Sat. Jul 8, 2017

E-Oral Presentation Area

JCK E-Oral Presentation

JCK E-Oral Presentation 1 (II-JCKEOP01)

Chair:Shiro Baba(Department of Pediatrics, Graduate School of Medicine, Kyoto University, Japan)
6:15 PM - 7:15 PM E-Oral Presentation Area (Exhibition and Event Hall)

[II-JCKEOP01-01] Differentiation of mesenchymal stem cells into cardiomyocytes is regulated by miRNA-1-2 via WNT signaling pathway

○Xing Shen[1], Bo Pan[1], Huiming Zhou[1], Lingjuan Liu[1], Tiewei Lv[1], Jing Zhu[1],Xupei Huang[2], Jie Tian[1] （1.Department of Cardiology, Chongqing Medical University ,Chongqing-City, China, 2.Department of Biomedical Science, Charlie E. Schmidt College of Medicine, Florida Atlantic University, Boca Raton, FL, USA）

6:15 PM - 7:15 PM

[II-JCKEOP01-02] Postoperative mortality and respiratory complications in CHD patients with heterotaxy and their relationship with ciliary dysfunction

○Weicheng Chen, Shuolin Li, Sida Liu, Guoying Huang, Bing Jia （Heart Center, Children's Hospital of Fudan University, Shanghai, China）

6:15 PM - 7:15 PM

[II-JCKEOP01-03] Cardiac function of the fetuses with positive maternal anti-SS-A antibody

○Yasuki Maeno, Yozo Teramachi, Akiko Hirose, Yasuyuki Kagiyama, Shintaro Kishimoto, Yusuke Koteda, Kenji Suda （Department of Pediatrics, Kurume University School of Medicine, Japan）

6:15 PM - 7:15 PM

[II-JCKEOP01-04] Improvement of portopulmonary hypertension after liver transplantation

○Shiro Baba[1], Daisuke Yoshinaga[1], Kouichi Matsuda[1], Kentaro Akagi[1], Shinji Uemoto[2], Takuya Hirata[1] （1.Department of Pediatrics, Kyoto University, Japan 2.Division Hepato-Billary-Pancreatic Surgery and Transplantation Department, Kyoto University, Japan）

6:15 PM - 7:15 PM

[II-JCKEOP01-05] Surgical outcomes of open surgical repair for coarctation of the aorta in adolescent and adult in the era of endovascular therapy

○Yoshikatsu Saiki, Osamu Adachi, Masatoshi Akiyama, Ichiro Yoshioka, Satoshi Kawatsu, Yukihiro Hayatsu,Yusuke Suzuki, Konosuke Sasaki, Kiichiro Kumagai, Shunsuke Kawamoto, （Division of Cardiovascular Surgery, Tohoku University, Sendai, Japan）

6:15 PM - 7:15 PM

[II-JCKEOP01-06] Perioperative effects of the Tolvaptan for the Fontan Patients

○Takeshi Konuma[1], Syuhei Toba[1], Ayano Hutuki[1], Kohsuke Kurihara[2], Noriko Yodoya[2], Hiroyuki Ohashi[2], Hirobumi Sawada[2], Yosihide Mitani[2], Jyunya Hirayama[2], Hideto Shinpo[1] （1.Departmemt of Thoracic and Cardiovascular Surgery, Mie University, Japan 2.Departmemt of Pediatrics, Mie University, Japan）

6:15 PM - 7:15 PM

[II-JCKEOP01-07] Enlargement of the pituitary gland in patients after Fontan operation: another portal circulation

○Yusaku Nagatomo, Jun Muneuchi, Mamie Watanabe, Ryohei Matsuoka, Hiromitsu Shirouzu, Seigo Okada, Chiaki Iida, Kunitaka Joo （Pediatrics, Kyushu Hospital, Kitakyushu city, Japan）

6:15 PM - 7:15 PM

[II-JCKEOP01-08] Utility of and hemodynamic association with alpha$_1$ -antitrypsin clearance in Fontan patients

○Satoshi Masutani, Yoichi Iwamoto, Hirotaka Ishido, Akiko Yana, Seiko Kuwata, Clara Kurishima, Hideaki Senzaki （Pediatric Cardiology, Saitama Medical University Saitama Medical Center, Japan）

6:15 PM - 7:15 PM

Sun. Jul 9, 2017

E-Oral Presentation Area

JCK E-Oral Presentation

JCK E-Oral Presentation 2 (III-JCKEOP02)

Chair:Atsuko Kato(Division of Cardiology, The Labatt Family Heart Centre, Department of Pediatrics, The Hospital for Sick Children, University of Toronto, Toronto, Canada)
Chair:Takaya Hoashi(Department of Pediatric Cardiovascular Surgery, National Cerebral and Cardiovascular Center, Suita, Japan)
1:00 PM - 2:00 PM E-Oral Presentation Area (Exhibition and Event Hall)

[III-JCKEOP02-01] Transcatheter closure of doubly committed VSDs - a 5 year single centre experience

○Tran Cong Bao Phung, Do Nguyen Tin （Cardiology Department, Children Hospital 1, Ho Chi Minh City, VietNam）

1:00 PM - 2:00 PM

[III-JCKEOP02-02] Effect of fenestration on ventricular-vascular coupling chronically after Fontan operation - Cardiac magnetic resonance study

○Yoichi Iwamoto, Seiko Kuwata, Akiko Yana, Hirotaka Ishido, Satoshi Masutani, Hideaki Senzaki （Division of Pediatric Cardiology, MFN Center, Saitama Medical University Saitama Medical Center, Japan）

1:00 PM - 2:00 PM

[III-JCKEOP02-03] Development of a New and Rapid 3D Printing System for Manufacturing Super Flexible Replicas of Congenital Heart Disease

