

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)

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

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

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[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)

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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 $(Qp-Qs) \times 100 / Qs$, 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.

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[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.

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[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.

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[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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[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.

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[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.

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[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.

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[III-APT4-08] TBA

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

TBA