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 bivetricular 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 Sano[1], Yasuhiro Kotani[2], Takahiro Eitoku[3], Kenji Baba[3], Shingo Kasahara[2]
（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 Sano[1], Yasuhiro Kotani[2], Takahiro Eitoku[3], Kenji Baba[3], Shingo Kasahara[2] (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 reapir) 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 Ebsein’ 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 venoveno 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.
3. Stulak, J.M., J.A. Dearani, and G.K.Danielson, *Surgical management of Ebstein's anomaly.* Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu, 2007: p. 105-11.
4. Dearani, J.A., et al., *Ebstein anomaly review: what's now, what's next?* Expert Rev Cardiovasc Ther, 2015. **13**(10): p. 1101-9.
5. Malhotra, S.P., et al., *Selective right ventricular unloading and novel technical concepts in Ebstein's anomaly.* Ann Thorac Surg, 2009. **88**(6): p. 1975-81; discussion 1981.
6. Raju, V., et al., *Right ventricular unloading for heart failure related to Ebstein malformation.* Ann Thorac Surg, 2014. **98**(1): p. 167-73; discussion 173-4.
7. Dearani, J.A., et al., *Anatomic repair of Ebstein's malformation: lessons learned with cone*

reconstruction. Ann Thorac Surg, 2013. **95**(1): p. 220-6; discussion 226-8.
8. Prifti, E., et al., *One and a half ventricle repair in association with tricuspid valve repair according to "peacock tail" technique in patients with Ebstein's malformation and failing right ventricle.* J Card Surg, 2014. **29**(3): p. 383-9.
9. Uribe, S., et al., *Hemodynamic assessment in patients with one-and-a-half ventricle repair revealed by four-dimensional flow magnetic resonance imaging.* Pediatr Cardiol, 2013. **34**(2): p. 447-51.

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.