○Isao Shiraishi[1], Kennichi Kurosaki[1], Suzu Kanzaki[2], Takaya Hoashi[3], Hajime Ichikawa[3] （1.Department of Pediatric Cardiology National Cerebral and Cardiovascular, Japan, 2.Department of Radiology, National Cerebral and Cardiovascular Center, Japan, 3.Department of Pediatric Cardiac Surgery, National Cerebral and Cardiovascular Center, Japan）

1:00 PM - 2:00 PM

[III-JCKEOP02-04] Minimally invasive epicardial implantable cardioverter-defibrillator placement in a young child

○Wakana Maki[1], Hiroko Asakai[1], Kazuhiro Shiraga[1], Takahiro Shindo[1], Yoichiro Hirata[1], Ryo Inuzuka[1], Tomoyuki Iwase[2], Tetsuhiro Takaoka[2], Akihiro Masuzawa[2], Yasutaka Hirata[2], Akira Oka[1] （1.Department of Pediatrics The University of Tokyo Hospital, Japan 2.Department of Cardiac Surgery The University of Tokyo Hospital,Japan）

1:00 PM - 2:00 PM

[III-JCKEOP02-05] Fetal Echocardiography Characteristics in Vietnam Population

○Tran Cong Bao Phung, Vu Minh Phuc, Do Nguyen Tin, Nguyen Tri Hao, Phan Hoang Yen, Phan Tien Loi （Cardiology Department, Children Hospital 1, Ho Chi Minh City, VietNam）

1:00 PM - 2:00 PM

[III-JCKEOP02-06] Mitral valve replacement using stented bovine jugular vein graft (Melody valve) in infants and small children

○Atsuko Kato[1], Osami Honjo[2] （1.Division of Cardiology, The Labatt Family Heart Centre, Department of Pediatrics, The Hospital for Sick Children, University of Toronto, Toronto, Canada, 2.Department of Cardiovascular surgery, The Labatt Family Heart Centre, The Hospital for Sick Children, University of Toronto, Toronto, Canada）

1:00 PM - 2:00 PM

[III-JCKEOP02-07] Long-term Surgical Outcome of Transposition of the Great Arteries with Intact Ventricular Septum and Left Ventricular Outflow Tract Obstruction

○Akihisa Furuta, Mitsugi Nagashima, Takahiko Sakamoto, Goki Matsumura, Kentaro Umezu, Jin Ikarashi, Junko Katagiri, Hironori Murakami （Department of Cardiovascular Surgery,

Tokyo Women's University, Japan）

1:00 PM - 2:00 PM

[III-JCKEOP02-08] Successful biventricular conversion late after primary one and one-half ventricle repair

○Takaya Hoashi[1], Masataka Kitano[2], Masatoshi Shimada[1], Kenichi Kurosaki[2], Hajime Ichikawa[1] （1.Department of Pediatric Cardiovascular Surgery, National Cerebral and Cardiovascular Center, Suita, Japan, 2.Department of Pediatric Cardiology, National Cerebral and Cardiovascular Center, Suita, Japan）

1:00 PM - 2:00 PM

JCK E-Oral Presentation

JCK E-Oral Presentation 1 (II-JCKEOP01)

Chair:Shiro Baba(Department of Pediatrics, Graduate School of Medicine, Kyoto University, Japan)
Sat. Jul 8, 2017 6:15 PM - 7:15 PM E-Oral Presentation Area (Exhibition and Event Hall)

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[II-JCKEOP01-02] Postoperative mortality and respiratory complications in CHD patients with heterotaxy and their relationship with ciliary dysfunction

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6:15 PM - 7:15 PM

[II-JCKEOP01-04] Improvement of portopulmonary hypertension after liver transplantation

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[II-JCKEOP01-05] Surgical outcomes of open surgical repair for coarctation of the aorta in adolescent and adult in the era of endovascular therapy

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[II-JCKEOP01-06] Perioperative effects of the Tolvaptan for the Fontan Patients

○Takeshi Konuma[1], Syuhei Toba[1], Ayano Hutuki[1], Kohsuke Kurihara[2], Noriko Yodoya[2], Hiroyuki Ohashi[2], Hirobumi Sawada[2], Yosihide Mitani[2], Jyunya Hirayama

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[II-JCKEOP01-07] Enlargement of the pituitary gland in patients after Fontan operation: another portal circulation

○Yusaku Nagatomo, Jun Muneuchi, Mamie Watanabe, Ryohei Matsuoka, Hiromitsu Shirouzu, Seigo Okada, Chiaki Iida, Kunitaka Joo （Pediatrics, Kyushu Hospital, Kitakyushu city, Japan）

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[II-JCKEOP01-08] Utility of and hemodynamic association with alpha$_1$-antitrypsin clearance in Fontan patients

○Satoshi Masutani, Yoichi Iwamoto, Hirotaka Ishido, Akiko Yana, Seiko Kuwata, Clara Kurishima, Hideaki Senzaki （Pediatric Cardiology, Saitama Medical University Saitama Medical Center, Japan）

6:15 PM - 7:15 PM

6:15 PM - 7:15 PM (Sat. Jul 8, 2017 6:15 PM - 7:15 PM E-Oral Presentation Area)

[II-JCKEOP01-01] Differentiation of mesenchymal stem cells into cardiomyocytes is regulated by miRNA-1-2 via WNT signaling pathway

○Xing Shen[1], Bo Pan[1], Huiming Zhou[1], Lingjuan Liu[1], Tiewei Lv[1], Jing Zhu[1],Xupei Huang[2], Jie Tian[1]
（1.Department of Cardiology, Chongqing Medical University ,Chongqing-City, China, 2.Department of Biomedical Science, Charlie E. Schmidt College of Medicine, Florida Atlantic University, Boca Raton, FL, USA）

