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）

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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）

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[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）

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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

○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）

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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[2], Hideto Shinpo[1] （1.Departmemt of Thoracic and Cardiovascular Surgery, Mie University, Japan 2.Departmemt of Pediatrics, Mie University, Japan）

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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）

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

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（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.

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[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

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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.

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

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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.

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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

○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.

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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.

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

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<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.

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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）

【 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.