Objective: To investigate the role of miR1-2 in differentiation of mouse BMSCs into cardiomyocyte-like cells and reveal the involved signaling pathways in the procedure. **Methods**: Mouse BMSCs were treated with miR1-2 and 5-azacytine (5-aza). The expression of cardiac cell markers: NKx2.5, cTnI and GATA4 in BMSCs were examined by qPCR, The apoptosis rate was detected by flow cytometry and the activity of the Wnt/β-catenin signaling pathway was evaluated by measuring the upstream protein of this signaling pathway. **Results**: After overexpression of miR1-2 in mouse BMSCs, the apoptosis rate was significantly lower than the 5-aza group, While the expressions of cardiac-specific genes: such as Nkx2.5 ,cTnI and GATA4 were significantly increased compared to the control group and the 5-aza group. Meanwhile, overexpression of miR1-2 in mouse BMSCs enhanced the expression of wnt11, JNK, β-catenin and TCF in the Wnt/β-catenin signaling pathway. Use of LGK-974, an inhibitor of Wnt/β-catenin signaling pathway, significantly reduced the expression of cardiac-specific genes and partially blocked the role of the miR1-2. **Conclusion**: Overexpression of miR1-2 in mouse BMSCs can induce them toward cardiomyocyte differentiation via the activation of the Wnt/β-catenin signaling pathway. Comparing with 5-aza, miR1-2 induced differentiation of BMSCs into cardiomyocytes is stronger and safer.

6:15 PM - 7:15 PM (Sat. Jul 8, 2017 6:15 PM - 7:15 PM E-Oral Presentation Area)

[II-JCKEOP01-02] Postoperative mortality and respiratory complications in CHD patients with heterotaxy and their relationship with ciliary dysfunction

○Weicheng Chen, Shuolin Li, Sida Liu, Guoying Huang, Bing Jia （Heart Center, Children's Hospital of Fudan University, Shanghai, China）

Recent studies have revealed an association among heterotaxy, congenital heart disease, and primary ciliary dyskinesia (PCD). Thus airway ciliary dysfunction (CD) similar to that of PCD may have relevance for increased respiratory complications in heterotaxy patients. Thus we explore the mortality and respiratory complications in heterotaxy patients as compared to CHD patients without heterotaxy in china.
A retrospective review of patients undergoing cardiac surgery was undertaken at our hospiatl between Jan, 1st, 2008 and Dec, 31th, 2014, which was performed on postsurgical outcomes of 137 patients with heterotaxy and congenital heart disease exhibiting the full spectrum of situs abnormalities associated with heterotaxy. As controls patients, 881 cardiac surgical patients with congenital heart disease, but without laterality defects, were selected.
We found the postsurgical deaths (16.9% vs 4.4%; OR, 3.2), mean length of postoperative hospital stay

(12.5 vs 9.2 days; OR, 3.1), mechanical ventilation(58 vs 41 hours; OR, 3.2) and ICU stay (92 vs 73 hours; OR, 2.3) were significantly increased in the heterotaxy patients. Also elevated were number of reintubation (1.6 vs 1.1 hours; OR, 3.3) ,salvage (11.4% vs 5.2%: OR, 2.2) and fever (66.4% vs 34.9%; OR, 3.7).
Our findings show heterotaxy patients had more postsurgical events with increased postsurgical mortality and risk for respiratory complications as compared to control patients. We speculate that some respiratory defects may contribute to the increased mortaliy and respiratory complications in heterotaxy patients.

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[II-JCKEOP01-03] Cardiac function of the fetuses with positive maternal anti-SS-A antibody

○Yasuki Maeno, Yozo Teramachi, Akiko Hirose, Yasuyuki Kagiyama, Shintaro Kishimoto, Yusuke Koteda, Kenji Suda （Department of Pediatrics, Kurume University School of Medicine, Japan）

Maternal anti SS-A antibody affects to the fetal heart and causes heart block and myocarditis. Detection of the damage to the atrioventricular node is well documented. However, the incidence of the myocardial damage is not well known. Therefore, we investigated the cardiac function in the fetuses with maternal anti SS-A antibody.
Method: This is a retrospective study in single center. We reviewed our clinical record of fetal echocardiography in 5 years from Jan. 2012 to Dec 2016. In this period, 24 cases had fetal echocardiography due to maternal positive anti SS-A antibody without fetal heart block. Of the 24, 18 cases had sequential examination from first trimester. Tei index of the right and left ventricle were measured.
Results: Of the 24, first examination were 18w3d to 38w1d (median 22w0d). During pregnancy, 2 cases (8%) had high Tei index >0.7, and other 10 (42%) had relatively high Tei index >0.6. Of the 12, 6 had Tei index >0.6 also at the latest examination before birth, but, the other had transient elevation. Of the 20 with postnatal data (born at 36w5d to 41w2d, median 39w2d; birth weight of 1918g to 3336g, mean 2778g), no case had clinical symptom of cardiac dysfunction, although one had transient tachypnea, one had small for gestational age, and the other had premature rupture of membrane.
Conclusions: Our data suggest high incidence of abnormal Tei index in the fetuses with maternal anti SS-A antibody. Although the cardiac dysfunction seems to be transient and subclinical in most of the cases, further study with postnatal follow-up are required.

6:15 PM - 7:15 PM (Sat. Jul 8, 2017 6:15 PM - 7:15 PM E-Oral Presentation Area)

[II-JCKEOP01-04] Improvement of portopulmonary hypertension after liver transplantation

○Shiro Baba[1], Daisuke Yoshinaga[1], Kouichi Matsuda[1], Kentaro Akagi[1], Shinji Uemoto[2], Takuya Hirata[1] （1.Department of Pediatrics, Kyoto University, Japan 2.Division Hepato-Billary-Pancreatic Surgery and Transplantation Department, Kyoto University, Japan）

Portopulmonary hypertension (POPH), associated with hepato-biliary diseases, is believed to improve after treatments of these liver diseases. However, not all patients improve their POPH even after the treatments. Thus, we retrospectively investigated 19 POPH patients (male: 8, female 11) in Kyoto university hospital whether their POPH improved after liver transplantation (LTx).
Diagnoses of these patients were biliary atresia (16), portosystemic shunt (2) and fulminant hepatitis (1). Accompanied diagnoses were tetralogy of Fallot (1), atrial septal defect (1), Alagille syndrome (1), Byler disease (1) and left isomerism (1). LTx was performed at the age of 4 months to 21 years old to all the patients, average was 8.6 years old. Mean pulmonary artery pressure (mPAp) and pulmonary artery resistance (Rp) at the timing of before LTx, 5.8±4.0 months and 6.5±4.9 years after LTx were 36.2±7.3, 30.3±7.5, 31.2±9.4mmHg、4.2±2.2, 5.4±2.6, 5.4±3.6U・m2, respectively. In these, only 5 patients' mPAp normalized, <25mmHg, after LTx. Previously, it is reported that reduction rate of mPAp of portosystemic shunt patients was 24.7±17.5% in 3 months after LTx. Comparing with this splendid results, mPAp of our patients reduced only around 15% after LTx and still most of the patients use anti-pulmonary hypertension drugs.
It is said that POPH improves after treatment of primary hepato-biliary diseases. But there are many patients whose mPAp still high even after radical treatments including LTx. For these patients, we have to pay more attention to re-worsening of POPH.

6:15 PM - 7:15 PM (Sat. Jul 8, 2017 6:15 PM - 7:15 PM E-Oral Presentation Area)

[II-JCKEOP01-05] Surgical outcomes of open surgical repair for coarctation of the aorta in adolescent and adult in the era of endovascular therapy

○Yoshikatsu Saiki, Osamu Adachi, Masatoshi Akiyama, Ichiro Yoshioka, Satoshi Kawatsu, Yukihiro Hayatsu,Yusuke Suzuki, Konosuke Sasaki, Kiichiro Kumagai, Shunsuke Kawamoto, （Division of Cardiovascular Surgery, Tohoku University, Sendai, Japan）

Background: Surgical repair for coarctation of the aorta (CoA) in adolescent and adult remains challenging since the lesion typically involves extensive area from the transverse arch to the proximal descending thoracic aorta. Although endovascular therapy has been evolving, contemporary surgical outcome has yet to be definitively determined. **Purpose:** We sought to evaluate our short and mid-tem outcomes after repair of adult CoA. **Patients and methods:** Between 2001 and 2016, 10 patients (mean 37.2 ys, range, 17-64 ys) underwent reparative procedures for CoA. Of those, primary CoA was diagnosed in 6 patients, Re-CoA in 2 and aortic aneurysm after previous patch aortoplasty in 2. Only one patient underwent TEVAR. The remaining 9 patients underwent open surgery. Left thoracotomy approach was employed when proximal aortic clamp was placed between the left carotid and subclavian arteries, whereas median sternotomy ± anterolateral thoracotomy with selective cerebral perfusion (SCP) were employed to perform total or partial aortic arch replacement. **Results**: Nine patients were associated with aneurysmal changes in the aorta. For those who underwent SCP (n=7), mean SCP time was 151.6 min (27-422 min). There were no hospital mortality, stroke or spinal cord injury. No aortic event or cerebrovascular complications were encountered during long-term follow-up. **Conclusions:** CoAs in adolescent and adult were frequently associated with aortic aneurysm necessitating the use of SCP; thus, require invasive surgery. However, the short and mid-tem outcomes are favorable and durable.

6:15 PM - 7:15 PM (Sat. Jul 8, 2017 6:15 PM - 7:15 PM E-Oral Presentation Area)

[II-JCKEOP01-06] Perioperative effects of the Tolvaptan for the Fontan Patients

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We have increasing an opportunity to give Tolvaptan in perioperative period, but a case of the Fontan patient is rarely reported. Recently we have some Fontan patients who improved prolonged pleural drainage after Tolvaptan administration.
Subject and Method
From Jan 2009 to 2016 Aug 14 patients underwent extracardiac TCPC procedure. Tolvaptan was administrated to the 5 patients whose urine output decreased and needed prolonged pleural drainage (Group T). And we compared another 7 patients as control (Group C). A dosage of Tolvaptan was 0.75-7.5mg (0.25mg/kg).
Results
Age at operation (y) : 3.5±0.5 Group T, 2.7±0.3 Group C (p=0.01), BW(kg),:12.1±0.9 Group T, 11.6±1.6 Group C (p= 0.5), Duration of pleural drainage (d) : 21.6±9.6 Group T, 4.6±1.8 Group C (p=0.0002). Age at the Fontan operation was higher and the duration of pleural drainage was longer in T group. There was no differences between two gropes in PA index, Rp(μ m^2), PA pressure(mmHg), and EF(%), EDP(mmHg). Four cases (80%) were extubated in operation room in Group T and 5 cases (55%) in Group C. ICU stay (d) was 3.6±3.5, Group C:2.0±1.7 (p=0.2). In group T, urine volume before and after Tolvaptan administration was 37:68 (ml/kg) (p=0.04), Pleural effusion drainage (ml/kg) was 10.8:6.8 (p=0.13). Urine volume and less pleural effusion were observed after administration. Duration of pleural drainage was 6.7 days（2-13） in average. Blood test showed after administration, TP: 3.8±0.5 Group T, 4.4±0.7 Group C (p=0.003), Na: 139.4±6.4 Group T, 138.0±23.9 Group C (p=0.7), K: 4.0±0.6 Group T, 3.9±0.5 Group C (p=0.2).
Conclusion
In Perioperative prolonged pleural drainage Fontan cases, urine volume increased and pleural effusion decreased after Tolvaptan administration. Chest drainage tube was extubated without delay after administration except for chylothorax case.
Any side effect, hypernatremia nor liver dysfunction was not observed.

6:15 PM - 7:15 PM (Sat. Jul 8, 2017 6:15 PM - 7:15 PM E-Oral Presentation Area)

[II-JCKEOP01-07] Enlargement of the pituitary gland in patients after Fontan operation: another portal circulation

○Yusaku Nagatomo, Jun Muneuchi, Mamie Watanabe, Ryohei Matsuoka, Hiromitsu Shirouzu, Seigo Okada, Chiaki Iida, Kunitaka Joo （Pediatrics, Kyushu Hospital, Kitakyushu city, Japan）

<Abstract>
Objective: There are two portal circulation systems in a human body, one is in the liver, and another is in the pituitary. Recently, it has been widely recognized about problems on the liver in patients after Fontan operation. However, it is not known about the pituitary. The aim of this study is to investigate the size of the pituitary after Fontan operation.
Methods: We investigated the size of the pituitary using the MRI brain imaging in the 40 adolescent patients after Fontan operation. We measured the height (H) and length (L) of the pituitary in sagittal section, and the width (W) of that in coronal section. We estimated the volume of that as follows: (H× L× W)×1/2. We compared them between the enrolled patients and the 60 age-matched control subjects without structural disorder.
Results: Fontan operation was performed at the median age of 3.3 years (1.6-5.7). The MRI was undertaken at the median age of 9.3 years (7.2-22.0). The size of the pituitary was larger than that of controls (H 7.5±1.3mm vs. 5.9±1.2mm, L 9.7±1.3mm vs. 8.0±1.3mm, W 12.9±1.8mm vs. 11.0±1.9mm, p<0.01, respectively). The volume of that was 935.0±216.8 mm3, which was enlarged nearly twice larger than the controls (534.5±208.7mm3) (p<0.01). When examining the correlation between each clinical data and pituitary volume, there were positive correlations between age (R=0.46), central venous pressure (R=0.51) and the pituitary volume.
Conclusion: Enlargement of the pituitary may develop due to high central venous pressure. We should pay attention to pituitary function in patients after Fontan operation.

6:15 PM - 7:15 PM (Sat. Jul 8, 2017 6:15 PM - 7:15 PM E-Oral Presentation Area)

[II-JCKEOP01-08] Utility of and hemodynamic association with alpha$_1$-antitrypsin clearance in Fontan patients

○Satoshi Masutani, Yoichi Iwamoto, Hirotaka Ishido, Akiko Yana, Seiko Kuwata, Clara Kurishima, Hideaki Senzaki （Pediatric Cardiology, Saitama Medical University Saitama Medical Center, Japan）

【 Background】 Prevention and early recognition of protein-losing enteropathy (PLE) are important in Fontan patients (F). α_1 -antitrypsin (AT) is an endogenous substance which is not secreted or reabsorbed in the intestine in normal condition. AT clearance (C_{AT}) increases as PLE advances. We assessed its relation with hemodynamics and detectability of a subtle protein losing which cannot be detected by serum albumin level (Alb).
【 Methods】 This study included 42 F （3-24, median 7.7 years） including 2 active PLEs. We obtained C_{AT} (mL/day): daily AT excretion (mg/day) measured by 3 days feces storage / plasma AT concentration (mg/dL) * 100. We compared C_{AT} with Alb, and hemodynamic variables obtained by simultaneous cardiac catheterizations.
【 Results】 C_{AT} were elevated (63.7, 90.6 mL/day) in 2 active PLE (both Alb 2.8 mg/dL). C_{AT} after remission of PLE (Alb 4.1, 3.8 mg/dL) were normal (2.2, 1.2 mL/day). After exclusion of active PLEs (range 1.1-21.4, median 6.0 mL/day), lower serum Alb correlated with lower C_{AT} (P< 0.01). All three patients with Alb<3.5 mg/dL showed normal C_{AT} <8 mL/day, and their ALB recovered without PLE therapy. All three patients with C_{AT} >13 mL/day had ALB >4 mg/d. C_{AT} did not correlate with the duration after F operation, CI, SaO2, Rp, HR, or CVP.
【 Discussion】 C_{AT} may be useful for the differential diagnosis of hypoalbuminemia. There should be the factors other than CVP which induce intestinal protein losing. Clinical significance of subtle protein losing deserves future investigations employing a large number of patients.

JCK E-Oral Presentation

JCK E-Oral Presentation 2 (III-JCKEOP02)

Chair:Atsuko Kato(Division of Cardiology, The Labatt Family Heart Centre, Department of Pediatrics, The Hospital for Sick Children, University of Toronto, Toronto, Canada)
Chair:Takaya Hoashi(Department of Pediatric Cardiovascular Surgery, National Cerebral and Cardiovascular Center, Suita, Japan)
Sun. Jul 9, 2017 1:00 PM - 2:00 PM E-Oral Presentation Area (Exhibition and Event Hall)

[III-JCKEOP02-01] Transcatheter closure of doubly committed VSDs - a 5 year single centre experience

○Tran Cong Bao Phung, Do Nguyen Tin （Cardiology Department, Children Hospital 1, Ho Chi Minh City, VietNam）

1:00 PM - 2:00 PM

[III-JCKEOP02-02] Effect of fenestration on ventricular-vascular coupling chronically after Fontan operation - Cardiac magnetic resonance study

○Yoichi Iwamoto, Seiko Kuwata, Akiko Yana, Hirotaka Ishido, Satoshi Masutani, Hideaki Senzaki （Division of Pediatric Cardiology, MFN Center, Saitama Medical University Saitama Medical Center, Japan）

1:00 PM - 2:00 PM

[III-JCKEOP02-03] Development of a New and Rapid 3D Printing System for Manufacturing Super Flexible Replicas of Congenital Heart Disease

○Isao Shiraishi[1], Kennichi Kurosaki[1], Suzu Kanzaki[2], Takaya Hoashi[3], Hajime Ichikawa[3] （1.Department of Pediatric Cardiology National Cerebral and Cardiovascular, Japan, 2.Department of Radiology, National Cerebral and Cardiovascular Center, Japan, 3.Department of Pediatric Cardiac Surgery, National Cerebral and Cardiovascular Center, Japan）

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[III-JCKEOP02-04] Minimally invasive epicardial implantable cardioverter-defibrillator placement in a young child

○Wakana Maki[1], Hiroko Asakai[1], Kazuhiro Shiraga[1], Takahiro Shindo[1], Yoichiro Hirata[1], Ryo Inuzuka[1], Tomoyuki Iwase[2], Tetsuhiro Takaoka[2], Akihiro Masuzawa[2], Yasutaka Hirata[2], Akira Oka[1] （1.Department of Pediatrics The University of Tokyo Hospital, Japan 2.Department of Cardiac Surgery The University of Tokyo Hospital,Japan）

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[III-JCKEOP02-05] Fetal Echocardiography Characteristics in Vietnam Population

○Tran Cong Bao Phung, Vu Minh Phuc, Do Nguyen Tin, Nguyen Tri Hao, Phan Hoang Yen, Phan Tien Loi （Cardiology Department, Children Hospital 1, Ho Chi Minh City, VietNam）

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[III-JCKEOP02-06] Mitral valve replacement using stented bovine jugular vein graft (Melody valve) in infants and small children

○Atsuko Kato[1], Osami Honjo[2] （1.Division of Cardiology, The Labatt Family Heart Centre, Department of Pediatrics, The Hospital for Sick Children, University of Toronto, Toronto, Canada, 2.Department of Cardiovascular surgery, The Labatt Family Heart Centre, The Hospital for Sick Children, University of Toronto, Toronto, Canada）

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[III-JCKEOP02-07] Long-term Surgical Outcome of Transposition of the Great Arteries with Intact Ventricular Septum and Left Ventricular Outflow Tract Obstruction

○Akihisa Furuta, Mitsugi Nagashima, Takahiko Sakamoto, Goki Matsumura, Kentaro Umezu, Jin Ikarashi, Junko Katagiri, Hironori Murakami （Department of Cardiovascular Surgery, Tokyo Women's University, Japan）

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[III-JCKEOP02-08] Successful biventricular conversion late after primary one and one-half ventricle repair

○Takaya Hoashi[1], Masataka Kitano[2], Masatoshi Shimada[1], Kenichi Kurosaki[2], Hajime Ichikawa[1] （1.Department of Pediatric Cardiovascular Surgery, National Cerebral and Cardiovascular Center, Suita, Japan, 2.Department of Pediatric Cardiology, National Cerebral and Cardiovascular Center, Suita, Japan）

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[III-JCKEOP02-01] Transcatheter closure of doubly committed VSDs - a 5 year single centre experience

○Tran Cong Bao Phung, Do Nguyen Tin （Cardiology Department, Children Hospital 1, Ho Chi Minh City, VietNam）

Background: There remains international debate regarding efficacy and safety of transcatheter closure of doubly committed VSDs (DCVSD). This study reports a 5 year single centre experience.
Methods: Retrospective review, October 2009 to Jul 2014; 44 patients underwent device closure of DCVSD. Selection criteria: Weight >10kg, no severe/moderate AR or cusp prolapse, defect <7mm, no other intra-cardiac abnormalities, plus evidence of pulmonary hypertension, left heart volume loading, or trivial/mild AR or cusp prolapse. Technique: Anterograde approach and AV loop, angiographic re-evaluation, echocardiographic evaluation of AR and outflow.
Results: Median age 63 months (10-170), weight 18kg (8-32), defect 3.7mm (2-6). Associated abnormalities: trivial to mild AR; 1 (2.3%), left heart dilation; 10 (22.7%), MR; 3 (6.8%), coronary cusp prolapse: 11 (25.0%). Devices used: PFM Coil: 10 (22.7%), ADO II: 21 (47.7%), PFM Coil and ADOII 13 (29.5%). Post-procedure murmur in 20: residual shunt disappeared on echo <48 hours in 18 (56,2%) the remainder after 6 months. Complications: haemolysis: 1 (2.3%) referred for surgery, embolization: 1 (2.3%), residual shunt 1 (2.3%) referred for surgery, RV outflow obstruction: 5 (11.4%) all resolved <3 months, AR increased: 4 (9.0%) 3 recovered to baseline <1 month, one referred for surgery. Mild LV outflow obstruction: 1 (2.3%) resolved <2 months.
Conclusion: Device closure of DCVSD is a controversial but effective and safe option in selected patients. Coils are associated with a greater incidence of haemolysis than ADOII or Coil &ADOII.

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[III-JCKEOP02-02] Effect of fenestration on ventricular-vascular coupling chronically after Fontan operation - Cardiac magnetic resonance study

○Yoichi Iwamoto, Seiko Kuwata, Akiko Yana, Hirotaka Ishido, Satoshi Masutani, Hideaki Senzaki （Division of Pediatric Cardiology, MFN Center, Saitama Medical University Saitama Medical Center, Japan）

[Background] We assessed how fenestration affects ventricular contractility and ventricular-vascular coupling (VVC) chronically after Fontan operation employing cardiac magnetic resonance imaging (cMRI).

[Methods] This study included consecutive 44 Fontan patients (8.8±4.2 years) with cMRI. EDVI, ESVI and stroke volume index (SVI) were measured by volumetry. Arterial effective elastance (Ea) was calculated as mean blood pressure (BP) divided by SVI. End-systolic elastance (Ees) was calculated by our developed single-beat method using BP, arm equilibrium pressure, and ESVI. We measured circulating blood volume by dye dilution method and calculated venous capacitance (VC) by blood volume and arm equilibrium pressure. We compared those in patent fenestration group (N=15, F group) with those in closed fenestration group (N= 29, non-F group).

[Results] F group had significantly lower central venous pressure (CVP) and higher VC than non-F group (9.4 vs. 11.4 mmHg, 3.5 vs. 2.5 ml/mmHg, p<0.05). F group had significantly higher EDVI and ESVI than non-F group (111.5 vs. 96.0 ml/m2 : p=0.02, 62.5 vs. 50.5 ml/m2 : p<0.01). F group had tendencies toward higher SVI and lower Ea than non-F group. There was no significant difference between F group and non-F group in Ees (2.3 vs. 2.4 mmHg/ml/m2), or in biomarker levels on renin-angiotensin-aldosterone system, heart failure, and fibrosis.

[Conclusion] Fenestration may be protective in suppressing venous maladaptation, keeping lower CVP and contributing to keep preload reserve and VVC in those chronically after Fontan operation.

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[III-JCKEOP02-03] Development of a New and Rapid 3D Printing System for Manufacturing Super Flexible Replicas of Congenital Heart Disease

○Isao Shiraishi[1], Kennichi Kurosaki[1], Suzu Kanzaki[2], Takaya Hoashi[3], Hajime Ichikawa[3] (1.Department of Pediatric Cardiology National Cerebral and Cardiovascular, Japan, 2.Department of Radiology, National Cerebral and Cardiovascular Center, Japan, 3.Department of Pediatric Cardiac Surgery, National Cerebral and Cardiovascular Center, Japan)

[Backgrounds] Recently, 3D printing technology has been applied for diagnosis and surgical simulation of congenital heart disease. We have shown a unique technology that manufactures precise and super flexible polyurethane replicas of congenital heart disease by means of stereolithography followed by vacuum casting. To improve limitations in time and cost of this technique, we here developed a novel and rapid 3D printing system in collaboration with several Japanese chemical and mechanical companies. [Materials and Methods] The new 3D printing machine consists of 4 inkjet heads with 200x500mm size that can reproduce not only child but also adult heart and thoracic aorta. Inkjet materials with similar texture to the human heart were also developed. We preliminary manufactured several different types of heart replicas including ASD, VSD, TOF, and ccTGA. [Results] After printing conditions have been optimized, super flexible replicas of congenital heart disease were manufactured with precise external surface and detailed internal structure of the atria and ventricles. The replicas exhibited similar texture to the real heart and allow surgeons simulation surgery by cutting and suturing. Whole process takes approximately 48 hours including initial 3D image processing. [Conclusions] We have successfully developed a new 3D inkjet printing technology based on industry -academia collaboration. Further improvements of ink materials and printing technologies are necessary to reduce time and cost of the replicas to be used all over the world.

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[III-JCKEOP02-04] Minimally invasive epicardial implantable cardioverter-defibrillator placement in a young child

○Wakana Maki[1], Hiroko Asakai[1], Kazuhiro Shiraga[1], Takahiro Shindo[1], Yoichiro Hirata[1], Ryo Inuzuka[1], Tomoyuki Iwase[2], Tetsuhiro Takaoka[2], Akihiro Masuzawa[2], Yasutaka Hirata[2], Akira Oka[1] (1.Department of Pediatrics The University of Tokyo Hospital, Japan 2.Department of Cardiac Surgery The University of Tokyo Hospital,Japan)

The use of implantable cardioverter-defibrillator (ICD) therapy for prevention of sudden cardiac death in the pediatric population has been increasing. However, the use of transvenous ICD lead systems is limited in younger children and in patients with congenital heart disease. Alternative techniques such as epicardial patch and subcutaneous systems require extensive surgery with often a full sternotomy or thoracotomy.
We report a case of successful minimally invasive pericardial ICD implantation in a 16kg child. The patient was a 6-year-old girl with left ventricular non-compaction. She had ventricular fibrillation (VF) arrest from which she was successfully resuscitated with an automated external defibrillator (AED). She recovered without any neurological consequences and ICD implantation was indicated for secondary prevention. Through a small subxiphoid incision, a transvenous ICD dual coil was advanced and screwed into the oblique sinus pericardium under fluoroscopic guidance. An additional sense-pace lead was sutured onto the RV apex, and the generator was placed in the upper abdominal wall through the same incision. Defibrillation threshold testing demonstrated successful defibrillation at 15J. Post implant, the patient had two episodes of appropriate shock due to VF. The ICD system continues to show stable impedance at 3 months follow-up. To our knowledge, this is the first case report in Asia of pericardial ICD placement with a minimally invasive subxiphoid approach.

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[III-JCKEOP02-05] Fetal Echocardiography Characteristics in Vietnam Population

○Tran Cong Bao Phung, Vu Minh Phuc, Do Nguyen Tin, Nguyen Tri Hao, Phan Hoang Yen, Phan Tien Loi (Cardiology Department, Children Hospital 1, Ho Chi Minh City, VietNam)

BACKGROUND:
To evaluate the incidence of fetal cardiac abnormalities and to establish normative reference ranges of fetal cardiac characteristics in Vietnamese population.
METHODS:
A cross_sectional study was undertaken on pregnancies examined at Thu Duc district hospital, Ho Chi Minh city from November 2012 to April 2013.
RESULTS:
Two hundred and twenty pregnant women were enrolled in this study.
1. Characteristics: mothers' mean age (years): 30.3 + 5.0 ; Mean gestational age (weeks) : 33.6 + 4.5 ; Population characteristics: intrauterine growth retardation (1.7%); diabetes (10.1%); genetic mutation (0.8%); abnormalities in routines echo scan (4.2%); twins (1.7%); previous child with congenital heart disease (0.8%); fetal tachycardia (0.8%); screening (78.2%).
2. Fetal cardiac characteristics: heart chest ratio (CTAR, %) = 31.4 + 4.8 . The mean cardiac dimension on 2D (mm): foramen oval = 4.9 + 1.4; mitral valve = 9.9 + 2.1; tricuspid valve = 10.4 + 2.4; aortic valve = 6.3 + 1.4; pulmonary valve annulus = 7.9 + 1.8; pulmonary trunk = 7.8+ 1.9; right pulmonary artery = 3.9 + 1.0 ; left pulmonary artery = 3.9 + 0.9 ; aortic isthmus = 4.9 + 1.0. The mean velocities : aortic

valve = 0.66 + 0.15(0.35-1.00); pulmonary valve = 0.61 + 0.14); aortic arch = 0.74 + 0.21
3.one fetus with ventricular septal defect (0.45%), one fetus with supra ventricular tachycardia (0.45%), one with sinusbradycardia (0.45%) and one with coarctaion (0.45%).
CONCLUSIONS:
This is the first report on normal ranges of fetal cardiac dimensions especially in the third trimester in Vietnam.

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[III-JCKEOP02-06] Mitral valve replacement using stented bovine jugular vein graft (Melody valve) in infants and small children

○Atsuko Kato[1], Osami Honjo[2] (1.Division of Cardiology, The Labatt Family Heart Centre, Department of Pediatrics, The Hospital for Sick Children, University of Toronto, Toronto, Canada, 2.Department of Cardiovascular surgery, The Labatt Family Heart Centre, The Hospital for Sick Children, University of Toronto, Toronto, Canada)

BACKGROUND: Melody® valve (Medtronic, MN) implantation in the mitral position is a novel procedure for the small mitral valve (MV) annulus, which does not require anticoagulation. We sought to analyze our initial experience with mitral valve replacement (MVR) with Melody® valve.
METHOD: The records of patients who underwent MVR using Melody® from 2014 to 2016 were retrospectively reviewed. The Melody® (22 mm) valve was prepared by sewing a 3.5 mm Gore-Tex tube graft at the middle of the stent as a cuff and resecting one or three zigs to avoid left ventricular outflow tract (LVOT) obstruction.
RESULTS: Five patients (age, 11 months, 5 - 16 months; weight, 6.8 kg, 4.6 - 8.6 kg) were included. All patient had dysplastic MV, including severe mitral stenosis (n=3) and/or regurgitation (n=3). Three patients had Melody® valve MVR as a salvage procedure: mechanical valve thrombosis (n=2) and tissue valve dysfunction (n=1). The valve was inflated to 18 mm (n=2) or 20 mm (n=3). Intraoperative echocardiography revealed trivial or none regurgitation in all patients with mean pressure Doppler gradient across the valve of 2 mmHg (1 – 4 mmHg), and peak LVOT gradient of 5 mmHg (0 – 12 mmHg). All the patients but one were discharged home. There was one patient who had sudden death at 3 months after surgery.
CONCLUSIONS: Melody® valve MVR is a viable alternative to mechanical MVR in small children. Early functional outcome of this procedure is excellent, whereas long-term outcome is to be investigated.

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[III-JCKEOP02-07] Long-term Surgical Outcome of Transposition of the Great Arteries with Intact Ventricular Septum and Left Ventricular Outflow Tract Obstruction

○Akihisa Furuta, Mitsugi Nagashima, Takahiko Sakamoto, Goki Matsumura, Kentaro Umezu, Jin Ikarashi, Junko Katagiri, Hironori Murakami （Department of Cardiovascular Surgery, Tokyo Women's University, Japan）

Objective
Left ventricular outflow tract obstruction (LVOTO) is sometimes combined with transposition of the great arteries (TGA) with intact ventricular septum (IVS). The purpose of this study is to evaluate the long-term surgical outcome of TGA with IVS and LVOTO.
Patients
Between 1980 and 2016, 13 patients who underwent the surgical repair for TGA with IVS and LVOTO (peak gradient on LVOT>30mmHg) were retrospectively reviewed. Type of LVOTO included subaortic in 7 and valvular in 6. Age at definitive repair was 37.5±42.5 month-old, and body weight was 6.8±3.8 kg. Definitive operation included Senning operation in 10, arterial switch operation in 1, truncal switch operation in 1 and modified Fontan operation in 1.
Results
Follow-up period was 19.6±7.4 years. There was no hospital death and 1 late death. The survival rate was 90.0% at 20 years. Re-intervention was performed in 4 patients. Freedom from re-intervention was 66.7 % at 20 years. Latest angiography revealed LVOT peak gradient of 8.2±8.4 mmHg, and systemic ventricular ejection fraction of 53.1±11.8 %. Latest echocardiography revealed moderate tricuspid valve regurgitation in 1, and LVOT flow of 1.7±0.4 m/s. Three patients showed atrioventricular rhythm disturbance in electrocardiogram.
Conclusions
Long-term surgical outcome of TGA with IVS and LVOTO was satisfactory in terms of the relief of LVOTO. However, the further careful observation is mandatory because some patients may present tricuspid valve regurgitation and rhythm disturbance associated with systemic ventricular dysfunction in the late period.

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[III-JCKEOP02-08] Successful biventricular conversion late after primary one and one-half ventricle repair

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（1.Department of Pediatric Cardiovascular Surgery, National Cerebral and Cardiovascular Center, Suita, Japan, 2.Department of Pediatric Cardiology, National Cerebral and Cardiovascular Center, Suita, Japan）

A six-year-old girl with unbalanced atrioventricular septal defect, hypoplastic right ventricle and severe common atrioventricular valve regurgitation developed patient-prosthetic mismatch. At six months old, she underwent primary one and one-half ventricle repair and replacement of left side atrioventricular valve. A catheter examination showed that her right ventricular end-diastolic volume increased from 39.4 ml/m^2 one year after the previous surgery, to 70 ml/m^2 at preoperative evaluation. Thus, at the timing of redo left side atrioventricular valve replacement, she was successfully converted to biventricular circulation. The postoperative course was uneventful, and the right atrial pressure was 7 mmHg before discharge.